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Cor et Vasa

SUPLEMENTUM 3
zaměřené na kazuistiky

Aneurysma vena poplitea u pacientky s plicní embolií

Double-vessel disease of acute myocardial infarction

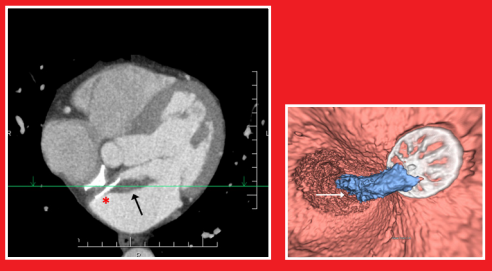
Off label treatment of intraabdominal dissection with iliac stent graft extension

***Brucella* prosthetic valve endocarditis**

Persistent left superior vena cava, permanent pacemaker implant

Chronic venous insufficiency in a 55-year-old female

Epicardial Pacing Lead Implantation in Complete Heart Block



CT scan showing a mass (→) on the left atrial disk (*) of the PFO occluder. Three-dimensional CT scan reconstruction (view from the left atrium) demonstrating a mass (→) on the left atrial disk of the PFO occluder. *(Dúbrava J, Hajzoková M, Drangová E, Gašparovič I. Late-onset acute limb ischemia after transcatheter patent foramen ovale closure with Occlutech Figulla Flex II PFO Occluder. Kazuistika, str. 89–93).*

Cor et Vasa

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Zvláštní suplementum *Cor et Vasa* zaměřené na kazuistická sdělení

(Special supplement of the *Cor et Vasa* journal focused on case reports)

Michael Aschermann

II. interní klinika, 1. lékařská fakulta Univerzity Karlovy a Všeobecná fakultní nemocnice v Praze, Praha, Česká republika

Kvalitní kazuistická sdělení patří ke standardním článkům kvalitních odborných medicínsky zaměřených časopisů. Náš časopis *Cor et Vasa* jim řadu let věnoval pozornost ve formě jednoho čísla ročně, které bylo věnováno právě kazuistikám. Bylo to pravidelně páté číslo každého ročníku v posledních deset letech. Výbor ČKS v minulém roce rozhodl, že kazuistická sdělení nebudou nadále publikována v časopisu *Cor et Vasa*, ale budou nově publikována pouze v druhém časopisu, který ČKS vydává – *Cor et Vasa Case Reports*. Vzhledem k tomu, že v redakčním systému *Cor et Vasa* bylo v době uvedeného rozhodnutí výboru ČKS již přijato více kazuistik, bylo dále rozhodnuto, že tyto kazuistiky budeme publikovat ve speciálním suplementu *Cor et Vasa*. Hlavním důvodem byl fakt, že uvedené kazuistiky byly přijaty do *Cor et Vasa* s impakt faktorem a jejich přesunem do *Cor et Vasa Case Reports* by tuto hodnotu ztrácely. Věříme, že čtenáři přijmou toto suplementum pozitivně, neboť zařazená kazuistická sdělení prošla standardním recenzním řízením a přinášejí velké množství zajímavých klinických případů.

Case reports of high quality are interesting and standard part of high quality medical journals. *Cor et Vasa* journal published case reports in special issue every year during last ten years. The Board of the Czech Society of Cardiology (ČKS) decided last year, that case reports will not be published in *Cor et Vasa* journal, but will be published only in the second journal, published by ČKS – *Cor et Vasa Case Reports* journal. Since at that time there were several case reports already accepted for publication in *Cor et Vasa* (therefore accepted for publication in the journal with impact factor), the decision was made to publish those case reports in this special supplementum of *Cor et Vasa* journal with impact factor. (In case of transfer of accepted manuscripts into the *Cor et Vasa Case Reports* journal, the impact factor will be lost.) We hope, that this special supplementum will be accepted well by readers, since it presents best of case reports which went through standard review process and offer very interesting cases.

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Brucella prosthetic valve endocarditis: A case study

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SOUHRN

Bakterie rodu *Brucella* jsou gramnegativní kokobacily, které typicky vyvolávají brucelózu projevující se nespecifickými symptomy, jako jsou horečka, únava a artralgie. Jedná se o multisystémové onemocnění, které může vést ke komplikacím, přičemž nejčastějším – i když vzácným – kardiovaskulárním projevem je brucelová endokarditida. Toto infekční onemocnění je odpovědné za 80 % případů úmrtí v souvislosti s brucelózou, často v důsledku srdečního selhání. K onemocnění primárně dochází po konzumaci nepasterizovaných mléčných výrobků. Vzhledem k rozmanitosti klinických projevů může diagnostika brucelózy představovat problém. Endokarditida umělé chlopně, vyvolaná bakteriemi rodu *Brucella*, se vyskytuje velmi vzácně a stanovení diagnózy brucelové endokarditidy a její léčba jsou obzvláště náročné. Popisujeme případ brucelové endokarditidy u muže ve věku nad 50 let s nevýznamnou anamnézou až na náhradu mitrální chlopně pro akutní revmatickou horečku. Po dobu déle než jeden rok trpěl recidivující horečkou, nočním pocením a úbytkem tělesné hmotnosti. Z laboratorních parametrů vykazovaly zvýšené hodnoty proteiny akutní fáze a jaterní enzymy, byla zjištěna hematurie a nízké hodnoty c3/c4, pozitivita antinukleárních protilátek a revmatoidního faktoru. Poslech srdce odhalil pansystolický šelest stupně 2/6 u mitrální chlopně, doprovázený slyšitelným zvukem kovové umělé chlopně. Byla provedena kultivace krevních vzorků odebraných v průběhu čtyř dnů z obou paží. První hemokultura prokázala růst gramnegativních bakterií po 72 hodinách, což vedlo k zahájení empirické léčby cefepimem a gentamicinem. Po pěti dnech byla prokázána přítomnost bakterie rodu *Brucella* a léčba byla změněna na podávání doxycyclinu, rifampicinu, a gentamicinu. Od zahájení empirické léčby nebyl v kulturách zjištěn žádný růst. Následně bylo provedeno vyšetření transezofageální echokardiografií, která odhalila útvar o velikosti 13 mm odpovídající vegetacím na umělé mitrální chlopni. Pacient byl operován a v pooperačním období nebyly pozorovány žádné komplikace.

Keywords:
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Infective endocarditis
PET/CT
Stroke

ABSTRACT

Brucella bacteria are Gram-negative coccobacilli that typically cause brucellosis, presenting with nonspecific symptoms like fever, fatigue, and arthralgia. It is a multisystem disease that can lead to complications, with brucella endocarditis being the most common cardiovascular manifestation, though rare. Brucella endocarditis accounts for 80% of brucellosis-related deaths, often due to heart failure. The disease is primarily contracted through unpasteurized dairy products. Diagnosing brucellosis can be challenging due to its diverse clinical features. Brucella-related prosthetic valve endocarditis is particularly rare and presents unique diagnostic and treatment difficulties. We present a case report of brucella endocarditis in a male patient in his 50s. His medical history was insignificant except a history of mitral valve replacement due to acute rheumatic fever. He had a history of relapsing fever, night sweat and weight loss over a year. There were diverse laboratory findings such as elevated acute phase reactants, liver enzymes, hematuria, low c3/c4, positive antinuclear antibody and rheumatoid factor. During cardiac auscultation, a pansystolic murmur grade 2/6 was detected at the mitral focus, accompanied by the audible sound of a metallic prosthetic valve. Blood cultures from both arms were taken over four days. The first culture showed Gram-negative bacterial growth at 72 hours, prompting empirical treatment with cefepime and gentamicin. After 5 days, *Brucella* was identified, and the treatment was adjusted to doxycycline, rifampicin, and gentamicin. No growth was seen in cultures taken after starting empirical treatment. Subsequently, a transesophageal echocardiogram was performed, revealing a 13 mm vegetative mass consistent with prosthetic mitral valve vegetations. The patient then underwent surgery, and no complications were observed in the postoperative period.

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Introduction

Brucella bacteria are aerobic, immobile, non-spore-forming, Gram-negative coccobacilli. Initially, brucellosis typically presents with nonspecific symptoms such as fever, fatigue, night sweats, and arthralgia.[1] Human brucellosis is a serious multisystem disease that can impact various organs. It may lead to some complications and relapses. Although cardiovascular complications are uncommon, endocarditis is the most frequently observed cardiovascular manifestation of brucellosis with 1.4%.[2] *Brucella* endocarditis accounts for 80% of *Brucella*-related deaths.[3] Death, particularly in cases where diagnosis is delayed, occurs as a result of heart failure. It can affect both natural and prosthetic heart valves. The consumption of unpasteurized milk and dairy products is a significant risk factor for brucellosis.[4] Diagnosing brucellosis can be difficult due to its systemic nature and diverse clinical presentations. *Brucella*-related prosthetic valve endocarditis is exceptionally uncommon and presents substantial diagnostic and therapeutic hurdles, especially concerning the optimal duration of treatment, selection of antibiotic regimens, and determining the timing or necessity of surgical intervention.[5]

In this case report, we will present a challenging case of *Brucella* prosthetic valve endocarditis characterized by a complex diagnostic process and extensive clinical and laboratory findings.

Presentation of case

A male patient in his 50s, residing in a rural area, presented with a year-long history of weight loss, night sweats, and nausea, accompanied by the recent onset of fever and back pain. Before his admission to our clinic, he had sought the opinions of various medical specialists at different hospitals regarding these constitutional symptoms. Over the past year, he underwent routine laboratory tests and thoracoabdominal computerized tomography (CT) scans, which revealed mild hepatosplenomegaly, anemia, mildly elevated liver enzymes, non-nephrotic proteinuria,

Table 1 – Laboratory findings

Test	Results	Reference range	Units
WBC	2.8	4–10.3	10^3/μL
Neutrophils	1.8	2.1–6.1	10^3/μL
Hemoglobin	9	12–16	g/dL
Hematocrit	26.6	36–46	%
MCV	86.2	80.7–95.5	fL
RDW	19.1	11.8–14.3	%
Platelets	120	156–373	10^3/μL
Creatinine	1.22	0.9–1.3	mg/dL
AST	47	0–50	U/L
ALT	26	0–50	U/L
GGT	267	0–55	U/L
ALP	272	30–120	U/L
LDH	279	125–220	U/L
Total bilirubin	1.08	0.30–1.20	mg/dL
Direct bilirubin	0.26	0–0.20	mg/dL
Total protein	10.39	6–8.3	g/dL
Albumin	2.62	3.5–5.2	g/dL
IgM	235	40–230	mg/dL
IgG	4694	700–1600	mg/dL
Ferritine	364	18–306	ng/ml
CRP	77	0.2–5	mg/L
Procalcitonin	0.67	0–0.05	ng/ml
ESR	93	0–15	mm/h
RF	626	0–14	IU/mL
C3	73.4	90–180	mg/dL
C4	7.49	10–40	mg/dL

ALP – alkaline phosphatase; ALT – alanine transferase; AST – aspartate transferase; CRP – C-reactive protein; ESR – erythrocyte sedimentation rate; GGT – gamma glutamyl transferase; LDH – lactate dehydrogenase; MCV – mean corpuscular volume; RDW – red cell distribution width; RF – rheumatoid factor; WBC – white blood cell.

microscopic hematuria, significantly high serum globulin levels, elevated erythrocyte sedimentation rate, positivity for antinuclear antibodies (ANA), increased level of rheumatoid factor (RF), low C3/C4 levels, elevated C-reactive protein (CRP) levels, and hypodense lesions in the liver and spleen. The patient had received empirical antibiotic therapy multiple times; however, while he experienced intermittent improvements, his symptoms never completely resolved. In summary, during this one-year period, the patient sought multiple consultations based on these findings: hematology for the evaluation of lymphoproliferative disorders and plasma cell dyscrasias, nephrology for proteinuria and hematuria, gastroenterology for hepatosplenomegaly and mildly elevated liver enzymes, and rheumatology for elevated acute phase reactants and positivity for autoantibodies. As a result, he had not undergone thorough investigations and struggled with maintaining consistent follow-up appointments until presenting to our clinic.

His medical history included a prosthetic mitral valve replacement (MVR) performed four years prior, as well as surgery six months earlier due to a hemorrhagic cerebrovascular event (CVE). He reported no known allergies or other significant medical conditions. The patient experienced elevated body temperature, which typically rose in the evenings and persisted for a duration of 4 to 5 days. Following this period, there were subsequent days characterized by an absence of fever. Upon physical examination, his temperature was 38 °C, heart rate was 96 beats per minute and rhythmic, and blood pressure was 128/81 mmHg. During cardiac auscultation, a pansystolic murmur grade 2/6 was detected at the mitral focus, accompanied by the audible sound of a metallic prosthetic valve. Traube's space exhibited dullness, and both the liver and spleen were palpable 4 cm below the costal margin. Additionally, petechial rashes were observed on the lower extremities, while the remainder of the physical examination findings were unremarkable.

Laboratory tests showed a white blood cell count of 2.8 10^3/μL, hemoglobin 9 g/dL, hematocrit 26.6%, platelet count 120 10^3/μL, erythrocyte sedimentation rate 93 mm/hour, total protein 10.39 g/dL, albumin 2.62 g/dL, lactate dehydrogenase (LDH) 279 U/L, alkaline phosphatase (ALP) 272 U/L, and gamma-glutamyl transferase (GGT) 267 U/L. Other laboratory tests were within normal limits (**Table 1**). The patient was admitted to the internal medicine department for further investigation.

Blood cultures were taken from both the right and left arms of the patient for four consecutive days. In the first blood culture, a signal indicating Gram-negative bacterial growth was detected at 72 hours. Given the patient's preliminary diagnosis of infective endocarditis, empirical treatment with cefepime and gentamicin was initiated. After the first doses of both antibiotics were administered, another blood culture was taken (as previously planned). Ultimately, *Brucella* growth was detected in the first culture after 5 days, and the treatment was adjusted to doxycycline 200 mg/day, rifampicin 600 mg/day, and gentamicin 3 mg/kg/day. Although *Brucella* signals were observed in all blood cultures taken before the initiation of empirical treatment, no growth was detected in the blood culture taken after the first doses of the empirical treatment. Serological tests were not requested because *Brucella* growth was already detected in the cultures. Additionally, serological tests are often positive in the Turkish population, even when brucellosis is not active, as Turkey is a high-risk endemic area for brucellosis.

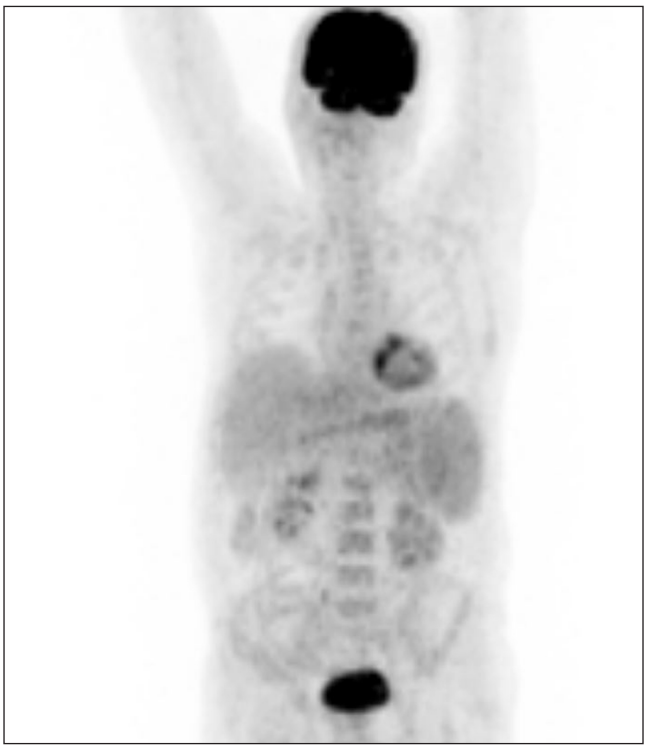

Fig. 1 – PET/CT showing increased FDG uptake around the mitral valve

An abdominal MRI was requested to further investigate the hypodense lesions, which were determined to be chronic infarct areas in the spleen. Additionally, when the FDG PET/CT images previously requested by hematology were reevaluated by our team, increased FDG uptake around the mitral valve was observed (**Fig. 1**). Despite the performance of three transthoracic echocardiograms, no vegetations were detected on the cardiac valves. Subsequently, a transesophageal echocardiogram was conducted, which revealed a 13 mm vegetative mass consistent with prosthetic mitral valve vegetations. The diagnosis of *Brucella* prosthetic valve endocarditis was established following the culture of *Brucella* from blood samples and the identification of vegetations on the prosthetic mitral valve via transesophageal echocardiography. A multidisciplinary council, comprising specialists from cardiology, cardiovascular surgery, infectious diseases, and internal medicine, convened and determined that the patient should undergo surgical intervention. Postoperative monitoring revealed no complications. The patient received antibiotic therapy for approximately six months, during which no relapses of brucellosis were observed. Upon investigation for brucellosis, it was discovered that both the patient and his wife had been consuming unpasteurized milk. The patient's spouse, who resides in the same household, reported exhibiting similar symptoms; however, the severity of her symptoms was relatively milder. Although the patient's wife tested positive

for Rose Bengal and Wright serological tests (1/640) for brucellosis, she exhibited no clinical signs of infective endocarditis. Her brucellosis was effectively managed with antibiotic treatment.

Discussion

Our patient presented numerous and varied clinical and laboratory findings. Due to a delay in diagnosis, these diverse symptoms progressively worsened. The following findings, which we believe are related to *Brucella* endocarditis in our case, can be listed as: recurrent fever, fatigue, and weight loss that had persisted for over a year (constitutional symptoms); an unexplained hemorrhagic stroke six months ago (the first clinical finding that brought the patient to the hospital); non-nephrotic proteinuria, hematuria, spontaneous ecchymoses, back pain, splenic infarctions, rheumatoid factor (RF) positivity, antinuclear antibody (ANA) positivity, reduced C3/C4 levels, normocytic anemia, elevated acute phase reactants, increased erythrocyte sedimentation rate, elevated serum immunoglobulin levels, rouleaux formation on peripheral blood smear, hepatosplenomegaly, and increased uptake around the mitral valve on FDG-PET/CT. Fadul et al. presented a case of *Brucella* endocarditis complicated by embolic stroke, similar to our case; however, our patient experienced a hemorrhagic stroke.[6] Immunological phenomena can be observed in endocarditis, such as in *Brucella* endocarditis, including positive RF, ANA, and endocarditis-related glomerulonephritis findings, as we presented in our case.[7] FDG-PET/CT has become an important tool for the evaluation and diagnosis of endocarditis in recent years, with a sensitivity of 86% for the diagnosis of prosthetic valve IE.[8] The FDG-PET/CT scan provided important findings for the diagnosis of IE in our case.

Considering that *Brucella* is transmitted through contact with animals and consumption of animal products such as milk, it is important to clinically evaluate and screen close contacts, such as household members, of individuals diagnosed with *Brucella*; as shown in this case, *Brucella* was also detected in the patient's wife.[4]

In our case, *Brucella* growth in the blood cultures taken before the initiation of empirical treatment appeared on the fifth day. However, no growth was observed in the blood culture taken after the first dose of empirical cefepime/gentamicin treatment. *Brucella* is a slow-growing infectious agent and more difficult to culture in blood compared to other pathogens. The sensitivity of blood cultures ranges from 10% to 90%.[9] Therefore, it is crucial to obtain blood cultures under appropriate conditions from patients with suspected brucellosis before initiating empirical treatments.

Sometimes, even when everything points to infective endocarditis, the diverse clinical and laboratory findings can obscure this reality. Our patient had presented to the hospital multiple times with different complaints and had been evaluated for various differential diagnoses before coming to us. However, if we had simplified the case to that of a patient with recurrent fever and a history of mitral valve replacement surgery, the first consideration would have been infective endocarditis. It is crucial to adopt a holistic approach to patients and to search for the most appropriate clinical pattern in a multifaceted disease such as infective endocarditis.

Conclusion

Brucellosis can present with a variety of clinical features, which can mislead physicians. A holistic approach, along with identifying the most appropriate pattern that explains the majority of findings, is crucial for the diagnosis of infective endocarditis due to brucellosis. *Brucella* typically infects humans through unpasteurized dairy products, and it is important to screen the households of index patients. As *Brucella* is a slow-growing bacterium in cultures, it is essential to collect blood samples for culture before starting empirical treatment.

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Conflict of interest
The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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Ethical statement
The present study followed international and national regulations and was in agreement with the Declaration of Helsinki, and ethical principles. Our institution does not require ethical approval for reporting individual cases or case series.

Informed consent
Written informed consent was obtained from the patient and his wife for their anonymized information to be published in this article.

Availability of data and materials
All are available upon reasonable request.

Authors' contribution
HD was a major contributor to the writing of the manuscript, and all authors (HD, AÜ, İEA, EÖ, MOT) commented on subsequent versions. All authors (HD, AÜ, İEA, EÖ, MOT) contributed to data collection, treatment, and patient follow-up. All authors (HD, AÜ, İEA, EÖ, MOT) read and approved the final manuscript.

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Infekční endokarditida vyvolaná neobvyklým původcem *Tropheryma whipplei* aneb Whippleova endokarditida

(Infective endocarditis with an atypical causative agent *Tropheryma whipplei* or Whipple's endocarditis)

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Aktinomyceta
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Ruptura sleziny
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Whippleova choroba

SOUHRN

Infekční endokarditida neboli zánětlivé postižení endokardu může být vyvolána obvyklými patogeny, jako jsou stafylokoky, streptokoky, enterokoky, ale také obtížně kultivovatelnými patogeny s negativními hemokulturami, tzv. BCNIE (blood culture-negative infective endocarditis).
Jedním z patogenů BCNIE je *Tropheryma whipplei*, která je původcem Whippleovy choroby. V případech, kdy dochází k izolovanému postižení endokardu, hovoříme o Whippleově endokarditidě. Oproti běžným infekčním endokarditidám bývá klinický nález atypický.
Popisovaný případ poukazuje na klíčovou úlohu polymerázové řetězové reakce (PCR) v diagnostice v případě podezření na atypického původce infekční endokarditidy při opakovaně negativních mikrobiologických nálezech a z toho plynoucí možnou příčinu nízké incidence Whippleovy endokarditidy.

Keywords:
Actinomyces
Infective endocarditis
Ruptured spleen
Tropheryma whipplei
Whipple's disease

ABSTRACT

Infective endocarditis (IE) is an inflammatory disease of the endocardium, most commonly caused by pathogens such as *Staphylococcus* spp., *Streptococcus* spp., and *Enterococcus* spp. However, in some cases, the causative organisms are difficult to culture, resulting in blood culture-negative endocarditis (BCNE). One of the rare pathogens associated with BCNE is *Tropheryma whipplei*, the bacterium responsible for Whipple's disease. When *T. whipplei* infects the endocardium without other systemic manifestations, the condition is referred to as Whipple's endocarditis.
Tropheryma whipplei is a rare cause of IE that often presents with atypical clinical features and persistently negative blood cultures. This case report underscores the crucial role of polymerase chain reaction (PCR) in establishing the diagnosis in patients with suspected IE and repeatedly negative blood cultures. Given the diagnostic complexity and often subtle clinical presentation, the true incidence of Whipple's endocarditis is likely underestimated.

Úvod

Termín infekční endokarditida (IE) definuje infekci vnitřního povrchu srdce (endokardu). Obvykle postihuje jednu nebo více srdečních chlopní (nativní, protetickou) nebo implantabilní intrakardiální materiál. Roční incidence IE dokumentovaná za rok 2021 se pohybuje okolo 5–14 případů na 100 000 obyvatel.

Mezi nejčastější původce IE patří *Staphylococcus aureus*, viridující streptokoky, enterokoky, koaguláza-negativní stafylokoky a bakterie skupiny HACEK (Haemophilus, Aggregatibacter [původně *Actinobacillus*], *Cardiobacterium*, *Eikenella*, *Kingella*) a BCNIE (blood culture-negative infective endocarditis). BCNIE je specifická skupina IE. Jedná se o špatně rostoucí nebo standardním způsobem nekultivovatelné patogeny (bakterie rodů *Bartonella*, *Brucella*, *Coxiella*, *Legionella*, *Mycobacterium*, *Mycoplasma*, *Tropheryma*, houby…), které tvoří 5–30 % všech IE. V našich zeměpisných podmínkách je nejčastějším původcem endokarditid skupiny BCNIE *Bartonella*. Zajímavostí je, že

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infekce způsobené bakterií *Tropheryma whipplei* bývají méně časté, ačkoli výskyt původce v našem prostředí není úplně vzácný.[1,2]

Tropheryma whipplei je intracelulární grampozitivní aktinomyceta, která je původcem tzv. Whippleovy choroby. V Evropě a ve Spojených státech amerických je dokumentována roční incidence Whippelovy choroby jeden až šest případů na 10 milionů obyvatel.

Infekce způsobené *Tropheryma whipplei* mohou mít čtyři odlišné projevy: klasickou Whippleovu chorobu, fokální onemocnění, akutní infekci a asymptomatické nosičství. Klasická Whippleova choroba se vyznačuje triádou klinických příznaků – artralgií, průjmy a hubnutím. U lokalizované formy je postižen např. kardiovaskulární nebo nervový systém, oči, kůže, klouby, plíce atd. Akutní onemocnění je nejčastěji kombinace bakteriemie a sepse, akutní gastroenteritidy nebo pneumonie. Typicky se jedná o první kontakt pacienta s bakterií. Nejčastěji se vyskytuje u dětí a jedná se pouze o přechodnou formu Whippleovy choroby. Posledním typem a zdaleka nejběžnějším je asymptomatické nosičství u zdravých jedinců, kdy předpokládáme výraznou roli genetické predispozice a imunitního systému k náchylnosti k onemocnění.

Chronické ložiskové onemocnění se nejčastěji projevuje jako subakutní, kultivačně negativní endokarditida a postihuje aortální nebo mitrální chlopeň, ojediněle trikuspidální a zcela výjimečně pulmonální chlopeň. Kromě endokarditidy byly zaznamenány případy, kdy *Tropheryma whipplei* způsobila rozvoj myokarditidy i perikarditidy.[3,4]

Odhadovaná míra případů Whippleovy endokarditidy se pohybuje okolo 2,6–7,1 % z celkového počtu BCNIE, kdy největší výskyt je popisován ve Francii, Španělsku, Německu a ve Švýcarsku.[5]

Příznaky Whippleovy endokarditidy nejsou typické pro infekční endokarditidu. Pacienti jsou ve většině případů afebrilní, ojediněle subfebrilní. Často trpí malátností, protrahovanými artralgiemi, gastrointestinálními obtížemi a ztrátou hmotnosti. Prvotním projevem onemocnění bývá často obraz srdečního selhání při závažném hemodynamickém postižení chlopně.[6]

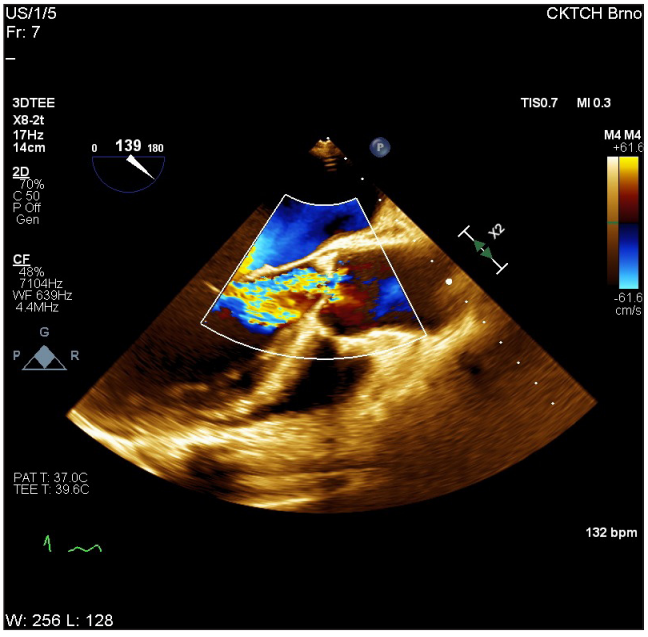
Ve většině případů nejsou splněna modifikovaná kritéria Duke. V laboratorních nálezech často vídáme nízké až negativní zánětlivé parametry a opakovaně negativní mikrobiologický nález v hemokulturách. Základní diagnostickou metodou je v tomto případě transtorakální a transezofageální echokardiografie, která poukáže na možnou infekční endokarditidu s destrukcí chlopně. Nicméně jedinou možností, jak získat informace o přesném původci, a tedy i stanovit diagnózu Whippleovy endokarditidy, je v současné době pouze molekulárněgenetická analýza metodou polymerázové řetězové reakce (PCR) z chirurgicky odebraného materiálu.[7] Základem úspěšné terapie je chirurgický zákrok na chlopni se správně zvolenou prolongovanou antibiotickou terapií.

Popis případu

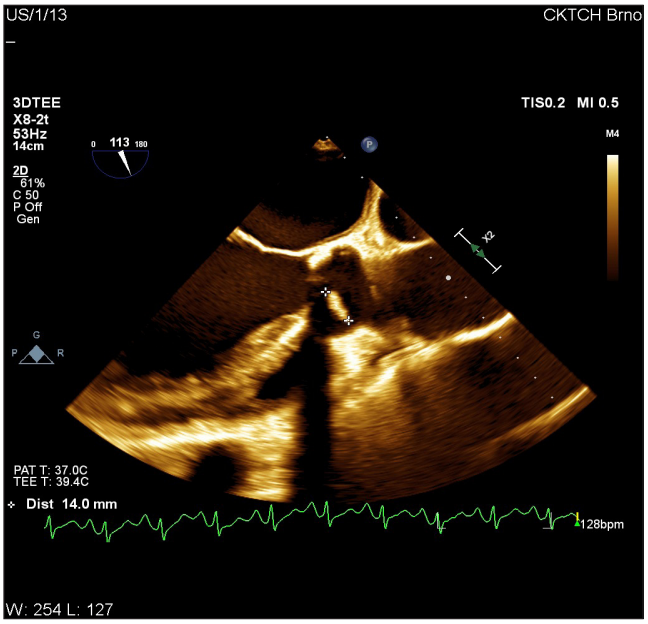
Jednapadesátiletý pacient byl hospitalizovaný na spádovém interním oddělení pro známky akutní bilaterální kardiální dekompenzace, postupně narůstající námahovou a později i klidovou dušnost, přibývání na váze, progresi otoků dolních končetin. V anamnéze pacient nebyl doposud sledován pro žádnou chronickou interní chorobu a nebyla u něj zavedena ani žádná dlouhodobá farmakoterapie. Pacient je chronický fumator (15 cigaret/den) a potator (denně průměrně 5 piv, tvrdý alkohol příležitostně), pracuje jako zedník.

Ve vstupních laboratorních vyšetřeních byla zaznamenána významná elevace N-terminálního fragmentu natriuretického propetidu typu B (NT-proBNP), lehká elevace zánětlivých parametrů (C-reaktivního proteinu [CRP] 20), nicméně bez přítomnosti leukocytózy.

Během klinického vyšetření byl u pacienta slyšitelný diastolický šelest nad aortální chlopní 3/6, oslabené dý-



Obr. 1 – TEE: barevné dopplerovské mapování průtoku (CFM) znázorněna aortální regurgitace. TEE – transezofageální echokardiografie.



Obr. 2 – TEE: vegetace na aortální chlopni. TEE – transezofageální echokardiografie.

chání bazálně vpravo a viditelné otoky dolních končetin ke kolenům. Pacient byl afebrilní, bez známek akutně probíhající infekce.

Dle rtg plic nebyly zachyceny infiltrativní ani ložiskové změny v plicním parenchymu, pouze popsán drobný pleurální výpotek vpravo. Elektrokardiogram byl s fyziologickým nálezem bez zjevné patologie. Echokardiograficky byly zaznamenány dilatované levostranné oddíly s globální hypokontraktilitou levé komory, s nízkou ejekční frakcí levé komory (EF LK 20 %). Na mitrální a trikuspidální chlopni byla detekována regurgitace do 1. stupně. Trojcípá aortální chlopeň byla s patrnými hyperechogenními a zbytnělými cípy a na jejím levém koronárním cípu nápadně vyvlávala vegetace maximální velikosti 14 mm, s regurgitací 2.–3. stupně (ze čtyřstupňové stupnice). Nebylo zjištěno extravalvární šíření infekce (píštěl či paravalvární absces) (**obr. 1, 2**).

Byl proveden mikrobiologický screening včetně opakovaně odebraných hemokultur, sérologie na atypické patogeny způsobující IE (*Legionella*, *Mycoplasma, Coxiella, Bartonella*), vše s negativním výsledkem, byla vyloučena jiná fokální či systémové infekce.

V rámci došetření etiologie dysfunkční levé komory byla provedena selektivní koronarografie s průkazem hladkostěnných věnčitých tepen. Následně byla doplněna magnetická rezonance srdce s detekcí pozánětlivých změn na spodní stěně levé komory.

Po maximalizaci farmakologické terapie srdečního selhání se sníženou ejekční frakcí LK (HFrEF) došlo ke kardiální kompenzaci pacienta a vymizení klinických obtíží.

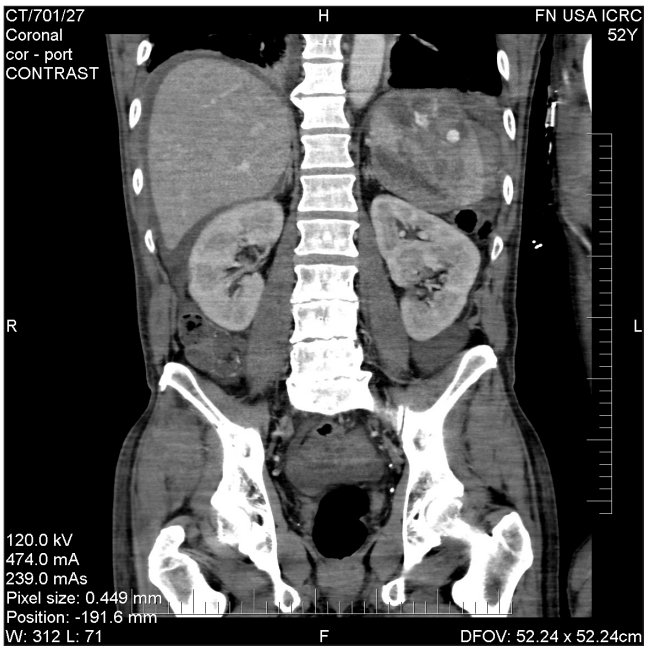
Za hospitalizace došlo k nárůstu zánětlivých parametrů (CRP 75, leukocyty 12), které nekorelovaly s klinickým nálezem. Pacient byl i nadále stran infekce zcela asymptomatický. Vzhledem k potvrzení infekční endokarditidy dle echokardiografického vyšetření byla po domluvě s antibiotickým střediskem nasazena empiricky dvojkombinace oxacillin s gentamicinem, která následně vedla k poklesu zánětlivých parametrů.

Vzhledem ke stacionárnímu nálezu vlající vegetace v souladu s guidelines kardiotým doporučil operační řešení aortální chlopně pro vysoké riziko systémové embolizace. V úvahu přicházela Rossova operace, popř. náhrada aortální chlopně mechanickou či biologickou protézou.

Patnáctý den od příjmu bylo přistoupeno k operačnímu výkonu, kdy po přihlédnutí k preferenci pacienta proběhla implantace bioprotézy Inspiris Resilia 25 mm do aortální pozice z ministernotomie. Perioperačně byla zjištěna verukózní vegetace s maximálním postižením pravého a nonkoronárního cípu. Výkon a časné pooperační období se obešly bez komplikací.

Výsledky kultivačního vyšetření z odstraněných cípů chlopně a vegetace byly negativní. Histologicky byly patrné ojediněle okrsky pěnivých makrofágů s intracytoplazmatickou přítomností PAS+/dPAS+ granul až tyčinkovitých formací rámcově kompatibilní s *Tropheryma whipplei*. Nicméně metodou PCR byla detekována jasná přítomnost *Tropheryma whipplei*. Na základě výsledků PCR došlo ke změně antibiotické terapie za ceftriaxon.

Pooperační kontroly proběhly s výborným výsledkem. Aortální bioprotéza byla bez detekce leaku či regurgitace. Systolická funkce a velikost levé komory se postupně znormalizovaly (EF LK 50 %).



Obr. 3 – AngioCT břicha: ruptura sleziny a kolekce tekutiny v dutině břišní.

Pátý pooperační den došlo k rozvoji prekolapsového stavu s hypotenzí. Proto byla provedena kontrolní echokardiografie perikardu s vyloučením srdeční tamponády. V krevním obraze došlo k poklesu hemoglobinu na 60 g/l. Subjektivně pacient udával dyspeptické obtíže a bolesti břicha. Objektivně byly patrné známky rozvíjejícího se hemoragického šoku, proto byla provedena akutní sonografie břicha s průkazem kolekce tekutiny v dutině břišní. Pro verifikaci byla doplněna výpočetní tomografie (CT) břicha (**obr. 3**), kde byla popsána ruptura sleziny s hemoperitoneem. Následně bylo přistoupeno k akutní splenektomii s revizí dutiny břišní. Histologicky v parenchymu sleziny byly zachyceny multifokálně infarkty sleziny jako komplikace systémových embolizací z vegetace.

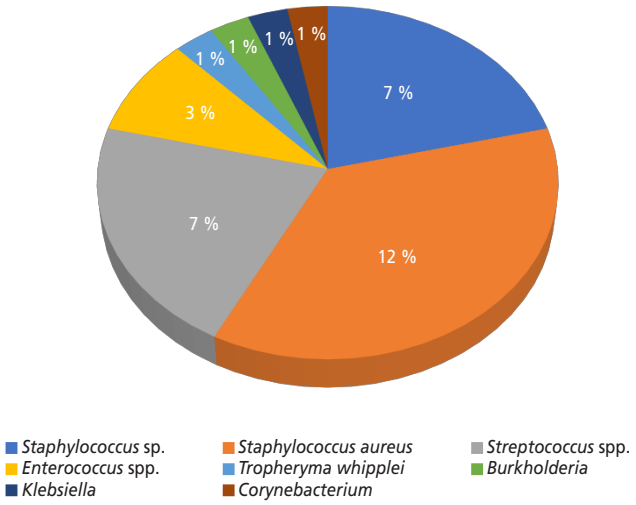
Po výkonu došlo k postupné klinické stabilizaci pacienta. V kontrolních laboratorních odběrech byla patrná normalizace krevního obrazu, zánětlivé parametry s klesajícím trendem (leukocyty 15, CRP 45). Opětovně bylo kontaktováno antibiotické středisko stran další strategie antibiotické terapie, kdy bylo doporučeno pokračovat v terapii ceftriaxonem do normalizace zánětlivých parametrů s následnou nutností dlouhodobé terapie doxycyklinem po dobu minimálně 12 měsíců pro vysoké riziko recidivy Whippleovy choroby.

Pacient byl dvanáctý pooperační den přeložen na spádové interní oddělení k pokračování v parenterální aplikaci ATB terapie.

Po roce byla provedena na naší klinice kontrola včetně echokardiografického vyšetření. Nález při echokardiografii je i nadále s vynikajícím výsledkem. Bylo patrno další zlepšení systolické funkce levé komory (tč. EF LK 68 %) bez detekce vegetace či chlopenní vady. Klinicky pacient nejevil známky infekce. Bylo doporučeno i nadále pokračovat v zavedené ATB terapii doxycyklinem (celkově v délce trvání 18 měsíců).

Diskuse

V průběhu roku 2023 se na našem pracovišti chirurgicky řešilo celkově 33 případů infekční endokarditidy. Převážně se jednalo o postižení nativních chlopní. Protetické infekce činily 20 % z celkového počtu případů. Převážně vyvolávajícími původci byly stafylokoky, streptokoky a enterokoky, které nejvíce postihovaly nativní aortální chlopeň (viz **obr. 4**). Pouze v jednom případě byla potvrzena *Tropheryma whipplei* jako vyvolávající agens.



Obr. 4 – Vyvolávající patogeny IE u operačně řešených pacientů na CKTCH za rok 2023.

Whippleova endokarditida postihuje dominantně aortální chlopeň. Nejčastěji se vyskytuje u 50letých mužů bělošského původu. Četnější výskyt je popisován u zemědělců, farmářů, pacientů s nižším hygienickým standardem. Určitou roli hraje i genetická predispozice a stav imunitního systému.

Pacient v námi popisovém případu je 51letý muž, bělošského původu, zaměstnáním zedník, dlouholetý fumator, potator, vstupně s kariézním chrupem. Vzhledem k socioekonomickému statusu, abúzu alkoholu a nikotinismu, práci na stavbách lze usuzovat možný horší stav imunitního systému, větší vystavení kontaktu s *Tropheryma whipplei* a z toho plynoucí vyšší riziko rozvoje Whippleovy endokarditidy.

Popisovaný případ je ukázkou specifického atypického průběhu fokální formy onemocnění s dlouhodobě negativními zánětlivými parametry a mikrobiologickým nálezem. *Tropheryma whipplei* není kultivovatelná na standardních půdách. Její diagnostika vyžaduje histologickou analýzu barvením PAS (periodic acid-Schiff) a testování metodou PCR z postižených explantátů, která má větší senzitivitu a specificitu v porovnání s PAS.

Díky prvotním příznakům primomanifestace HFrEF byla na základě echokardiografického vyšetření zjištěna vegetace na aortální chlopni. Na základě splnění indikačních kritérií, v tomto případě pro vysoké riziko systémové embolizace, bylo přistoupeno k náhradě aortální chlopně a následně k verifikaci původce molekulární analýzou.

Metoda PCR hraje klíčovou úlohu při diagnostice Whippleovy choroby. K diagnostice lze využít tkáňové vzorky z biopsie zasaženého orgánu, např. duodenální sliznice, plicní či mozkové tkáně, synoviální tekutiny, mozkomíšního moku, kostní dřeně, endokardu, kůže… atd. V rámci neinvazivního PCR vyšetření lze použít také vzorek stolice, slin, moči či krve, které mohou sloužit jako relativně snadný, citlivý a specifický screening. Nicméně v rámci prvotní diagnostiky je nutnost verifikace agens PCR z bioptického vzorku. Hlavní nevýhodou zůstává i nadále nutnost získání tkáňového vzorku. Proto v případě nesplnění indikačních kritérií k operačnímu řešení chlopenní vady je velmi obtížné získání informace o vyvolávajícím původci a z toho vyplývající adekvátní antibiotická terapie.[4,5]

Mezi nejčastější komplikace infekční endokarditidy patří systémová embolizace, která dle dostupných dat činí 20–40 % případů. Nejčastěji bývají postiženy septickými embolizacemi mozek (50 %), slezina (5–12 %), dále plicní systém, ledviny, končetiny, kůže, jejichž výskyt je dosti variabilní. Infarkt sleziny je ve většině případů asymptomatický, v 5 % dochází k rozvoji abscesu. Ve většině případů je spojen s infekcemi způsobenými patogeny *Staphyloccus aureus*, *Streptococcus bovis* či mykotickými původci. Výskyt ruptury sleziny při infekční endokarditidě je spíše raritní. Nicméně mortalita se v případě potvrzení ruptury sleziny pohybuje okolo 58 % případů, a proto je nezbytné myslet na možnou rupturu a včas ji diagnostikovat.[8–10] Dle dostupných dat prozatím nebyl zjištěn vyšší výskyt ruptury sleziny při Whippleově chorobě. Nicméně atypický průběh onemocnění a z toho plynoucí prodlení v zahájení antibiotické terapie a s tím spojené rozsáhlejší vegetace mohou být určitou predispozicí k většímu výskytu embolických komplikací.

Podle dostupných dat a informací prozatím neexistuje jasná standardní léčba Whippleovy endokarditidy, včetně guidelines, kde je uvedena prozatím jako empirická. Můžeme pouze vycházet z doporučení České kardiologické společnosti (ČKS), Evropské kardiologické společnosti (ESC), American Heart Association (AHA), popř. z jednotlivých kazuistik.

Základem terapie je chirurgický zákrok na chlopni se správně zvolenou antibiotickou terapií. V souladu s guidelines v případě potvrzení Whippleovy endokarditidy má být zahájena terapie kombinací doxycyklin s hydroxychlorochinem per os, alternativně parenterální aplikace ceftriaxonu na dva až čtyři týdny, následovaná perorálním podáváním trimetropinu v kombinaci se sulfametoxazolem. Vždy je doporučena dlouhodobá léčba, nicméně optimální délka trvání není známá (minimálně po dobu jednoho roku).

Někteří autoři nedoporučují podávání kombinace trimetropin a sulfametoxazol pro větší riziko relapsů a vyšší výskyt rezistence.[3,4] Malé observační studie zase udávají z dlouhodobého hlediska možnost užívání pouze doxycyklinu bez hydroxychlorochinu.[7] Jiné studie zase poukazují, že v případě operačního řešení je možné zkrácení doby užívání dvojkombinace doxycyklin a hydroxychlorochin. V obou případech nedošlo k většímu výskytu relapsů.[4,5]

V našem případě na základě doporučení antibiotického střediska bylo přistoupeno ke čtyřtýdenní terapii ceftriaxonem s následným přechodem na dlouhodobou terapii doxycyklinem. Nyní vyvstává otázka, zda z dlou-

hodobého hlediska monoterapie doxycyklinem po provedené náhradě aortální chlopně bude dostačující či povede ke zmíněnému relapsu. Výsledek naší léčby si ověříme v rámci sledování pro případné riziko relapsu.

Závěr

Tropheryma whipplei je sporadickým původcem infekční endokarditidy. Vzhledem k obtížné diagnostice, negativním hemokulturám a ve většině případů nespecifickým projevům infekční endokarditidy může být incidence významně podceněna. Proto v případě výskytu především známek subakutní či chronické endokarditidy, protrahovaných artralgií nebo gastrointestinálních příznaků bychom měli mít na paměti tohoto původce. V případě přistoupení k operačnímu výkonu je vždy třeba využít analýzu PCR k verifikaci. V budoucnu je možné, že s přibývajícím využíváním molekulární diagnostiky dojde k nárůstu incidence výskytu tohoto patogenu. Při včasné a vhodně zvolené terapii má toto onemocnění velmi dobrou prognózu. Základem je dlouhodobá antibiotická terapie a operační zákrok na chlopni.

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Všichni autoři prohlašují, že nemají střet zájmů.

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Informovaný souhlas
Práce byla vypracována s informovaným souhlasem pacienta.

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Silent Armor Around the Heart: Calcific Pericarditis in a Young Man: A Case Report

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SOUHRN

Na oddělení urgentního příjmu byl přivezen 48letý muž pro bolesti žaludku a únavu. Netrpěl žádnou známou komorbiditou, jednalo se o nekuřáka a v jeho rodinné anamnéze nebylo žádné srdeční, ani jiné významnější onemocnění. Výsledky EKG a rentgenového vyšetření byly normální, zato vyšetření hrudníku výpočetní tomografií odhalilo malý perikardiální výpotek, ztluštění a kalcifikaci na bázi srdce. Transtorakální echokardiografie prokázala malý výpotek kolem perikardu, jeho ztluštění a zvýšenou echogenitu. Vyšetření srdce magnetickou rezonancí potvrdilo výpotek a ztluštění perikardu; nález odpovídal konstriktivní perikarditidě. Popisujeme tento zajímavý případ rozsáhlé kalcifikující perikarditidy nijak neupozorňující na závažnost stavu a bez přítomnosti dyspnoe, otoků nebo bolesti na hrudi.

Keywords:
Cardiac magnetic resonance
Chronic constrictive pericarditis
Computerized tomography

ABSTRACT

A 48-year-old male patient was admitted to the emergency room complaining of stomach pain and fatigue. He had no known comorbid diseases, was a non-smoker, and had no family history of heart disease or any other disease. Electrocardiography and X-ray were normal, but a thoracic computerized tomography scan revealed mild pericardial effusion, thickening, and calcification at the base of the heart. Transthoracic echocardiography showed mild pericardial effusion around the pericardium, thickening, and increased echogenicity. Cardiac magnetic resonance imaging confirmed pericardial effusion and thickening. The findings were consistent with constrictive pericarditis. We present the story of this interesting case due to the grand calcific pericarditis with silent complaint and no dyspnea, edema, or chest pain.

Introduction

Constrictive pericarditis involves the formation of granulation in the pericardial tissue and a loss of pericardial elasticity due to calcium deposition. In this condition, the pericardium envelops the heart like armor, and myocardial relaxation is disrupted during diastole. Patients typically present with vague complaints, with diastolic heart failure and indications of right heart failure being most prominent.

A 48-year-old male patient was admitted to the emergency room complaining of stomach pain and fatigue. He had no known comorbid diseases, was a non-smoker, and had no family history of heart disease or any other disease.

The head and neck examinations were normal, and the heart sounds were deep. Lung sounds are normal, and there are no pathological findings on abdominal examination. There was no pretibial edema on her legs.

Laboratory: hemoglobin was 14.1 g/dL, white blood count was 7 × 10^3, C-reactive protein was 4 mg/dl, troponin I was 4 nanogram/l (within normal limits), creatine level was 0.8 mg/dL. Liver tests were normal.

Electrocardiogram: The rhythm was normal sinus rhythm, and the heart rate was 75/min beat. The T wave was negative in D2,3 and aVF derivations, and all derivations had low voltage findings (**Fig. 1A**).

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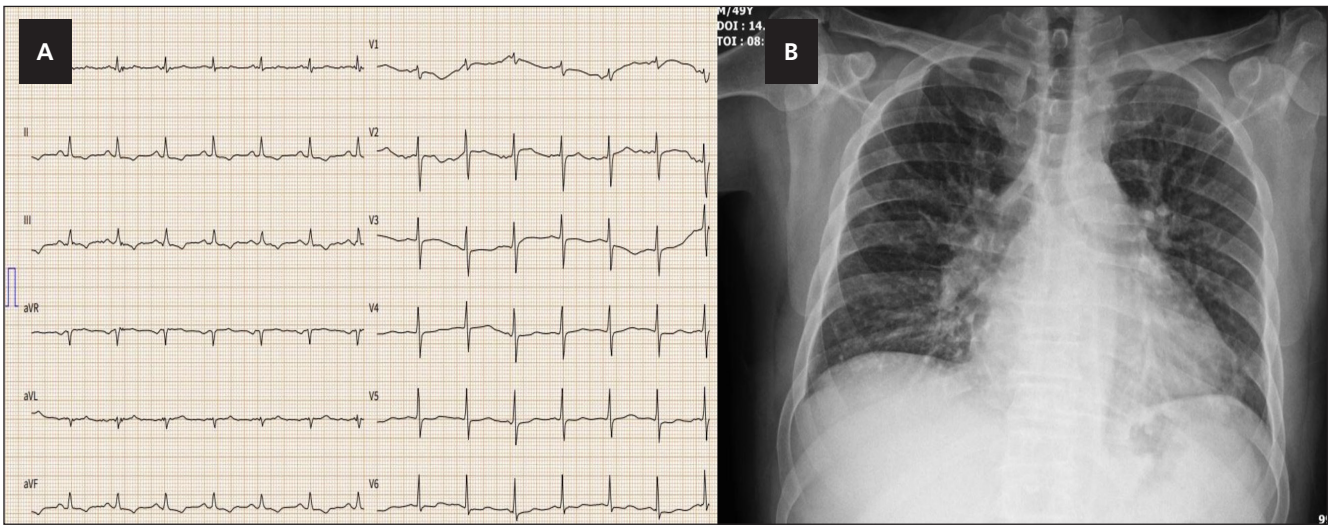

Fig. 1 – Electrocardiography and chest X-ray of the patient on admission to the emergency department.

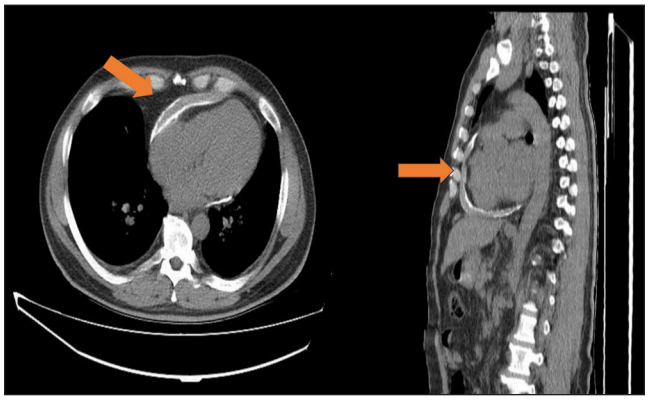

Fig. 2 – Calcification around the pericardium in the computerized tomography taken due to the patient's complaints.

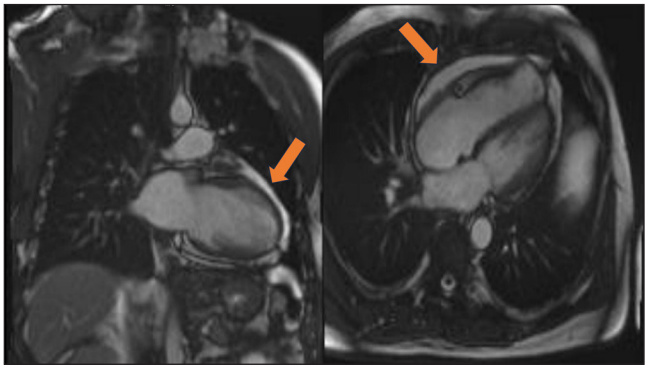

Fig. 3 – Cardiac magnetic resonance images showed pericardial thickening and calcification.

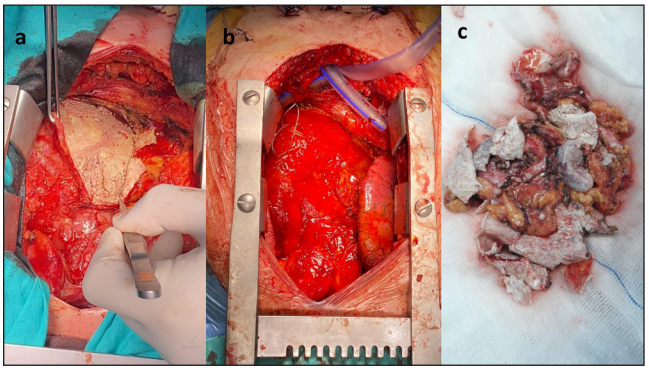

Fig. 4 – Surgical demonstration of the patient's calcified pericardium (A), the postoperative image of the heart with the calcification removed (B), and the image of the removed calcified pericardium (C).

Chest X-ray showed the heart size was normal width, and no obvious calcification was observed around the heart (**Fig. 1B**).

Thoracic computerized tomography scan revealed mild pericardial effusion, pericardial thickening, and pericardial calcification at the base of the heart (**Fig. 2**).

Transthoracic echocardiography was performed in an emergency room. Ejection fraction was normal, mild pericardial effusion around the pericardium, pericardial thickening, and increased echogenicity were observed.

Cardiac magnetic resonance ımaging showed pericardial effusion (14 mm) and pericardial thickening, as well as a decrease in left ventricular EF (LVEF 55%) and an increase in left atrial volume. The findings were consistent with constrictive pericarditis (**Fig. 3**).

Coronary angiography demonstrated coronary arteries were normal, left ventricular and diastolic pressures were increased, and in right heart catheterization, there was an increase in right ventricular end-diastolic pressure. Pulmonary artery wedge pressure was increased.

When all the patient's findings were combined, it was concluded that he had calcific constrictive pericarditis. A consensus decision was made considering all the results and the relevant latest guideline recommendations, and pericardiectomy was considered appropriate.

The decision for surgery was shared with the patient, and verbal and written consent was obtained.

The patient underwent pericardiectomy due to constrictive pericarditis (**Fig. 4 A, B**). Thickened pericardial tissue containing intense calcification was observed in the surgical materials (**Fig. 4C**).

The pericardial biopsy result was interpreted as compatible with the findings of inflamed fibrinous material.

In the materials taken for the differential diagnosis of tuberculosis, no bacilli were detected, and no tuberculosis-specific pathological necrosis was observed.

The patient was not symptomatic during the postoperative check-up, follow-up echocardiograms were normal, and he was discharged with recommendations. There have been no problems with follow-up visits for regular check-ups for 3 years.

Discussion

Constrictive pericarditis is characterized by the formation of granulation in the pericardial tissue. There is a loss of pericardial elasticity, and this leads to restriction in ventricular filling.[1] The leading cause of constrictive pericarditis worldwide is tuberculosis, and it can occur even in patients receiving antituberculous therapy. In developed countries, idiopathic or post-viral infections are the most common causes. Other important causes may occur as complications after heart surgery and after the radiation therapy to the mediastinum. It may also be associated with connective tissue diseases. Often, as in our patient, it is very difficult to determine a definitive etiological cause. It is often thought to be an asymptomatic attack of viral pericarditis.[2,3]

Constructive calcific pericarditis is a clinical condition that is rare in young patients and is particularly characterized by right heart failure findings. According to the latest guideline, the first method recommended for the diagnosis of constrictive pericarditis is echocardiography.[4] At the time of the first admission to our hospital, a chest X-ray was requested due to shortness of breath, and then a thorax CT was requested. Upon observation of pericardial calcification on thorax CT, the patient underwent echocardiography under emergency conditions. Afterward, cardiac magnetic resonance was performed to confirm the diagnosis and elucidate the etiology. All guideline recommendations were completed in the patient, who also underwent cardiac catheterization.

Since our patient did not develop signs of right heart failure, there were no findings suspicious for constrictive pericarditis in the physical examination. Patients with constrictive pericarditis often have vague complaints such as fatigue, weakness, and shortness of breath. They may also present with high fever, palpitations, paroxysmal nocturnal dyspnea, sweating, and swelling in the feet.

Low voltage, T wave changes, and even atrial fibrillation can often be seen in the ECGs of these patients. Our patient also had low voltage and T-wave changes. In echocardiography, minimal effusion and thickening were observed only in the pericardium. The patient was diagnosed with constrictive pericarditis after imaging examinations and a surgical method was recommended for the treatment of the disease and clarification of the etiology.

A pericardiectomy is the accepted standard treatment for patients with chronic constrictive pericarditis with persistent and significant symptoms.[4]

Surgical removal of the pericardium has a mortality rate ranging from 6–12%. End-stage patients benefit little from pericardiectomy, and operative mortality is increased. Recently, cachexia, atrial fibrillation, low flow rate findings, hypoalbuminemia due to protein loss enteropathy, cardiac cirrhosis, and impaired liver tests have been observed.[5]

If the underlying cause is known, medical treatment can be initiated (e.g., tuberculosis or other infectious diseases, connective tissue diseases). Although no etiological cause can be found in our hospital, the patient comes for regular check-ups after surgery without needing medical treatment.

Conflict of interest
The authors declare no conflict of interest.

Funding
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Ethical statement
This case report includes both verbal and written patient consent. No experiments on humans are performed. It complies with the ethical standards of our institution's committee or the Declaration of Helsinki and its revisions.

Informed consent
Written informed consent was obtained from the patient for the publication of this article.

AI disclosure
This case report did not use artificial intelligence (AI)-powered technologies (such as Large Language Models [LLMs], chatbots, or image generators).

Author contribution
Concept/design: SU, SSP. Data analysis/interpretation: Drafting article: SU, SKO. Critical revision of the article: SU, EK. Approval of article: SU, EAD. All authors discussed the results and contributed to the final manuscript. All authors provided critical feedback and helped shape the research, analysis, and manuscript. All authors read and approved the final version of the paper.

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Effusive-constrictive pericarditis following mild COVID-19: a case of combined surgical and medical therapy

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Perikardiální výpotek

SOUHRN

Dnes je již prokázáno, že infekční onemocnění SARS-CoV-2, zprvu považované za respirační onemocnění, způsobuje kardiovaskulární komplikace včetně vzácných případů efuzivně-konstriktivní perikarditidy. Popisujeme případ 45letého muže, u něhož došlo po překonání infekčního onemocnění covid-19 s mírným průběhem k rozvoji efuzivně-konstriktivní perikarditidy. Pacient byl původně přijat pro akutní perikarditidu s malým perikardiálním výpotkem léčeným ibuprofenem a colchicinem. O dva týdny později se muž dostavil k vyšetření znovu pro zhoršující se symptomy srdečního selhání včetně ortopnoe, otoků a únavy. Vyšetření srdce echokardiograficky a magnetickou rezonancí (cardiac magnetic resonance, CMR) ukázala na přítomnost efuzivně-konstriktivní perikarditidy a subakutní myokarditidy. Přes protizánětlivou léčbu a podávání diuretik symptomy přetrvávaly, a vyžádaly si tak perikardiektomii. Perikard byl silně ztluštělý s rozsáhlými srůsty. Symptomy srdečního selhání přetrvávající po operaci a nález fibrózního perikardu zobrazovacími metodami vedly k zahájení léčby kortikosteroidy s následným vymizením symptomů. Tento případ názorně ukazuje náročnost diagnostiky a léčby efuzivně-konstriktivní perikarditidy v kontextu infekčního onemocnění covid-19. Identifikaci aktivního zánětu perikardu a vedení léčby napomohlo použití neinvazivních zobrazovacích metod, zvláště CMR. Zatímco perikardiektomie představuje i nadále rezolutní intervenci v jinak neřešitelných případech, popsaný případ ukazuje na potenciální přínos imunosupresivní léčby pro vymizení reziduálních symptomů. Vzhledem k omezenému množství literatury na téma perikarditidy v souvislosti s infekčním onemocněním covid-19 je pro optimalizaci výsledku léčby a směřování dalšího výzkumu nutný multidisciplinární přístup.

Keywords:
Constrictive pericarditis
COVID-19
Pericardial effusion

ABSTRACT

SARS-CoV-2 infection, initially perceived as a respiratory illness, is now recognized to cause cardiovascular complications, including rare cases of effusive-constrictive pericarditis. We present a case of a 45-year-old male who developed effusive-constrictive pericarditis following mild COVID-19. The patient was initially admitted with acute pericarditis and moderate pericardial effusion, treated with ibuprofen and colchicine. Two weeks later, he returned with worsening symptoms of heart failure, including orthopnea, edema, and fatigue. Echocardiography and cardiac magnetic resonance (CMR) revealed findings consistent with effusive-constrictive pericarditis and subacute myocarditis. Despite anti-inflammatory and diuretic therapy, symptoms persisted, necessitating pericardiectomy. The pericardium was severely thickened with extensive adhesions. Postoperatively, persistent heart failure symptoms and imaging findings of fibrotic pericardium led to initiation of corticosteroid therapy, which resolved symptoms. This case underscores the diagnostic and therapeutic challenges of effusive-constrictive pericarditis in the context of COVID-19. Non-invasive imaging, particularly CMR, was instrumental in identifying active pericardial inflammation and guiding treatment. While pericardiectomy remains a critical intervention in refractory cases, this case highlights the potential benefit of immunosuppressive therapy in resolving residual symptoms. Given the limited literature on COVID-19-associated pericarditis, multidisciplinary management is essential for optimizing outcomes and guiding future research.

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Introduction

Initially regarded as a respiratory illness, SARS-CoV-2 has been associated with extrapulmonary manifestations, including cardiovascular disorders like myocardial injury, arrhythmias, acute coronary syndrome, thromboembolism, pericarditis and myocarditis. The risk of constrictive pericarditis is usually low for viral or idiopathic acute pericarditis, while it is more frequent for specific etiologies, including bacterial and tuberculous pericarditis.[1]

There are only a few cases reported of SARS-CoV-2 related effusive-constrictive pericarditis.

Case description

A 45-year-old male, diagnosed with mild COVID-19 in the previous week, was presented with acute pericarditis with moderate pericardial and bilateral pleural effusions. Admission tests revealed elevated NT-proBNP (221pg/ml, N 0–125 pg/mL) and C-reactive protein (15 mg/dL, N 0–0.5 mg/dL), normal troponin I levels and sinus tachycardia and diffuse negative T waves on ECG. Echocardiogram showed circumferential pericardial effusion (11 mm), without hemodynamic compromise; normal biventricular systolic function; no valve alterations; dilated inferior vena cava (IVC) without respiratory variation. The patient started colchicine and ibuprofen and was discharged under this treatment after reduction of the effusions. One week later, colchicine was discontinued due to abdominal discomfort.

Two weeks after discharge, the patient returned with orthopnea, edema of the lower limbs, fatigue, and decrease in urinary output in the previous three days. On examination, he exhibited heart rate of 104/ min, normal blood pressure, auscultation with normal heart sounds with no rub or murmurs and decreased breath sounds on the right, jugular venous distension, ascites and significant lower limb swelling.

Laboratory results revealed elevated NT-proBNP (587 pg/mL), C-reactive protein (4,17 mg/dL), alkaline phosphatase (135 U/L), gamma GT (84 U/L), and hyperbilirubinemia (1.6 mg/dL), with normal albumin and INR. ECG revealed sinus tachycardia, chest X-ray showed right pleural effusion, and echocardiogram revealed mild pericardial effusion (7 mm, circumferential), no signs of hemodynamic compromise but with apparent septal bounce, normal global biventricular systolic function; no valve alterations and dilated IVC.

He was admitted to investigate polyserositis, while maintaining ibuprofen and starting furosemide.

Thoracocentesis revealed a transudate according to light criteria and diagnostic tests for autoimmune, bacterial, and viral causes were negative (including IGRA). Echocardiography confirmed mild pericardial effusion, pericardial thickening, septal bounce, and respiration-related variation of atrioventricular transvalvular flow, without cardiac chambers collapse. The patient was discharged, with anti-inflammatory ther-

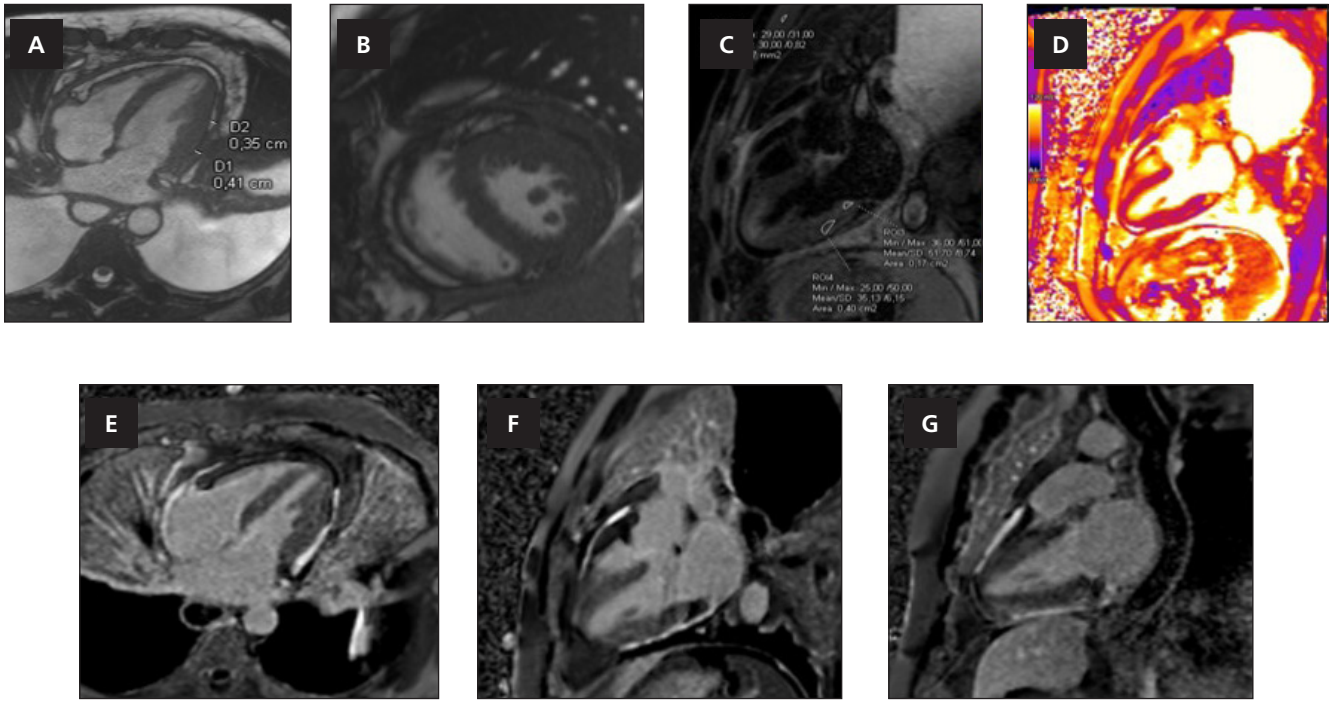

Fig. 1 – First cardiac magnetic resonance, performed on the day 49 after diagnosis of SARS-CoV-2 infection and after 39 days of anti-inflammatory therapy. Source: Clinical records of the patient. (A and B) Ventricular interdependence signs diffuse pericardial thickening and mild pericardial effusion. (C) T2 STIR without hypersignal. (D) T2 mapping with focally increased values in the basal inferolateral segment. (E, F) Late gadolinium enhancement with diffuse hypersignal of the pericardium and focal midventricular enhancement.

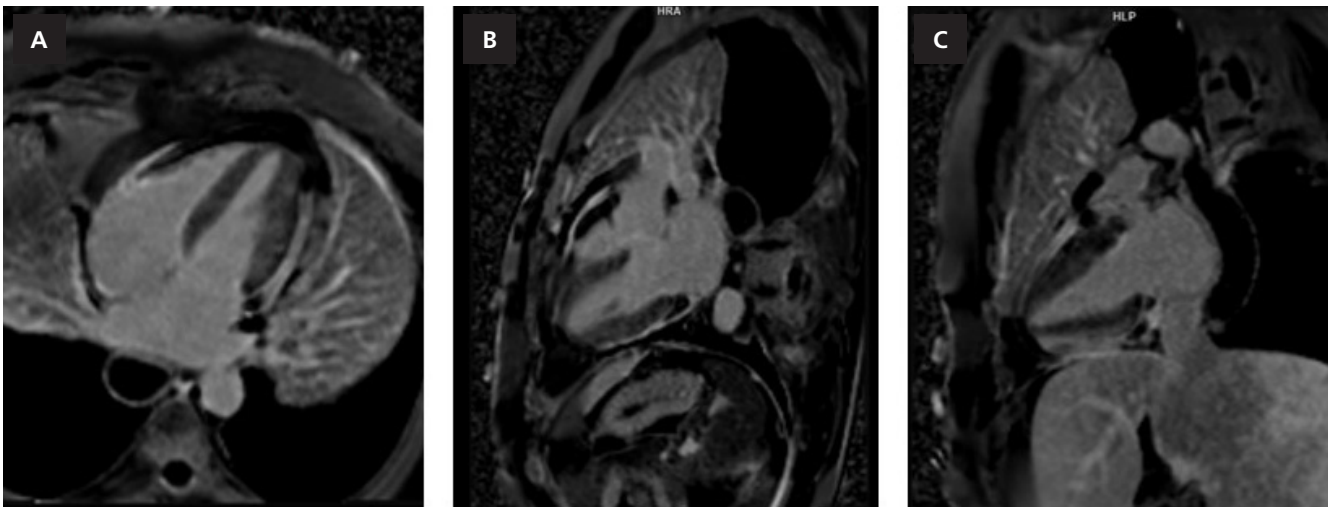

Fig. 2 – Second cardiac magnetic resonance, performed on the day 70 after diagnosis of SARS-CoV-2 infection and after 60 days of anti-inflammatory therapy. Source: Clinical records of the patient. (A–C) Reduced extent of the pericardial late gadolinium enhancement.

apy and furosemide, awaiting cardiac magnetic ressonance (CMR).

The CMR, performed on the 39th day of anti-inflammatory therapy showed signs of ventricular interdependence with septal bounce and respiration-related shift of the interventricular septum; myocardium with increased extracellular volume, focal increase of T2 mapping values in the basal inferolateral segment of the left ventricle, diffuse areas of midventricular late gadolinium enhancement (basal and middle segments of anterolateral, inferolateral and inferior walls, basal anterior, middle anteroseptal and infero-septal and apical inferior, lateral and septal); pericardium with diffuse thickening, mild pericardial effusion and diffuse late gadolinium enhancement; moderate bilateral pleural effusion; preserved systolic function of both ventricles; compatible with subacute myocarditis and effusive-constrictive pericarditis (**Fig. 1**). Symptoms worsened despite therapy, and he was readmitted.

He maintained anti-inflammatory therapy with ibuprofen, restarted colchicine and needed high doses of IV diuretics and sequential nephron block for right heart failure compensation. Cardiac catheterization performed on the 52nd day of anti-inflammatory therapy showed atypical right ventricle pressure curve without typical features of constriction.

CMR was repeated on the 60th day of therapy, with reduced extent of pericardial late gadolinium enhancement and less marked signs of ventricular interdependence (**Fig. 2**). After intensive intravenous diuretic therapy and hydro saline restriction, he showed progressive improvement, allowing discharge to the outpatient clinic.

The case was discussed with cardiac surgery, and the patient was subsequently submitted to pericardiectomy, revealing severely thickened pericardium (8 mm) with extensive adhesions.

Post-surgery CMR showed absence of pericardium in anterior topography, maintaining extensive areas of thickened pericardium with diffuse late enhancement with clear hyposignal that suggests a concomitant fibrotic process and thin lamina of pericardial effusion.

Persistent heart failure symptoms necessitated corticosteroid therapy (prednisolone 1 mg/kg/day), leading to symptom resolution.

Relevance and final considerations

Effusive-constrictive pericarditis diagnosis typically relies on intrapericardial pressure measurements,[2] but non-invasive imaging may be equally useful for its diagnosis.

CMR is critical for identifying active pericardial inflammation. The reversibility of the condition is thought to be related by the underlying pathophysiological mechanisms, which includes inflammation, fibrin deposits, and edema, whereas fibrosis and calcification indicate poorer outcomes.[3]

Limited data exist on COVID-19-associated pericarditis, with only two reported cases of effusive-constrictive pericarditis, one requiring pericardiectomy.[4,5]

Further research on immunosuppressive therapy and optimal timing for surgical intervention is needed. A multidisciplinary approach involving infectious disease specialists, cardiologists, and surgeons is essential for management.

Conflict of interest
Nothing to disclose.

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The patient provided informed written consent for the publication of the study data.

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A multifaceted prosthetic valve infective endocarditis

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Infekční endokarditida
Transezofageální echokardiografie
Zánětlivý stav

SOUHRN

Infekční endokarditida představuje významný problém veřejného zdraví s vysokou mortalitou a morbiditou. Do nemocnice byl s bolestí na hrudi dopraven 79letý muž po implantaci umělé aortální chlopně. Elektrokardiogram prokázal **přední infarkt myokardu s elevací úseku** ST. Urgentní koronarografie odhalila kritickou stenózu přední sestupné větve levé koronární tepny, která byla řešena angioplastikou a implantací lékového stentu. Transtorakální a transezofageální echokardiografie prokázala nozokomiální endokarditidu na biologické aortální chlopni a významný periprotetický absces. Kultivací krevních vzorků byla zjištěna přítomnost multirezistentní bakterie *Staphylococcus haemolyticus*. Po opakovaném transezofageálním vyšetření se pacient rozhodl – přes doporučení chirurgů – pro zahájení léčby antibiotiky během pobytu v nemocnici a v domácí péči. Tato kazuistika popisuje vzácný případ úspěšné konzervativní léčby řešící paravalvulární absces jako situaci obvykle vyžadující chirurgickou intervenci. Tento vynikající výsledek dokládá možnost nechirurgického řešení ve vybraných případech. Během sledování zůstává pacient v dobrém zdravotním stavu, což podtrhuje význam multidisciplinární péče a důsledného monitorování.

Keywords:
Acute myocardial infarction
Infectious endocarditis
Inflammatory state
Outpatient parental antimicrobial therapy
Transesophageal echocardiography

ABSTRACT

Infective endocarditis is a significant public health challenge, with high mortality and morbidity. A 79-year-old man, post-aortic bioprosthetic implantation, presented with chest pain. An electrocardiogram indicated anterior ST-elevation myocardial infarction. Urgent coronary angiography showed critical left anterior descending artery stenosis, treated with angioplasty and a drug-eluting stent. Transthoracic and transesophageal echocardiograms revealed nosocomial endocarditis on the aortic bioprosthetic and a significant peri-prosthetic abscess. Blood cultures identified multidrug-resistant *Staphylococcus haemolyticus*. Despite surgical recommendations, the patient opted for intra-hospital antibiotic therapy and home care after repeated transesophageal echocardiograms. Remarkably, this case underscores the rare and significant success of conservative therapy in resolving a paravalvular abscess, a condition typically necessitating surgical intervention. This extraordinary outcome highlights the potential for non-surgical management in selected cases. The patient remains in good health upon follow-up, emphasizing the importance of multidisciplinary care and vigilant monitoring.

Introduction

The pervasiveness of infective endocarditis (IE) underscores its substantial impact on public health, marked by considerable mortality and morbidity.[1] These trends find explanation in the overarching demographic shift towards an aging population, augmented utilization of valvular prostheses – encompassing percutaneous interventions – and the widespread deployment of implantable devices and central venous catheters.[2] Furthermore, the escalating challenge of antibiotic resistance contributes significantly to the complexity of managing this condition. Currently, a global incidence of IE is approximately 13.8 cases per 100,000 individuals, with more 66 000 fatalities.[3] The complications of endocarditis can be local, such as abscess formation or fistula, often necessitating cardiac surgery, or they can include arrhythmic complications. Endocarditis can lead to systemic embolization, particularly affecting the brain and spleen. Notably, aortic valve endocarditis can embolize into the coronary vessels, resulting in ischemia.

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Case presentation

A 79-year-old male presented to the Emergency Department (ED) with nocturnal-onset chest pain, complicated by hemodynamic compromise: non-invasive blood pressure was 80/50 mmHg, heart rate was 150 bpm, and oxygen saturation was 85% in room air. Three months prior, he underwent aortic bioprosthesis (Intuity 23) implantation for severe aortic stenosis. Pre-intervention coronary angiography revealed diffuse atheromatous plaques without significant stenosis. His medical history included paroxysmal atrial fibrillation, hypertension, and chronic obstructive pulmonary disease. The electrocardiogram depicted high ventricular response atrial fibrillation with ST segment elevation from V_1 to V_5 and reciprocal ST depression in inferior leads (**Fig. 1**). Arterial blood gas analysis revealed pH 7.25 and lactate 6 mmol/L. Prompt initiation of inotropic therapy and ventilatory support occurred, activating the catheterization lab. Coronary angiography unveiled a 90% stenosis of the mid-left anterior descending artery (**Fig. 2**), treated with angioplasty and coronary stent placement. Transthoracic echocardiography showed global left ventricular hypokinesis (ejection fraction [EF] 50%) linked to apical akinesia and aortic bioprosthesis with thickened leaflets suggestive of multiple vegetations. Transesophageal echocardiography (TOE) confirmed nosocomial IE, complicated by mild-to-moderate stenosis and a large abscess peri-prosthetic (**Fig. 3A** and **3B**). The abscess was confirmed on contrast-enhanced computed tomography (**Fig. 4**). Extensive patient interrogation revealed the onset of evening fever, weight loss, and general malaise over the past two weeks. The suspicion of nosocomial endocarditis on the aortic bioprosthesis was confirmed. The atherosclerotic plaque on the anterior interventricular branch increased in size, leading to a reduction in downstream flow. Together with the elevated heart rate and septic state, this resulted in the onset of a mixed cardiogenic and septic shock. A possible hypothesis is that the local and systemic inflammatory state induced by endocarditis may have contributed to the plaque rupture/erosion and shock state with low blood pressure, atrial fibrillation with high

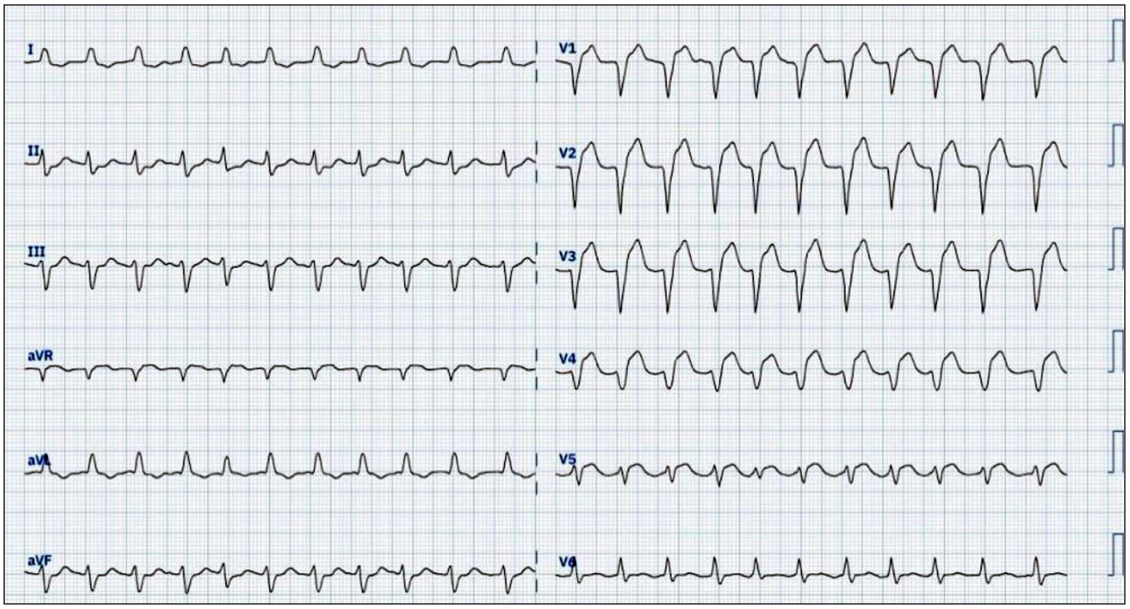

Fig. 1 – Electrocardiogram showing evidence of acute coronary syndrome with elevation of the anterior ST segment.

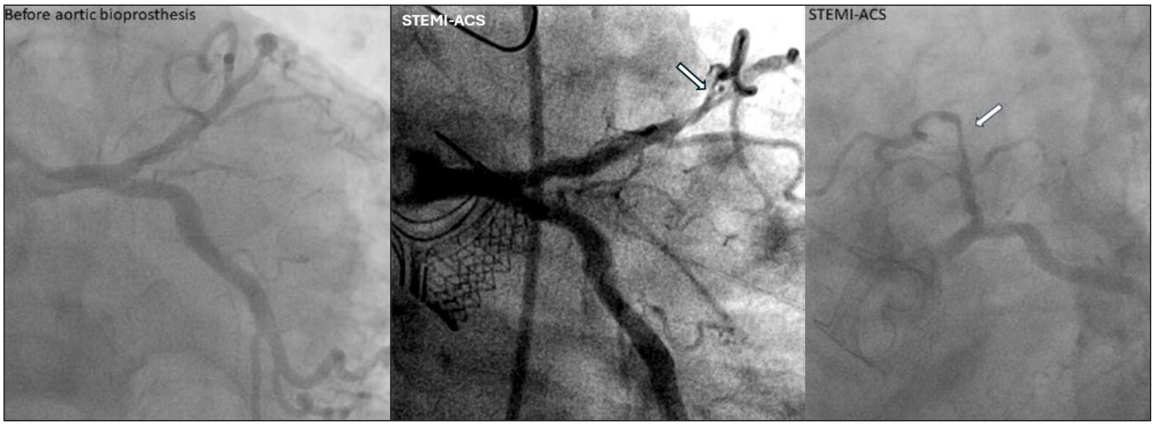

Fig. 2 – The two coronary angiographies compared.

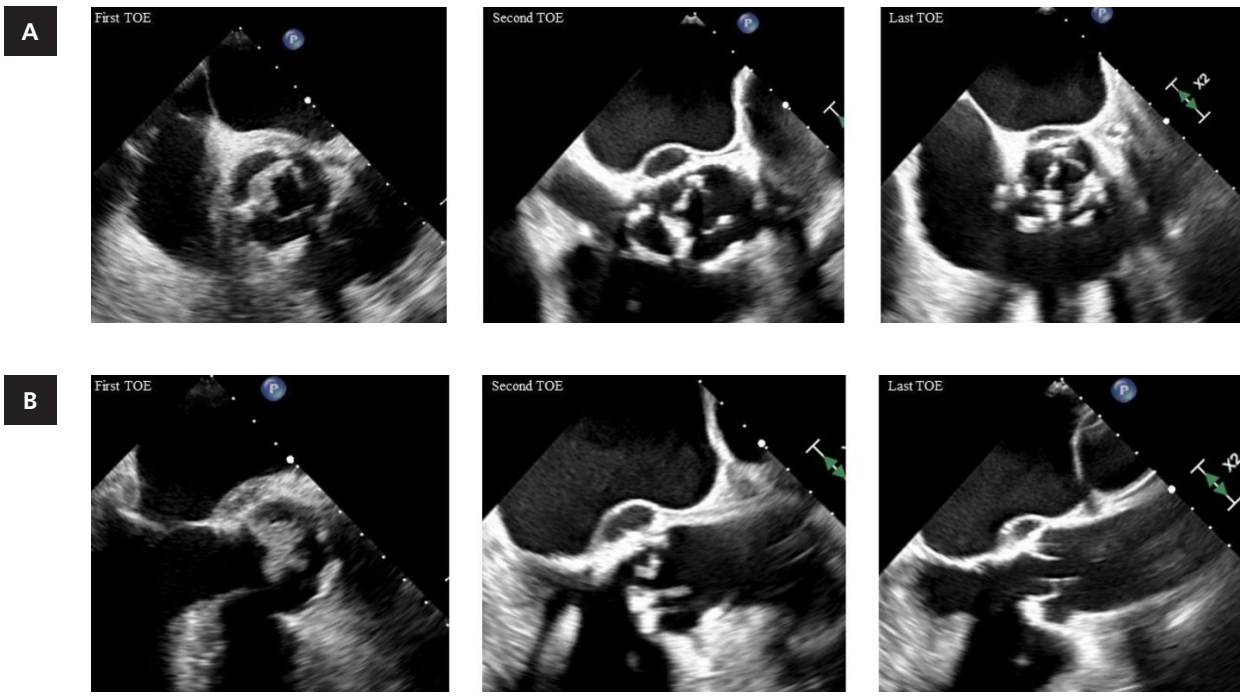

Fig. 3 – (A) Comparative transesophageal images: 45° aortic valve short-axis. (B) Comparative transesophageal images: 120° aortic valve long-axis.

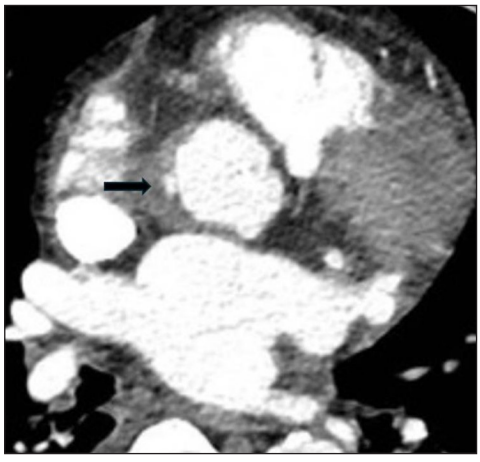

Fig. 4 – Abscess formation (arrow) on cardiac computed tomography in axial projection.

ventricular response could contribute to acute coronary syndrome. Blood cultures identified multidrug-resistant *Staphylococcus haemolyticus*, and meropenem was initiated as antibiotic therapy. Despite the recommendation for cardiac surgery, the patient and his family opted against it due to the high operative risk. Follow-up TOE, after approximately six weeks of intravenous antibiotic therapy, showed reduced vegetations on the bioprosthetic valve leaflets and partial resolution of abscess (**Fig. 3A** and **3B**). With hemodynamic stability achieved, the patient was discharged home while continuing intravenous antibiotic therapy with Dalbavancina, under the outpatient parenteral antimicrobial therapy (OPAT) protocol. At the one-month post-discharge follow-up, the patient remained asymptomatic and hemodynamically stable, and a new TOE demonstrated normally mobile bioprosthetic valve leaflets without vegetations and a reduced abscess without paravalvular regurgitation (**Fig. 3A** and **3B**).

Discussion

In synthesizing these clinical and epidemiological insights, our case report underscores the multifaceted nature of IE, accentuating the need for a holistic and individualized approach to its prevention, diagnosis, and management. IE poses a dual threat to coronary health, with embolic events and inflammatory processes contributing to the intricate relationship between inflammation and myocardial infarction. The embolization of endocardial vegetations into the coronary lumen, albeit rare, is the most recognized cause of myocardial infarction in infective endocarditis.[4] However, the systemic and local inflammatory state due to IE can play a significant pro-atherosclerotic role.[5] Chronic inflammation, characterized by dysfunctional immune responses, destabilize atherosclerotic plaques to their rupture and subsequent myocardial infarction. The inflammation contributes to the formation of coronary thrombi, thereby increasing the risk of acute coronary syndromes. Conditions characterized by pro-inflammatory states have consistently been linked to a higher risk of atherosclerosis. The intricate interplay between inflammation, atherosclerosis, and their impact on coronary health is emphasized by the recognition that inflammatory immune-mediated disorders, such as rheumatic diseases, inflammatory bowel diseases, rheumatoid arthritis, and systemic lupus erythematosus are closely associated with acute cardiovascular events indepen-

dently of traditional cardiovascular risk factors.[6,7] Indeed, patients affected by rheumatoid arthritis undergoing treatment with anti-inflammatory medications exhibit a lower incidence of acute coronary syndrome compared to those not utilizing such medications.[8] The dangerousness of untreated inflammation has recently been confirmed in patients with ischemic heart disease under optimized medical therapy.[9,10] The recent guidelines from the European Society of Cardiology have recommended an anti-inflammatory treatment in these patients.[11] In our case, pre-implantation coronary angiography revealed extensive atheromatosis. In our case, extensive atheromatosis was observed during pre-implantation coronary angiography. Given the patient's ongoing and highly effective statin therapy, it's improbable that this atheromatous condition would have deteriorated significantly within a mere three months without additional external factors. Therefore, the inflammatory state accelerated the atherosclerotic process, ultimately culminating in plaque rupture. Another challenging aspect of the patient's condition was the extent of perivalvular disease, marked by the formation of extensive abscess cavities, which warranted consideration for cardiothoracic surgical intervention. Nosocomial endocarditis is often highly destructive, exhibiting a diverse course depending on the pathogenicity of the involved microorganism. In this case, the course was subacute, as evidenced by positive blood cultures for coagulase-negative staphylococci (CoNS). Given the patient's refusal of surgery, a two-phase antibiotic therapy, proved successful in eradicating the infection. Notably, the infection resolved without causing dysfunction of the aortic bioprosthesis. While the OPAT protocol is typically reserved for stable patients without extensive disease,[2] the decision to continue antibiotic therapy at home was made after weeks of antibiotic treatment and improvement seen on a follow-up TOE. This choice proved judicious, impacting both hospitalization costs and the patient's overall well-being positively. The conservative antibiotic strategy resulted in the resolution of the paravalvular abscess, representing one of the few cases documented in the literature with a long-term follow-up demonstrating the patient's clinical well-being.

Conclusion

In conclusion, IE not only causes coronary embolic events but also profoundly influences coronary atherosclerosis through its inflammatory state. These mechanisms should be better understood to provide new therapeutic options. The conservative antibiotic therapy resolved the paravalvular complication, an exceedingly rare event in clinical practice. Furthermore, the management of endocarditis and its complications always requires a multidisciplinary effort.

Conflict of interest
The authors declare they have no competing interests.

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Ethical statement
Due to the nature of this report, ethical approval was not applicable.

Informed consent
The patient has given his consent for the use of his medical data.

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Successful Implantation of a Dual-Chamber Permanent Pacemaker in a Patient with Persistent Left Superior Vena Cava and Absence of Right Superior Vena Cava: Tips and Tricks

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Challenging pacemaker implant
Persistent left superior vena cava

SOUHRN

Ageneze pravostranné horní duté žíly při perzistující levostranné horní duté žíle (persistent left superior vena cava, PLSVC) představuje vzácnou a obecně asymptomatickou vrozenou malformaci. Obvykle se zjistí náhodně během implantace kardiostimulátoru. V této kazuistice popisujeme naše zkušenosti s implantací dvoudutinového kardiostimulátoru u pacienta s tak složitými anatomickými poměry a vyzdvihujeme klinický význam venografie pro jednoznačný popis anatomie konkrétního žilního systému na operačním sále. Zdůrazňujeme zvláště metody použité pro zajištění správného umístění elektrody.

ABSTRACT

An absent right superior vena cava associated with persistent left superior vena cava (PLSVC) is a rare and generally asymptomatic congenital malformation. It is usually discovered incidentally during pacemaker (PM) implantation. In this report, we describe our experience with implanting a dual-chamber pacemaker in a patient with such complex anatomy and the clinical value of peripheral phlebography for clearly describing venous anatomy in the surgical room. In particular, we highlight the methods used to ensure correct lead positioning.

Introduction

Persistent left superior vena cava (PLSVC) is a rare vascular anomaly that is usually identified incidentally during cardiovascular imaging or procedures. This anomaly arises because of the failure of the left anterior cardinal vein to regress during embryonic development, which occurs in 0.3–0.5% of healthy individuals.[1,2] Generally, the PLSVC drains into a dilated coronary sinus (CS); however, in rare instances, it can drain into the left atrium via an unroofed CS or, less commonly, directly. Almost 90% of cases present with bilateral superior venae cavae; conversely, the absence of the right superior vena cava (ARSVC) is rare, with a prevalence of 0.09% to 0.13% in the population.[3]

PLSVC might be associated with abnormal electrophysiological function resulting from anatomical and structural heart abnormalities, presenting clinically as either tachycardia or bradyarrhythmia.[4,5] Although PLSVC is often benign, it can complicate invasive procedures, such as pacemaker (PM)/Implantable Cardioverter-Defibrillator (ICD) implantation, central venous access, and cardiothoracic surgery. We report successful and uncomplicated implantation of a dual-chamber PM in a patient with PLSVC and ARSVC who experienced a complete heart block.

Case presentation

A previously healthy 46-year-old male patient was referred to our hospital with complaints of shortness of breath, dizziness, and weakness. He had no significant medi-

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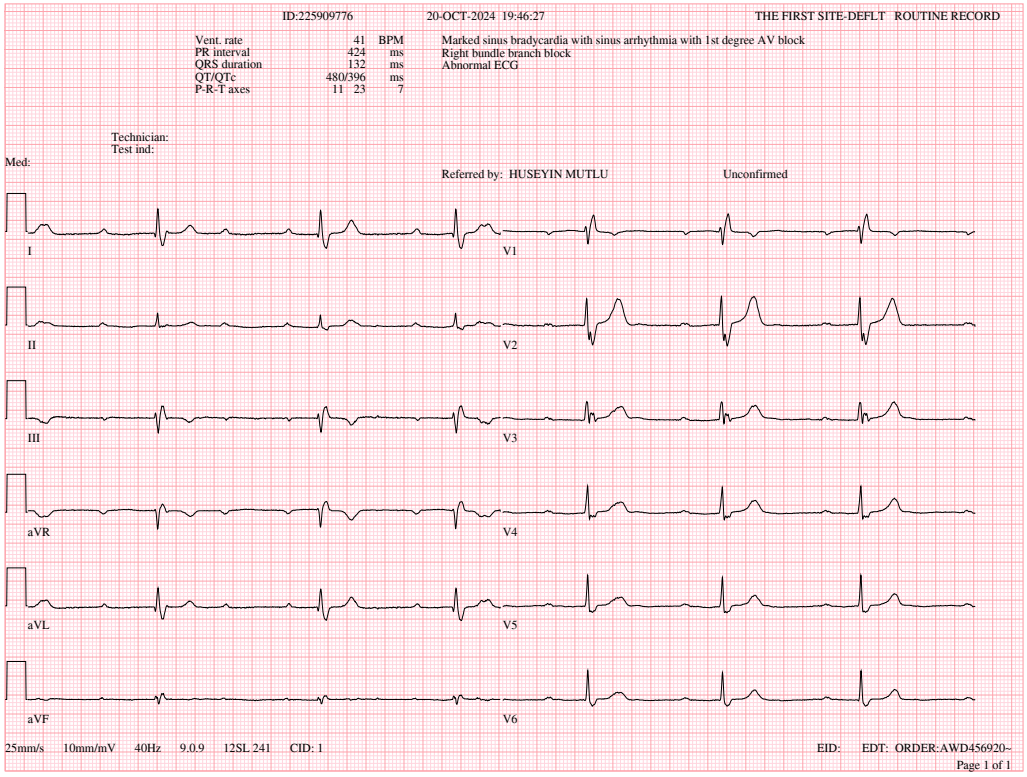

Fig. 1 – A 12-lead ECG showed a ventricular rate of 4 1/min with atrioventricular dissociation.

cal history and denied experiencing angina, palpitations, syncope, or any related symptoms. On examination, his blood pressure was 148/66 mmHg with a regular pulse of 38 beats per minute, with no clinical evidence of heart failure. His examination was otherwise unremarkable. A 12-lead electrocardiogram revealed a complete atrioventricular block with a ventricular rate of 34 beats per minute and right bundle branch block morphology (**Fig. 1**). Transthoracic echocardiography revealed normal left ventricular size and function. There was no laboratory evidence of either a metabolic or ischemic cause of the conduction disease. Coronary angiography revealed normal coronary arteries, and dual-chamber PM implantation was scheduled.

The procedure was initially performed using the left subclavian approach with a left pectoral incision. Following the left subclavian puncture, resistance was encountered while attempting to advance the lead, leading to its retraction. A venogram revealed a PLSVC draining into the right atrium (RA) via the CS and its acute angle of emergence (**Video 1**). Therefore, we decided to place the lead contralaterally via the right subclavian vein. The right subclavian vein was cannulated under fluoroscopy. Unexpectedly, the venogram revealed the ARSVC, with the right brachiocephalic vein draining directly into the PLSVC.

The placement of the right atrial pacing lead into the RA cavity was not particularly challenging; however, positioning it within the right atrial appendage required careful manipulation. After entering the RA cavity, the straight stylet of the RA pacing lead was replaced with the curved "J" stylet, and clockwise torque was required to advance the lead anteriorly and place it in the adjacent atrial appendage location.

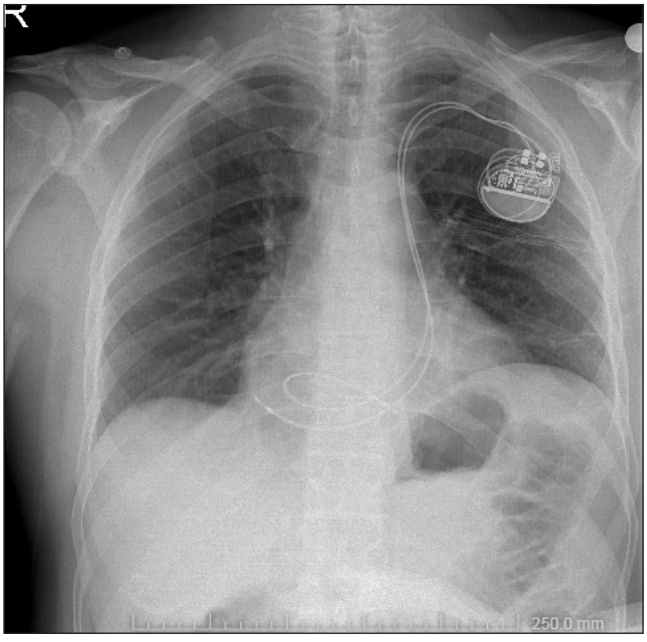

Fig. 2 – Radiographic view of the final placement of the right atrial and ventricular leads via persistent left superior vena cava and coronary sinus.

Positioning the right ventricular (RV) pacing lead was more challenging since the pacing lead gained atrial access through the CS ostium, which was very close to the RV inlet, but the CS ostium directed the lead away from

the RV inlet toward the lateral free wall of the RA. The stylet was shaped into a semicircle to facilitate entry into the RV and then pushed into the RV by making a loop against the lateral wall of the RA. Retracting the stylet a few centimeters was crucial to direct the tip of the RV pacing lead from the RA-free wall into the RV and ensure that the lead tip was no longer stiff, thus minimizing the risk of atrial perforation. The lead was positioned near the RV apex, which was confirmed by fluoroscopy (**Fig. 2, Video 2**). Care was taken to form an adequate loop in the RA to avoid dislodgement in the future. Both leads exhibited excellent parameters, with thresholds of 0.9 mV for each and resistances of less than 750 ohm. A dual-chamber PM was connected to the leads and implanted in a deep pocket. The wound was then closed in layers. The recovery was uneventful, and the patient was discharged after 2 days.

Discussion

In the current case, we reported our experience with PM implantation in a patient with a rare venous anomaly consisting of an absent right and a PLSVC.

PLSVC is the most common benign thoracic venous anomaly and is usually discovered incidentally. Nevertheless, the presence of PLSVC affects the structure of the heart and vessels. The PLSVC drains approximately 20% of the total venous return and significantly dilates the CS. Draining directly into the left atrium is often associated with complex cardiac pathologies, such as atrial septal defect and aortic coarctation.[1] Therefore, timely identification of PLSVC, which is associated with conduction abnormalities and congenital heart disease, is crucial for patient prognosis.

The PLSVC can be identified before PM implantation. This anomaly may be suspected during a routine chest X-ray performed after central venous access is established. Moreover, echocardiography that shows a dilated CS should also be considered. These findings should encourage clinicians to use preoperative venous imaging. Understanding the anatomy facilitates the procedure and allows for more complex interventions because it is associated with the use of techniques to overcome pathological angulations.

The potential for a PLSVC should be considered during device implantation if the guiding wire moves in a left downward direction or if resistance is felt while trying to push it forward. In this situation, performing venography before the procedure is an effective and simple method. Injecting 50 cc of contrast agent into each subclavian vein allows for precise mapping of the thoracic venous system, and the ARSVC can be easily documented.

The PM implantation with PLSVC remains challenging. The large size and thin wall of the CS increase the risk of injury during lead implantation, raising safety concerns. In addition, accessing through the PLSVC is technically complex, resulting in a higher likelihood of failed lead implantation and difficulties in achieving the optimal lead position. Hand-shaped styles and active fixation leads might help to overcome technical difficulties and achieve successful long-term results.

Conclusion

The ARSVC and PLSVC is an extremely rare congenital anomaly encountered unexpectedly during routine PM implantation. Intraprocedural venography is highly effective in verifying the ARSVC and in accurately determining the CS route. An experienced operator can overcome anatomical challenges by using shaped styles and active fixation leads.

Acknowledgements
None.

Conflict of interest
None.

Ethical statement
We declare that the case report was conducted following the applicable ethical standards and guidelines, as outlined in the Declaration of Helsinki.

Informed consent
Appropriate permissions, including written informed consent, were obtained.

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Epicardial Pacing Lead Implantation in Complete Heart Block as a Manifestation of Cardiac Mass: Case Report

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Epikardiální kardiostimulátor
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Obstrukce útvarem v srdci

SOUHRN

Kontext: Abnormální atrioventrikulární (AV) převod se může projevovat širokým spektrem klinických nálezů od asymptomatických případů až po těžké symptomy spojené s bradykardií. Zatímco transvenózní implantace kardiostimulátoru je obecně ceněna pro svůj minimálně invazivní charakter a účinnost v klinické praxi, tento přístup může být kontraindikován u pacientů s anatomickými anomáliemi nebo s významnými komorbiditami. V této kazuistice přinášíme popis vzácného případu intrakardiálního útvaru tvořícího překážku převodní dráhy srdeční, přičemž klasický transvenózní přístup nebyl proveditelný. V takových složitých scénářích se jako nejvhodnější a nejúčinnější intervence jeví implantace epikardiálního kardiostimulátoru.
Popis případu: Do nemocnice byl přivezen 48letý muž s postupným úbytkem tělesné hmotnosti a dyspnoe po dobu šesti měsíců, přičemž v předchozím týdnu došlo k exacerbaci symptomů. Transtorakální echokardiografie odhalila intrakardiální útvar v pravé síni, mezisíňové přepážce a v levé síni. Následně provedené vyšetření výpočetní tomografií (computed tomography, CT) potvrdilo maligní zvětšující se solidní útvar v mezisíňové přepážce, který zasahoval do pravé i levé síně, pravé plicní žíly a dolní duté žíly a připomínal primární srdeční tumor. Okraje útvaru byly nepravidelné a samotný útvar měl rozměry přibližně 6,0 × 6,0 × 8,2 cm. Útvar přerušil převodní AV dráhu a způsobil kompletní srdeční blokádu s následnou hemodynamickou nestabilitou. Vzhledem k velikosti útvaru a jeho anatomickému uložení v pravé síni bylo transvenózní zavedení stimulační elektrody považováno za neproveditelné pro potenciální riziko jejího chybného umístění, perforace nádoru nebo embolizace; proto byla provedena chirurgická implantace epikardiálního kardiostimulátoru. Výkon byl úspěšný a bez komplikací; u pacienta bylo zaznamenáno významné zlepšení hemodynamiky. Pooperační zotavení proběhlo bez zvláštních příhod a pacient byl propuštěn pět dní po operaci s výrazným zlepšením klinického stavu.
Diskuse: Tento případ zdůrazňuje vzácný a komplexní projev velkého intrakardiálního útvaru vyvolávajícího systémové symptomy, jako je úbytek tělesné hmotnosti a dyspnoe postupně se zhoršující po dobu šesti měsíců. Případ byl z hlediska diagnostiky a léčby náročný obzvláště pro skutečnost, že útvar zasahoval do několika srdečních struktur včetně pravé síně, mezisíňové přepážky a levé síně. Vzhledem k tomu, že útvar již značně prorostl srdeční a žilní struktury, nebylo možné provést transvenózní implantaci kardiostimulátoru; takový výkon by byl navíc riskantní. V tomto kontextu představovalo bezpečnou a účinnou alternativu zavedení epikardiální elektrody, jež by umožnilo vyhnout se rizikům spojeným s transvenózním přístupem. Popsaný přístup vyzdvihuje zásadní úlohu implantace epikardiálního kardiostimulátoru při léčbě pacientů s obstruujícími útvary v srdci a zdůrazňuje účinnost a bezpečnost tohoto postupu v situacích, kdy klasické transvenózní přístupy nelze použít.
Závěr: Epikardiální zavedení elektrody je nutno zvážit v situacích, kdy nelze provést primární transvenózní umístění elektrody ve složitých situacích, jakou je právě popsaný případ, kdy srdeční hmota obstruovala dráhu AV převodu.

ABSTRACT

Background: Atrioventricular (AV) conduction abnormalities can manifest with a wide spectrum of clinical presentations, ranging from asymptomatic cases to severe bradycardia-associated symptoms. While transvenous pacemaker implantation is widely regarded for its minimal invasiveness and clinical efficacy, this approach may be contraindicated in patients with anatomical anomalies or significant comorbidities. This report presents a rare case of an intracardiac mass obstructing the AV conduction pathway, where the conventional

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transvenous approach was not feasible. In such complex scenarios, epicardial pacemaker implantation emerges as the most suitable and effective intervention.

Case description: A 48-year-old male presented with a six-month history of progressive weight loss and dyspnea, with symptom exacerbation over the preceding week. Transthoracic echocardiography revealed an intracardiac mass involving the right atrium, interatrial septum, and left atrium. Subsequent computed tomography (CT) imaging confirmed a malignant, enhancing solid mass occupying the interatrial septum, with extensions into the right atrium, left atrium, right pulmonary vein, and inferior vena cava, suggestive of a primary cardiac tumor. The mass exhibited irregular borders and measured approximately 6.0 × 6.0 × 8.2 cm. This mass disrupted the AV conduction pathway, leading to complete heart block and resulting in hemodynamic instability.

Given the mass's size and its anatomical position within the right atrium, transvenous pacemaker lead placement was deemed unfeasible due to the potential risk of lead misplacement, tumor perforation, or embolization. Thus, surgical epicardial pacemaker implantation was performed. The procedure was completed successfully without complications, resulting in significant improvement in the patient's hemodynamic status. Postoperative recovery was uneventful, and the patient was discharged five days after surgery with marked clinical improvement.

Discussion: This case underscores a rare and complex presentation of a large intracardiac mass causing systemic symptoms such as weight loss and dyspnea, progressively worsening over six months. The involvement of multiple cardiac structures, including the right atrium, interatrial septum, and left atrium, posed significant diagnostic and therapeutic challenges. The mass's extensive involvement of cardiac and venous structures rendered transvenous pacemaker implantation impractical and potentially hazardous. In this context, epicardial lead placement provided a safe and effective alternative, circumventing the risks associated with transvenous approaches. This case highlights the critical role of epicardial pacemaker implantation in managing patients with obstructive cardiac masses, emphasizing its efficacy and safety in scenarios where conventional transvenous methods are not viable.

Conclusion: Epicardial lead placement should be considered when primary transvenous lead placement cannot be performed in challenging cases, such as in the case where a cardiac mass obstructs the AV conducting pathway.

Keywords:
Cardiac mass obstruction
Complete atrioventricular block
Epicardial pacemaker
Intracardiac mass

Background

Patients with atrioventricular (AV) conduction abnormalities can present with a wide spectrum of clinical manifestations, ranging from asymptomatic cases to severe bradycardia-induced symptoms.[1] Numerous conditions, both congenital and acquired, can impair the AV conduction system, leading to varying degrees of AV block. In rare instances, structural abnormalities such as cardiac masses may disrupt the integrity of the AV conduction pathway, resulting in complete heart block and necessitating permanent pacing.[2] While transvenous pacemaker implantation remains the standard approach due to its minimally invasive nature and proven efficacy, it may be contraindicated in cases with anatomical alterations or significant comorbidities. This report describes a rare case of a cardiac mass obstructing the AV conduction pathway, rendering the conventional transvenous approach unfeasible. In such complex clinical scenarios, epicardial pacemaker implantation offers a safe and effective alternative, ensuring optimal patient outcomes when traditional methods are not viable.

Case description

A 48-year-old male presented with a six-month history of progressive weight loss and dyspnea, which had significantly worsened over the preceding week. Initial transthoracic echocardiography, as illustrated in **Figure 1**, revealed an intracardiac mass involving the right atrium, interatrial septum, and left atrium. Further evaluation with computed tomography (CT) imaging confirmed the presence of a malignant, enhancing solid primary cardiac mass measuring approximately 6.0 × 6.0 × 8.2 cm. The mass exhibited irregular borders, occupied the interatrial septum, and extended into the right atrium, left atrium, right pulmonary vein, and inferior vena cava. This extensive involvement disrupted the atrioventricular (AV) conduction pathway, resulting in complete heart block and subsequent hemodynamic instability, as demonstrated in **Figure 2**.

Due to the tumor's considerable size and critical location within the right atrium, safe transvenous pacemaker lead placement was deemed impractical. The mass's involvement of the right atrium and adjacent venous structures posed a significant risk of procedural complications, such as lead misplacement, tumor perforation, or embolization. As a result, surgical epicardial lead implantation was identified as the most appropriate and effective intervention for this patient.

Intraoperatively, the pericardium was carefully opened, revealing dense adhesions between the epicardial surface and the underlying cardiac wall, likely secondary to tumor infiltration or chronic inflammation. The pacemaker lead was meticulously affixed to the visceral epicardium using 4-0 polypropylene sutures to ensure stable lead positioning. Epicardial sensitivity testing was conducted at the anatomical right ventricle (RV) site using a unipolar pacemaker lead, as illustrated in **Figure 3**. The obtained ventricular lead parameters demonstrated optimal function, with a pacing threshold of 1.0 V, sensing amplitude of 4.8 mV, and impedance measuring 310 ohms, confirming the lead's appropriate placement and functionality.

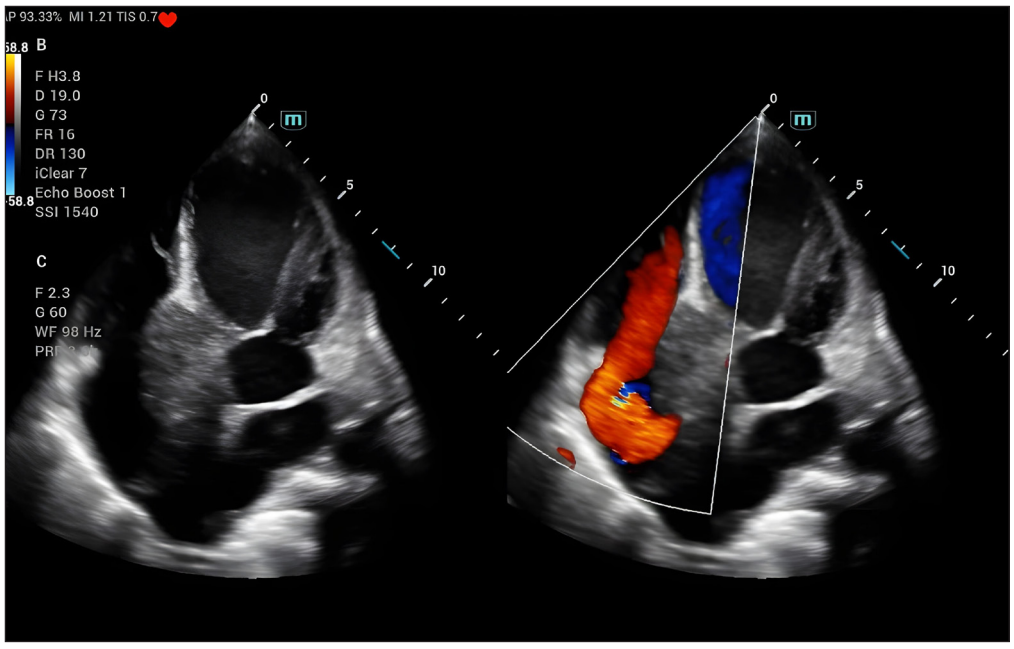

Fig. 1 – 2D transthoracic echocardiography showing mass in right atrium, interatrial septum, and left atrium.

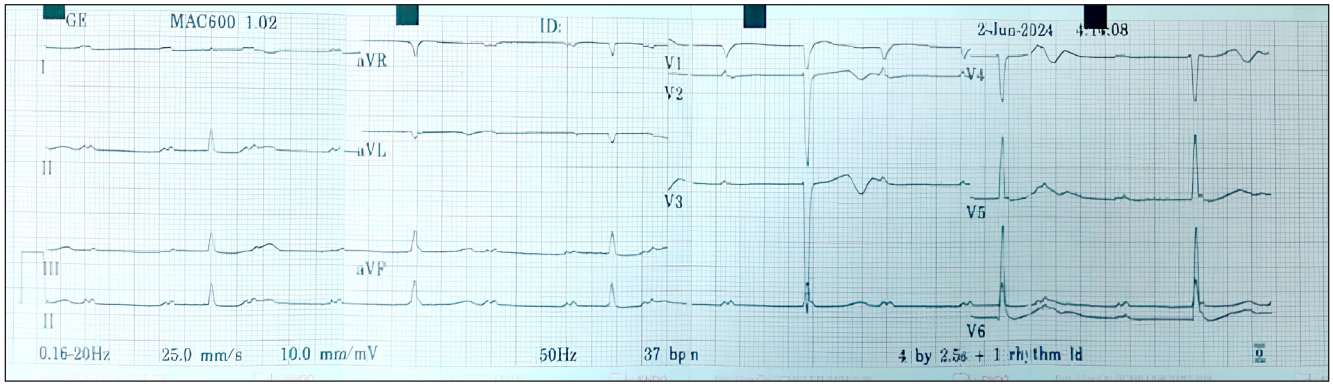

Fig. 2 – Pre-procedural ECG.

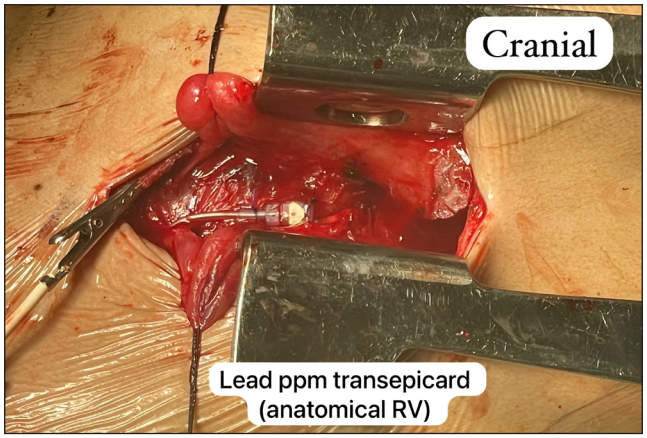

Fig. 3 – Epicardial lead implanted PPM through left anterolateral thoracotomy.

The procedure was completed without complications, leading to significant improvement in the patient's hemodynamic status. After confirming satisfactory pacing and sensing parameters, an incision was made in the upper left quadrant of the abdomen to create a generator pocket, as shown in **Figure 4**. The pacemaker lead was carefully tunneled from the abdominal pocket to the left hemithorax region to facilitate connection. The ventricular lead was then attached to the pulse generator, set in VVI mode, and securely positioned to ensure optimal device function. Additionally, a pericardial biopsy was performed to further assess the histopathological characteristics of the cardiac mass. Following the completion of device implantation, the left anterior thoracotomy incision was closed meticulously using layered suturing techniques to promote optimal wound healing and minimize postoperative complications. Due to the presence of a left pleural effusion, a 28 Fr chest tube was inserted into the left hemithorax for effective drainage.

The pacemaker implantation was successfully completed without any intraoperative complications, and the patient's hemodynamic parameters improved significantly. Post-procedural electrocardiogram (ECG) findings confirmed restored ventricular pacing, as demonstrated in **Figure 5**. The patient's postoperative course was stable, and he was discharged from the hospital five days after surgery, exhibiting marked clinical improvement.

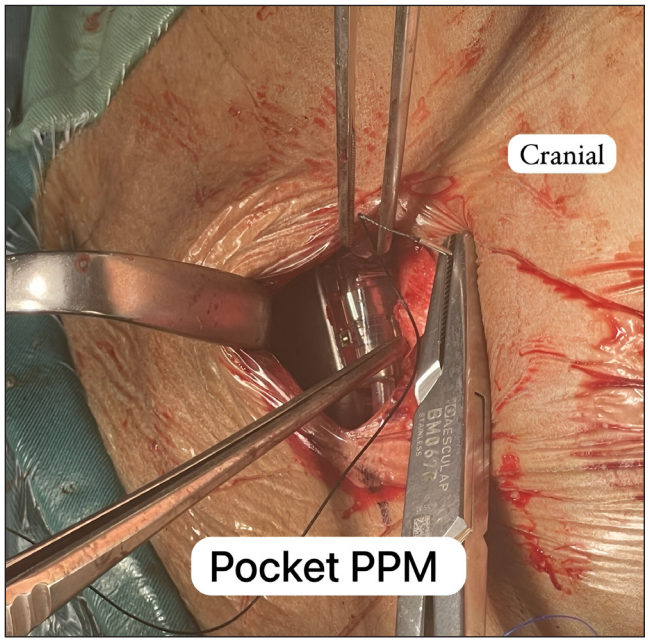

Fig. 4 – Generator pocket in the upper left quadrant of the abdomen.

Discussion

This case highlights a rare and complex presentation of a large intracardiac mass, manifesting with systemic symptoms such as progressive weight loss and dyspnea over six months. The extensive involvement of multiple cardiac structures, including the right atrium, interatrial septum, and left atrium, posed significant diagnostic and therapeutic challenges. Cardiac masses are broadly categorized as neoplastic or non-neoplastic, with neoplastic lesions further classified into primary benign, primary malignant, and metastatic tumors.[3,4] Primary cardiac tumors are exceedingly rare, with an autopsy-reported incidence ranging from 0.0017% to 0.33%. Among these, approximately 75% are benign, with cardiac myxomas being the most common, accounting for nearly half of all primary cardiac tumors.[3–6] The rarity and complexity of this case underscore the need for a multidisciplinary approach in the diagnosis and management of intracardiac masses, particularly when standard therapeutic strategies, such as transvenous pacemaker placement, are rendered unfeasible due to anatomical constraints. In the present case, computed tomography (CT) imaging revealed a solid mass with an attenuation of 30 Hounsfield Units (HU), characterized by spiculated margins, well-defined borders, and irregular edges. The mass measured approximately 6.0 × 6.0 × 8.2 cm and was located within the interatrial septum. Post-contrast imaging demonstrated enhancement of the mass, with attenuation increasing to 51 HU. The mass extended partially into the lumens of the right atrium, left atrium, right pulmonary vein, and inferior vena cava, with additional infiltration into the medial mediastinum. The central axis of the mass remained intracardiac. Furthermore, multiple pulmonary nodules were identified in both lungs, with the largest measuring approximately 1.0 × 1.1 × 0.9 cm in the lateral segment of the right middle lobe. A subpleural nodule, measuring 1.4 × 1.3 × 0.9 cm, was observed in the left hemithorax. These findings raised suspicion of a malignant, enhancing solid mass within the interatrial septum, suggestive of a primary cardiac tumor. The presence of multiple pulmonary nodules and a subpleural lesion indicated a potential metastatic process. A biopsy of the patient's tumor was obtained, and the initial histopathological result indicated a diagnosis of lipoma. However, due to the challenging nature of obtaining an adequate tissue sample through the available access, the quality of the specimen may not be optimal for definitive pathological evaluation. Therefore further biopsy and diagnostic assessment are required to confirm the diagnosis with greater accuracy.

Complete heart block necessitates pacemaker implantation due to the risk of serious complications, including hemodynamic instability and sudden cardiac events.[7] In this case, the tumor's considerable size and its critical location within the right atrium rendered transvenous lead placement impractical. The tumor's proximity to major venous structures significantly increased the risk of procedural complications, such as tumor perforation or embolization, during the advancement of transvenous leads. Given these anatomical and procedural challenges, surgical epicardial lead implantation was deemed the safest and most effective intervention.

Epicardial permanent pacemaker implantation is specifically indicated in cases where traditional transvenous pacing is unfeasible or contraindicated, such as in patients with complex congenital heart anomalies, lim-

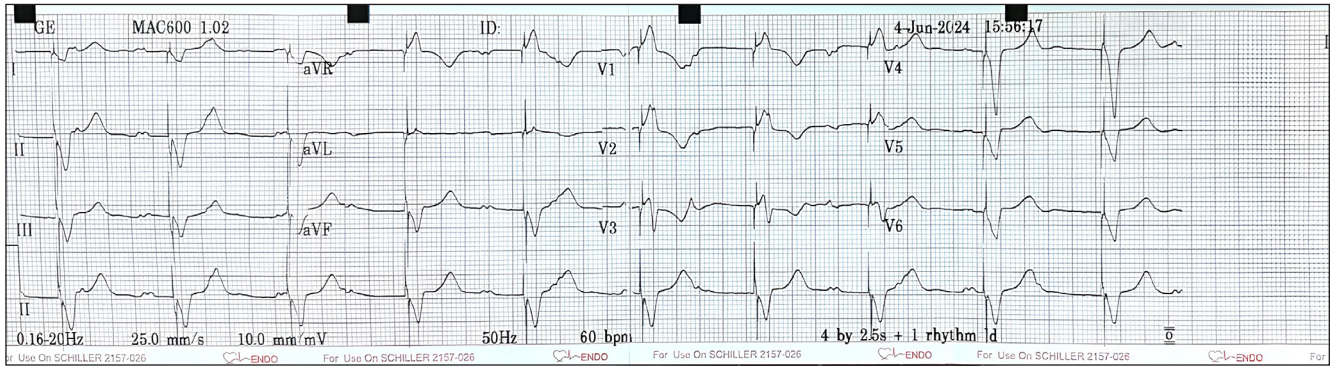

Fig. 5 – Post-procedural ECG.

ited venous access due to superior vena cava or subclavian vein occlusion, intracardiac mass in right atrial and right ventricle near the tricuspid, tricuspid atresia, or the presence of a mechanical tricuspid valve prosthesis. Additionally, it is preferred in patients at risk for recurrent infections, including those with a history of infective endocarditis involving the tricuspid valve.[7] Although epicardial pacemakers are more commonly utilized in pediatric populations and individuals with congenital cardiac abnormalities, their role extends to adult patients with structural heart disease where conventional transvenous approaches pose significant risks.[7,8] Unlike the standard transvenous technique, which involves threading leads through the venous system into the heart, epicardial pacing requires a more invasive surgical approach, with the leads being directly affixed to the epicardial surface of the myocardium.[8] This technique ensures reliable pacing in anatomically complex cases, reinforcing its value as an essential alternative when transvenous lead placement is not viable.

Various surgical approaches can be employed for epicardial pacemaker implantation, with the choice of technique depending on anatomical considerations, procedural complexity, and patient-specific factors. The most commonly utilized approaches include sternotomy, which provides optimal cardiac exposure through a standard midline incision, or a limited lower mini-sternotomy for a less invasive alternative.[9,10] Thoracotomy, performed via a left anterior or anterolateral approach, allows direct access to the epicardial surface and is frequently used in cases where sternotomy is not preferred.[11] A subcostal approach, involving a subxiphoid incision within the upper rectus sheath, serves as another viable option for pericardial access with minimal disruption to thoracic structures.[11] Additionally, video-assisted thoracic surgery (VATS) has emerged as a minimally invasive alternative, utilizing thoracoscopic guidance to facilitate lead placement while minimizing surgical trauma and postoperative recovery time.[12] In this case, a left anterolateral thoracotomy was chosen as the preferred approach, providing direct visualization and access to the myocardial surface for lead placement.

The epicardial pacemaker leads were secured using an active fixation mechanism, which incorporates a small screw (helix) at the lead tip to anchor it directly to the cardiac tissue.[1] Active fixation offers several advantages over passive fixation, including greater flexibility in lead placement, particularly in regions where passive fixation may not be feasible, such as the atrial wall or interventricular septum. Additionally, the screw mechanism provides enhanced lead stability, reducing the risk of dislodgement and ensuring long-term pacing reliability. However, this approach also carries potential drawbacks, such as an increased risk of myocardial trauma or perforation during lead placement. Moreover, lead extraction is inherently more complex, with a higher likelihood of cardiac tissue damage, making future revisions or removals more challenging.[13]

Patients with total atrioventricular block (TAVB) typically require permanent pacemaker (PPM) implantation, with the choice between dual-chamber (DDD) or ventricular (VVI) pacing modes depending on clinical and logistical factors. DDD pacing is considered more physiological, as it maintains atrioventricular synchrony and reduces the risk of pacemaker syndrome.[14] However, VVI pacing is often employed when dual-chamber pacing is not feasible due to anatomical restrictions, device availability, or financial constraints. In this case, due to the unavailability of a dual-chamber pacemaker and economic limitations, the VVI mode was selected as the most practical option.

Differentiating primary cardiac tumors from metastatic cardiac involvement is essential in guiding appropriate treatment strategies. Primary benign cardiac tumors are typically managed with surgical resection, whereas metastatic cardiac tumors often require systemic therapies such as chemotherapy, targeted therapy, or immunotherapy, depending on the primary malignancy. Accurate diagnosis is critical, as treatment decisions depend on tumor origin, histopathological characteristics, and the extent of cardiac involvement.[5,6]

In this case, the tumor's considerable size and extensive involvement of multiple cardiac structures, including the right atrium, interatrial septum, and left atrium, presented significant challenges in both diagnosis and management. The extensive infiltration of cardiac chambers raised concerns regarding the feasibility of surgical excision and the potential risk of hemodynamic compromise.[5] Given these complexities, a multidisciplinary approach involving cardiology, cardiothoracic surgery, and oncology is paramount in determining the optimal therapeutic strategy while ensuring patient safety and improving clinical outcomes.

Conclusion

Epicardial lead placement serves as a viable alternative when transvenous pacing is not feasible, particularly in complex cases where anatomical abnormalities, such as a cardiac mass obstructing the AV conduction pathway, prevent standard lead placement. For patients with severe AV conduction disorders, including advanced or complete heart block, pacemaker implantation is critical to preventing life-threatening complications such as syncope or sudden cardiac death. Although dual-chamber pacing (DDD) is generally preferred due to its ability to maintain atrioventricular synchrony and optimize hemodynamic function, VVI mode was selected in this case due to financial constraints and the limited availability of dual-chamber pacing hardware.

The tumor's considerable size and location within the right atrium posed significant risks, making transvenous lead implantation impractical. The close association of the tumor with major venous structures increased the likelihood of procedural complications, including tumor perforation or embolization. Given these challenges, surgical epicardial pacemaker implantation emerged as the safest and most effective alternative, allowing stable pacing without the risks associated with a transvenous approach.

Additionally, differentiating primary cardiac tumors from metastatic involvement is essential for guiding an appropriate multidisciplinary treatment strategy. While benign primary tumors are often managed surgically,

metastatic involvement typically necessitates systemic therapies such as chemotherapy or targeted treatments. In complex cases such as this, a multidisciplinary team approach, involving specialists in cardiology, cardiothoracic surgery, and oncology, is crucial for optimizing patient outcomes.

Conflict of interest
The authors declare no conflicts of interest.

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Informed consent
Written informed consent for publication, including accompanying images, has been obtained from the patient.

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Catheter ablation of a right lateral accessory pathway using ibutilide: a case report

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Ibutilid
Katetrizační ablace
Wolffův–Parkinsonův–Whiteův syndrom

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Accessory pathway
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Ibutilide
Wolff–Parkinson–White syndrome

SOUHRN

Úvod: Katetrizační ablace akcesorní dráhy může být u pacientů s recidivujícími epizodami fibrilace síní náročná. Použití antiarytmik může významně interferovat s fungováním akcesorní dráhy, což znemožní mapování pomocí katétru.
Metody: V této kazuistice je popsána aplikace ibutilidu v prevenci fibrilace síní během mapování a ablace pravé laterální akcesorní dráhy pomocí endokardiálního katétru u mladého pacienta, aniž by došlo k významné interferenci s fungováním akcesorní dráhy.
Závěr: Úspěšné mapování a ablace pravé laterální akcesorní dráhy pomocí katétru s aplikací ibutilidu u mladého pacienta s epizodami fibrilace síní.

ABSTRACT

Introduction: Catheter ablation of an accessory pathway can be challenging in patients with recurrent episodes of atrial fibrillation. The use of antiarrhythmics can significantly interfere with the accessory pathway, making its catheter mapping impossible.
Methods: This case report describes the use of ibutilide to prevent atrial fibrillation during endocardial catheter mapping and ablation of a right lateral accessory pathway in a young patient without significantly interfering on accessory pathway.
Conclusion: Successful catheter mapping and ablation of a right accessory pathway in a young patient with episodes of atrial fibrillation using ibutilide.

Introduction

Radiofrequency (RF) ablation is a safe and effective therapy for the treatment of recurrent atrio-ventricular tachycardia (AVRT).[1] In these cases, RF is delivered on the annulus of tricuspid and mitral rings aiming to eliminate accessory pathways (AP), which can be in almost any site along the valvular rings. The most common causes of failed AP ablations are inability to access the target site (25%), catheter instability (23%), mapping errors because of oblique AP orientation (11%), epicardial AP (8%) and recurrent atrial fibrillation (AF) (3%).[2] The occurrence of AF and flutter can preclude mapping and require cardioversion. Antiarrhythmic drugs can alter conduction on the accessory pathway and the electrical cardioversion cannot be performed too often. In a young patient with a symptomatic ventricular preexcitation, after an unsuccessful ablation attempt, the RF was performed using small doses of ibutilide (0.1 mg to maximum of 1–2 mg). Indeed, this antiarrhythmic drug may prevent atrial arrythmias without altering significantly AP conduction and refractoriness maintaining ventricular pre-excitation (delta wave).[3,4]

Case report

An 18-year-old male patient of Ivorian origin, without structural heart disease, history of palpitations, was refe-

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rred to our hospital for an electrophysiological (EP) study. His resting surface electrocardiogram (ECG) suggested a right lateral accessory pathway with a PR interval of 87 ms and a QRS duration of 172 ms (**Fig. 1**). In November 2023, the patient signed an informed consent form for an electrophysiological study (EPS) and catheter ablation (CA). Antiarrhythmic medications, including flecainide and beta blockers, were discontinued for three days before the intervention. Electroanatomic mapping (EAM) was adopted to avoid the use of ionizing radiation by preferring the zero-X-ray procedure and to allow a more precise localization of the accessory pathway. The EAM was performed using the EnSite NavX mapping system (Abbott, St. Paul, MN, USA). After establishing femoral vascular access, a steerable decapolar catheter (Inquiry, Abbott) was positioned in the coronary sinus, while a quadripolar non-steerable catheter (Supreme, Abbott) was placed in the right ventricle. An irrigated ablation catheter was then positioned near the tricuspid annulus. During catheter placement, orthodromic atrioventricular reentrant tachycardia was observed, with a tachycardia cycle length of 380 ms (**Fig. 2**). During mapping, the earliest ventricular activity was recorded at the lateral tricuspid annulus. However, several sustained episodes of atrial fibrillation (AF) occurred, necessitating electrical cardioversion (EC). The electroanatomic mapping of the tricuspid annulus was difficult due to the numerous AF episodes which required intravenous flecainide. After resolution of the arrhythmic episodes, the ventricular preexcitation disappeared, making it impossible to continue mapping the accessory pathway, leading to the decision to abort the procedure.

In June 2024, a second attempt was made. As before, during mapping of the tricuspid annulus, several AF episodes recurred. Intravenous ibutilide was administered (up to 1 mg), which converted AF to atrial flutter, and eventually sinus rhythm. This status allowed us to induce and maintain the atrioventricular reentrant tachycardia (AVRT), allowing us to map the tricuspid annulus and identification of the area of greatest V-A fusion near the lateral wall. Due to poor contact and stability of the ablation catheter, we utilized a deflectable introducer (Agilis, Abbott). In this region, radiofrequency (RF) energy was applied (35 W, 43 °C) using a 3.5-mm irrigated tip ablation catheter (Tacticath Contact Force, Abbott), resulting in permanent loss of preexcitation without any complications (**Fig. 3**). The procedure was terminated after 30 minutes, with no recovery of accessory pathway (AP) conduction observed during testing with atrial and ventricular pacing, as well as adenosine infusion.

Discussion

During electrophysiological studies to locate the accessory pathway, episodes of AF or AFL may occur, potentially prolonging the procedure or hindering accurate mapping if these arrhythmias are frequent or prolonged. These arrhythmias can be managed with electrical or pharmacological cardioversion. Electrical cardioversi-

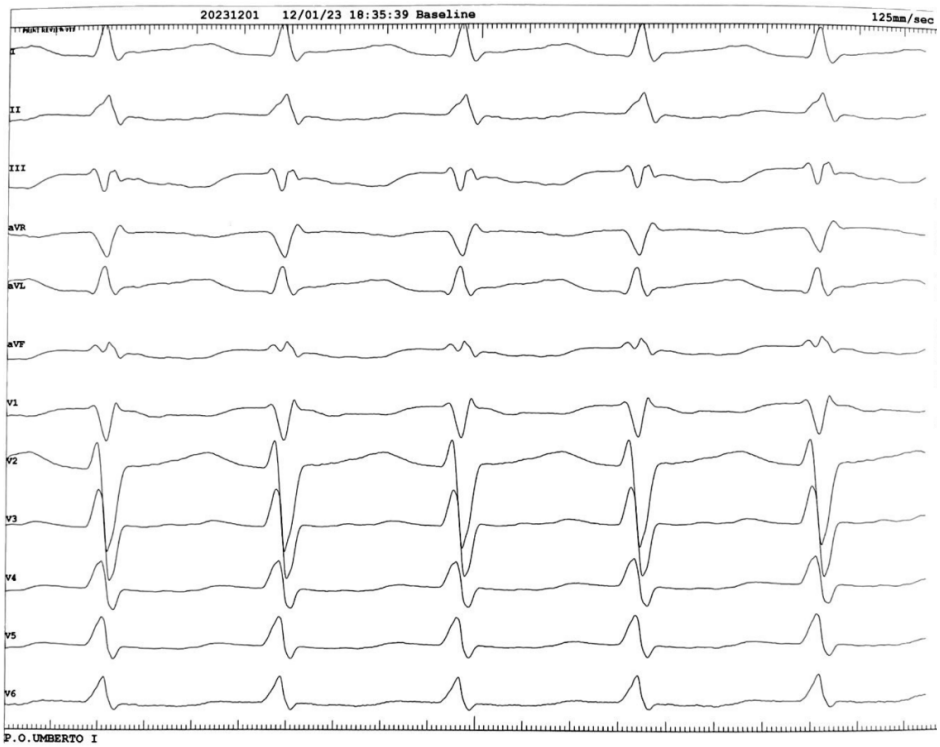

Fig. 1 – Surface EKG showed a sinus rhythm with a short PR interval, evidence of full ventricular preexcitation with left bundle branch block (LBBB) morphology suggestive of a right lateral accessory pathway.

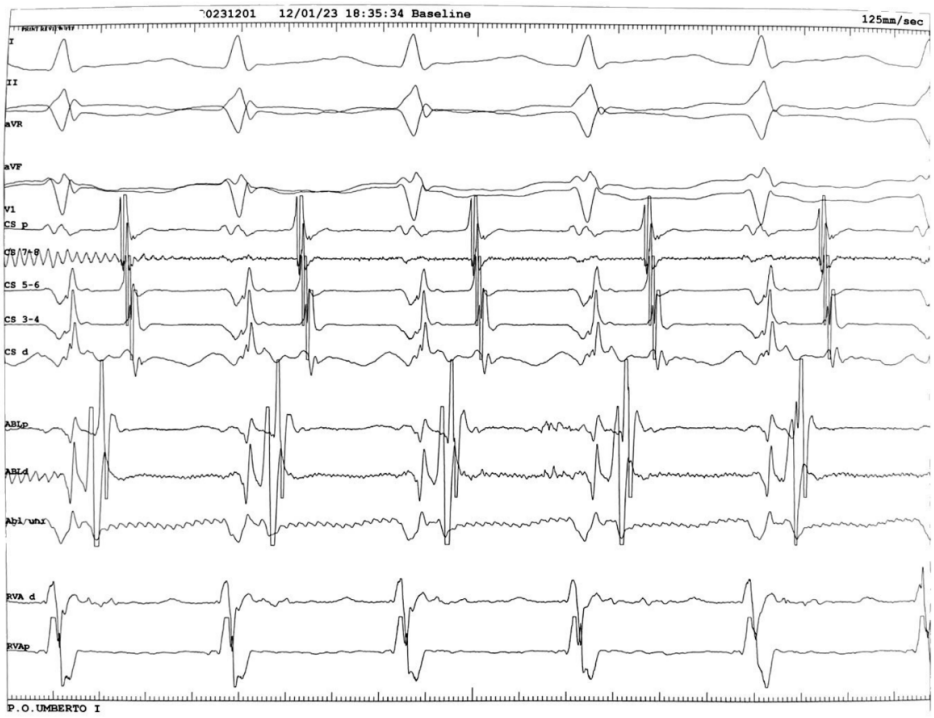

Fig. 2 – Atrioventricular reentrant tachycardia (AVRT) and then the continuation of the mapping of the tricuspid annulus, identifying the area of the greatest V-A fusion near the lateral wall.

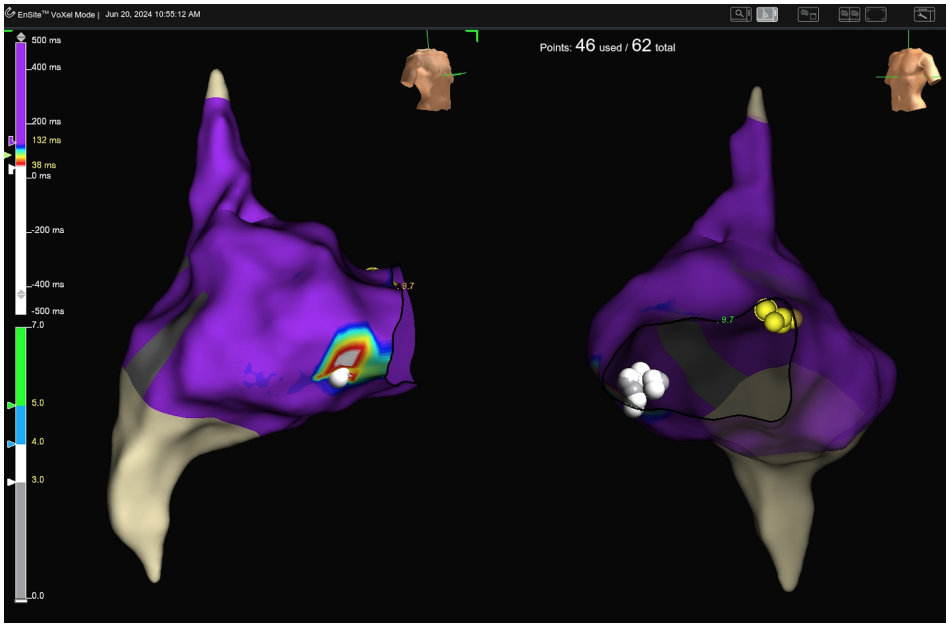

Fig. 3 – In lateral tricuspid annulus RF energy was applied (35 W, 43 °C) with a 3.5-mm irrigated tip ablation catheter (Tacticath Contact Force, Abbott) led to permanent loss of pre-excitation.

on is preferred for hemodynamically unstable patients, while pharmacological cardioversion can be considered for stable patients, though it is generally less effective. In patients with AF and an accessory pathway, drugs like procainamide, propafenone, or flecainide may be used, but they can interfere with accessory pathway conduction, causing loss of the pre-excitation (delta wave). Ibutilide, however, is recommended by the 2019 ESC Guidelines for supraventricular tachycardia, as it resolves arrhythmias without affecting accessory pathway conduction, preventing hypotension and allowing the procedure to proceed.[6] In our case, flecainide in the first attempt led to loss of conduction along the accessory pathway, halting the procedure. In the second attempt, considering the results of Glatter et al.,[3] ibutilide successfully resolved AF/AFL episodes, preserved conduc-

tion along the accessory pathway, and allowed accurate mapping and ablation of the pathway.

Conclusion

We presented the case of a young patient with a right-sided accessory pathway that was nearly impossible to map due to frequent episodes of atrial fibrillation. After the infusion of ibutilide, which helped maintain sinus rhythm without significantly affecting the conduction and refractoriness of the accessory pathway, the procedure became more manageable. This allowed for successful mapping and effective ablation of the pathway. One month later, the patient has not experienced any arrhythmic recurrences.

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MD: Data curation, conceptualization, writing – original draft.
MLP: Data curation, methodology, conceptualization.
RB: Data curation, methodology, conceptualization.
GL: Writing – original draft.
MB: Writing – review & editing.
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Double-vessel disease of acute myocardial infarction in a 27-year-old young female: a case report

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Postižení dvou tepen
Žena

SOUHRN

S akutním infarktem myokardu (AIM) se u mladých lidí nesetkáváme **často**. Kromě toho se vzhledem k obtížnému rozpoznávání symptomů, méně přesně definované „atypické" bolesti na hrudi i vzhledem k některým normálním výsledkům angiografie uvádějí mnohem nižší počty **žen** s AIM. Studie provedené v nedávné době však upozornily na zvýšenou incidenci AIM s postižením jedné tepny jako nejčastějším angiografickým nálezem u mladších pacientů. Akutnímu infarktu myokardu u mladých **žen** se dosud věnovala menší pozornost, zvláště při postižení dvou tepen. Cílem této kazuistiky je rozšířit současné poznatky pomocí detailní analýzy případu mladé **ženy**, která prodělala AIM a u níž byla stanovena diagnóza postižení dvou tepen.
Sedmadvacetiletá Indonésanka byla převezena do naší nemocnice se synkopou, jíž předcházela akutní bolest na hrudi. Elektrokardiografický záznam prokázal elevaci segmentu ST v předních svodech (V_3–V_6) i zadních svodech. Podle vyšetření krevních vzorků pacientka trpěla hypertriglyceridemií (262 mg/dl) a měla abnormální hodnotu glukózy v plazmě (516 mg/dl), což ukazovalo na možnou přítomnost diabetes mellitus 2. typu. Výsledky koronarografického vyšetření ukázaly uzávěry dvou tepen, 99% ve střední **části** r. circumflexus (RC) a 70% v proximální – střední **části** r. interventricularis anterior (RIA). Na RC jako nejkritičtějším místě uzávěru byla provedena perkutánní koronární intervence (PCI). Přítomnost diabetu 2. typu představovala u pacientky faktor zvyšující riziko.
Pro poznání příčiny rozvoje AIM v nízkém věku je důležité odebrat rodinnou i osobní anamnézu.

Keywords:
Acute myocardial infarction
Double-vessel disease
Female
Diabetes
Obstructive CAD

ABSTRACT

Acute myocardial infarction (AMI) is an uncommon occurrence in young people. Furthermore, females suffering from AMI reported much lower rates due to poor symptom recognition, less defined "atypical" chest pain, and some normal angiographic findings. However, recent studies have highlighted an increase in the incidence of AMI in younger patients with single-vessel disease, which is the most prevalent angiographic feature. To date, AMI in young females has received less study, especially in cases of double-vessel disease. The present case study aims to contribute to the existing body of knowledge by offering a detailed analysis of a young female patient who has experienced an AMI and has been diagnosed with double-vessel disease. A 27-year-old Indonesian female was transferred to our hospital with a syncope presentation, preceded by acute chest pain. An electrocardiogram shows ST elevation in anterior leads (V_3–V_6) and inferior leads. From lab examination, the patient has hypertriglyceridemia (262 mg/dL) and abnormal plasma glucose (516 mg/dL), with the possibility of type 2 diabetes mellitus. Coronary angiogram results revealed double-vessel occlusions at the middle right circumflex artery (RCA) at 99% and the proximal-middle left anterior descending coronary artery (LAD) at 70%. Percutaneous coronary intervention (PCI) was conducted at RCA as the most critical occlusion site. Type 2 diabetes was the contributing risk factor for the patient. History taking of family and personal diseases is important to comprehend the cause of AMI occurrence at an early age.

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Introduction

Acute myocardial infarction (AMI) is the predominant cause of cardiovascular events globally, with an increasing incidence in patients under 35 years of age.[1,2] Although infrequent, several studies indicate that the prevalence of these AMIs among the young ranges from 2% to 10% of overall AMI cases.[3,4] The age of onset is progressively decreasing due to various risk factors, including gender, smoking habits, obesity, prior coronary artery disease, familial history of premature acute myocardial infarction (AMI), non-obstructive coronary artery disease (CAD), genetic disorders associated with blood coagulation or fibrinogen, hypertension, diabetes, elevated low-density lipoprotein cholesterol (LDL-C) levels, and increased uric acid levels.[2,5,6]

Gender considerations significantly influence the prevalence of acute myocardial infarction across all age groups. Female experiencing acute myocardial infarction reported much lower rates.[7] Males have a higher incidence, accounting for approximately 70% of all AMI cases and having 7 to 10 years earlier onset than females. Similar to the older age, young AMI patients aged 18–35 were characterized as males with previously mentioned risk factors.[8] However, females experience a greater one-year mortality rate (OR 1.6).[9] This mortality burden in women may be due to a lack of symptom recognition by clinicians and patients. The "atypical" chest pain described by women is less likely to be defined than in men, reducing timely and evidence-based interventions for AMI symptoms in women.[7] Females with AMI also tend to present to the hospital later after symptom onset than males, which may indicate a possible lack of dangers or acute myocardial symptomology knowledge in the general population.[10]

Furthermore, recent reports have indicated that 20.4% of female patients with AMI exhibited angiographically normal coronary arteries.[11] These findings suggest that female patients may not be experiencing an acute coronary syndrome (ACS) and could be presenting with symptoms due to other underlying conditions. These findings suggest that the prevalence of acute coronary syndrome (ACS) in female patients may be underestimated, and that other underlying conditions may be responsible for the observed cases of female acute myocardial infarction.[6] However, the same study also reported that 45.5% of female AMI cases had obstructive coronary artery disease, with single-vessel disease being the most common in the young group. Due to the chronicity of AMI, double-vessel disease (DVD) or multiple-vessel disease was rarely seen in young patients.

To date, the prevalence of acute myocardial infarction (AMI) in young females has received comparatively less attention, especially in the context of dual-vessel disease (DVD). Consequently, this paper presents a case report of AMI in a young female with DVD, accompanied by a concise literature review.

Case report

A 27-year-old female was admitted to a referring hospital following an incident of syncope. The aforementioned symptoms were characterised by cold sweat and palpitation, and were preceded by acute chest pain. The patient regained consciousness within minutes of admission. The intensity of the chest pain was measured using a visual analogue scale, with a rating of 10/10, indicating severe discomfort. The patient described the sensations as pressure, tightness, heaviness, or a burning feeling. The patient reported that these symptoms were new and previously unobserved. The patient was subsequently transferred to our hospital due to the persistent mild chest pain and the ECG findings suggestive of acute myocardial infarction. A detailed history revealed that the patient does not consume tobacco or alcohol. The patient reports no previous history of similar symptoms or cardiovascular diseases. The patient is employed as a treasury employee at a small convenience store. No family history of similar symptoms or cardiovascular diseases were reported.

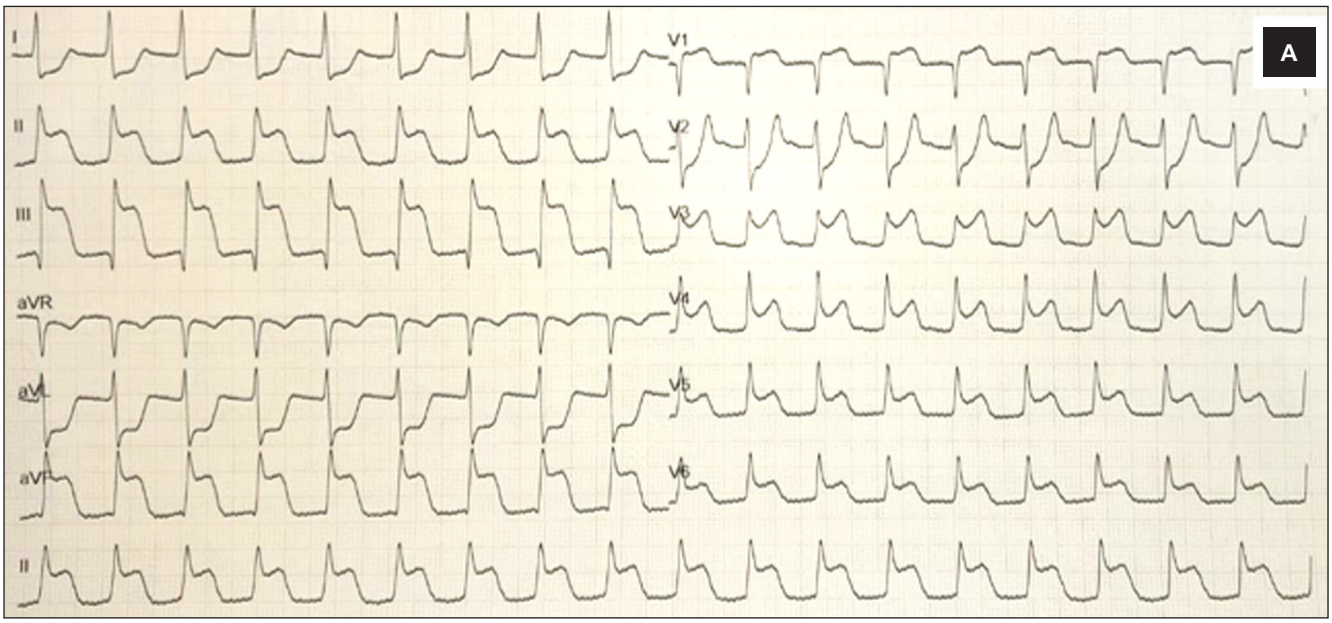
Despite the pain, her vital signs were normal, with a blood pressure of 90/63 mm Hg, a heart rate of 75 regular beats per minute, a body temperature of 36.5 °C, an oxygen saturation of 98% with 3 lpm nasal cannula support, and a respiratory rate of 20/min. She is fully conscious with a score of 15 on the Glasgow Coma Scale (E4V5M6). She still has mild chest pain with a visual analogue scale score of 6/10.There is nothing else wrong when we examine her body, including her skin and nervous system. The patient weighs 41 kg and is 145 cm tall, with a body surface area of 1.29 m³ and a BMI of 19.5 kg/m^2.

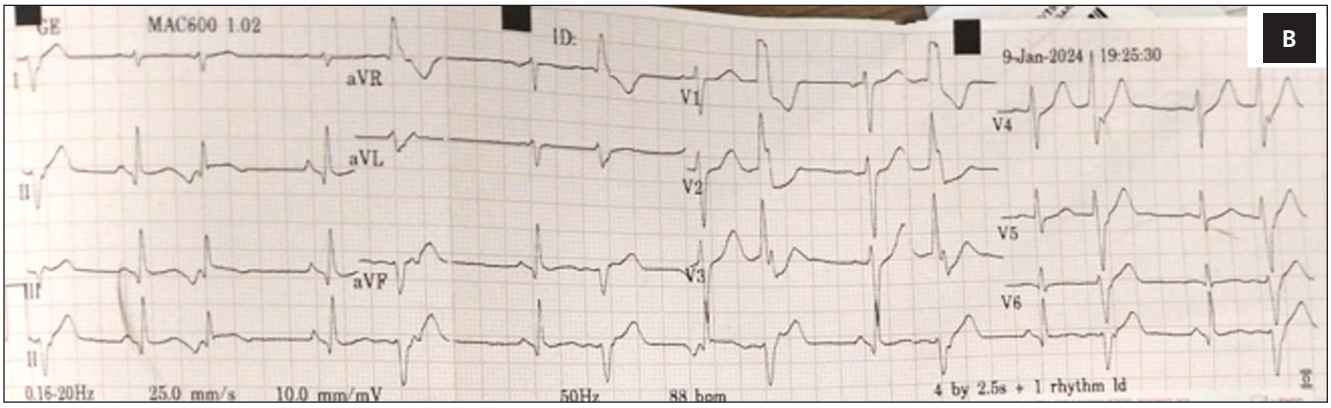
A normal blood test taken one day after the chest pain started in our hospital showed higher levels of certain cells and chemicals involved in fighting infection, as well as higher levels of glucose (sugar), triglycerides,

Table 1 – Laboratory results

Parameter (unit)	Measured value	Normal value
Haemoglobin (g/dL)	11.8	12.1–15.1 (female)
White blood cells (× 10^3/µL)	16.05*	3.9–9.7
Platelets (× 10^3/µL)	333	150–450
Aspartate transaminase (AST/SGOT) (IU/L)	222*	5–37
Alanine aminotransferase (ALT/SGPT) (IU/L)	180*	6–43
Albumin (g/dL)	3.65**	3.8–5.2
Blood urea nitrogen (mg/dL)	8.9	8–21
Creatinine (mg/dL)	0.7	0.6–1
Plasma glucose (mg/dL)	516*	65–109
Low-density lipoprotein cholesterol (mg/dL)	98	70–139
Triglyceride (mg/dL)	262*	30–149
High-sensitivity troponin T (ng/L)	>14*	0–14
Sodium (Na) (mEq/L)	137	135–145
Potassium (K) (mEq/L)	3.3	3.5–5
Chloride (Cl) (mEq/L)	109	96 - 107
Activated partial thromboplastin time (APTT) (s)	27.1	23–36
Partial thromboplastin time (PTT) (s)	11.2	25– 35

* Higher than normal value, ** lower than normal value.





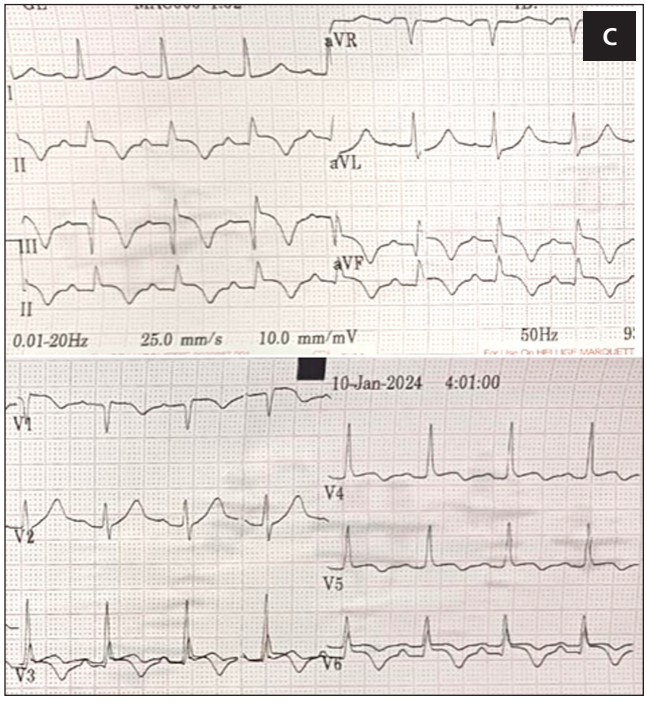


Fig. 1. ECG result. (A) A junctional rhythm at 110 regular beats/min with ST elevation in anterior leads and inferior leads from ECG result taken from referred hospital. (B) A serial ECG of PVC bigeminy features to pathological Q (C) accompanied by T inversion in similar lead taken from transferred hospital.

and a protein involved in heart damage. Albumin levels, which are often used as a marker of overall health, were lower. The results of other blood tests (see **Table 1**) were normal. The plasma glucose was exceptionally high (516 mg/dL), suggesting the patient has type 2 diabetes mellitus. An ECG result taken at a different hospital (four hours after the start of symptoms) showed a junctional rhythm at 110 regular beats per minute with ST elevation in the V_3–V_6 and II, III, and aVF leads. A second ECG at our hospital showed signs of a more serious problem, with the ST-T segment showing an upward shift. This was from PVC bigeminy features (a type of irregular heartbeat) to pathological Q (a type of arrhythmia) with T inversion in II, III, aVF, and V_3–V_6 (see **Fig. 1**). A chest X-ray result was normal, with a heart that takes up 47% of the chest. Transthoracic echocardiography shows the left ventricle is dilated with LVH (LVDMi 111 g/m^2; RWT 0.307). A further analysis showed that the left ventricle was not working

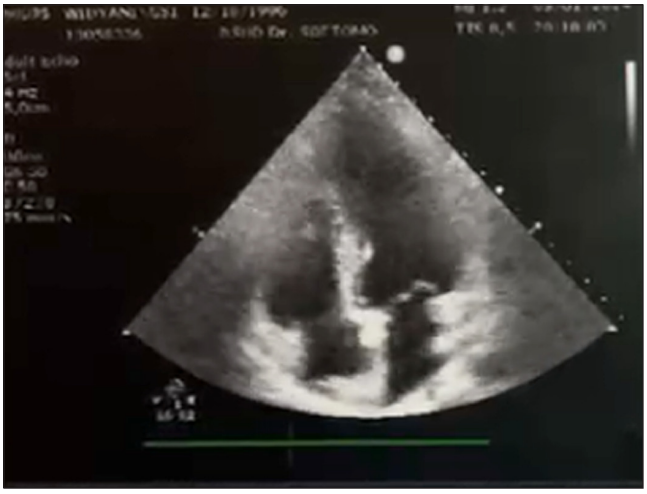


Fig. 2 –Transthoracic echocardiography result. A dilatated and hypokinetic (inferoseptal [B-M], inferior [B-M-A]) left ventricular dimensions with eccentric LVH.

well (inferoseptal [B-M], inferior [B-M-A]) but the other parts were working normally. The left ventricle was able to pump blood at 48.1% (by TEICH), which is normal. This shows that the left ventricle is working well during diastasis (**Fig. 2**).

A coronary angiogram (CAG) was conducted, using the right femoral artery approach with a 6 French sheath, and it revealed occlusions at the middle right circumflex artery (RCA) of 99% and proximal-middle left anterior descending coronary artery (LAD) of 70%. No abnormal findings, such as coronary artery aneurysm, were confirmed through the CAG. We establish the diagnosis of STEMI in the anterior (70% occlusion in LAD) and inferior (99% critical occlusion in RCA) with type 2 diabetes mellitus.

Percutaneous coronary intervention (PCI) was conducted at the middle right circumflex artery to expand the stenosis through plain old balloon angioplasty. The procedure installs a third-generation drug-eluting stent (the Ultimaster 3.5 mm diameter/24 mm length sirolimus eluting coronary stent) (**Fig. 3B**). The duration from arrival

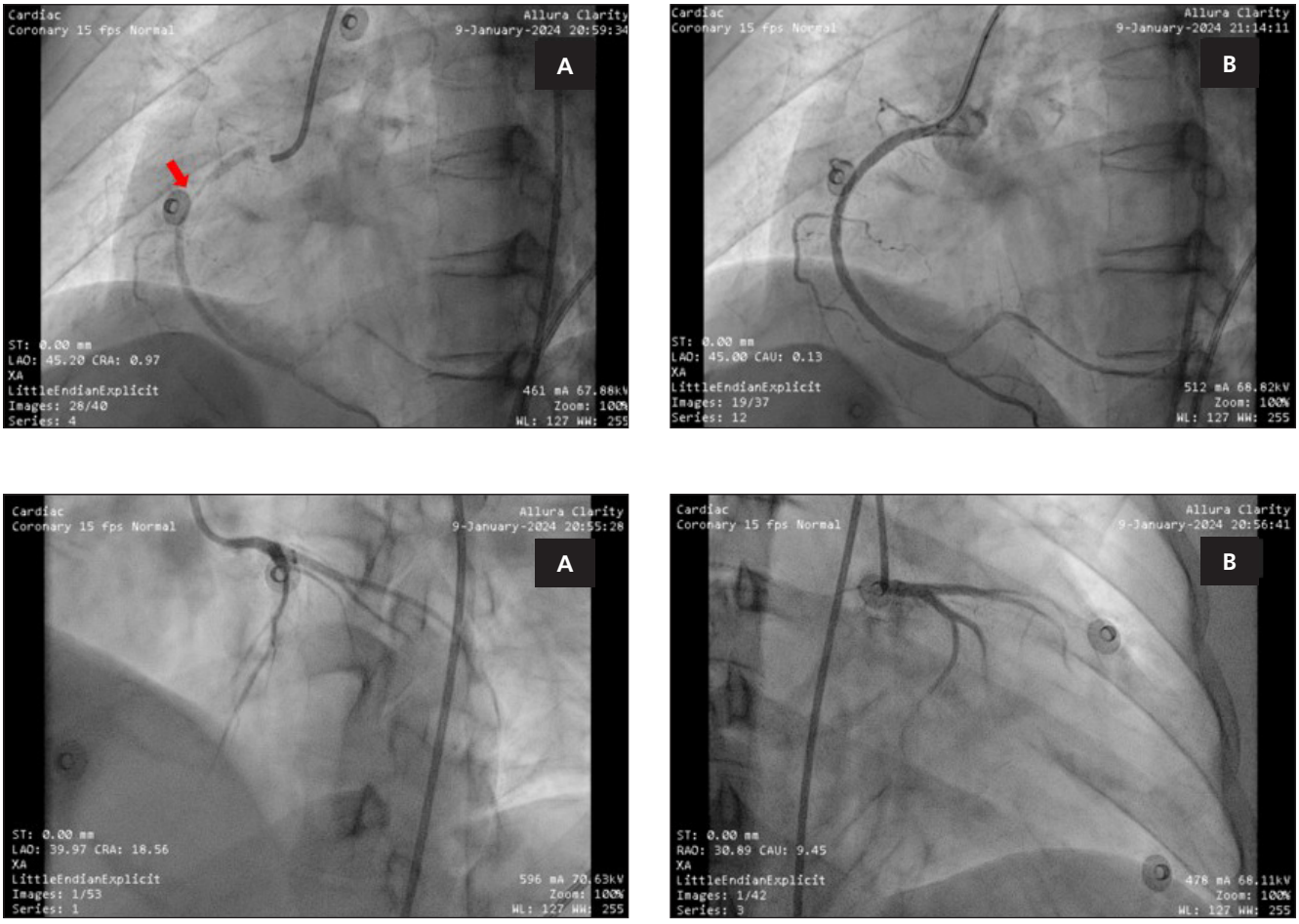


Fig. 3 – (A) An occlusion at mid RCA. Percutaneous coronary intervention (PCI) at mid RCA to expand the stenosis up to 0%. (B) Plain old balloon angioplasty procedure installed the third-generation drug eluting stent (the Ultimaster 3.5 mm diameter/24 mm length sirolimus eluting coronary stent). (C) The occlusion at prox-mid LAD was then scheduled for revascularization within 45 days after the first PCI. (D) The LCx was normal.

at our ED to coronary artery recanalization, or "door to balloon time", was 2 h, and the duration from the onset of symptoms to coronary artery recanalization was 36 h and 50 min. The patient was stable after the procedure and started a routine of 100 mg of aspirin, 3.75 mg of prasugrel, 2.5 mg of bisoprolol fumarate, and 40 mg of atorvastatin 1 day after PCI. The patient also went to an endocrinologist for type 2 diabetes mellitus. And the patient was scheduled for revascularization of the prox-mid LAD within 45 days after the first PCI.

Discussion

We present the ST-elevation acute myocardial infarction cases in a young female with syncope presentation. Based on a large prospective study of AMI in young patients in India (n = 1116; maximum age of 30), 4.9% of cases were female and 2.3% reported syncope as a clinical presentation.[3] A number of risk factors has been identified as being of particular importance in cases of acute myocardial infarction (AMI) in young females. Previous case reports have indicated that a combination of six risk factors is often present: obesity, hypertension, renal dysfunction, hyperuricemia, hypercholesterolemia, high LDL-C level, hypertriglyceridemia, and abnormal glucose tolerance, in addition to a family history of hypertension.[2] Of all the risk factors under consideration, familial hypercholesterolemia (FH) has been observed to be associated with an earlier age of onset for acute myocardial infarction (AMI).[12] Individuals diagnosed with familial hypercholesterolemia (FH) exhibit a twofold elevated risk of cardiovascular incidents in comparison to those not afflicted with this condition. The second most common and modifiable risk factor of acute myocardial infarction (AMI) in young patients is smoking.[2,13] As indicated in other case reports of a young female patient in Indonesia, smoking was identified as a primary factor, in conjunction with a clinical history of type 2 diabetes and peripartum cardiomyopathy (PPCM).[13] In this case report the patient has no history of FH nor diabetes, however, the patient has hypertriglyceridemia (262 mg/dL) and abnormal plasma glucose (516 mg/dL), with the possibility of type 2 diabetes mellitus.

The present literature suggests an increased risk of diabetes and hypertension in young adult females.[14] In the case of CAD females, the age at which symptoms manifest is advanced relative to other demographics. Furthermore, a greater number of risk factors, including but not limited to type 2 diabetes mellitus, are present in this demographic.[7,15] Patients diagnosed with type 2 diabetes have an increased likelihood of experiencing silent or unrecognised myocardial infarction (MI), irrespective of their sex. The presence of high fasting glucose levels, prediabetes, and type 2 diabetes has been demonstrated to enhance the probability of unrecognised MI.[7,16] In addition to the previously mentioned poor recognition and less defined "atypical" in female chest pain,[10] the challenges posed by AMI in young females in daily practice are attributable to a paucity of conducted evidence-based studies.

A coronary angiogram (CAG) showed a double-vessel occlusion at the middle right circumflex artery (RCA) and middle left anterior descending coronary artery (LAD). Obstructive CAD presents the most common angiographic characteristic of AMI in young patients (80.6%), however, single-vessel occlusion at LAD was the most frequent condition reported (58.1%).[3] Double vessel disease (DVD) presents a later chronic stage of CAD, which is rarely seen in young female patients. A previous study on angiographic characteristics in female patients shows a greater prevalence of DVD and triple-vessel disease (TVD) in the elderly group.[11] Previously mentioned large prospective cohort study in India stated only 6.4% of young AMI cases were double vessel, specifically in LAD and RCA as the most common location.[3] We failed to gain the history of previous chest pain symptoms, however, based on these findings, there is a possibility that the patient had experienced similar symptoms before. Multiple-vessel disease increases the risk of a fall in systemic blood pressure (BP) resulting in a decrease in global cerebral blood flow, which is in line with this patient syncope presentation.[17] Diabetes, obesity, and dyslipidemia were the risk factors mentioned for DVD.[11]

Percutaneous coronary intervention (PCI) was conducted at RCA as the most critical occlusion site (99%). PCI is still the most warranted treatment for obstructive CAD with multi-vessel occlusion, as it decreases the readmission risk in AMI.[7] The likelihood of readmission in young patients was higher for women and for patients who had a prior AMI, increased depressive symptoms, longer inpatient stay, or diabetes. Several readmission predictors were reported to be psychosocial characteristics, rather than AMI severity markers.[15] percutaneous coronary intervention for the other significant lesion, in this case at proximal-mid LAD, can be done either during the index PCI or later within 45 days to achieve complete revascularization.[17] Patients with a young age coronary incident have a strikingly high long-term prognosis of repeat occurrences in later life. Approximately 65% of the 165 young patients with ACS who underwent PCI at a young age and had a long-term mean follow-up of 9.1±4.6 years experienced at least one endpoint of mortality or significant CV event, according to recent studies.[18] One of the most important factors in preventing the young patient's subsequent CV event is early intervention, which focuses on treating hypertension, changing lifestyle habits, and receiving the best medical care possible.

As a study limitation, this case report constitutes a solitary instance of a young female patient with an uncertain clinical and family history. Consequently, the actual situation and nature of the disease may deviate from the outcomes of the literature review, owing to the presence of reporting bias. Moreover, a biopsy on the occlusion was not conducted in this particular case report.

Conclusion

We present the ST-elevation acute myocardial infarction cases with double-vessel disease in a young female. Type 2 diabetes was identified as a contributing risk factor for the patient. A comprehensive medical history, encompassing both familial and personal diseases, is imperative in order to elucidate the underlying causes of AMI in early-aged individuals.

Conflict of interest
The authors declare no competing interests.

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The authors declare that no financial resources were obtained during the preparation of this manuscript.

Ethical statement and consent to participate
The subjects provided proper informed consent to participate in the study.

Consent for publication
The patient has given informed consent for the case details and any associated medical images to be published in this case report.

Data availability
No datasets were generated or analyzed during the current study.

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The RAC, bleb, and crossed aorta signs: retroaortic anomalous coronary artery visualization by transthoracic echocardiography

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SOUHRN

Transtorakální echokardiografie představuje užitečný nástroj pro screening vrozených anomálií koronárních tepen. Jednoznačně vyvrátila názor, že anatomii koronárních tepen nelze zkoumat neinvazivně. Uznávají se tři echokardiografické znaky související s anomáliemi levé koronární tepny. Tyto znaky jsou: znak retroaortálního r. circumflexus (retroaortic circumflex, RAC), znak bublinky (bleb sign) a znak překřížené aorty (crossed aorta sign). Znak RAC je vysoce specifický a na popisu echokardiogramu jednoznačně ukazuje na anomální koronární tepnu. Je však naprosto nezbytné ho odlišit od podobně vypadajících struktur. Proto se doporučuje jako primární metody pro ověření známých anomálií koronárních tepen nebo podezření na ně použít koronární CT angiografii.

ABSTRACT

Transthoracic echocardiography is a useful screening tool for identifying congenital coronary artery (CA) anomalies. It has effectively dispelled the notion that CA anatomy cannot be identified noninvasively. There are three recognized echocardiographic signs associated with anomalous left coronary artery. These signs are the retroaortic circumflex (RAC) sign, the blew sign, and the crossed aorta sign. The RAC sign is highly specific and should strongly indicate an anomalous coronary artery on echocardiogram reports. However, it is crucial to distinguish it from similar-looking structures. Therefore, coronary computed tomography angiography is recommended as the primary method for assessing known or suspected coronary anomalies.

Introduction

Identification of congenital coronary artery (CA) anomalies using transthoracic echocardiography (TTE) as a screening tool has become routine and has helped dissipate the notion that CA anatomy cannot be identified noninvasively. In the last years, coronary computed tomography angiography (CCTA) has replaced cardiac catheterization as the primary tool for the characterization of most congenital CA anomalies.[1,2]

TTE is a highly recommended modality in clinical practice due to its versatility, safety, cost-effectiveness, and non-invasive nature. It can also be performed easily in any clinical setting, making it a widely available option for many patients.[3,4] Three signs are recognized echocardiographic features well linked with anomalous left coronary artery, specifically associated with left circumflex (Lcx) artery abnormalities. These signs are: 1) The retro-aortic course of the Lcx (RAC) sign, described as a "tubular shape"[3] in TTE, 2) the "bleb sign" described by Wierzbowska–Drabik et

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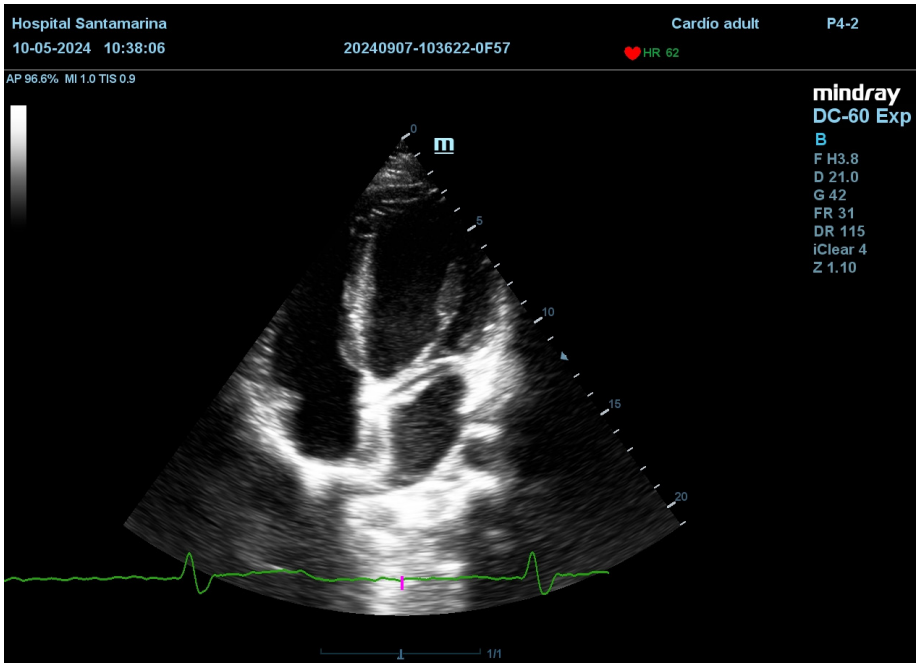

Fig. 1 – Apical four chambers view showed the retroaortic circumflex (RAC) sign.

al. by transesophageal echocardiography (TEE) studies, as a round structure in the mitro-aortic angle in the aortic long-axis view,[5] and 3) the 'crossed aorta sign', a binary structure that appears to cross the aorta perpendicular to its longitudinal axis in an apical five-chamber view.[6,7]

Case presentation

An asymptomatic 16-year-old male underwent a routine cardiac evaluation. RAC sign was incidentally detected by TTE. In the apical 4 chambers (A4C) view, tilting anterior demonstrated the RAC sign, a highly echogenic tubular structure in the retro-aortic region above the mitral valve (**Fig. 1**, and **Video 1**). Additionally, in the parasternal long-axis view, a round structure next to the aorto-mitral curtain, known as the bleb sign, was also identified (**Video 2**). The RAC sign was also evident in the short-axis (SAX) as a tunnel-shaped structure located behind the aortic root. This structure is also an orthogonal view of the "bleb sign". An anomalous LCx was confirmed with CCTA (**Figs 2**, and **3**).

Discussion

Anomalous origin and course of the LCx from the right sinus of Valsalva is the most frequent anomaly (0.39%).[8] Traditionally, coronary angiography has been required for diagnosis; however, these tests are expensive and invasive. Currently, CCTA is considered superior to conventional angiography in diagnosing and classifying coronary anomalies[9,10] since it provides useful information regarding the anatomy and course of anomalous coronary arteries.

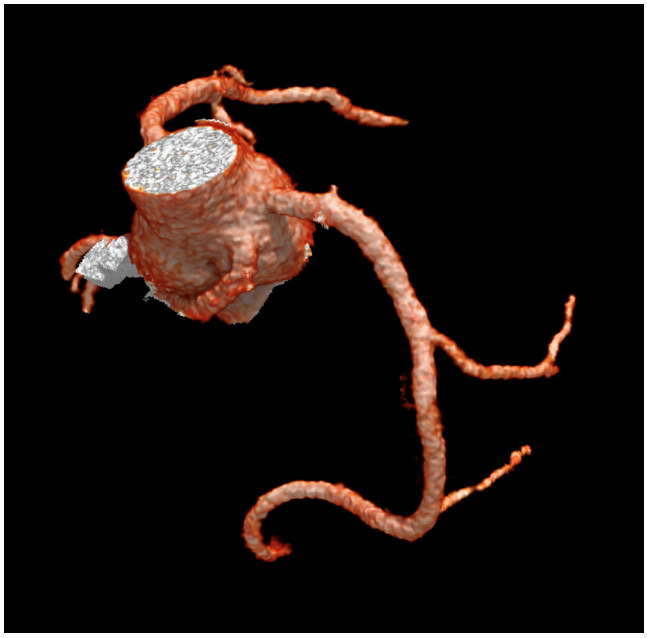

Fig. 2 – Coronary computed tomography angiography (CCTA). Three-dimensional reconstruction showing the origin of the coronary arteries.

In 2018 Witt et al. reported that RAC sign was seen in 63.3% of patients with confirmed coronary anomalies and in 6.1% of patients with normal coronary anatomy.[3] They also reported that the RAC sign had a sensitivity of 63.3% and a specificity of 93.9%. Additionally, these authors demonstrated a significant association between the RAC sign and the presence of a retro-aortic anomalous coronary artery (p <0.001).[3] When RAC and bleb

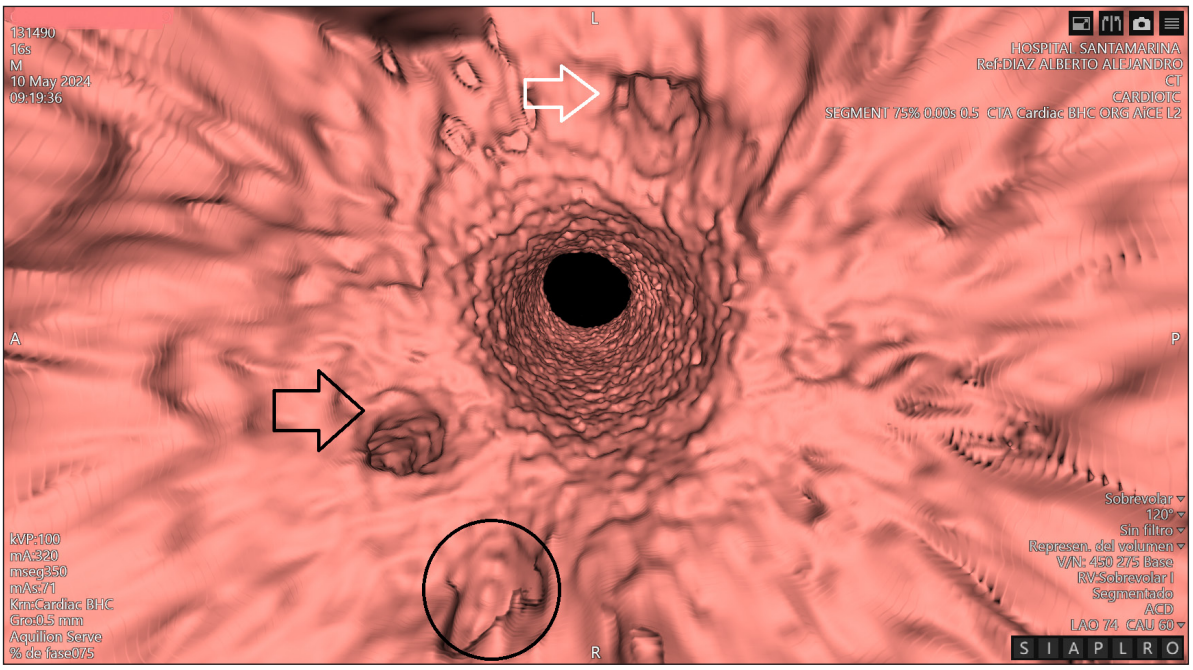

Fig. 3 – Virtual coronary endoscopy created by computed tomography angiography. Visualization of right coronary ostium (white arrow) left coronary ostium (black arrow), and independent circumflex ostium (black circle).

signs are present in an echocardiographic routine study, it's important to consider other more common findings as possible differential diagnoses. In the A4C view, the RAC sign may be mistaken for the coronary sinus, which is typically located further back. Other possible causes of confusion include catheters, coronary calcifications, and calcifications of the aortic valve or mitral annulus. In addition, the bleb sign can be differentiated from an abscess of the mitral-aortic fibrosa by observing the retro-aortic tubular appearance of the structure in the SAX view.[3,4,6,7] The use of CCTA is a reliable noninvasive tool for defining anomalous coronary arteries and provides detailed three-dimensional anatomic information that may be difficult to obtain with invasive coronary angiography.[9]

Conclusion

The RAC, bleb, and crossed aorta signs offer a noninvasive means of identifying an anomalous LCx. The RAC sign also has a high specificity, signifying that its presence could be documented as strongly suggestive of an anomalous coronary artery on echocardiogram reports. This case emphasizes the significance of maintaining a high level of suspicion of coronary anomalies when RAC and/or bleb signs are present in routine echocardiographic studies. The usefulness of these signs should not be overlooked or misinterpreted, as being aware of their significance can prevent confusion. CCTA has been recommended as the first-line method for the assessment of known or suspected coronary anomalies.

Conflict of interest
The authors declare that they have no conflicts of interest.

Funding
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Ethical statement
All ethical matters have been fully considered to protect the rights of the patient. This case report was exempted from approval by the Institutional Ethics Committee.

Informed consent
Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

Credit authorship contribution statement
Diaz Alejandro: conceptualization, writing original draft, review & editing. Victoria Vannoni and Sofia Inclán: conceptualization, writing original draft.
Jose Guillen and Daiana Cattaneo: methodology, writing, review & editing.

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Rare Cause of Acute Coronary Syndrome in a Young Female Patient: Coronary Embolism

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Perkutánní balonková angioplastika

SOUHRN

Kontext: Koronární embolizace u pacientů s mechanickou chlopní jako příčina uzávěru koronární tepny je považována za vzácnou příhodu s neznámou incidencí. Kromě toho chybí shoda na účinné léčbě a způsobu odstranění koronárních embolů v dané situaci.
Přehled případu: Šestadvacetiletá žena se dostavila k lékaři s bolestí na hrudi trvající jednu hodinu v důsledku non-ST akutního koronárního syndromu vyvolaného uvolněním embolu z mechanické mitrální chlopně v důsledku nedostatečné adherence k antikoagulační léčbě. U pacientky byla do 60 minut od příjezdu na oddělení akutního příjmu provedena srdeční katetrizace. Angiografické vyšetření prokázalo totální uzávěr středního segmentu první levé marginální větve; po provedení perkutánní angioplastiky této větve došlo ke zlepšení koronární perfuze, okamžitému úplnému ústupu bolesti na hrudi a k ústupu změn úseku ST a vlny T na EKG záznamu. Další den bylo provedeno vyšetření transezofageální echokardiografií, které prokázalo dobře fungující mechanickou mitrální chlopeň s dvěma útvary nasedajícími na atriální stranu mechanické mitrální chlopně. U pacientky se pokračovalo s terapeutickou antikoagulací do dosažení cílové hodnoty mezinárodního normalizačního poměru (INR). Následné transezofageální echokardiografické vyšetření prokázalo zmenšení útvarů.
Diskuse: Tento případ prokázal, že koronární embolizaci může zkomplikovat mechanický trombus se suboptimálními hodnotami INR a lze jej úspěšně léčit perkutánní angioplastikou.

Keywords:
Coronary angiography
Mechanical mitral prosthesis
Percutaneous balloon angioplasty

ABSTRACT

Background: Coronary embolization in patients with mechanical prosthesis as a cause of coronary artery occlusion has been considered a rare condition with unknown incidence. Moreover, there is a lack of consensus for effective treatment and management of coronary emboli in this setting.
Case summary: A 26-year-old female patient presented with chest pain of one hour duration due to non-ST acute coronary syndrome caused by embolization from mitral mechanical prosthesis due to inadequate adherence to anticoagulant therapy. The patient underwent cardiac catheterization within 60 minutes of arrival to emergency room. Angiography revealed total occlusion of the mid segment of first obtuse marginal branch. Percutaneous angioplasty was done to the obtuse marginal branch with improvement of coronary perfusion, immediate complete resolution of chest pain and regression of ST–T wave changes in electrocardiogram. Transesophageal echocardiography was done one day later and revealed well-functioning mitral mechanical prosthesis with two masses seen attached to atrial aspect of mitral mechanical prosthesis. Patient was kept on therapeutic anticoagulation and achieved target international normalized ratio (INR). Follow-up transesophageal echocardiography showed regression of the size of masses.
Discussion: This case demonstrated coronary embolization may complicate mechanical thrombus with suboptimal INR levels and can be successfully treated with percutaneous angioplasty.

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Introduction

Coronary embolization in patients with mechanical prosthesis as a cause of coronary artery occlusion has been considered a rare condition with unknown incidence. Moreover, there is lack of consensus for effective treatment and management of coronary emboli in this setting. The purpose of this report is to show the successful management of case of coronary embolism with primary angioplasty using balloon without stenting.

Case report

A 26-year-old female with history mitral mechanical prosthesis was admitted to our hospital on May 7, 2022 complaining of sudden onset of severe typical chest pain that began one hour prior to presentation in the emergency room. She had no history of hypertension, diabetes mellitus, or coronary artery disease. She underwent mitral valve replacement in August 2008 for severe mitral regurgitation due to rheumatic heart disease. She had another Redo mitral valve replacement in October 2016 on top of malfunctioning mitral mechanical prosthesis after her 2nd pregnancy. She was not adherent to her anticoagulants, and she stopped warfarin for the past three weeks. On admission to hospital, physical examination revealed that the patient was anxious, pale with heart rate 90/minute and blood pressure of 110/70 mmHg. Cardiac examination revealed well heard click replacing 1st heart sound with no murmurs heard. Chest examination revealed normal vesicular breathing with no adventitious sounds. ECG

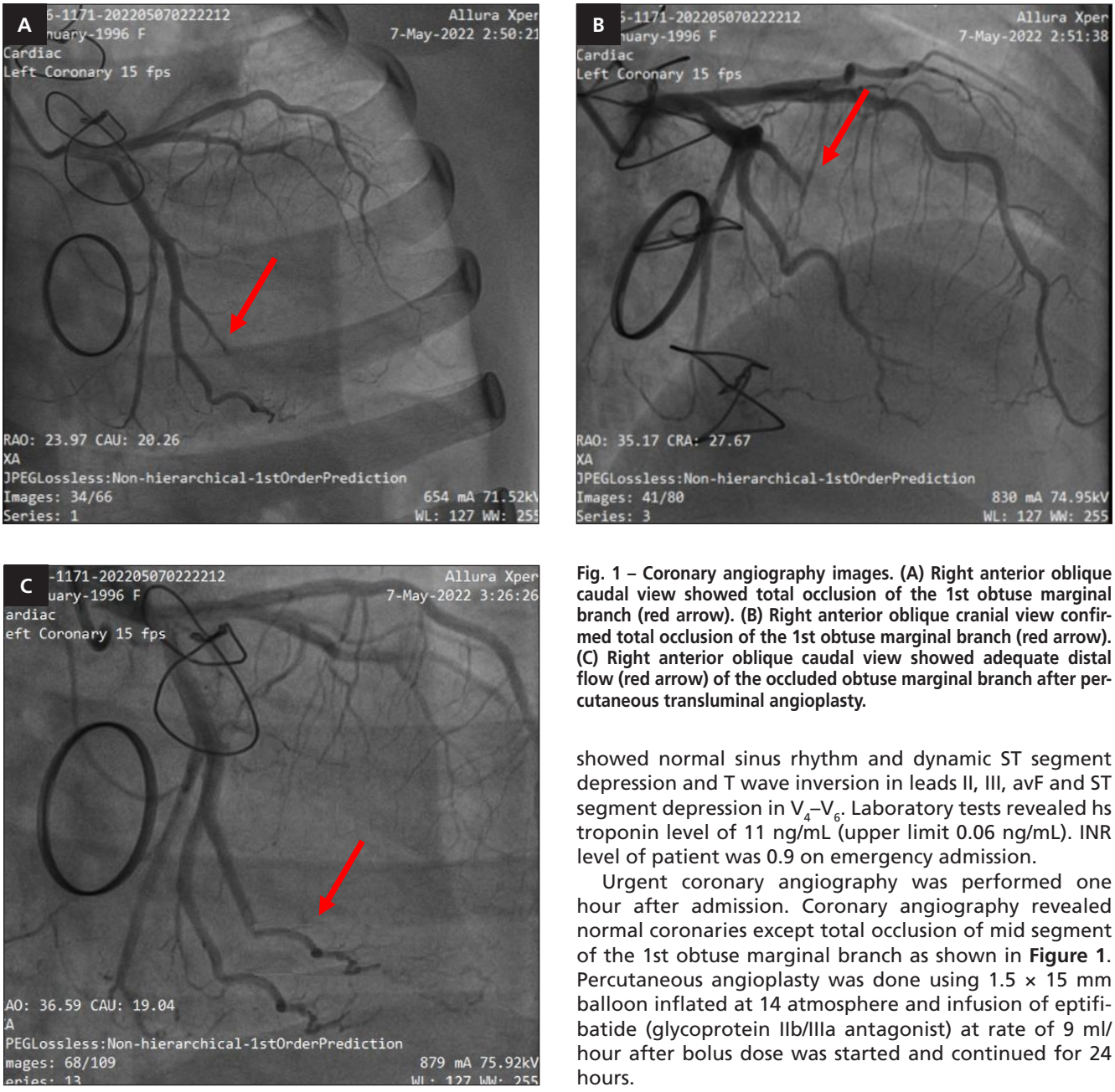

Fig. 1 – Coronary angiography images. (A) Right anterior oblique caudal view showed total occlusion of the 1st obtuse marginal branch (red arrow). (B) Right anterior oblique cranial view confirmed total occlusion of the 1st obtuse marginal branch (red arrow). (C) Right anterior oblique caudal view showed adequate distal flow (red arrow) of the occluded obtuse marginal branch after percutaneous transluminal angioplasty.

showed normal sinus rhythm and dynamic ST segment depression and T wave inversion in leads II, III, avF and ST segment depression in V_4–V_6. Laboratory tests revealed hs troponin level of 11 ng/mL (upper limit 0.06 ng/mL). INR level of patient was 0.9 on emergency admission.

Urgent coronary angiography was performed one hour after admission. Coronary angiography revealed normal coronaries except total occlusion of mid segment of the 1st obtuse marginal branch as shown in **Figure 1**. Percutaneous angioplasty was done using 1.5 × 15 mm balloon inflated at 14 atmosphere and infusion of eptifibatide (glycoprotein IIb/IIIa antagonist) at rate of 9 ml/hour after bolus dose was started and continued for 24 hours.

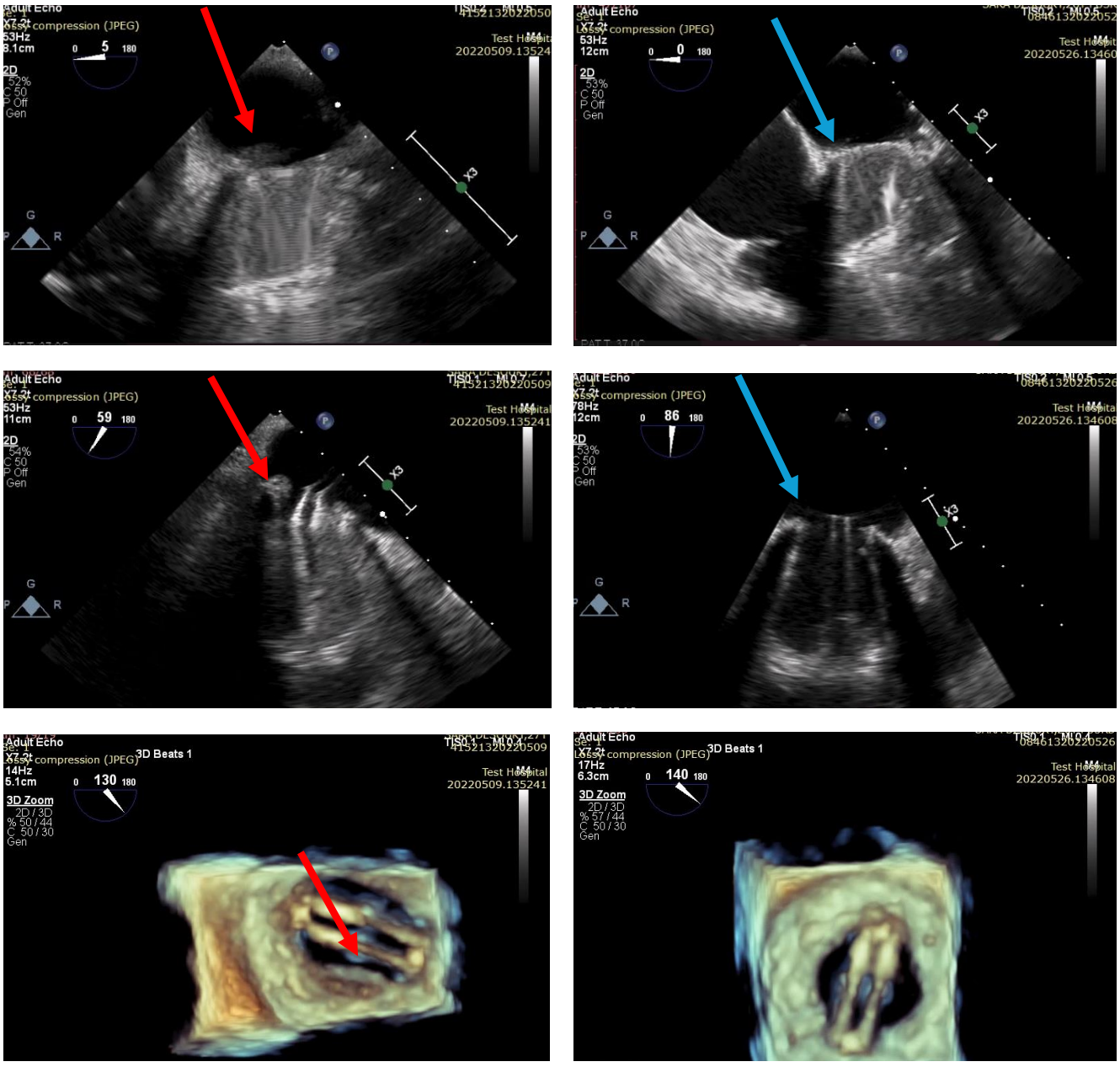

Fig. 2 – Transoephageal images. Left panel images showed thrombus (red arrows) related to one of the occluder of mitral mechanical prothesis. Right panel images showed follow-up transoesophageal echocardiography images revealing resolution of the thrombus (blue arrows) after anticoagulation therapy

Angiogram after percutaneous angioplasty revealed TIMI III coronary blood flow was restored as shown in **Figure 1**. The patient was shifted to the coronary care unit where transthoracic echocardiography was done showing normal left ventricle dimensions and mildly impaired systolic function; in addition, there were suspicious masses attached to mitral prosthesis. On the second day, transesophageal echocardiography (TEE) was done and revealed well-functioning mitral mechanical prosthesis with two masses seen attached to atrial aspect of mitral mechanical prosthesis, largest measure 1.3 × 0.6 cm as shown in **Figure 2** (left panel). Infective endocarditis was excluded with clinical, and lab work up. Heparin infusion was administered for three days, and warfarin 4 mg was started. Follow-up transesophageal echocardiography was done two weeks later and revealed resolution of the thrombus after achieving INR of 3.0 as shown in **Figure 2** (right panel). The patient was discharged on warfarin, aspirin, bisoprolol, and proton pump inhibitor therapy. The patient was informed about the importance of adherence to anticoagulant and timing of next coagulation profile.

Discussion

Virchow described the first case of coronary embolism in 1856.[1] In earlier studies, diagnosis was based on auto-

psy or coronary angiography findings, reported that 4% to 7% of AMI patients did not have atherosclerotic coronary disease.[2] Another autopsy study reported that 55 of 419 patients (13%) had coronary artery embolic infarcts.[3] Infective endocarditis was considered the most common cause of coronary embolism in earlier studies,[4] while atrial fibrillation was the most common underlying cause followed by cardiomyopathy and valvular heart disease in recent series.[5] Prosthetic cardiac valves were considered new disease process with the first case of coronary embolism arising from a mitral prosthesis in 1964.[6] The size of embolus and size of the lumen of the artery were the most important predictors of consequences of coronary embolism.[7] There is no consensus for the treatment of coronary embolism. Different treatment options as percutaneous interventions like catheter-aspiration embolectomy, percutaneous transluminal coronary angioplasty, stent placement, and medical approach with thrombolytic agents may be considered for management of coronary embolism.[8,9] Steinwender et al. reported success of intravenous application of bivalirudin for resolution of coronary embolus.[10] Qinn et al. reported successful management with intracoronary urokinase and intravenous abciximab following angioplasty procedure in complete resolution of the thrombus in a coronary embolism case.[11] Atmaca et al. reported the successful management of coronary embolism with half-dose tissue-type plasminogen activator (tPA) and tirofiban given intravenously in a patient with mitral valve prosthesis.[12] We experienced restoration of TIMI III coronary blood flow with percutaneous transluminal angioplasty and continued glycoprotein IIb/IIIa antagonist for 24 hours.

Conclusion

Coronary embolism secondary to prosthetic valve is a rare cause of myocardial infarction. Percutaneous balloon angioplasty without stenting and eptifibatide administered intravenously may be an effective treatment of this condition.

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Conflict of interest
The authors had no conflict of interest to declare.

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Ethical statement
This case report was conducted in accordance with the principles of the Declaration of Helsinki.

Informed consent
The patient provided an informed consent to present this case report.

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Aneurysma vena poplitea u pacientky s plicní embolií

(Popliteal vein aneurysm in a patient with pulmonary embolism)

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SOUHRN

Plicní embolie v rámci manifestace žilního tromboembolismu je třetím nejčastějším akutním kardiovaskulárním onemocněním. Obvyklým zdrojem plicní embolizace je hluboká žilní trombóza dolních končetin. V určitém procentu případů není trombóza ultrazvukovým vyšetřením identifikována, nicméně může být zjištěna jiná anomálie žilního systému jako pravděpodobný zdroj tromboembolu. V případě kazuistiky naší pacientky s manifestním tromboembolismem se jedná o rozsáhlé, dosud zcela asymptomatické aneurysma vena poplitea. Jde o vzácnou, málo zdokumentovanou žilní patologii s nejednotným terapeutickým postupem. V našem případě bylo přistoupeno k operačnímu řešení, díky čemuž bylo následně možné vysadit antikoagulační terapii. V rámci dalšího sledování u pacientky dosud nedošlo k opětovné významné redilataci popliteální žíly ani k recidivě tromboembolické nemoci.

Keywords:
Deep vein thrombosis
Popliteal vein aneurysm
Pulmonary embolism

ABSTRACT

Pulmonary embolism as a manifestation of venous thromboembolism is the third most common acute cardiovascular disease. The most common source of pulmonary embolism is deep vein thrombosis of the lower extremities. In a certain percentage of cases, this is not identified by ultrasound examination; however, another anomaly of the venous system may be detected as a probable source of embolus. In the case of our patient with manifest thromboembolism, it is a large, so far completely asymptomatic aneurysm of the popliteal vein. It is a rare, poorly documented venous pathology with a non-uniform therapeutic approach. In our case, a surgical solution was approached, which subsequently made it possible to discontinue anticoagulation therapy. As a part of further follow-up, there has been no recurrence of thromboembolic disease or significant redilatation of the popliteal vein.

Úvod

Žilní tromboembolická nemoc (TEN), klinicky se prezentující jako hluboká žilní trombóza (HŽT) nebo plicní embolie (PE), je celosvětově třetím nejčastějším akutním kardiovaskulárním onemocněním, hned za infarktem myokardu a cévní mozkovou příhodou.[1] Zdrojem PE je nejčastěji proximální HŽT dolních končetin, nicméně u části pacientů nemusí být zdroj embolizace identifikován. Může tomu tak být z důvodu kompletní embolizace trombu nebo jeho atypické lokalizace (v úvahu pak připadá dolní dutá žíla, splanchnické nebo pánevní řečiště).

V případě naší kazuistiky představujeme případ 69leté pacientky s PE, u které bylo v rámci došetřování zdroje plicní embolizace sonograficky zjištěno dosud zcela asymptomatické rozsáhlé aneurysma vena poplitea. Jedná se o vzácnou, málo zdokumentovanou abnormalitu žilního systému dolních končetin, jejíž přesná prevalence, stejně jako jasná korelace mezi velikostí aneurysmatu a rizikem tromboembolismu s ním spojeným není známá. Vzhledem k raritnosti tohoto nálezu stále neexistuje jeho jasná definice a není stanoven obecně uznávaný konsenzus ohledně terapeutického přístupu.

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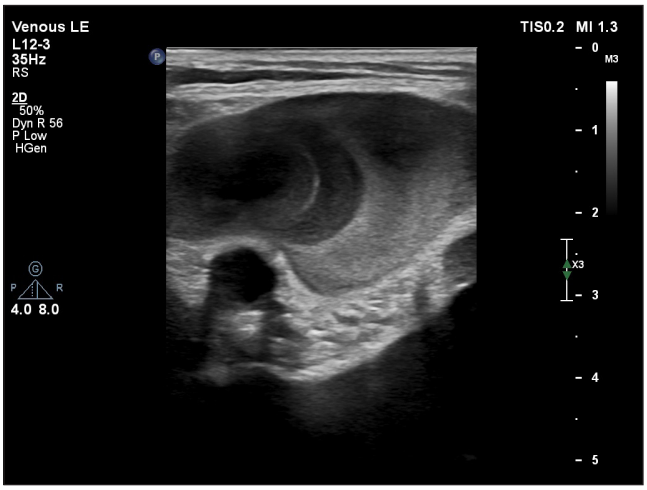
Kazuistika

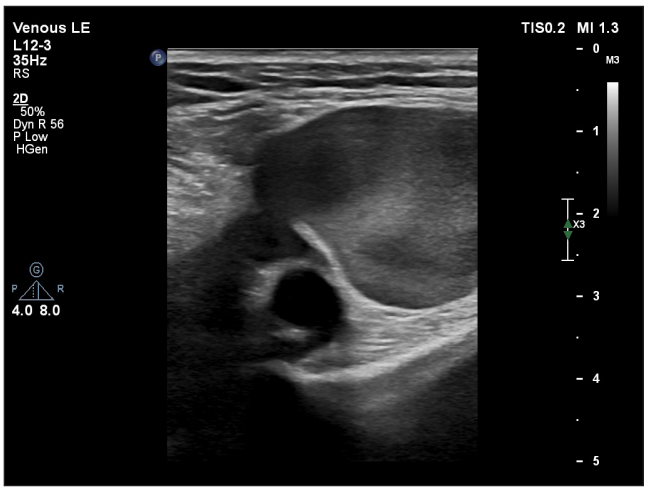
Devětašedesátiletá pacientka, exkuřačka, dosud bez zásadnějších interních či jiných komorbidit, byla přijata k hospitalizaci na naše interní oddělení pro CT angiograficky verifikovanou, bilaterální segmentální, nízkorizikovou plicní embolii, skóre PESI II. Jednalo se o primomanifestaci TEN. Recentně byla nemocná v předchorobí hospitalizována na infekčním oddělení pro bilaterální pneumonii vyvolanou covidem-19 s hraničními saturacemi, byla léčena remdesivirem, dále probíhala symptomatická terapie respiračního infektu včetně parenterálně podávaných kortikosteroidů a zvýšené preventivní dávky nízkomolekulárního heparinu (LMWH).

V rámci naší hospitalizace a terapie PE bylo zahájeno podávání plné dávky LMWH dle hmotnosti nemocné s následným převodem na přímá perorální antikoagulancia (DOAC), v našem případě apixaban v plné terapeutické dávce. V průběhu hospitalizace bylo doplněno echokardiografické vyšetření, kde pravá komora srdeční nebyla dysfunkční, nebyly prokázány známky klidové plicní hypertenze. Dále proběhl základní onkologický screening (sonografické vyšetření břicha s přiměřeným nálezem, gynekologické vyšetření bez suspekce na malignitu). Laboratorní vyšetření, až na pozitivní D-dimery, bylo v normě bez významnějších odchylek. V rámci pátrání po zdroji plicní embolizace bylo provedeno duplexní ultrazvukové vyšetření (DUS) žilního systému dolních končetin. Zde nebyla prokázána hluboká ani povrchová žilní trombóza, ale bylo zjištěno objemné aneurysma vena poplitea na pravé dolní končetině o rozměrech 40 × 20 mm, bez průkazu trombotizace, nicméně s výrazným denzním spontánním echokontrastem (**obr. 1, 2, 3**). Toto aneurysma bylo dosud klinicky zcela němé.

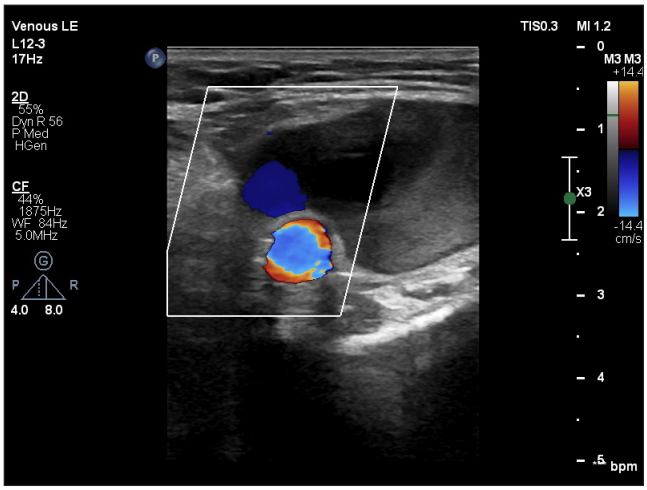
Přítomnost rozsáhlého aneurysmatu vena poplitea s obsahem hmot s velkým protrombogenním potenciálem se ve světle recentně prodělané bilaterální pneumonie způsobené covidem-19 s nutností hospitalizace, terapie antivirotiky a kortikosteroidy jevila jako vysoce pravděpodobný zdroj tromboembolu i přesto, že žilní trombóza nebyla jednoznačně prokázána. TEN jsme tedy hodnotili jako indukovanou infektem covid-19 v terénu predisponující patologie žilního systému.



Obr. 1 – Sakulární aneurysma vena poplitea se spontánním denzním echokontrastem.



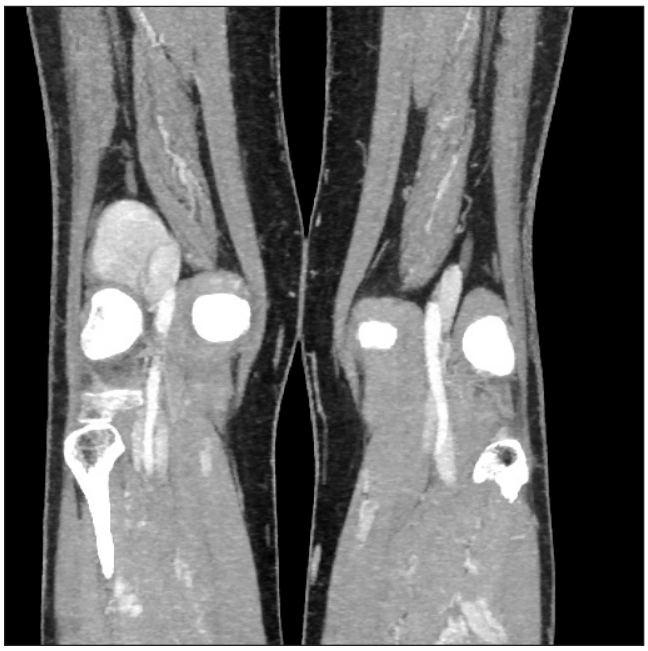
Obr. 2 – Vena poplitea s krčkem navazujícího sakulárního aneurysmatu.



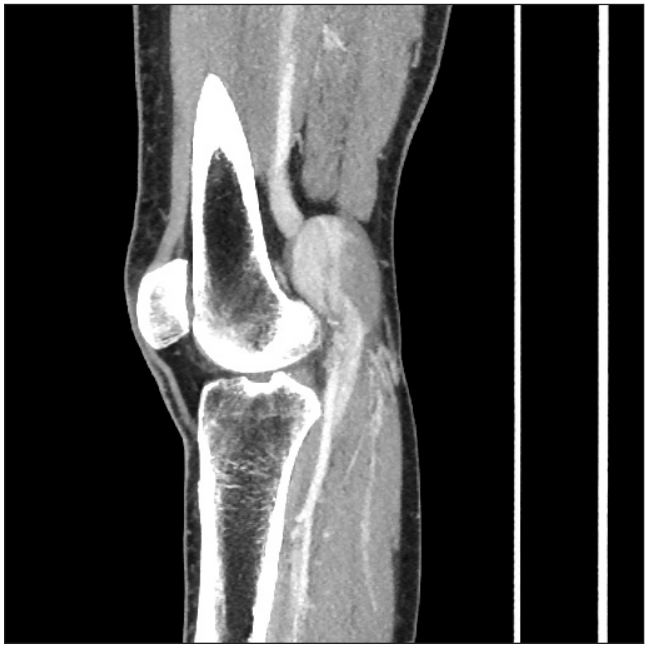
Obr. 3 – Aneurysma vena poplitea v intimním kontaktu s arteria poplitea, barevné dopplerovské zobrazení.

Po pětidenní hospitalizaci byla nemocná propuštěna s nastavenou terapií (včetně venotonik, kompresní terapie dolních končetin a poučení o režimových opatřeních) v celkově dobrém stavu a se zajištěnou následnou sonografickou dispenzarizací.

Při angiologické kontrole s odstupem šesti měsíců bylo provedeno kontrolní ultrazvukové vyšetření (došlo k další mírné dilataci aneurysmatu vena poplitea na 50 × 40 × 35 mm s přetrvávajícím denzním echokontrastem, bez průkazu formovaného trombu) a byla zvažována další antikoagulační strategie. Vzhledem k tomu, že se jednalo o primomanifestaci TEN indukovanou infektem covid-19, bylo by možné antikoagulační terapii ukončit, nicméně přítomnost rozsáhlého aneurysmatu vena poplitea s přetrvávajícím trombogenním potenciálem byla trvalým rizikovým faktorem recidivy HŽT. Z tohoto důvodu byla indikována dlouhodobá antikoagulační terapie a následné operační řešení aneurysmatického rozšíření popliteální žíly s výhledem možnosti ukončení antikoagulace. Antikoagulační terapii jsme do plánovaného výkonu ponechali v dávce sekundární prevence HŽT (apixaban 2,5 mg



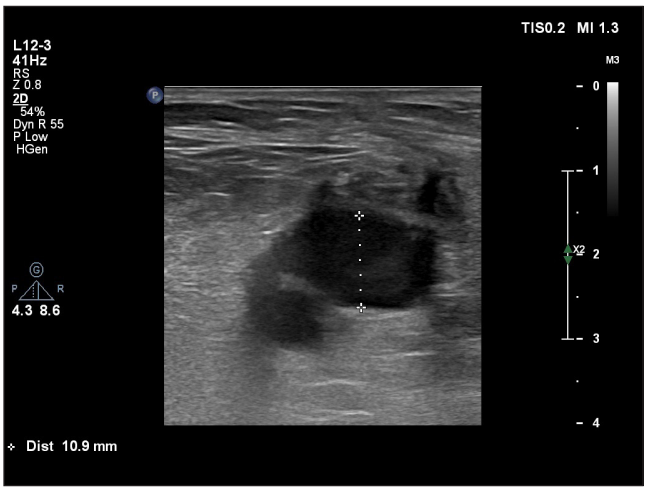
Obr. 4 – CT venografie se zaměřením na aneurysma vena poplitea pravé dolní končetiny – koronární řezy.



Obr. 5 – CT venografie se zaměřením na aneurysma vena poplitea pravé dolní končetiny – sagitální řezy.

ráno a večer). Byla doplněna CT venografie dolních končetin pro účely konzultace s cévním chirurgem a k ozřejmění detailních anatomických poměrů v zákolenní jámě.

Dle CT venografie se jednalo o objemné sakulární aneurysma vycházející z popliteální žíly, kdy vak aneurysmatu byl lokalizován v laterální části zákolenní jamky mezi femurem (ventrálně), šlachami musculus semitendinosus, musculus semimembranosus (laterálně) a musculus biceps femoris (mediálně) nad úrovní kolenní kloubní štěrbiny. Krček aneurysmatu byl obtížně detekovatelný, šíře aneu-



Obr. 6 – Sonografická kontrola vena poplitea po resekci aneurysmatického vaku.

rysmatického vaku kraniokaudálně činila 25 mm, vlastní aneurysma pak dosahovalo velikosti 38 × 40 × 78 mm s nehomogenní náplní kontrastem, kdy dle popisu rentgenologa nebylo možno vyloučit i nástěnnou trombózu (**obr. 4, 5**).

Nález byl indikován k chirurgické resekci a plikaci. Výkon i pooperační období proběhlo bez komplikací. Následná ultrazvuková kontrola byla s příznivým nálezem. V antikoagulační léčbě jsme pokračovali dalších šest měsíců od výkonu. Aktuálně je v medikaci ponechán pouze sulodexid a venotonika, zavedena kompresní terapie a režimová opatření. Pacientka dále dochází na pravidelné sonografické kontroly. Subjektivně je bez obtíží, objektivně je končetina bez otoku, jizva zhojena *per primam*. Sonograficky je vena poplitea po resekčním výkonu lehce dilatovaná, bez další progrese v čase sledování, bez spontánního echokontrastu a bez trombózy (**obr. 6**).

Diskuse

Periferní žilní aneurysmata jsou vzácným onemocněním. Údaje o nich lze proto čerpat pouze z menších sérií případů, jednotlivých kazuistik a systematických přehledů.

Samotná definice žilního aneurysmatu není jednotná. Nejčastěji je používána definice McDevitta a spol., kteří označují jako aneurysma perzistentní izolovanou dilataci žíly na dvojnásobek jejího normálního průměru, který je většinou mezi 5–7 mm.[2] Jiné práce definují venózní aneurysma jako 1,5násobek průměru dané žíly[3] či rozšíření nad 20 mm.[2]

Prevalence aneurysmat žil dolních končetin není zcela známá, především kvůli tomu, že od doby, kdy bylo v roce 1968 prvně popsáno, byly následně publikovány jen kazuistiky a malé série případů.[3,4] Aneurysma popliteální žíly je nejčastěji popsaným aneurysmatem v rámci hlubokého žilního systému dolních končetin. Vyskytuje se nejčastěji v její proximální části.[3–5] Méně častá jsou pak aneurysmata ileofemorální, aneurysmata v oblasti žil horních končetin a aneurysmata jugulární žíly. Recentní německý soubor s 39 případy udává prevalenci popliteálního aneurysmatu 0,06 % u pacientů vyšetřovaných primárně pro jiné žilní

onemocnění.[6] Podle některých prací se vyskytují popliteální aneurysmata stejně často u obou pohlaví,[4] jiné práce uvádějí vyšší četnost u mužů,[7] další naopak u žen.[2] Zdá se, že častěji je postižena levostranná končetina.[7] Aneurysma popliteální žíly se může vyskytovat takřka v jakémkoliv věku.[8] Průměrný věk výskytu bývá udáván v 5.[8] až 6.[7,9] dekádě života. Aneurysmata mohou být vícečetná a mohou se vyskytovat i v koincidenci s aneurysmaty arteriálními.[9]

Venózní aneurysmata obecně můžeme dle etiologie vzniku dělit na primární a sekundární. Za primární považujeme ta, která nejsou výsledkem traumatu, arteriovenózní fistule, rozšíření žíly vzniklého v důsledku infekce, portální hypertenze nebo kongenitálních vaskulárních abnormalit.[5] Etiologie vzniku primárního žilního aneurysmatu není plně objasněna, jedná se pravděpodobně o kombinaci vrozené nedostatečnosti žilní stěny, prodělaného traumatu v dané oblasti, lokálních degenerativních změn cévní stěny,[4] zánětu, rheologických a hemodynamických faktorů.[2]

Jsou popisovány dvě morfologické formy popliteálních aneurysmat – sakulární a fuziformní. Která morfologická varianta obecně převažuje, není možno s určitostí říci. V některých pracích se vyskytovalo více sakulárních,[8,9] v jiných naopak více fuziformních aneurysmat.[3]

Klinicky se popliteální aneurysma může manifestovat jako rezistence v podkolenní jamce, otok končetiny či bolest vyvolaná útlakem okolních struktur samotnou výdutí.[10,11] Často se ale jedná o náhodný nález v rámci ultrazvukového vyšetření žilního systému dolní končetiny z jiného důvodu. Někdy je první manifestací až plicní embolie.[8]

Popliteální aneurysma je ve většině případů diagnostikováno pomocí duplexní sonografie, nejčastěji v rámci zobrazení žilního systému dolních končetin při vyšetření pro suspekci na chronickou žilní nedostatečnost, v rámci pátrání po zdroji plicní embolizace nebo při sonografickém vyšetření z jiného důvodu. Dalšími možnými diagnostickými metodami jsou CT venografie, MR venografie či invazivní venografie, které jsou ale v rámci primární diagnostiky zastoupeny v menšině. Někteří autoři uvádějí nutnost venografie k lepšímu ozřejmění anatomických poměrů před jakoukoliv operační intervencí,[4] nicméně tyto další předoperační zobrazovací modality nad rámec duplexní sonografie ve srovnání s pouhým sonografickým vyšetřením neprokázaly lepší pooperační výsledky.[3]

Vzhledem k poruše žilního toku krve je v oblasti dilatované žíly zvýšené riziko vzniku rozsáhlé hluboké žilní trombózy a její následné embolizace. Riziko plicní embolizace je obtížné stanovit vzhledem k nejasné prevalenci aneurysmatu vena poplitea. V některých pracích se udává toto riziko mezi 25 % a 50 %[4,5] a plicní embolie může být primárním symptomem až v 80 % případů.[4] Mnohdy nemusí být ani zdroj v podobě HŽT následně detekován a je nalezena pouze aneurysmatická dilatace popliteální žíly. Existuje představa, že aneurysma vena poplitea funguje jako generátor trombů, které jsou neprodleně uvolňovány do plic.[2] Velká sakulární aneurysmata s turbulentním tokem[6] jsou náchylnější k tromboembolickým komplikacím[2] než aneurysmata menší nebo fuziformní. V práci z roku 2022 byl zjištěn statisticky signifikantní rozdíl ve velikosti aneurysmat prezentujících se s tromboembolickými komplikacemi či bez nich. Průměrná velikost popliteálních žilních aneurysmat s přítomnou HŽT či PE byla 3,8 cm, bez TEN 2,5 cm.[3]

Kvůli raritnosti této patologie popliteální žíly nebyl do dnešní doby stanoven žádný obecně akceptovaný konsenzus ohledně jejich léčby.[3] Terapie popliteálních aneurysmat může být buď konzervativní, nebo operační.[5] Použití endovaskulární léčby je popsáno v léčbě žilních aneurysmat v jiných lokalizacích, než je popliteální žíla.[12,13]

Konzervativní přístup může být zvolen v případě, že pacient nesouhlasí s operačním zákrokem, pokud není operační zákrok pro pacienta únosný vzhledem k jeho celkovému stavu a komorbiditám nebo pokud je žilní aneurysma asymptomatické, menších rozměrů, s nízkým tromboembolickým rizikem.

Konzervativní postup je doporučován některými autory pro fuziformní aneurysmata < 20 mm[8] či pro aneurysmata < 20 mm nebo ty bez turbulentního toku.[6] Vzhledem k vysokému trombogennímu potenciálu je v rámci konzervativní terapie u větších aneurysmat indikována plná antikoagulace a pravidelné sonografické sledování. Nicméně u malých (< 20 mm) fuziformních aneurysmat bez dalších rizikových faktorů vzniku tromboembolismu a anamnézy TEN Sessa a spol. antikoagulační léčbu nepodávali.[2] Dle dostupných zdrojů jsou ale i přes dobře vedenou antikoagulační terapii u rozměrnějších výdutí dokumentovány opakované tromboembolické příhody. Kazuistiky a malé soubory případů ukazují více než 60% riziko rekurence tromboembolismu v případě konzervativní terapie.[4] Antikoagulaci jako samostatnou terapii u pacientů s anamnézou PE nedoporučuje práce autorů Sessa a spol. (retrospektivní analýza na 25 pacientech) z důvodu vysokého rizika rekurence (až 80 %).[2] Dle jiné retrospektivní analýzy autorů Noppeney a spol. s 39 pacienty, z nichž 8 nebylo řešeno operačně, ale byli jen sledováni a dlouhodobě antikoagulováni, oproti tomu nedošlo k žádné komplikaci ve smyslu TEN.[6] Stejně tak práce autorů Patel a spol. z roku 2019 a 2022 dokládá, že v průběhu sledování pomocí DUS u primárně konzervativně řešených asymptomatických malých aneurysmat nebyla zachycena žádná tromboembolická komplikace.[3,4]

Většina autorů doporučuje, aby byla symptomatická aneurysmata řešena operačně,[3] byť ani v tom nepanuje naprostá shoda. Autoři Davidson a spol. doporučují pouze zvážit chirurgické řešení u pacientů se symptomy obstrukce a rozměrem > 3 cm.[7] U asymptomatických nemocných lze nalézt různá doporučení. Mendes a spol. operovali nemocné s velikostí aneurysmatu > 20 mm či s murálním trombem,[9] Sessa a spol. doporučují operovat všechna sakulární aneurysmata a fuziformní s velikostí > 20 mm.[2,8] Noppeney a kol. doporučují operovat nemocné s rozměrem výdutě > 20 mm (ve vzpřímené poloze) a turbulentním tokem.[6] Dle autorů Patel a spol. představuje rozměr 2,5 cm hranici, nad kterou by operace měla být nabídnuta; přítomnost nástěnného trombu obturující lumen > 25 % či sakulární morfologie by dle nich také mohly představovat další indikaci k operaci.[3] Donaldson a spol. jsou konzervativnější – doporučují zvážit operaci až při recidivě tromboembolické nemoci či při perzistenci trombu po třech až šesti měsících antikoagulační léčby.[7]

Operační řešení žilních aneurysmat dolních končetin obecně vede k redukci rekurence tromboembolismu. Po operačním výkonu na aneurysmatu nebyl dokumentován žádný případ rekurence plicní embolizace.[3,4]

V rámci invazivního řešení se využívají různé operační postupy. Patří k nim tangenciální aneurysmektomie

s laterální venorhafyií (nejčastěji prováděná a doporučovaná pro sakulární aneurysmata) a resekce aneurysmatu s interpozicí žilního štěpu (ke štěpu jsou využívány vena saphena parva [VSP], vena saphena magna [VSM] či superficiální femorální žíla). Dále se používá excize a ligace či resekce s end-to-end anastomózou. Lze využít i endovaskulárně asistovaného postupu.[14] Nejčastější komplikací u výkonů byl pooperační hematom v ráně,[3] přechodná paréza peroneálního nervu[15] a infekce v ráně.[2]

Jako rozumná alternativa operačního řešení či dlouhodobé antikoagulace se pro starší a křehké pacienty, kteří nejsou schopni podstoupit invazivní výkon nebo jež není možno dlouhodobě z určitého důvodu plně antikoagulovat, jeví implantace kaválního filtru.[2]

Nutnost pooperační antikoagulace zůstává nejasná. Doporučuje se ponechání krátkodobé antikoagulace (ve formě warfarinu či DOAC) po dobu tří až šesti měsíců po operačním zákroku a kompresní bandáže dolních končetin.[16] Jiní autoři doporučují LMWH po dobu tří týdnů a následnou perorální antikoagulaci na tři měsíce u pacientů s rizikovými faktory, předchozí TEN či po komplexní chirurgické rekonstrukci.[8] Celková doba antikoagulační léčby po operačním zákroku je značně individualní. Doporučuje se nošení elastických punčoch či bandáže.[8]

Následné sledování pacientů k vyloučení rekurence aneurysmatu či trombotické komplikace probíhá většinou pomocí DUS v obvykle šestiměsíčních intervalech,[17] v jiných souborech byli pacienti sledováni v delších 12měsíčních intervalech.[4]

Závěr

Naše kazuistika představuje pacientku bez interních komorbidit, u které došlo k primomanifestaci tromboembolické nemoci pravděpodobně indukované infektem covid-19. V průběhu běžného došetření bylo pomocí DUS, v rámci pátrání po zdroji plicní embolizace, zjištěno objemné, dosud zcela asymptomatické aneurysma vena poplitea. V aneurysmatu nebyla sonograficky detekována žilní trombóza. Pacientka byla léčena standardní perorální antikoagulační terapií. Vzhledem k tomu, že se jednalo o primomanifestaci TEN, navíc zjevně indukovanou, byla by primárně indikována jen dočasná antikoagulační terapie. Nicméně nález aneurysmatického rozšíření vena poplitea znamenal v případě volby konzervativního postupu antikoagulaci dlouhodobou, resp. časově neomezenou. S přihlédnutím ke známým faktům z dostupných kazuistik a lokálnímu ultrazvukovému nálezu bylo navíc u naší nemocné (kde byl v oblasti aneurysmatické části popliteální žíly přítomen výrazně denzní spontánní echokontrast a turbulentní krevní tok) i přes dobře vedenou antikoagulační terapii nemalé riziko recidivy TEN. Po domluvě s pacientkou jsme proto indikovali operační řešení žilní výdutě. Výkon i pooperační průběh byl nekomplikovaný a následně bylo možno vysadit antikoagulační terapii. Nyní je v medikaci zatím ještě ponechán sulodexid, venotonika a zavedená komprese končetiny. Sonografické kontroly jsou příznivé, dosud nedošlo k recidivě TEN ani k významné redilataci podkolení žíly a díky operačnímu řešení nebude pacientka dále zatížena antikoagulační terapií se všemi jejími případnými komplikacemi.

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Off label treatment of intraabdominal dissection with iliac stent graft extension. A case report

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SOUHRN

K disekci aorty omezené na abdominální aortu dochází poměrně vzácně; představuje pouhých 1–4 % všech případů disekce aorty. Pokud se týče optimální léčby symptomatické izolované disekce abdominální aorty (isolated abdominal aortic dissection, IAAD), je k dispozici málo důkazů. Dvěma hlavními intervenčními metodami jsou otevřená operace a endovaskulární přístup. Léčba onemocnění aorty – v nepřítomnosti aneurysmatu – klasickými stentgraftovými systémy je vzhledem ke „konkurenci" jejích jednotlivých větví ve zúženém distálním segmentu aorty náročná. Tato kazuistika popisuje naše klinické zkušenosti s off-label (mimo schválené indikace) endovaskulární léčbou asymptomatické disekce infrarenální části aorty doprovázené těžkou aterosklerózou aorty. U pacientky byla indikována katetrizační implantace aortální chlopně (transcatheter aortic valve implantation, TAVI) a pro zajištění potřebného přístupu pro výkon byla prodloužena větev aortálního endograftu.

Keywords:
Aortic dissection
Endovascular management of abdominal aortic dissections
Isolated abdominal aortic dissections

ABSTRACT

Aortic dissection limited to the abdominal aorta is an uncommon condition, accounting for only 1% to 4% of all aortic dissections. There is limited evidence on the optimal management of symptomatic isolated abdominal aortic dissections (IAAD). The two primary interventional methods are open surgery and the endovascular approach. Treating nonaneurysmal aortic disease is challenging with conventional stent-graft systems due to limb competition in a narrow distal aorta. This case report describes our clinical experience with an off-label endovascular treatment for an asymptomatic infrarenal aortic dissection, accompanied by severe aortic atherosclerosis, in a female patient. The patient was scheduled for a transcatheter aortic valve implantation (TAVI) procedure, and an aortic endograft limb extension was used to ensure an adequate pathway for the procedure.

Introduction

Aortic dissection limited to the abdominal aorta is an uncommon clinical condition, accounting for only 1% to 4% of all aortic dissections.[1,2] The causes of isolated abdominal aortic dissection (IAAD) can be spontaneous, traumatic, or iatrogenic. Most affected individuals are men, with a median age of 60 years, and they often present with high blood pressure.[3] The disease can present either acutely with a sudden onset of symptoms or chronically, with symptoms appearing 14 days after onset.[3] Due to its rarity, documented mostly through case reports and small case series, the natural history and treatment options for IAAD are not well understood. The most common symptom is abdominal or back pain, while claudication and lower limb ischemia are rare.[2] Asymptomatic cases with a non-dilated aorta are usually managed conservatively, whereas symptomatic cases are treated with either open or endovascular repair, depending on anatomical considerations and the surgeon's expertise. This case report discusses the management of an asymptomatic infrarenal aortic dissection, combined with severe aortic atherosclerosis in a female patient, who was scheduled for a transcatheter aortic valve implantation (TAVI) procedure to ensure an adequate pathway for the procedure.

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Case report

A 78-year-old female patient was admitted to the cardiology department due to worsening chronic heart failure. Her medical history included severe aortic valve stenosis, aortic and mitral valve regurgitation, persistent atrial fibrillation, dyslipidemia, and hypertension. A cardiac CT scan was performed, and the patient was scheduled for TAVI. However, CT angiography of the distal vessels revealed a severely diseased infrarenal abdominal aorta with a stenotic and severely dissected lumen (**Fig. 1**), which was asymptomatic. Performing TAVI in this compromised abdominal aorta posed a risk of worsening the dissection, causing distal embolization, or resulting in aortic rupture. To create a suitable pathway for the TAVI procedure, an off-label iliac extension stent graft 16/82mn (Endurant Medtronic) was placed in the diseased infrarenal aorta. The final result was excellent, with no complications and normal flow of the contrast agent (**Fig. 2**). A CT scan one month later confirmed a patent stent graft with no residual stenosis (**Fig. 3**). Moreover, the patient underwent TAVI procedure three months after, with no complications.

Discussion

Aortic dissection confined to the abdominal aorta is an uncommon condition, accounting for only 1% to 4% of all aortic dissections.[1–3] The existing literature offers limited guidance on the optimal treatment approach, primarily consisting of small patient series. In 2009, Jonker et al.[4] conducted a review and meta-analysis of clinical data involving 92 patients with isolated abdominal aortic

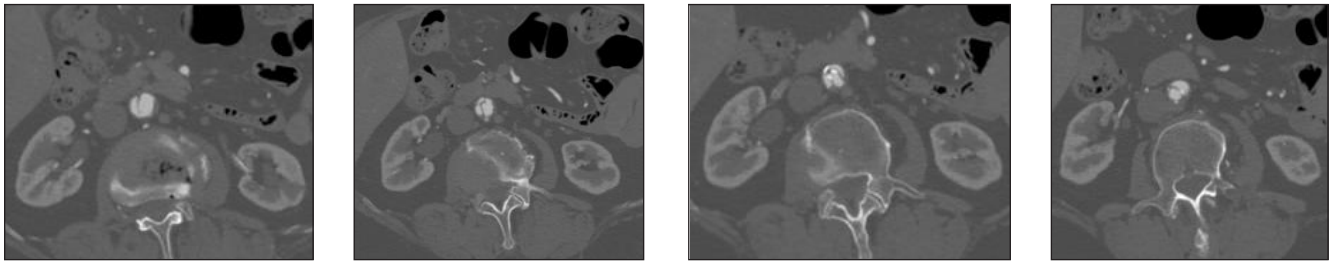

Fig. 1 – Dissection of infrarenal aorta

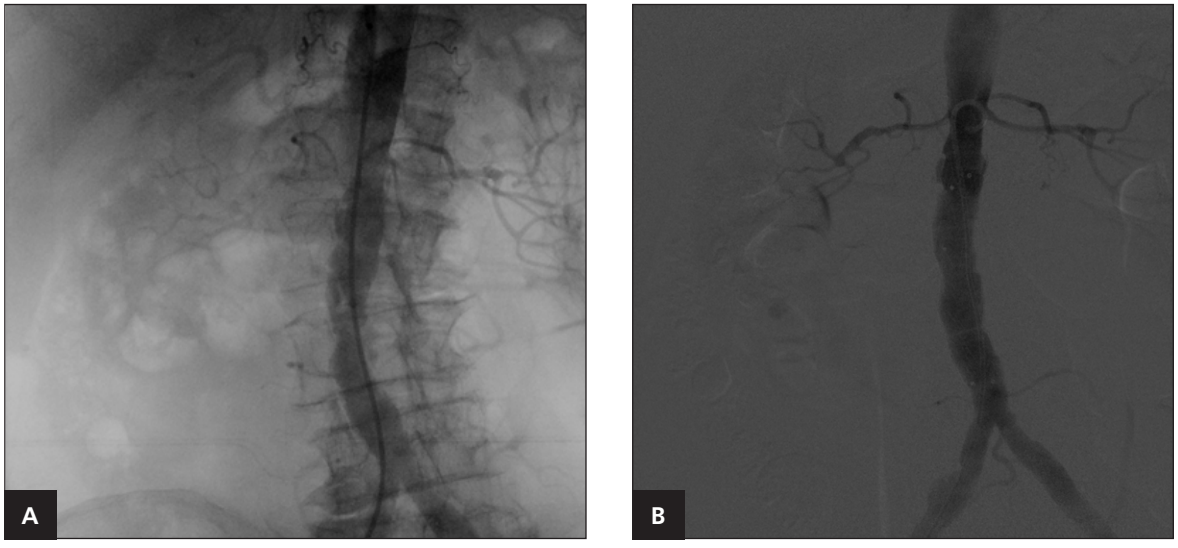

Fig. 2 – (A) Initial angiogram showing and ulcerated and dissected abdominal aorta. (B) Excellent result after covered stent placement.

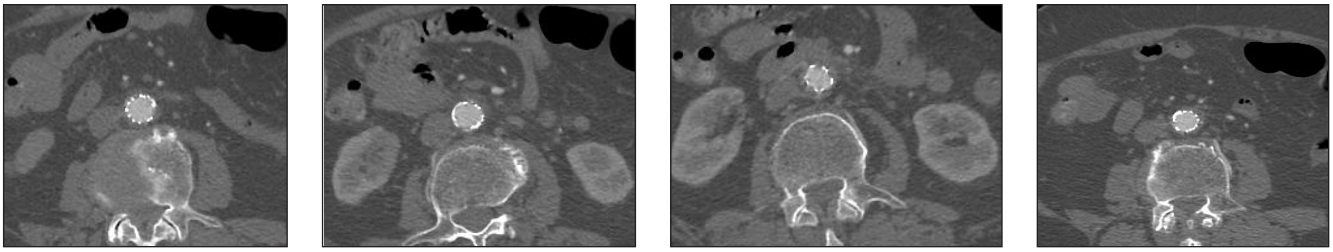

Fig. 3 – CT scan after stent placement showing no residual stenosis.

dissections. Most of these patients received conservative treatment or underwent traditional surgical procedures, with only 19 (21%) undergoing endovascular repair using a variety of stents, stent-grafts, and balloons. Due to the heterogeneity of the cases, short-term follow-up, and the small number of patients treated, no definitive conclusions could be drawn regarding the best therapeutic strategy for these dissections.

In a more recent study, Jawadi et al.[5] presented the long-term outcomes of 21 patients (18 men; mean age 72 ± 18 years, range 34–90) with IAAD who underwent endovascular treatment between January 2000 and December 2012. Fourteen patients had spontaneous abdominal dissections, while 7 had iatrogenic dissections. The average dissection length was 45 ± 12 mm (range 18–98). Ten patients received tube stent-grafts, 9 had bifurcated endografts, one received an aorto-uni-iliac (AUI) endoprosthesis, and one was treated with a bare metal stent. All patients achieved complete aortic remodeling over a 12-year period. The study found no mortality, few complications, and a rare need for secondary interventions with endovascular IAAD treatment.

Surgical treatment of IAAD typically carries a significant risk of in-hospital mortality (4%) and complications (9%).[4] In a meta-analysis by Jonker et al. surgery was chosen as the treatment option for 50% of the 92 patients analyzed. Among them, one patient (2.2%) died during the hospital stay, and six patients (13%) experienced complications such as neurological issues in the lower limbs (n = 4), bleeding (n = 1), or renal failure (n = 1). In contrast, endovascular techniques offer a less invasive approach to IAAD treatment with minimal or no mortality and a low complication rate.[4] Based on our clinical experience with aortic wall lesions, including PAUs, intramural hematomas, and dissecting saccular aneurysms, we advocate for an early and aggressive approach to treating these conditions, given the progressive degeneration of the aortic wall that can ultimately lead to rupture.

There are no definitive guidelines favoring one type of endovascular device over another. If there is no thrombus formation in the true lumen, the risk of peripheral embolization is minimal. In such cases, bare metal stents can be considered the first choice to maintain perfusion in collateral pathways and lumbar arteries. However, if there is a risk of peripheral embolization or a co-existing abdominal aortic aneurysm (AAA), stent-grafts are recommended. In the case presented, although there was no evidence of aortic intraluminal thrombus, the heavily calcified dissected lesion posed a high risk of aortic rupture, particularly during post-dilatation aimed at increasing the luminal diameter. For this reason, we opted for a covered stent (balloon expandable) or stent-graft. Before stent placement, we were uncertain about the potential residual stenosis we might encounter, which led us to choose a self-expandable stent-graft for better alignment with the vessel wall. After the initial stent implantation, a high-grade stenosis was observed, which we treated with aggressive balloon post-dilatation to achieve better lumen expansion. The outcome was excellent, with minimal risk of aortic leakage.

Conclusion

Endovascular treatment of IAADs appears to be a safe approach, delivering lasting results over the long term. The procedure's minimally invasive nature is linked to significant clinical advantages, particularly in reducing mortality and morbidity. In this clinical case, we successfully achieved complete aortic remodeling using an off-label application of an iliac endograft limb extension in the aortic position, without encountering any peri- or post-procedural complications. Moreover, we secured a pathway for a future TAVI procedure which yielded excellent results with no complications.

Conflict of interest
The authors declare no conflict of interest.

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Ethical statement

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Chronic Venous Insufficiency in a 55-Year-Old Female: Highlighting Overlooked Cases for Increased Awareness

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Kompresní terapie

SOUHRN

Úvod: Chronická žilní insuficience (chronic venous insufficiency, CVI) je časté onemocnění s postižením toku krve žilami vedoucí k žilní hypertenzi. Tato dysfunkce je často výsledkem selhání funkce chlopní v důsledku varixů (křečových žil), poškození hlubokých žil nebo slabých žilních stěn v důsledku nedostatku kolagenu a elastinu. Chronická žilní insuficience se projevuje dilatací žil, otoky dolních končetin, bolestí a změnami kůže. Přes tyto významné důsledky zůstává CVI nedostatečně diagnostikována a léčena, proto často progreduje do postflebitického syndromu a tvorby žilních vředů. Cílem této studie bylo zdůraznit prevalenci CVI u této vysoce rizikové populace a popsat účinná preventivní opatření a léčebné strategie pro snížení počtů komplikací a zlepšení výsledného stavu pacientů.
Popis případu: V únoru 2024 se k lékaři dostavila 55letá žena s progredujícími otoky a ulcerací na levé noze; tento stav se zhoršoval již tři měsíce. Otoky zhoršující se při delším stání a zmírňující se při zvednutých dolních končetinách byly doprovázeny bolestí a pocitem těžkých nohou. Podobné symptomy měla nemocná již o šest měsíců dříve; ty se zmírnily krátkodobou léčbou. Fyzikální vyšetření prokázalo tuhý edém a odhalilo vřed o rozměrech 10 × 10 cm na levé dolní končetině. Hodnota indexu tělesné hmotnosti (BMI) pacientky byla 28,23, krevní tlak 130/80 mm Hg a kromě popsaných obtíží nebyla anamnéza ženy ničím významná. Ultrazvuk prokázal žilní nedostatečnost v nepřítomnosti obstrukce.
Diskuse: Chronická žilní insuficience vzniká na podkladě inkompetence žilních chlopní a vede k žilní hypertenzi a poškození tkání. Postižení progreduje z žilní stáze do změn kůže a tvorby vředů. Symptomy pacientky odpovídaly typickým projevům CVI a zhoršovaly je nadváha a skutečnost, že se jednalo o ženu. Účinná léčba je založena na konzervativních opatřeních, jako jsou používání kompresních punčoch, zvedání dolních končetin a změny životosprávy spolu s pokročilými léčebnými metodami, jako jsou skleroterapie, termální ablace a v případě potřeby chirurgická intervence. Popsaný případ podtrhuje význam časného stanovení diagnózy a komplexní léčby pro prevenci progrese a zlepšení výsledného stavu pacienta.
Závěr: Chronická žilní insuficience, která je často nerozpoznána, vyžaduje časnou intervenci, aby se předešlo závažným komplikacím. Popsaný případ podtrhuje nutnost zvýšené pozornosti a proaktivní léčby, zvláště u vysoce rizikových populací. Pro účinnou léčbu a zlepšení výsledného stavu pacientů jsou nezbytné komplexní léčebné strategie včetně úpravy životosprávy a použití pokročilých léčebných metod.

Keywords:
Case report
Chronic venous insufficiency
Compression therapy

ABSTRACT

Introduction: Chronic venous insufficiency (CVI) is a prevalent condition characterized by impaired venous flow leading to venous hypertension. This dysfunction often results from valve failure due to varicose veins, deep vein damage, or vein wall weakness influenced by altered collagen and elastin. CVI manifests as dilated veins, leg edema, pain, and skin changes. Despite its significant impact, CVI remains underdiagnosed and undertreated, often progressing to postphlebitic syndrome and venous ulcers. This study aims to highlight the prevalence of CVI in this high-risk population and to provide insights into effective preventive measures and management strategies to reduce complications and improve patient outcomes.

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Case illustration: A 55-year-old female presented in February 2024 with progressive swelling and ulceration of the left leg, worsening over three months. The swelling, exacerbated by prolonged standing and relieved by elevation, was accompanied by a heavy, painful sensation. An earlier episode of similar symptoms six months prior had improved with short-term treatment. Physical examination revealed non-pitting edema and a 10 × 10 cm ulcer on the left leg. The patient had a BMI of 28.23, blood pressure of 130/80 mmHg and no significant medical history aside from the current condition. Ultrasound showed venous insufficiency without obstruction.
Discussion: CVI is caused by venous valve incompetence leading to venous hypertension and tissue damage. This condition progresses from venous stasis to skin changes and ulcer formation. The patient's symptoms align with CVI's typical presentation, exacerbated by her overweight status and gender. Effective management includes conservative measures like compression stockings, leg elevation, and lifestyle changes, along with advanced treatments such as sclerotherapy, thermal ablation, and, when necessary, surgical intervention. The case underscores the importance of early diagnosis and comprehensive management to prevent progression and improve patient outcomes.
Conclusion: CVI, often under-recognized, requires timely intervention to prevent severe complications. This case highlights the need for heightened awareness and proactive treatment, especially in high-risk populations. Comprehensive management strategies, including lifestyle modifications and advanced therapies, are essential for effective treatment and improved patient outcomes.

Introduction

Chronic veins insufficiency (CVI) is a condition where venous flow is disrupted which causes venous hypertension.[1] This is caused by failure of venous valve function due to varicose veins or damage to the deep veins secondary to venous thrombosis, trauma or venous obstruction.[2] Another study shows that apart from valve failure, weakness of vein's walls is more influential in causing CVI which is caused by changes in the composition of collagen and elastin.[3] The major clinical manifestations of chronic venous insufficiency (CVI) are dilated veins such as telangiectasias, reticular veins, varicose veins, leg edema, pain/achiness/heaviness, and skin alterations.[4] Despite its potentially fatal impact, CVI has been underdiagnosed and undertreated for a long time which is usually progressive and leads to postphlebitic syndrome and venous ulcers.[1,2] The prevalence of CVI is higher in women, 25–40% and in contrast 10–20% in men, with an annual incidence of 2–6% in women and 1.9% in men.[5] This is believed to occur due to various factors, including the influence of the hormone estrogen and pregnancy.[3,4] Other risk factors for this condition include older age over 30, heavy lifting, multiple pregnancies, oral contraceptive, obesity, sedentary lifestyle, prolonged standing occupation, history of leg trauma, family history, and prior history of deep venous thrombosis.[1,2,4] Furthermore, hereditary factors are also considered to play an important role in the development of CVI, such as the genetic disorders Klippel–Trenaunay and Parkes–Weber.[1]

Initial management of CVI is conservative treatment, such as compression stockings with a tension between 30–50 mmHg which can improve pain, edema and pigmentation.[1,2,4,6] Advanced CVI that damages the surface of the skin/ulcers can be treated with wound compression bandaging systems, topical moisturizers, and other wound care.[1,2,4,6] Other conservative treatments include leg elevation, weight management, and exercise.[1,4] Sclerotherapy using sclerosing agents include the hypertonic solution of sodium chloride (23.4%), polidocanol, sodium iodide, chromated glycerin, sodium tetradecyl sulfate, and sodium morrhuate is beneficial in treating telangiectasias, reticular veins, varicose veins (1–4 mm diameter), and veins with reflux.[1,6] It can be used as a primary treatment or in combination with other treatments.[1,6] For patients with GSV reflux, ablation utilizes thermal energy through radiofrequency or laser results in complete obliteration in 85% of patients after two years.[6] For iliac vein stenosis and occlusion, endovascular stenting can be used.[6] Sulodexide and pentoxifylline for adjuvant to compression therapy is an effective pharmacological therapy which targets multiple sites involved in the pathogenesis of CVD.[2,4,7] In individuals with persistent discomfort and disability after pharmacological or endovenous therapy, surgical surgery for CVI may be considered in addition to compression stockings.[1,2,6] The surgical method used depends on the pathophysiological process underlying the CVI, and may consist of stripping, simple ligation/division, and venous valve reconstruction.[1,2,6]

Case description

In February 2024, a 55-year-old outpatient female with complaints of a wound and swelling on the left lower leg for three months was brought to the RS Universitas Airlangga. The swelling was localized to the left leg, with no involvement of the right leg. It began gradually, initially affecting the dorsum of the left foot and progressively extending upward below the knee over the last two months. The wound appeared suddenly and has progressively enlarged over the past month. The swelling worsens when the patient stands for extended periods or walks, and reduces when she sits and elevates her leg. Initially, the swelling would decrease to nearly match the size of her right leg, but over time it became persistent and difficult to reduce. The patient reported that her left leg feels heavier, with pain worsening as the swelling increased, making walking difficult. There was no associated redness, skin discoloration, or additional wounds on the affected limb.

Six months earlier, the patient had a similar episode of swelling and pain in the left leg, although the symptoms were milder. At that time, an ultrasound examination identified venous issues without obstruction. The patient was treated for five days, resulting in improve-

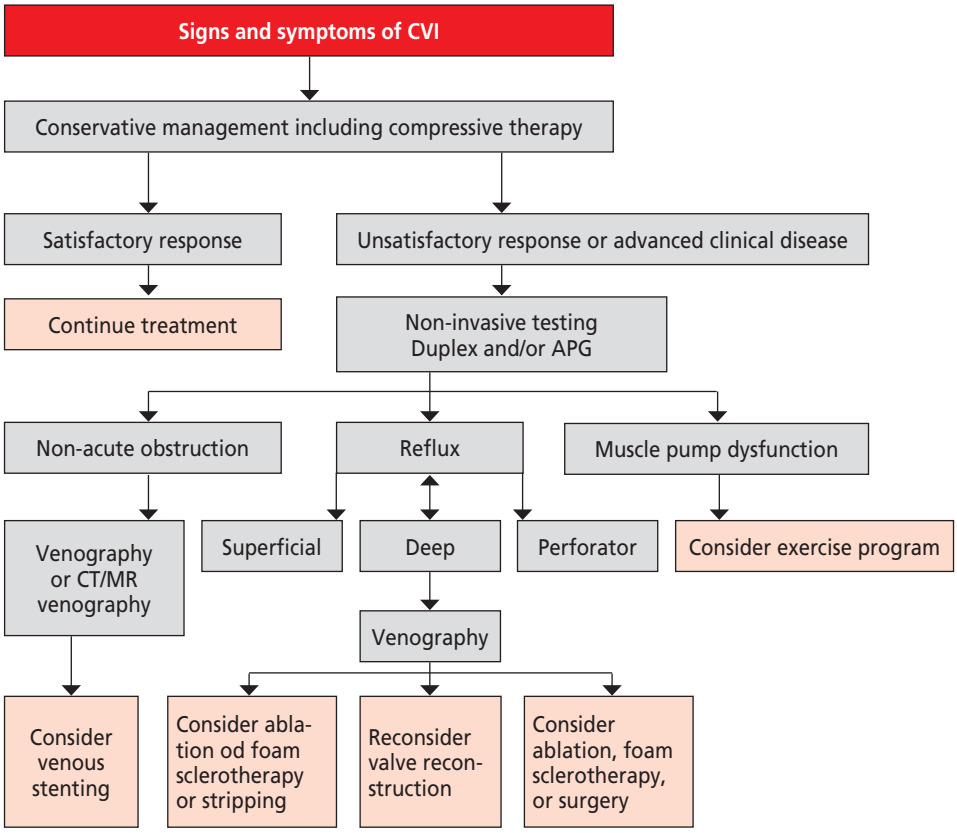

Fig. 1 – Algorithm for diagnosing and treating CVI.[2]

ment, after which she was discharged. However, she has not been on any routine medications for this condition. The patient's medical history is otherwise unremarkable, with no history of diabetes mellitus, hypertension, stroke, heart disease, or kidney disease.

On physical examination, the patient appeared moderately ill, with a Glasgow Coma Scale score of 15. Her vital signs were stable, with a blood pressure of 130/80 mmHg, pulse rate of 88 beats per minute, respiratory rate of 18 breaths per minute, and a body temperature of 36.8 °C. The patient had a BMI of 28.23 (overweight). The general examination revealed a normocephalic head, evenly distributed black hair, and symmetrical facial features. Eye examination showed no eyelid edema, normal conjunctiva and sclera, clear cornea, and equal, reactive pupils. The ears and nose were normal, with no hearing loss or septal deviation. The oral cavity and pharynx were unremarkable, and there was no thyroid enlargement or tracheal deviation. Jugular venous pressure was within normal limits. Thoracic examination revealed normal chest configuration, with vesicular breath sounds bilaterally and no adventitious sounds. The heart had a regular rhythm with normal S1 and S2 sounds, and no murmurs. The abdomen was flat, with normal bowel sounds, no masses, and no tenderness. The extremities were warm, with non-pitting edema more prominent in the left leg. A 10x10 cm ulcer with irregular edges was present on the left lower leg, with no associated warmth, erythema, or varicose veins. Capillary refill time was less than 2 seconds, and there were no signs of cyanosis or varicosities. This case highlights a recurrent venous disorder with persistent non-pitting edema and an ulcer, requiring further evaluation and management.

Discussion

Pathophysiology

Chronic venous insufficiency (CVI) is a complex and progressive condition characterized by the inability of the venous system, particularly in the lower extremities, to efficiently return blood to the heart, caused venous to reflux. This inefficiency is largely due to venous valve incompetence, which leads to venous hypertension, venous stasis, and, over time, a cascade of pathological changes.[8,9] Venous hypertension is a critical factor in the development of CVI, as it results in increased pressure within the superficial veins. Over time, this leads to damage to the venous endothelium, capillary leakage, and subsequent inflammation, ultimately culminating in tissue damage, skin changes, and ulcer formation, as seen in the patient discussed here.[8–10] The impaired valve function allows blood to pool in the lower extremities, which increases the pressure within the veins and leads to distention and leakage into the surrounding tissues. This is compounded by a failure in the calf muscle pump, which is essential for

venous return, especially during activities such as walking.[9,11]

The resultant inflammatory response involves leukocyte activation and adhesion, increased expression of inflammatory mediators (e.g., cytokines like TNF-α and IL-6), and matrix metalloproteinases (MMPs), which degrade the extracellular matrix and contribute to the breakdown of the skin and subcutaneous tissue. Persistent venous hypertension also leads to capillary permeability, causing protein-rich fluid to accumulate in the interstitial spaces (edema), which is a hallmark of CVI. Over time, this can lead to lipodermatosclerosis, atrophie blanche, and ultimately venous ulceration, as observed in the patient.[10,12]

Anamnesis

The patient, a middle-aged female, presented with a history of unilateral lower limb swelling and ulceration that had progressively worsened over a three-month period. The symptoms were exacerbated by prolonged standing and relieved by leg elevation, a classic pattern in CVI. Notably, the patient had a history of similar, albeit milder, symptoms six months prior, suggesting a chronic and relapsing course typical of CVI. The ultrasound findings of venous insufficiency without evidence of occlusion align with the diagnosis of CVI, where valvular incompetence rather than thrombotic obstruction is the primary pathology.

The patient's demographic profile (female gender, middle age) and her body mass index (BMI of 28.23, indicative of overweight status) are significant as they are known risk factors for CVI. Hormonal factors, particularly in women, contribute to venous wall relaxation and valve dysfunction, which may explain the higher prevalence of CVI in females.[13,14] Additionally, obesity is a well-documented risk factor due to increased intra-abdominal pressure and its contribution to venous hypertension.[14]

Symptoms and manifestations

Chronic venous insufficiency is a condition in which the veins in the lower extremities have a decreased ability to effectively return blood to the heart, leading to a buildup of blood in the leg veins. This results in common symptoms such as leg discomfort, swelling, varicose veins, and skin changes or ulceration.[15] As CVI progresses, patients typically experience venous leg discomfort characterized by heaviness, throbbing, dull pain, and swelling in the legs, which worsens with prolonged standing and is often accompanied by a sensation of pressure; relief is generally achieved through measures that reduce venous pressure, such as elevating the legs, using compression stockings, or walking.[15,16] However, approximately 20% of patients with other clinical features of CVI do not experience leg discomfort, while around 10% of patients may have leg discomfort as their only clinical symptom.[15]

Patients with CVI may present without any complaints, but often show signs such as simple telangiectasia (<1 mm in diameter), reticular veins (1–3 mm in diameter), and varicose veins (>3 mm in diameter).[15] Varicose veins are dilated superficial veins that become increasingly tortuous and enlarged. They can develop episodes of superficial thrombophlebitis, which are identified by painful, hardened, and inflamed areas along the affected vein.[17] Edema initially manifests in the perimalleolar region and progresses upward along the leg due to dependent fluid accumulation. This swelling can evolve from pitting edema to more induration, resulting in increased hardness and stiffness of the tissues.[16,17]

Evaluation of the legs in patients with CVI may reveal skin changes such as hyperpigmentation, stasis dermatitis, erythema, eczema, and venous ulcers (VLU). Hyperpigmentation is thought to be caused by hemosiderin or melanin deposition due to venous hypertension. VLU represents a significant medical issue caused by CVI, driven by increased venous pressure, which may result from venous occlusion and/or varicose veins. The healing process for VLUs can span from a few months to several years, with approximately 25% remaining unhealed. These ulcers are the most prevalent chronic leg ulcers among the elderly population.[8] Advanced skin manifestations of CVI include atrophie blanche (ulcers with white scars), corona phlebectatica (widening of the superficial veins around the ankle), and ulceration around the medial and lateral malleolus.[15,17] Additionally, patients with CVI often exhibit lipodermatosclerosis, a fibrotic process affecting the dermis and subcutaneous fat, which is linked to an increased risk of cellulitis, leg ulceration, and delayed wound healing.[15,17]

Furthermore, CVI may lead to the development of lymphedema, with Stemmer's sign being a notable clinical feature of this condition. Approximately one-third of CVI cases result in secondary lymphedema, known as phlebolymphedema, which may improve or resolve with the correction of the underlying CVI.[16,17]

While the CEAP scoring system provides a valuable framework for classifying the severity of CVI, it does not offer a specific algorithm to definitively distinguish CVI from other conditions.[17] This lack of a precise diagnostic tool often leads to delayed diagnosis and suboptimal treatment. In the future, it is crucial to refine diagnostic algorithms to distinguish CVI from other conditions, especially those identifiable through straightforward physical examination methods. This focuses on distinctive, easily detectable signs could enhance for early diagnosis and treatment.

Conclusions

Chronic venous insufficiency is a prevalent but often under-recognized vascular disorder with significant clinical implications, including the development of venous ulcers and chronic leg swelling. This case study highlights the importance of early recognition, accurate diagnosis, and comprehensive management of CVI to prevent progression and complications. The patient's presentation, characterized by recurrent leg swelling and ulceration, underscores the progressive nature of untreated CVI. Effective management strategies, including the use of compression therapy, lifestyle modifications, and when necessary, advanced interventions like sclerotherapy or surgical procedures, are crucial in improving patient outcomes. Given the higher prevalence of CVI in women and those with risk factors such as obesity, targeted awareness, and proactive treatment approaches are essential to

reduce the burden of this condition. This study emphasizes the need for increased awareness among clinicians and patients alike to ensure timely intervention and to bridge the gap between seen and unseen cases of CVI.

Conflict of interest
The authors declare no conflicts of interest.

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Ethical statement
The work was carried out in accordance with the Declaration of Helsinki.

Informed consent
The patient gave consent to publishing of the case report.

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Surgical correction of partial anomalous pulmonary venous return in a patient with persistent left superior vena cava: cannulation techniques and challenges

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Technika kanulácie
Vrodená srdcová chyba

SÚHRN

Parciálny anomálny návrat pľúcnych žíl asociovaný so sínusovým venóznym defektom predsieňového septa a perzistujúcou ľavou hornou dutou žilou predstavuje významnú chirurgickú výzvu. Tieto vrodené anomálie si vyžadujú dôkladné plánovanie mimotelového obehu a stratégii venóznej kanulácie.
Tridsaťosemročná pacientka sa prezentovala s anamnézou supraventrikulárnej tachykardie a hemodynamicky významným ľavo-pravým skratom (Qp : Qs = 1,87) v dôsledku sínusového venózneho predsieňového septa a parciálny anomálny návrat pľúcnych žíl. Predoperačné zobrazovacie vyšetrenia preukázali drenáž pravej hornej pľúcnej žily do perzistujúcej ľavej hornej dutej žily. Pacientke bola úspešne vykonaná chirurgická korekcia metódou dvojitej náplasti s podporou mimotelového obehu, pričom sa využila komplexná stratégia kanulácie oboch horných dutých žíl. V pooperačnom období sa pacientka zotavila bez známok reziduálneho skratu alebo významnej chlopňovej dysfunkcie.
Prípad poukazuje na chirurgický prístup pri zriedkavej kombinácii vrodených anomálií a diskutuje alternatívne možnosti kanulácie pri mimotelovom obehu u pacientov s perzistujúcou ľavou hornou dutou žilou.

Keywords:
Cannulation technique
Congenital heart disease
Partial anomalous pulmonary venous return
Persistent left superior vena cava
Sinus venosus atrial septal defect
Surgical correction

ABSTRACT

Partial anomalous pulmonary venous return (PAPVR) associated with a sinus venosus atrial septal defect (SVASD) and a persistent left superior vena cava (PLSVC) presents significant surgical challenges. These anomalies require careful planning of cardiopulmonary bypass (CPB) and venous cannulation strategies.
A 38-year-old woman presented with a history of supraventricular tachycardia (SVT) and a hemodynamically significant left-to-right shunt (Qp : Qs = 1.87) due to SVASD and PAPVR. Preoperative imaging revealed drainage of the right upper pulmonary vein into the PLSVC. The patient underwent successful surgical correction using the double-patch technique with cardiopulmonary bypass support via a complex cannulation strategy involving both superior venae cavae. Postoperatively, the patient recovered well, with no evidence of residual shunt or significant valvular dysfunction.
The case highlights the surgical approach to a rare combination of anomalies and discusses alternative cannulation strategies for CPB in patients with PLSVC.

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Introduction

Sinus venosus atrial septal defect (SVASD) with partial anomalous pulmonary venous return (PAPVR) represents a rare congenital anomaly with significant implications for cardiac function. Its prevalence is up to 0.5% of the general population and 3% to 10% in patients with congenital heart disease.[1,2] These defects are often accompanied by additional venous anomalies, such as a persistent left superior vena cava (PLSVC), complicating the surgical approach. The double-patch technique, a well-established method for addressing PAPVR, becomes particularly relevant in such scenarios.[3] This case report describes the surgical management of a patient with these anomalies, emphasizing the challenges in cardiopulmonary bypass (CPB) setup and highlighting alternative venous cannulation options.

Case presentation

A 38-year-old woman presented with recurrent episodes of supraventricular tachycardia (SVT), diagnosed as atrioventricular nodal reentrant tachycardia (AVNRT) in 2005 and 2023. She had no significant comorbidities but reported exertional dyspnea and occasional chest pressure. Transthoracic echocardiography (TTE), transesophageal echocardiography (TEE), and cardiac CT confirmed a sinus venosus defect (20 × 15 mm) with associated PAPVR. Imaging revealed anomalous drainage of the right upper pulmonary vein into the right superior vena cava (RSVC) and partial drainage into the left atrium (LA). Additionally, a persistent left superior vena cava (PLSVC) was identified, draining into the coronary sinus (**Fig. 1**). Hemodynamic studies showed a left-to-right shunt with Qp : Qs = 1.87, with preserved systolic function and no pulmonary hypertension. On October 31, 2024, the patient underwent surgical correction. Cannulation for CPB was performed using separate cannulas for the RSVC and PLSVC

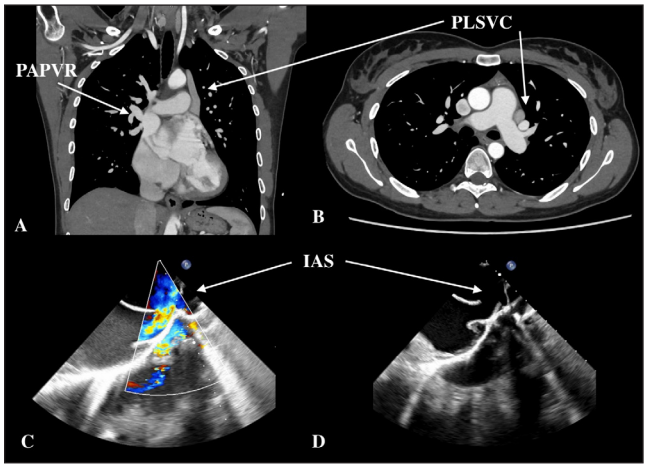

Fig. 1 – (A) Computed tomography coronal view showing the partial anomalous pulmonary venous return. (B) Computed tomography axial view showing the persistent left vena cava superior. (C, D) Transesophageal images showing the interatrial septum. IAS – interatrial septum; PAPVR – partial anomalous pulmonary venous return; PLSVC – persistent left vena cava superior.

to ensure effective drainage, and antegrade cardioplegia was administered. A Foley catheter was temporarily placed into the coronary sinus for PLSVC drainage. The double-patch technique was employed to redirect the anomalous pulmonary venous flow to the left atrium. The sinus venosus defect was closed with a pericardial patch. Intraoperative transesophageal echocardiography confirmed no residual shunt and unobstructed flow through the reconstructed venous pathways.

The patient experienced an uneventful recovery. Echocardiography prior to discharge showed preserved left and right ventricular function, absence of residual atrial septal defects, and no significant valvular dysfunction. She was discharged on the ninth postoperative day in a stable condition.

Discussion

SVASD with PAPVR and PLSVC represents a unique surgical challenge due to the anatomical complexity of venous drainage and potential difficulties in establishing adequate venous return during CPB. The presence of a PLSVC often necessitates modifications to the standard cannulation strategy, as the anomalous vessel may impede venous drainage if not properly addressed.

PLSVC is the most common venous anomaly of the thoracic circulatory system, with an incidence of 0.5% of the general population, and in 90% of the cases it is drained into the coronary sinus, like our patient. These patients remain asymptomatic and the diagnosis is mostly an incidental finding. Only in 10% of the patients it is drained into the left atrium, resulting in a right-to-left shunt, with systemic hypoxemia and an increased risk of paradox embolization.[4] Its presence requires some consideration in the management of myocardial protection and intraoperative conduct. It is a contraindication to retrograde cardioplegia, since the flow may result in cardioplegia washout. In order to overcome the aforementioned problems, PLSVC can be momentarily occluded in case the anonymus vein is present. However, even antegrade cardioplegia may fail, due to the steal effect by the hemiazygos venous system linked to PLSVC.[5] In our case, antegrade cardioplegia was used without any complications.

It is more common in patients undergoing surgical procedures congenital heart disease, and if it is suspected, computed tomography (CT), magnetic resonance imaging (MRI), venous angiography or a digital substraction venogram can be performed for the correct diagnosis.6 Moreover, TTE and TEE can be performed for the diagnosis, but sometimes these studies do not necessarily reveal a PLSVC that drains to the left atrium unless a bubble study with agitated saline is performed. In some cases where the PLSVC is identified intraoperatively, the cannulation technique should be reconsidered and managed accordingly.[4]

In our case report, the PLSVC was identified preoperatively by CT so the cannulation strategy and planning were decided before the operation.

Different cannulation techniques for the PLSVC are proposed. Direct cannulation of the PLSVC is an effective method and maybe feasible when properly visualized in-

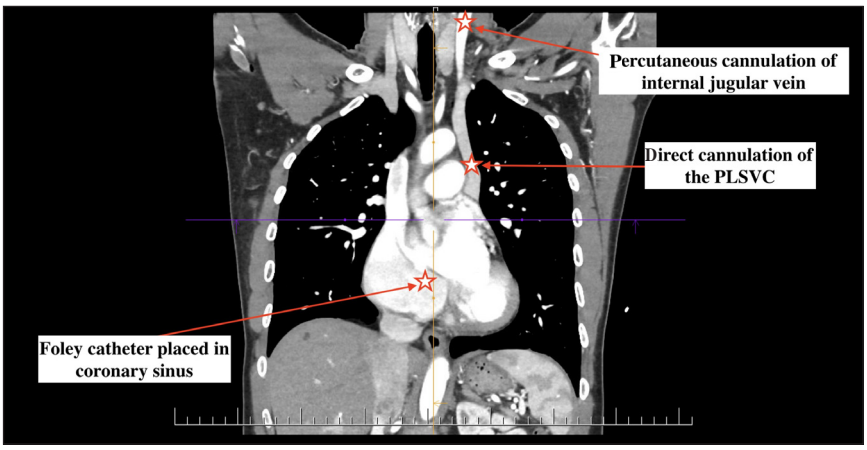

Fig. 2 – Proposed cannulation techniques.

side the pericardium. Based in our experience, we propose using Foley catheter placed in coronary sinus to be used for venous drainage during CPB. This method is simple, can be performed easily without any complications, and can provide adequate drainage of the PLSVC. Another method as proposed by G. Bianchi et al[5] is a hybrid cannulation by direct percutaneous cannulation of internal jugular vein. The proposed cannulation techniques are shown in **Figure 2**.

The surgical correction using the double-patch technique is safe approach for addressing PAPVR with associated SVASD. This technique allows effective redirection of anomalous pulmonary venous flow to the left atrium while preserving normal venous pathways. The main advantage of the two-patch technique is the lower incidence of SVC stenosis on long-term follow-up, however, as many as 55% may have sinus node dysfunction while incising the SVC anteriorly. This may be directly related to the sinus node itself, as well as its supply artery.[7] The patient's postoperative rhythm disturbances, including junctional rhythm, were transient and resolved with supportive care. This highlights the importance of vigilant postoperative monitoring, particularly in patients with preoperative arrhythmias.

Conclusion

This case underscores the importance of individualized surgical planning for complex congenital cardiac anomalies. The successful use of the double-patch technique, combined with a tailored cannulation strategy, ensured optimal outcomes. Awareness of alternative CPB cannulation approaches is crucial for addressing anatomical variations, such as PLSVC, and minimizing intraoperative complications.

Conflict of interest

The authors declare no conflict of interest.

Funding

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Ethical statement

This case report was conducted in accordance with the principles of the Declaration of Helsinki. The authors affirm that all methods were carried out in accordance with relevant guidelines and regulations.

Informed consent

The patient provided an informed consent to present this case report.

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Thoracic aortic aneurysm with an aberrant right subclavian artery and truncus bicaroticus

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Kľúčové slová:
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Debranching aorty
TEVAR

SÚHRN

Úvod: Arteria lusória, alebo aberantná pravá arteria subclavia (ARSA), je anomália supraaortálnych vetiev aorty s výskytom 0,2 – 1,7 percenta. U 8 % pacientov s ARSA sa rozvinie aneuryzma hrudnej aorty, v porovnaní s výskytom 4,4 % v populácii bez anomálie aortálneho oblúka.
Kazuistika: Kazuistika prezentuje 56-ročného muža bez pridružených komorbidít, ktorý mal anomáliu aortálneho oblúka pozostávajúcu z truncus bicaroticus a aberantnej pravej arteria subclavia bez Komerellovho divertikla, ktorá mala retroezofageálny priebeh, v spojení s rozsiahlo kalcifikovanou sakulárnou aneuryzmou zostupnej hrudnej aorty. Stav bol diagnostikovaný ako náhodný nález. Pacient podstúpil multidisciplinárnu dvojstupňovú operáciu skladajúcu sa z debranchingu aorty a extraanatomického bypassu, nasledovaného uzatvorením ARSA a implantáciou TEVAR za účelom exklúzie aneuryzmy.
Diskusia a záver: Pacient bol prepustený na 15. pooperačný deň v dobrom stave. Hybridné prístupy v chirurgickej liečbe anomálií aortálneho oblúka ponúkajú bezpečnú a efektívnu terapeutickú možnosť.

Keywords:
Aberrant right subclavian artery
Aortic debranching
Arteria lusoria
TEVAR

ABSTRACT

Introduction: Arteria lusoria, or aberrant right subclavian artery (ARSA), is an anomaly of origin of the supra-aortic branches with an estimated incidence of 0.2–1.7%. 8% of these patients with ARSA go on to develop a thoracic aortic aneurysm, in contrast to 4.4% of the population without an aortic arch anomaly.
Case report: This case report presents a 56-year-old male, without associated comorbidities, who presented with an aortic arch anomaly composed of a truncus bicaroticus with a concomitant aberrant right subclavian artery without Komerell diverticulum and with a retroesophageal course, in association with an extensively calcified saccular aneurysm of the descending thoracic aorta, found incidentally on imaging performed for another reason. The patient underwent a multidisciplinary 2-stage operation consisting of an aortic debranching and extra-anatomical bypass followed by Amplatzer occlusion of ARSA and TEVAR exclusion of the aneurysm.
Discussion and conclusion: The patient was finally discharged on the 15th post-operative day in good condition. Hybrid approaches in surgical treatment of aortic arch anomalies offer a safe and effective therapeutic option.

Introduction

Arteria lusoria, or aberrant right subclavian artery (ARSA), is an anomaly of origin of the supra-aortic branches with an estimated incidence of 0.2–1.7%.[1] It was first documented by Hunauld in 1735, while David Bayford reported the initial case of dysphagia lusoria in 1794. In its most classical form, the right subclavian artery arises as a separate branch from the aortic arch distal to the left subclavian artery (G-1 subtype), but several distinct anatomical configurations have led to the development of the Adachi–Williams classification. Another possible configuration includes both ARSA and truncus bicaroticus (H-1 subtype) (**Fig. 1**), with an incidence of 28% among all cases of ARSA.[2] In this particular subtype, the ARSA occurs in conjunction with a common trunk that gives rise to both common carotid arteries.

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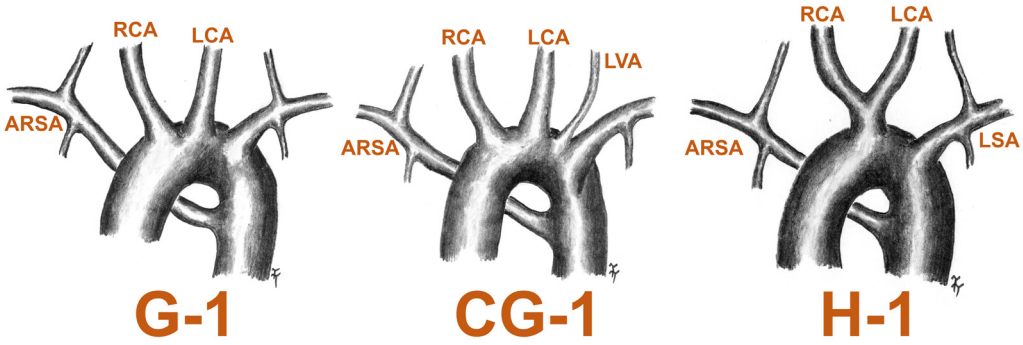

Fig. 1 – Adachi–Williams classification of arteria lusoria subtypes. ARSA – aberrant right subclavian artery; LCA – left carotid artery; LSA – left subclavian artery; LVA – left vertebral artery; RCA – right carotid artery.

In normal development of the right subclavian artery, this blood vessel is ultimately formed by the fusion of the right fourth aortic arch, the right proximal dorsal aorta, and the right seventh intersegmental artery. In addition, involution of the right distal dorsal aorta ultimately gives the right brachiocephalic artery its normal course.[4] In the case of ARSA, regression of the right fourth vascular arch and the proximal right dorsal aorta, as well as the continuation of the seventh intersegmental artery from the right distal descending thoracic aorta, leads to the arteria lusoria taking its aberrant path (**Fig. 2**).[5]

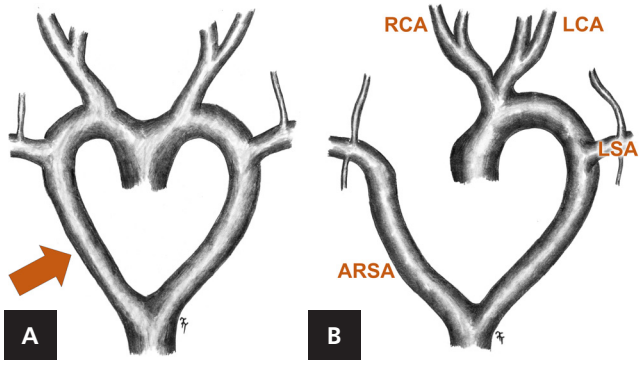

Fig. 2 – Depiction of embryonic development of ARSA. (A) Normal development of the aortic arch according to the double aortic arch hypothesis by Edwards and (B) degeneration of the right 4th aortic arch and proximal right dorsal aorta leading to ARSA. ARSA – aberrant right subclavian artery; LCA – left carotid artery; LSA – left subclavian artery; RCA – right carotid artery.

Due to the persistence of the distal right dorsal aorta, in conjunction with the right 7th intersegmental artery, the ARSA courses retroesophageally in most cases (80%). In other instances, it may course between the trachea and esophagus (15%) or pretracheally (5%).[6] Although most patients with ARSA remain asymptomatic lifelong, the abnormal vascular course may compress the esophagus and the trachea, leading to a myriad of symptoms resulting from compression of intramediastinal structures. Compression of the esophagus may lead to symptoms of dysphagia and pyrosis, classically described as dysphagia lusoria, whereas tracheal compression may lead to dyspnoea and chronic cough. In addition, compression of intramediastinal structures may also present as retrosternal pain.[7] This abnormal blood vessel is frequently linked with additional irregularities, like the non-recurrent laryngeal nerve and the bicarotid trunk, and with conditions such as aneurysms, congenital heart abnormalities, and even genetic syndromes.[8]

Anomalies of the aortic arch, apart from leading to possible symptoms of compression, have also been proven to be associated with a higher occurrence of thoracic aortic aneurysms, which is the main interest of this study. As a general rule, research concluded that aortic arch anomalies are associated with a threefold increase in thoracic aortic aneurysms, but with distinct levels of association according to each different aortic arch anomaly. In the case of left-sided aortic arch with ARSA, 8% of patients develop a thoracic aortic aneurysm, in contrast to 4.4% of the population without an aortic arch anomaly.[9] The underlying reason for this association, either alterations of CT tissue elements in patients with congenital arch anomalies or abnormal flow due to abnormal aortic arch configurations causing aneurysm formation, remains elusive.

Descending thoracic aortic aneurysms are particularly dangerous due to their usual progression, which advances from asymptomatic to sudden aortic dissection or aortic rupture. This is especially true for saccular thoracic aneurysms, which are generally treated invasively at smaller diameters due to a perceived belief by surgeons of higher propensity for rupture and dissection.[10]

Case report

We report the case of a 56-year-old obese male, without associated comorbidities, who presented with an aortic arch anomaly composed of a truncus bicaroticus with a concomitant aberrant right subclavian artery without Komerell diverticulum and with a retroesophageal course, in association with an extensively calcified saccular aneurysm of the descending thoracic aorta, found incidentally on imaging performed for another reason. The patient was completely asymptomatic and finally underwent aortic debranching and TEVAR after a decision from the vascular surgery team due to the large size of the descending aortic aneurysm, which was our main concern from a surgical standpoint.

According to measurements from a CT angiography, the saccular aneurysm was located at the transition point

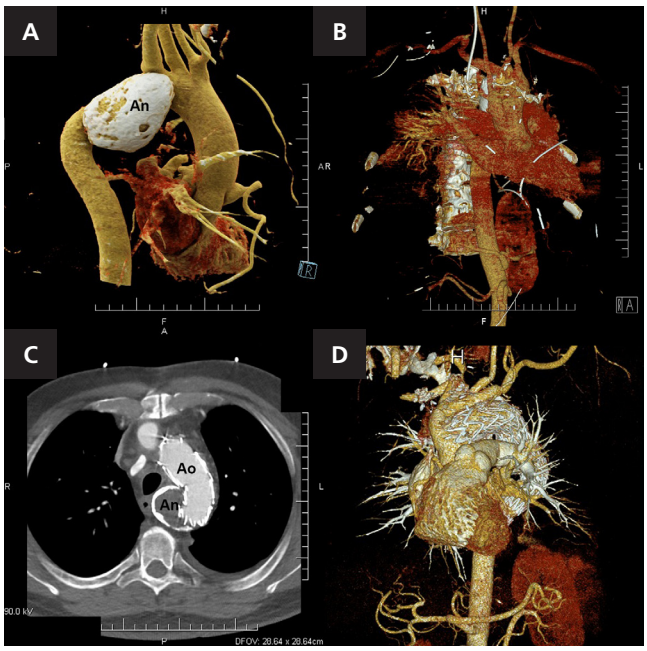

Fig. 3 – CT angiography images depicting the course of treatment. (a) Pre-operative 3D-reconstructed image, dorsal view, (B) 3D-reconstructed image after aortic debranching and before TEVAR implantation, ventral view, (C) image after TEVAR implantation, axial view and (D) 3D-reconstructed image after TEVAR implantation, ventral view. An – aneurysm; Ao – aorta.

from the distal aortic arch to the descending thoracic aorta, just beyond the origin of the ARSA (**Fig. 3**). The neck of the saccular aneurysm measured 45mm in width, and the maximum size of the aneurysm sac, facing dorsolaterally to the right, measured 43 × 52 × 74 mm. The wall of the aneurysm, apart from being extensively calcified, also slightly compressed the trachea and the esophagus, causing slight dislocation of these structures towards the right side without any symptoms of dysphagia or dyspnea. The proximal anchoring zone was 4–5 mm wide, and the descending aorta was 2.2 x 2.3 cm wide.

In this particular case, the surgical team opted for a 2-stage operation consisting of a surgical debranching plus TEVAR exclusion of the descending aortic aneurysm in order to avoid surgical supra-aortic branch and descending thoracic aorta reconstruction.

In the first operative stage, an aortic debranching technique with 3 vessel anastomoses and a trans-pleural extra-anatomical bypass to the distal limb of the arteria lusoria was performed completely off-pump. Using a 4-limbed Dacron graft (**Fig. 4B**), the proximal anastomosis to the ascending thoracic aorta using a side-clamp was done first, followed by distal anastomoses of the individual carotid arteries (aorta-right common carotid and aorta-left common carotid bypasses) and aorta-left subclavian artery bypass in order of left to right (**Fig. 4A**). Localisation of also the ARSA in an effort to perform a 4th distal anastomosis was attempted, but its deep intramediastinal course made it unfeasible to debranching. Due to this surgical unfeasibility, the arteria lusoria was localized within the right pleural cavity by means of a right infraclavicular approach, and finally an extra-anatomical, trans-pleural bypass using a Dacron graft from the ascending thoracic

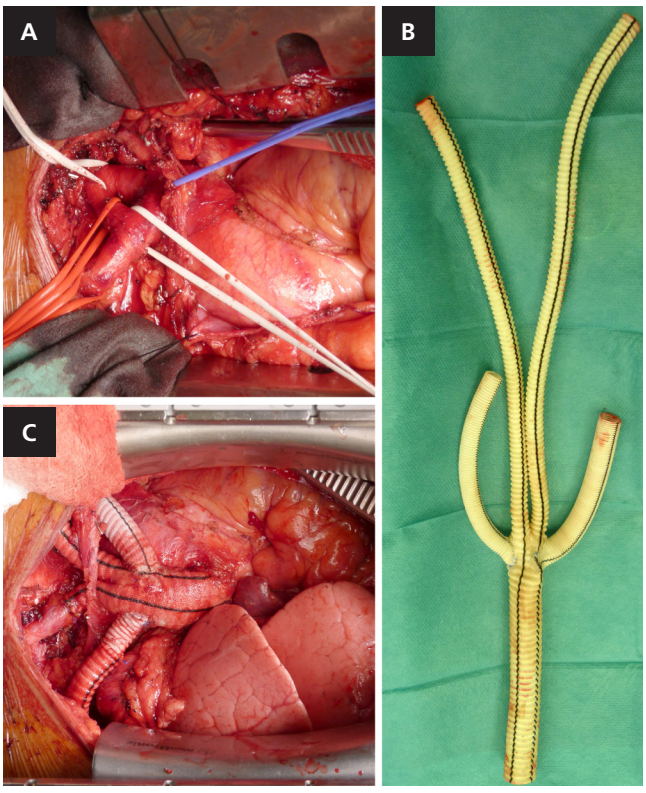

Fig. 4 – Images depicting surgical course of the first stage of treatment. (A) Intra-operative division of the supra-aortic branches using vessel loops, (B) 4-limbed Dacron graft used to construct the anastomoses and (C) post-anastomotic result of aortic debranching plus trans-anatomical bypass.

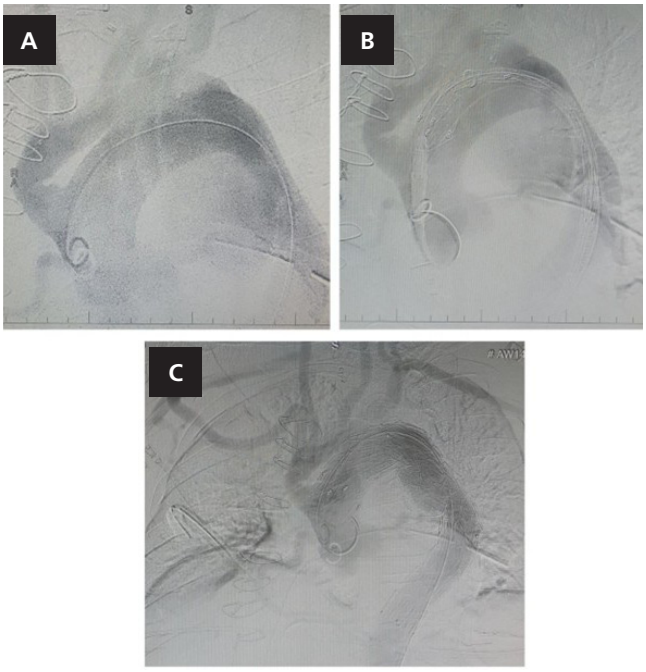

Fig. 5 – Images depicting course of the second stage of treatment under DSA guidance. (A) Pigtail catheter advancement into the ascending aorta, (B) stent graft advancement into the aortic arch, (C) final TEVAR result after stent graft deployment and ARSA occlusion.

aorta to the arteria lusoria, which provided retrograde blood flow to it, was performed (**Fig. 4C**).

In the second stage, an occlusion of the ARSA with an Amplatzer occluder followed by TEVAR of the descending thoracic artery with a Valiant thoracic stent graft was performed (**Fig. 5**). The reasoning behind occluding the ARSA with an Amplatzer occluder first was to prevent any possible type 2a endoleak, which could potentially develop with time after stent graft deposition, from the retrograde blood flow arising from the extra-anatomical bypass due to the proximity of the aneurysm to the ARSA.

Discussion

When posed with the surgical challenge of managing a descending thoracic aneurysm with this particular aortic arch configuration, 3 distinct surgical strategies are possible and must be kept in the armamentarium of the surgical team: on one hand, it is possible to perform a purely surgical replacement of the aneurysmatic descending thoracic aorta; another possibility is to perform a 2-stage hybrid approach consisting of an aortic debranching followed by endovascular aneurysm exclusion by TEVAR; and a final possibility is a purely endovascular approach. Purely endovascular repair refers to the use of fenestrated stent grafts, snorkel techniques, chimney grafts, or laser fenestration to preserve aortic branch perfusion.[11]

When choosing to perform a purely surgical descending thoracic aortic replacement, it is imperative to bear in mind that due to the proximity of the ARSA, the left subclavian artery, and possibly the truncus bicaroticus, it may be necessary to reconstruct these branches using a branched conduit, possibly necessitating hypothermic circulatory arrest. In addition to this, surgical experience with this procedure is imperative, as research has proven the overall in-hospital mortality of thoracic aortic surgery to be catastrophic in low-volume centers (21.9% in low-volume centers vs. 8.6% in high-volume centers).[12] Another technical factor that was classically believed to favor TEVAR was post-operative risk of paraplegia after open surgery, which has decreased due to the introduction of neuroprotective methods such as minimization of spinal ischemic time, reimplantation of spinal arteries (particularly at the T8–L1 level), drainage of the cerebrospinal fluid, regional hypothermia of the epidural space, intra-operative monitoring of evoked potentials, and use of left-sided heart bypass.[13] Thanks to the implementation of these neuroprotective methods, it is estimated that the rates of post-procedural paraplegia are similar between open surgery and TEVAR.[14,15]

On the other hand, when opting for a hybrid 2-stage approach, several factors must be considered. Among these, the possibility of having to reintervene due to the limited durability of stent grafts or possible stent graft complications such as endoleaks and the fact that technically open surgery is considered the main method of treatment to this date due to the lack of long-term follow-up on patients after TEVAR because of its relative novelty stand out. Another technical factor to bear in mind is that in order for the TEVAR procedure to be possible, a suitable anatomy is required, as unfavorable anatomy is considered the main contraindication for TEVAR. This means that the femoral and iliac arteries cannot have a high degree of tortuosity that could pose an impediment to the advancement of the catheter, and that a suitable landing zone proximally and distally to the aneurysm is necessary to ensure proper fixation of the stent graft, as otherwise, a type 1 endoleak could arise.[11]

Joo et al performed a study comparing outcomes of open surgery vs. a hybrid approach in the treatment of descending thoracic aortic aneurysms and concluded that open repair had greater reliability due to higher freedom from reintervention, whereas hybrid repair had similar perioperative mortality but decreased pulmonary complications. The difference in in-hospital mortality between open repair (10.1%) and hybrid repair (6.5%) was demonstrated to be statistically insignificant. The difference in the occurrence of paraplegia between open repair (2.5%) and hybrid repair (0.0%) was also proven insignificant. However, freedom from reintervention at 10 years showed a marked improvement for open repair (85.2%±7.1%) as opposed to hybrid repair (46.3%±11.0%).[16]

Conclusion

In conclusion, due to a demonstrated relationship in occurrence between congenital aortic arch anomalies and thoracic aneurysms, it is imperative that the surgical team be aware of the anatomy of the aortic arch and the surgical armamentarium available to correct such defects. This case report proves that a hybrid approach consisting of an aortic debranching plus a trans-pleural extra-anatomical bypass to the ARSA, followed by a TEVAR exclusion of the descending thoracic aneurysm, is a suitable strategy for the approach of patients with a descending saccular thoracic aneurysm in association with an arteria lusoria with truncus bicaroticus. With this case report, we not only reiterate the association between aortic arch anomalies and thoracic aorta aneurysm formation, but we focus especially on new and applicable trends in cardiac surgery for the management of congenital aortic arch anomalies.

Conflict of interest
The authors declare no conflict of interest.

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Ethical statement
The case report was conducted in accordance with the Declaration of Helsinki and approved by the Ethics Committee of East Slovak Institute for Cardiovascular Diseases (protocol code A5012025; date of approval: 7 January 2025).

Informed consent
The patient signed informed consent for examinations, hospitalization, and surgery. Due to the retrospective character of the manuscript, the ethics committee did not indicate the need for obtaining additional consent for publication. Informed consent is available for the editor.

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A Rare Case of Carotidynia: Aneurysm and Kinking of the Left Internal Carotid Artery

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Surgical management

SOUHRN

Chronická bolest v oblasti krku může mít různou etiologii včetně karotidynie, což je poměrně vzácný stav charakterizovaný bolestí v oblasti nad karotickou tepnou, u níž se předpokládá původ v postižení cév. V této kazuistice popisujeme případ 47leté pacientky s chronickou bolestí v oblasti krku, u níž byla stanovena diagnóza aneurysmatu a zalomení levé vnitřní karotické tepny. Byla provedena úspěšná chirurgická rekonstrukce bez neurologických komplikací. Tato kazuistika upozorňuje na význam časné diagnostiky a odpovídajícího chirurgického řešení přítomnosti aneurysmatu a zalomení levé vnitřní karotidy v léčbě karotidynie.

ABSTRACT

Chronic neck pain can be attributed to various etiologies, including carotidynia, a relatively rare condition characterized by pain in the region overlying the carotid artery attributed to underlying vascular pathologies. In this case report, we present the case of a 47-year-old female patient who experienced chronic neck pain and was diagnosed with left internal carotid artery aneurysm and kinking. Surgical reconstruction was performed, resulting in a successful outcome without neurological complications. This case highlights the importance of early diagnosis and appropriate surgical intervention of an aneurysm and kinking of the left internal carotid artery for the management of carotidynia.

Introduction

Chronic neck pain is a prevalent symptom encountered in clinical practice, often attributed to musculoskeletal or cervical spine pathology. However, in some cases, neck pain can be attributed to carotidynia, a rare condition characterized by pain in the carotid artery region. The etiology of carotidynia necessitates a thorough investigation of the region to rule out vascular diseases such as carotid dissection, stenosis, or occlusion, as its underlying causes and management remain poorly understood.[1]

Aneurysms and kinking of the carotid arteries are infrequent yet intriguing vascular disorders that typically manifest as parapharyngeal pulsatile masses.[2] They may exhibit partial or complete thrombosis, consequently leading to embolization or compression of neural vasculature, with additional complications including ruptures and ischemic events. Consequently, the mortality rate among non-operated patients is notably elevated.[3] The primary etiological factor implicated in this condition is atherosclerosis, with occasional instances attributed to trauma. Surgical intervention is strongly advised for symptomatic patients across all age groups.[4]

This case report presents a unique case of carotidynia in a 47-year-old female patient and describes the successful surgical intervention for the associated left internal carotid artery aneurysm and kinking, emphasizing the importance of accurate diagnosis and appropriate management in such intriguing clinical scenarios.

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Case report

A 47-year-old female patient presented with chronic, debilitating neck pain that had persisted for an extended duration. The pain was localized to the region overlying the left carotid artery. The patient described the discomfort as dull and aching, exacerbated by neck movement and palpation over the left carotid artery region. She had no associated neurological symptoms, such as weakness or sensory changes. Initial clinical evaluation revealed a tender and palpable swelling overlying the left carotid artery. The patient's medical history was unremarkable, with no history of trauma, recent infections, or inflammatory conditions. Her family history did not include any vascular disorders. Routine laboratory tests, including inflammatory markers, were within normal limits.

Given the atypical presentation of chronic neck pain and the localized tenderness over the left carotid artery, further imaging studies were pursued to investigate the underlying pathology. A Doppler USG indicated enlargement of the left carotid artery. A CT angiography was performed, which unveiled two significant findings: left internal carotid artery aneurysm measuring 2.5 cm in diameter as well as kinking and superficial pouching of the vessel (**Fig. 1**). These findings explained the patient's chronic neck pain, ultimately leading to the diagnosis of carotidynia attributed to a left internal carotid artery aneurysm and kinking.

Upon identifying the pathology, the necessity for surgical intervention was established to alleviate the patient's pain and mitigate potential complications associated with the left internal carotid artery aneurysm and kinking. The patient was informed about the risks and benefits of the treatment in detail, and upon receiving consent the patient was scheduled for surgery.

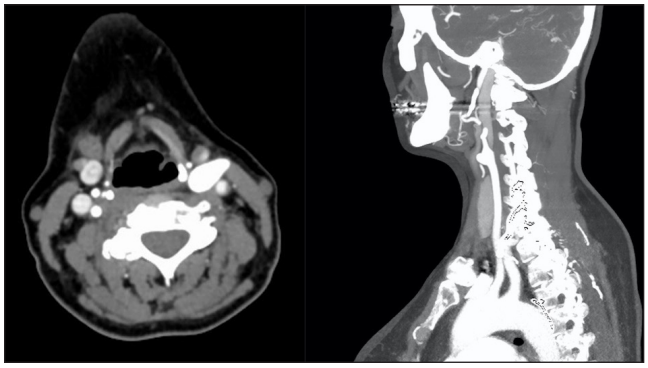

Fig. 1 – Preoperative CT angiography indicating carotid aneurysm. Note the corresponding contralateral carotid artery sizes.

Surgical management

Under regional anesthesia, the operation was initiated with a linear incision to the left cervical region anterior to the sternocleidomastoid muscle. The left facial vein was prepared for use as a patch. The left common carotid artery (CCA), internal carotid artery (ICA), and external carotid artery (ECA) were explored (**Fig. 2**), and the vessels were temporarily clamped using tape. A local anesthetic was injected into the carotid sinus to ameliorate perioperative hemodynamic instability. Following heparinization, the left ICA was clamped, and the patient's neurological functions were examined which remained stable. Following confirmation of absence in neurological and motor deficits, the CCA, and ECA were clamped as well. The aneurysmal segment of the left ICA was resected at

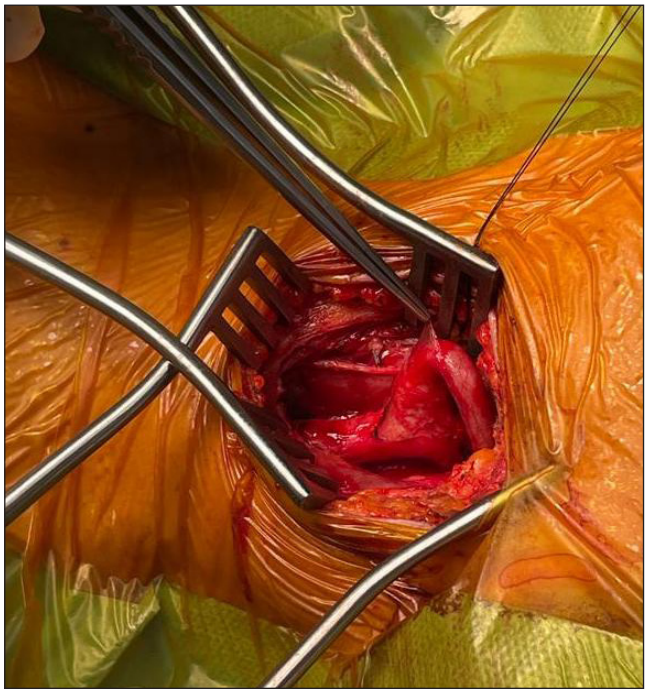

Fig. 2 – Perioperative view of the dilated and kinked carotid artery.

Fig. 3 – The arterial anatomy following resection of the aneurysm, reconstruction of the common carotid artery, proximal and lateral re-implantation of the internal carotid artery to the common carotid with venous patch augmentation.

the point of bifurcation. The CCA was primarily closed with Prolene sutures. An atherosclerotic segment with intimal hyperplasia was present at the orifice of the left ICA which was resected. The area where the ICA would be anastomosed to the CCA was prepared with a punch. During the end-to-side anastomosis of the left ICA to the CCA, the facial vein was employed as a patch to augment the arteriotomy (**Fig. 3**). This approach served as a prophylactic measure to prevent restenosis. The clamps were removed, and no neurological deficit was observed. The operation was concluded following appropriate surgical protocol. Postoperative imaging confirmed adequate blood flow through the reconstructed carotid artery, and the patient reported significant relief from her neck pain. The success of this surgical intervention was evident, as the patient was discharged on the second day post-surgery without any neurological deficits.

Discussion

Carotidynia, originally described by Temple Fay in 1927, is characterized by localized neck pain, often elicited by palpation over the carotid artery, without any clear signs of inflammation or infection. It remains a diagnosis of exclusion, as there are no specific diagnostic criteria, and its pathophysiology is not fully understood. The challenge in diagnosing carotidynia lies in differentiating it from more sinister conditions such as atherosclerotic disease or carotid artery dissection.[5] In our case, the diagnostic workup with a CT angiography revealed a 2.5 cm left internal carotid artery aneurysm and kinking. These imaging findings provided a clear explanation for the patient's persistent neck pain. This unique presentation highlights the importance of considering vascular etiologies in the evaluation of chronic neck pain, especially when accompanied by unusual or focal symptoms.

Kinks in the internal carotid artery can manifest as acquired or developmental anomalies. A common cause is the excessive length of the carotid artery which may result in redundancy and tortuosity of the vessel.[6] Notably, kinks are observed to be four times more prevalent in women than in men. However, the precise incidence remains uncertain due to many patients remaining asymptomatic. Symptoms associated with kinks in the internal carotid artery tend to parallel those of atherosclerotic diseases affecting the carotid artery, encompassing conditions such as transient ischemic attacks, strokes, and amaurosis.[7] The underlying pathophysiology often revolves around hemodynamic and structural abnormalities within the kinked carotid artery, leading to turbulent flow, intimal ulcerations, and subsequent embolization.[8]

Most cases of internal carotid artery aneurysms occur spontaneously, with atherosclerosis serving as the primary etiological factor, or they are a consequence of blunt trauma affecting the neck region and the high cervical segment of the internal carotid artery.[9] Blunt or nonpenetrating trauma, although more commonly associated with thrombosis of the injured vessel, can also give rise to the development of a false aneurysm. Such blunt injury to the carotid artery can lead to various abnormalities, including spasms, intimal and medial tears, dissection, and partial or complete severance of the vessel.[10] Disruption of the arterial wall's continuity emerges as the primary causal factor in the formation of an aneurysm.

Internal carotid artery aneurysms exhibit the potential to undergo partial or complete thrombosis, leading to adverse consequences such as distal embolization, compression of adjacent structures, or even rupture.[11] Consequently, patients managed conservatively face a notably elevated mortality and morbidity rate. Nevertheless, surgical intervention plays a pivotal role in averting permanent neurologic deficits arising from thromboembolism or atheroembolism. Employing advanced vascular surgical techniques, the mortality rates associated with surgical treatment are now less than 2%.

The choice of surgical technique is essential in ensuring optimal patient's outcomes. The preferred surgical treatment approach for an aneurysm of carotid arteries involves the resection of the aneurysm with subsequent restoration of blood flow. When feasible, the optimal method for addressing an aneurysmal carotid pathology is internal carotid re-implantation. Other surgical alternatives encompass resection coupled with saphenous vein interposition, internal carotid re-implantation into the common carotid artery, and extended patch angioplasty. Notably, there has been a growing utilization of endoluminal stents and stented grafts in recent times for the management of these lesions.[12]

In our case, surgical intervention was deemed necessary to address the underlying vascular abnormalities. The chosen surgical approach involved the resection of the left internal carotid artery aneurysm at the point of bifurcation, followed by the pull-down and end-to-side anastomosis of the left internal carotid artery to the common carotid artery. Additionally, patch augmentation using the facial vein was performed as a prophylactic measure to prevent restenosis. This approach aims to address both the aneurysm and the kinking of the common carotid artery, reducing the risk of complications.

While the etiology of carotidynia remains elusive, cases such as ours demonstrate the clinical significance of thorough diagnostic evaluation, particularly in atypical presentations of neck pain. This case underscores the importance of early diagnosis and appropriate surgical intervention in managing carotidynia when underlying vascular abnormalities are identified. Although our case had a successful outcome, it is crucial to recognize that carotidynia is a heterogeneous condition, and treatment strategies may vary depending on the individual patient's presentation and the underlying vascular pathology. Further research is needed to establish standardized diagnostic criteria and treatment guidelines for carotidynia.

Conclusion

This case report emphasizes the need for a comprehensive evaluation of chronic neck pain, including consideration of vascular etiologies including aneurysms and kinking. Early diagnosis and appropriate surgical intervention, as demonstrated in this case, can lead to favorable outcomes and improved quality of life for affected individuals. Further research and collaboration are essen-

tial to advance our understanding of this rare condition and refine treatment strategies for optimal patient care.

Conflict of interest
None.

Funding
None.

Ethical statement
The work was carried out in compliance with the Declaration of Helsinki.

Informed consent
The patient provided written informed consent for the publication of this case report and any accompanying images. All identifying information has been removed to protect the patient's privacy and confidentiality, in accordance with the Declaration of Helsinki.

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From weak knees to a troubled heart – a case of severe autonomic dysfunction in Guillain–Barré syndrome

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Klíčová slova:
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Autonomní dysfunkce
Guillainův–Barrého syndrom

SOUHRN

Guillainův–Barrého syndrom (GBS) je autoimunitní zánětlivé onemocnění postihující periferní nervový systém, jehož rozvoji často předchází infekce respiračního nebo gastrointestinálního traktu. Závažnou komplikací je dysfunkce autonomních systémů, zvláště kardiovaskulárního systému, projevující se kolísáním krevního tlaku, arytmiemi a změnami na EKG záznamu. K rozvoji infarktu myokardu dochází vzácně; jeho diagnostika v dané situaci je obtížná.

Popisujeme případ 67letého muže s nedávno prodělanou infekcí horních cest dýchacích, který byl dopraven k lékaři s progredující slabostí dolních končetin. Rychle u něj došlo k rozvoji tetraparezie, dysfagie, respiračního selhání a akutního otoku plic, doprovázených změnami na EKG záznamu odpovídajícími akutnímu koronárnímu syndromu, se zvýšenými hodnotami troponinu. Koronarografické vyšetření nicméně nepotvrdilo obstrukční postižení, zatímco echokardiografické vyšetření prokázalo tranzitorní dysfunkci levé komory.

Pacientovi byl intravenózně aplikován imunoglobulin a současně byla přijata intenzivní podpůrná opatření. Neurologické vyšetření a vyšetření mozkomíšního moku později potvrdily GBS. Průběh onemocnění se prodloužil a zkomplikoval se rozvojem nozokomiálních infekcí, u pacienta však došlo k částečnému zotavení z neurologického hlediska a k úplné obnově systolické funkce levé komory srdeční.

Popsaný případ upozorňuje na význam důsledného monitorování kardiovaskulární funkce u pacientů s GBS, protože dysfunkce autonomních systémů může mít podobné projevy jako akutní koronární syndrom a může významně ovlivnit prognózu.

Keywords:
Acute coronary syndrome
Autonomic dysfunction
Guillain–Barré syndrome

ABSTRACT

Guillain–Barré Syndrome (GBS) is an autoimmune inflammatory disease of the peripheral nervous system, often preceded by respiratory or gastrointestinal infection. A serious complication is autonomic dysfunction, particularly cardiovascular, manifesting as blood pressure fluctuations, arrhythmias, and electrocardiographic changes. Myocardial infarction is rare and its diagnosis may be challenging in this setting.

We report the case of a 67-year-old man with a recent upper respiratory infection who presented with progressive weakness of the lower limbs. He rapidly developed tetraparesis, dysphagia, respiratory failure, and acute pulmonary edema, accompanied by electrocardiographic changes consistent with acute coronary syndrome and elevated troponin. However, coronary angiography revealed no obstructive disease, while echocardiography showed transient left ventricular dysfunction.

The patient was treated with intravenous immunoglobulin and intensive supportive measures. Neurophysiological and cerebrospinal fluid studies later confirmed GBS. His course was prolonged, complicated by nosocomial infections, but he achieved partial neurological recovery and full recovery of left ventricular systolic function. This case highlights the importance of careful cardiovascular monitoring in patients with GBS, as autonomic dysfunction may present with acute coronary syndrome-like features and significantly influence prognosis.

Introduction

Guillain–Barré Syndrome (GBS) is an inflammatory autoimmune disease that affects the peripheral nervous system, characterized by symmetric ascending paralysis and areflexia of sudden onset, generally preceded by a respiratory or gastrointestinal infection in the weeks before.[1,2] A common and serious complication of GBS is autonomic dysfunction, which frequently involves the cardiovascular

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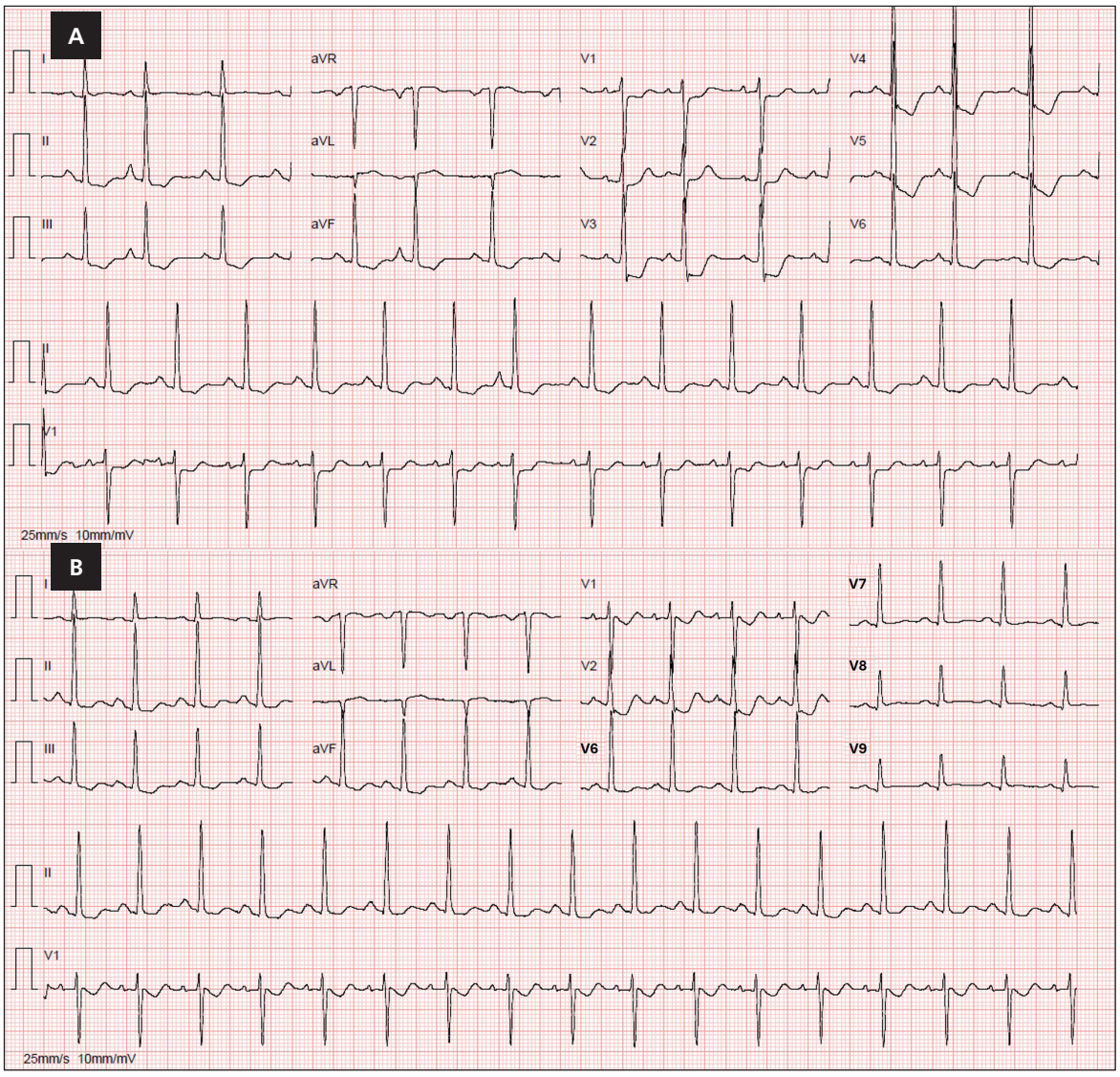

Fig. 1 – ECG during acute pulmonary edema. (A) sinus rhythm, HR 89/min, ST depression from V_2 to V_6; (B) same ECG with ST elevation in the posterior leads.

system, namely with fluctuations in blood pressure, arrhythmias, and electrocardiographic changes.[3] Myocardial infarction is rare, with only a few cases reported, and its diagnosis can be challenging due to the presence of electrocardiographic changes in patients with GBS.

Case report

A 67-year-old man, with a history of mild upper airway infection 3 weeks earlier, presented to the emergency room (ER) due to weakness in the lower limbs. Upon admission, he had a blood pressure (BP) of 169/90 mmHg and heart rate (HR) of 80/min, he was conscious and oriented, with no changes in cardiac and pulmonary auscultation, strength grade 3/5 in the lower limbs and 5/5 in the upper limbs. Sensitivity and reflexes were not tested. The blood tests (including troponin I), arterial blood gas analysis, ECG and head CT were normal. During his stay in the ER, he progressively developed weakness of the upper limbs, dysphagia and dyspnea. He maintained high blood pressure despite captopril. He suddenly developed acute pulmonary edema, diaphoresis, and cognitive impairment, with a BP of 204/110 mmHg, HR 150/min, with crackles on lung auscultation, and SpO_2 65% with FiO2 31%. Arterial blood gas analysis was performed with type 2 respiratory failure and respiratory acidemia: pH 7.28, pC_{O2} 53.1 mmHg, pO_2 45.6 mmHg, Lac 1.6 mmol/L, HCO_3 –24.6 mmol/L. The ECG showed sinus rhythm, HR 89/min, ST depression from V_2 to V_6 and ST elevation in posterior leads (**Fig. 1**). Repeated blood tests showed leukocytosis (19.7 G/L, N 4.1–11.1) with neutrophilia (17.8 G/L, N 2–7.5), normal renal function, elevated troponin I (2030 pg/mL, N <47.3) and NT- proBNP (928 pg/mL, N 0–125). An echocardiogram was performed which showed moderate depression of the left ventricular ejection fraction, with anterolateral and inferolateral akinesia. He was stabilized with non-invasive ventilation and intravenous furosemide and was sent for emergency

coronary angiography, which ruled out obstructive coronary disease.

After the procedure, approximately 12 hours after admission, his deficits worsened, being anarthric, unable to protrude the tongue, strength grade 0/5 in the lower limbs and 1/5 in the upper limbs, arereflexic and with weak chest expansion, with worsening respiratory failure and failure of non-invasive ventilation, and was then intubated and ventilated.

The most likely diagnostic hypothesis was Guillain–Barré syndrome with rapidly progressive onset, with severe respiratory failure and cardiac involvement.

He was accepted into the intensive care unit of the referral hospital for emergent plasmapheresis, however, after discussion with immunochemotherapy, it was considered unsafe to carry out this procedure due to high cardiovascular risk, which is why he was started on immunoglobulin 30 g/day for 5 days. He needed vasopressor support with norepinephrine, which was suspended on the 3rd day of hospitalization. He later developed a hypertensive profile, requiring antihypertensive therapy.

Neurophysiological and cerebrospinal fluid (CSF) examinations were consistent with the diagnosis of GBS. CSF examination showed an elevated protein level (45.7 mg/L, N 15–45) with normal cells (1.6/mm^3). The EMG on the 9th day of hospitalization showed severe, acquired, acute motor demyelinating polyneuropathy. She had a repeat EMG a week later, which showed acute motor axonal polyneuropathy.

The patient had a prolonged hospitalization, punctuated by nosocomial infections. Despite this, he showed slow and partial recovery from the deficits, and full recovery of left ventricular systolic function, and ended up being discharged to a rehabilitation unit.

Discussion

Autonomic dysfunction with cardiovascular involvement of GBS is widely recognized.[1,4–7] The most common electrocardiographic changes reported are sinus tachycardia, followed by ST segment changes, with ST segment elevation being relatively uncommon. Isolated elevation of troponin has also been observed.[4] These abnormal patterns could, in part, be explained by catecholamine-associated myocardial injury theories: a disorder of catecholamine uptake around myocytes,[8] redistribution of coronary blood flow,[9] or denervation hypersensitivity of the myocardium.[3]

An observational study reported that acute coronary syndrome (ACS) developed in 2.08% of patients with GBS.[4] Coronary vasospasm in patients with GBS, as a result of dysautonomia accompanied by increased catecholamines, also contributes to ACS. In our case, vasospasm could justify the patient's presentation, namely ST elevation, high troponin and segmental changes on echocardiogram, the without obstructive coronary disease. Cardiovascular complications are usually short-term, but can be fatal in GBS, being one of the determining factors in clinical outcome.[2,3] Thus, their assessment and treatment is of the utmost importance.

Conflict of interest
Nothing to disclose.

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Informed consent
The patient provided informed written consent for the publication of the study data.

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Giant Right Ventricle Myxoma Presenting as Right Heart Failure – a Case Report

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Kľúčové slová:
Myxóm pravej komory
Náhrada trikuspidálnej chlopne
Obštrukcia výtokového traktu pravej komory
Operácia srdca
Zlyhanie pravého srdca

SÚHRN

Úvod: Primárne nádory srdca sú všeobecne veľmi zriedkavým ochorením, s výskytom iba 0,0001 % až 0,3 % nekrektómií. Napriek tomu, že myxóm je najčastejším primárnym nádorom srdca, v komorách srdca sa vyskytuje v menej ako 5 %.
Kazuistika: 19-ročný pacient bez inej komorbidity bol prezentovaný pre jednoročnú anamnézu námahovej dušnosti, únavy, opuchu tváre a palpitácií. Echokardiografické vyšetrenie zaznamenalo útvar veľkosti 60 × 70 mm v pravej komore, spontánny echokontrast v pravej predsieni, maximálnu rýchlosť prietoku krvi v pľúcnom kmeni 3 m/s, zväčšenie pravej predsiene a zachovanú ejekčnú frakciu ľavej komory. Pacient podstúpil resekciu útvaru so súčasnou náhradou trikuspidálnej chlopne biologickou protézou za použitia mimotelového obehu. Pooperačne bol priebeh komplikovaný atrioventrikulárnym blokom, pre ktorý bol implantovaný kardiostimulátor s epikardiálnou elektródou. Histologická analýza dokumentovala srdcový myxóm s rozmermi 110 × 90 × 50 mm. Kontrolné echokardiografické vyšetrenie tri mesiace po operácii poukázalo na dobrú funkciu oboch srdcových komôr, s normalizáciou parametrov pravej predsiene a pravej komory.
Diskusia a záver: Nádory pravých oddielov srdca môžu viesť k obštrukcii výtokového traktu pravej komory, s príznakmi zlyhania pravého srdca. Napriek dobrým perioperačným výsledkom a nízkemu riziku recidívy je skorá diferenciálna diagnostika a včasný chirurgický zákrok kľúčový pre optimálnu prognózu pacientov.

Keywords:
Heart surgery
Right heart failure
Right ventricle myxoma
Right ventricle outflow tract obstruction
Tricuspid valve replacement

ABSTRACT

Introduction: Primary cardiac tumors are generally a very rare condition, with an incidence of only 0.0001% to 0.3% in autopsies. Despite the fact that myxoma is the most common primary heart tumor, fewer than 5% of myxomas, which vary in size, occur in the ventricles.
Case report: A 19-year-old patient with an unremarkable medical history presented with a one-month history of exertional dyspnea, fatigue, facial edema, and palpitations. Echocardiography recorded a mass measuring 60 × 70 mm in the right ventricle, spontaneous echo contrast in the right atrium, a maximum blood flow velocity in the pulmonary trunk of 3 m/s, enlargement of right atrium and preserved left ventricle ejection fraction. The patient underwent a resection of the mass with concomitant tricuspid valve replacement with a biological prosthesis under cardiopulmonary bypass. Postoperatively, the condition was complicated by atrioventricular block, and an epicardial electrode and pacemaker were implanted. Histological analysis revealed a cardiac myxoma of diameter 110 × 90 × 50 mm. The follow-up echocardiographic examination three months after surgery documented good function of both the left and right ventricles, with normalization of the right atrial and right ventricle parameters.
Discussion and conclusion: Right-sided cardiac tumors can lead to right ventricular outflow tract obstruction, with symptoms of right-sided heart failure. Despite good perioperative outcomes and a low risk of recurrence, early differential diagnosis and early surgical intervention are critical for patients to have an optimistic prognosis.

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Introduction

Primary cardiac tumors are generally a very rare condition, with an incidence of only 0.0001% to 0.3% in autopsies.[1–4] On the other hand, metastases from other tumors to the pericardium, myocardium, coronary arteries or great vessels are found in 0.7% to 3.5% of the general population and in up to 9.1% of patients with known oncological disease.[5] Metastases from other tumors to the heart most commonly originate from the lungs, breast, skin, thyroid gland and kidneys. The spread of other tumors to the heart can occur via four pathways: direct expansion, hematogenous spread, the lymphatic system or invasion through the vena cava or pulmonary veins.[6] On the other hand, cardiac malignancies may also manifest with secondary metastases.[1] The most frequent primary tumor of the heart is myxoma, which is typically located in the atria (more often the left atrium), arising from the place of the fossa ovalis.[1,5,7] Other tumors observed in the heart include sarcoma, angiosarcoma, rhabdomyosarcoma, papillary fibroelastoma, lipoma, hemangioma, mesothelioma and rhabdomyoma.[5] The distribution of different types of tumors in childhood is significantly different. The most common cardiac tumor in children is rhabdomyoma (42–75%), followed by fibroma (6–25%). Myxomas represent only 5 to 10% of cardiac tumors in the pediatric population.[3,8]

Cardiac myxomas in adults usually arise as a solitary in the left atrium (75%) or right atrium (10%–20%).[1,9,10] The occurrence of a myxoma is approximately 1.5 per one million population.[9,10] The presence of a myxoma is more common in women and most frequently occurs between the age of 30 and 60.[8,10] The presence of myxomas can also be associated with Carney complex, which is defined by the presence of cardiac and cutaneous myxomas, endocrine overactivity and distinctive pigmented lesions of the skin and mucosal surfaces.[9] Familial occurrence of myxomas is observed in only 10% of these tumors.[4,10] These familial myxomas often have multiple and atypical locations.[7] From the perspective of all cardiac tumors, it can be stated that intramural tumors are less symptomatic than intracavitary tumors.[3] The literature also reports secondary occurrence of myxoma in the fossa ovalis region following a catheter-based intervention in this area.[7] Clinical manifestation of myxomas consist of the triad of constitutional symptoms, embolization, and intracardiac obstruction.[1,2] Constitutional symptoms include fever, weight loss, infections and so on, which may be associated with the release of pro-inflammatory molecules, particularly interleukin 6 (IL-6). Obstructive symptoms are generally related to obstruction of a cardiac ventricle, atrium or a valve. The manifestation of obstructive and embolic symptoms is differentially distinct in left-sided and right-sided tumors.[2,3] The clinical presence of a myxoma varies significantly from case to case and depends primarily on its size, location and mobility.[2]

Case report

A 19-year-old patient with an unremarkable medical history presented with a one-month history of exertional dyspnea, fatigue, facial edema, and palpitations. Echocardiography revealed a mass measuring 60 × 70 mm in the right ventricle, causing obstruction of both the inflow and outflow tracts of the right ventricle (**Fig. 1**). Spontaneous echo contrast was observed in the right atrium. The left ventricular ejection fraction (LVEF) was preserved at 60%, with the mitral and aortic valves functioning normally. There was a moderate pericardial effusion and enlargement of the right atrium, which measured 34 cm². The maximum blood flow velocity in the pulmonary trunk was 3 m/s. Computed tomography corroborated the presence of a tumor in the right ventricle and noted a subsegmental embolism in a branch of the right pulmonary artery (**Fig. 2**). Additionally, CT revealed hepatomegaly and the presence of free fluid within the abdominal cavity. An urgent surgical intervention was recommended by the heart team.

The patient underwent a resection of the mass with concomitant tricuspid valve replacement with a biological prosthesis under cardiopulmonary bypass. Medial sternotomy was chosen as the surgical approach. Cardiopulmonary bypass was administered by cannulation

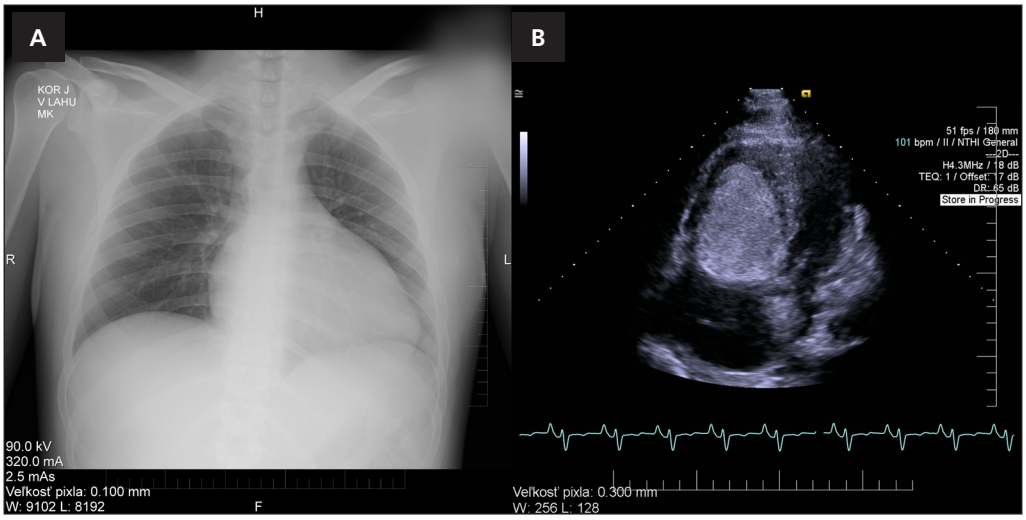

Fig. 1 – (A) X-ray image of the patient's thorax prior to surgery; (B) echocardiographic imaging of the tumor in the right ventricle.

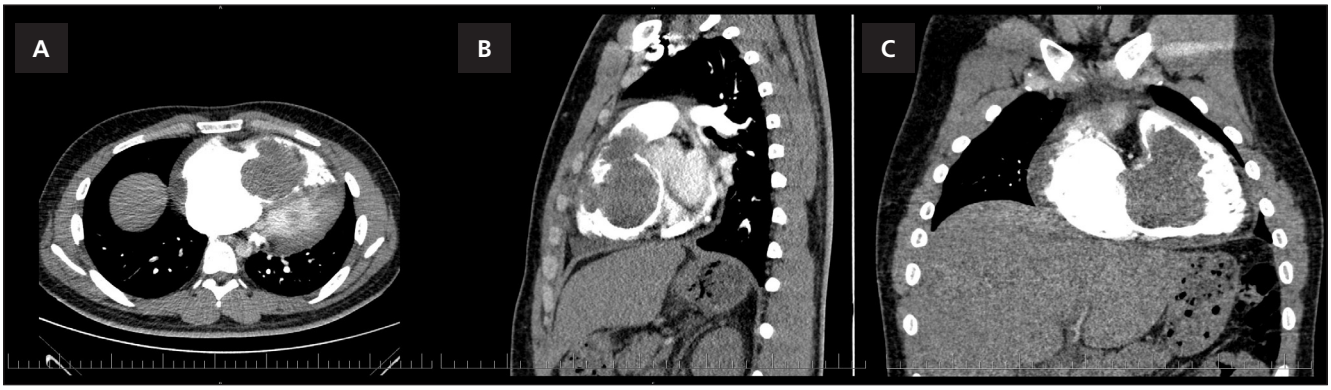

Fig. 2 – Computed tomography of the cardiac tumor. tumor. (A) Axial plane; (B) sagittal plane; (C) coronal plane.

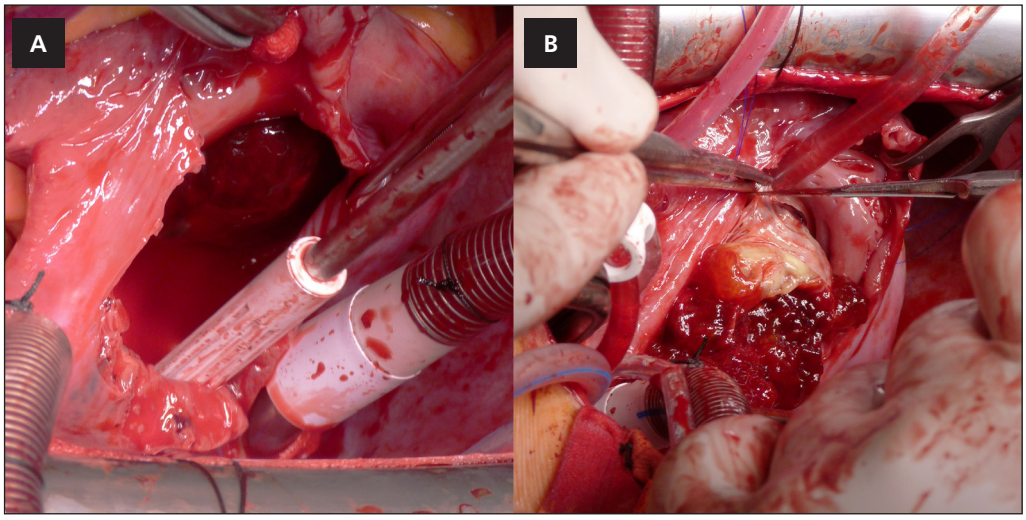

Fig. 3 – Intraoperative photographs: (A) view of the right atrium with a tumor obstructing nearly the entire annulus of the tricuspid valve; (B) dissection of the tumor's attachment from the leaflets of the tricuspid valve.

of the aortic arch and upper and lower vena cava separately. The tumor was exposed by incision of the pulmonary trunk and right atrium wall and was found to be obstructing nearly the entire orifice of both the pulmonary and tricuspid valves, occupying the entirety of the right ventricle and originating directly from the anterior leaflet of the tricuspid valve. Complete excision of the tumor was performed along with the anterior and posterior leaflets of the tricuspid valve, followed by replacement of the tricuspid valve with a bioprosthesis (**Fig. 3**). The total duration of cardiopulmonary bypass was 95 minutes, with an aortic cross-clamp time of 61 minutes.

The resected tumor measured 110 × 90 × 50 mm. Histological analysis revealed a cardiac myxoma with chronic inflammatory cell infiltration and dystrophic calcifications. Immunohistochemical studies demonstrated isolated S100 positivity, focal CD31 and CD34 positivity, and positivity for AE1/3, calretinin and SMA. No malignant transformations were observed in the excised tissue (**Fig. 4**).

On the first postoperative day, a reduction in the area of the right atrium to 22 cm² was observed, with the LVEF remaining stable at 60% and the maximum blood flow velocity in the pulmonary trunk at 0.88 m/s. Due to the ongoing presence of a third-degree atrioventricular block (AVB), epicardial pacing electrodes and a pacemaker were implanted on the seventh postoperative day. The patient was discharged to outpatient care on the 13th postopera-

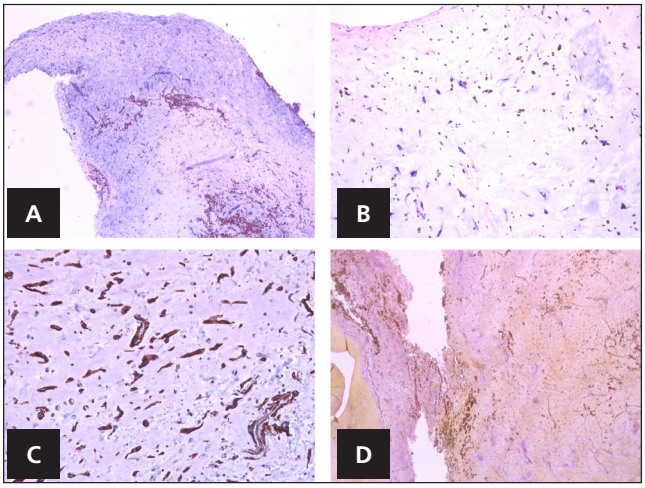

Fig. 4 – Histological appearance of the resected right ventricular myxoma. (A, B) Hematoxylin & eosin staining; (C) CD34 antibody; (D) mucicarmine staging; (A, D – 25×; B, C – 100×).

tive day. At a three-month follow-up echocardiographic assessment, the right atrial area measured 22 cm² with a mean tricuspid valve gradient of 6.8 mmHg; the LVEF remained at 60%. The right ventricle/left ventricle basal diameter ratio was 0.7, with a right ventricle basal diameter of 36 mm and a mid diameter of 35 mm. The patient reported feeling well.

Discussion

Despite the fact that myxoma is the most common primary heart tumor, fewer than 5% of myxomas, which vary in size, occur in the ventricles.[9] The localization of myxoma in the ventricle leads to the presence of an incomplete right bundle branch block more frequently, and ST/T wave changes on the electrocardiography (ECG) can also be found in the presence of a tumor in the right heart chambers.[9] Moreover, right-sided myxomas, compared to left-sided ones, more frequently lead to serious complications, including arrhythmias, pulmonary embolism, syncope and even sudden death.[10] While obstructive symptoms in left-sided heart tumors lead to pulmonary congestion and systemic ischemia, resulting in dyspnea or syncope, the obstructive consequences of a tumor in the right chambers of the heart lead to peripheral edema, ascites or superior vena cava syndrome.[2] Right-sided tumors may further cause pulmonary embolism, which was also documented by computer tomography (CT) examination in the presented case report.[2] Tumors leading to obstruction of the right atrium can also manifest as hepatomegaly, ascites, and edema of the lower extremities.[1,3] Sudden death due to the presence of an intracardiac tumor occurs as a result of coronary, or systemic embolism, or as a consequence of obstruction of blood flow at the mitral or tricuspid valve.[1] Obstruction of the right ventricular outflow tract (RVOT) in right-sided myxomas is often manifested as right-sided heart failure.[2] Initially, a myxoma may be accompanied by peripheral edema due to right heart failure, which can be misdiagnosed as nephrotic syndrome.8 The growth of myxomas is difficult to monitor, due to the immediate indication for their resection as soon as possible after diagnosis. The literature reports a common growth rate of myxomas at 3 mm per month.[2,11] Given the size of the tumor in the presented case, it can be hypothesized that the findings were already present three years prior to its manifestation. Deebis et al. present a cardiac tumor measuring 9 × 4.6 × 3.7 cm, with the patient reporting symptoms for two years prior to the diagnosis of the tumor.[3] Myxomas larger than 110 mm are extremely rare in the literature.[9] Compared to other reports of myxomas presented in the literature, the presented case is rare, particularly due to the size of the tumor and its location in the right ventricle of the heart. Furthermore, the literature contains only one case report presenting tumors in the right ventricle with dimensions that led to nearly complete obstruction of both the atrioventricular valve and the RVOT.[10] Another study presents a tumor in the right ventricle measuring 9.5 × 5.0 cm obstructing the RVOT.[2]

Dysfunction of the right ventricle is associated with high incidence of morbidity and mortality. Obstruction of the right ventricular outflow tract (RVOTO) is one of the rare causes of right ventricle failure. The normal pressure gradient between the right ventricle and the pulmonary trunk should be less than 6 mmHg.[12] If RVOTO is presented, right ventricle dysfunction is caused by a pressure gradient between the right ventricle and the pulmonary trunk. A hemodynamically significant RVOTO is presented when the maximum pressure gradient reaches 25 mmHg.[12] Other authors define RVOTO based on a blood flow velocity >2.0 m/s through the RVOT using continuous-wave Doppler and transthoracic echocardiography (TTE).[13] In the presented case study, the preoperative blood flow velocity in the pulmonary trunk was 3 m/s.

RVOTO can be divided based on several factors related to the RVOT and the cardiac cycle. Based on the anatomical relationship of the underlying cause with the RVOT, we distinguish between intrinsic and extrinsic RVOTO. Intrinsic causes account for 58% of RVOTO, while extrinsic causes represent 42% of the RVOTO cases reported in the literature.[12] Depending on its manifestation during the cardiac cycle, RVOTO can present continuously (constant) or only during systole or diastole (dynamic).[12] A constant cause of RVOTO, manifesting throughout the entire cardiac cycle, accounts for 90% of the RVOTO cases reported in the literature.[12] The etiology of RVOTO can be divided into congenital, iatrogenic and non-congenital and non-iatrogenic.[12] Congenital RVOTO includes conditions such as a subpulmonary membrane.[14] The most common etiology of RVOTO is non-congenital, non-iatrogenic.[12] Non-congenital and non-iatrogenic RVOTO are caused by a primary cardiac tumor in 40% of cases, by an extracardiac tumor due to the presence of metastases or compression in 23% of cases and by an aneurysm of the Valsalva sinus in 18% of cases.[12] The most common etiology of intracavitary processes leading to RVOTO or left ventricular outflow tract obstruction (LVOTO) is thrombosis.[6] The incidence of myxoma or a primary neoplasia of the pulmonary trunk as the etiology of RVOTO is comparable, at 28% and 25%, respectively.[12] A significantly different pathophysiology of RVOTO is present in what is known as suicidal RVOTO. This typically arises after lung transplantation, surgical procedures, and heart catheter interventions. Hypertrophic cardiomyopathy of the right ventricular outflow tract is generally considered to be a thickening of the right ventricular wall greater than 5 mm. If the etiology of RVOTO is hypertrophic cardiomyopathy, conservative medication therapy can be considered. Conservative therapy can be implemented by administering myosin inhibitors.[15] Pathophysiologically, RVOTO is caused by changes in pressure gradients in the right heart chambers and lungs, leading to collapse and subsequent obstruction of the RVOT. This is usually a dynamic obstruction.[12] Due to the rapid growth of a myxoma, this finding is not common after the surgical excision of a myxoma from the RVOT. In the presented case, surgical intervention led to an early normalization of pressure gradients in the right heart chambers, with a significant reduction in the diameter of the right atrium, resulting in a positive outcome for the patient. This was likely caused by a combined obstruction of the RVOT and the tricuspid valve, leading to a significant reduction in right ventricular preload.

The diagnosis of RVOTO can be performed by transthoracic echocardiography, transesophageal echocardiography, computed tomography, magnetic resonance, epicardial echocardiography or cardiac catheterization. Cardiac catheterization measures the pressure gradient by monitoring the blood pressure in the right ventricle and pulmonary artery. It is crucial for the diagnosis of RVOTO, but not for determining its etiology.[12] Echocardiography can visualize the anatomical position of a tumor and its relation to the surrounding tissue throughout the cardiac cycle. When located posteriorly, transesophageal echocardiography provides a better image and more accurate anatomical correlations with surrounding structures.[5] Computed tomography (CT) and magnetic resonance imaging (MRI) are additional imaging modalities. Conventional CT provides superior visualization of calcified masses in the heart.[5] The administration of a contrast agent allows for visualization of the vascularity of the tissue. Neoplastic masses, whether primary or secondary, manifest on a CT examination as areas of contrast hyper- or hypoenhancement, whereas a thrombus shows no enhancement.[6] CT guided by electrocardiogram (ECG) offers comparably high-quality imaging of the soft tissues of the heart, similar to that of magnetic resonance imaging.[5] Contrast enhancement in early and delayed gadolinium images during cardiac MRI is also highly effective in differentiating a cardiac tumor from a non-enhancing thrombus.[3] Larger tumors may undergo central necrosis, central hemorrhage or a decrease in perfusion at the tumor's center. This can lead to reduced enhancement on contrast-enhanced CT as well as MRI. Therefore, for larger tumors, this differential diagnostic criterion may be misleading.[3]

The differential diagnosis of an intracardiac mass should focus on distinguishing between thrombus, tumor, and vegetation in infectious endocarditis. A thrombus is more likely if the ECHO image fails to visualize a stalk, the atrium is enlarged, and cardiac output is low. A large, organized thrombus intimately adherent to the endocardium and unresponsive to thrombolysis can closely resemble a tumor in differential diagnosis. The diagnosis of a thrombus may then be supported by the coincidence of lupus erythematosus, antiphospholipid syndrome or the patient's autoimmune condition.[5] Vegetations in infectious endocarditis can also mimic a tumor during echocardiographic examination. They occur due to bacterial or fungal infections. Vegetations are usually irregular, arising from the valve leaflets, and they more commonly affect the left chambers of the heart. Larger sizes are more indicative of a fungal infection.[5] Differential diagnosis is more challenging with atypical findings of a tumor in the right heart chambers. Suspicion of an oncological process often arises only when there is no response to thrombolytic and/or antibiotics therapy.[16] Histologically, a tumor in the right heart chambers causing RVOTO may also be a metastasis from a distant carcinoma.[5]

Currently, there is no effective pharmacological therapy for myxomas.[1] Resection is the only definitive therapy possible for myxoma.[2] Early surgical intervention is critical to prevent obstruction at the level of the atria, atrioventricular valve or ventricular outflow tract.[3,4] Even in cases of diagnostic uncertainty, biopsy of a tumor or mass in the chamber is not recommended. A biopsy could lead to catastrophic consequences, and primary resection is the preferred choice for both histological diagnosis and therapy.[4] In general, approaches to tumors of the right ventricle may include right atriotomy, right ventriculotomy, and pulmonary arteriotomy.[3] When the tumor is located in the right ventricle, direct visualization via ventriculotomy is not preferred. According to the literature, a more favorable approach is atriotomy and tumor visualization through the atrioventricular orifice.[9] However, the literature also reports the effective use of right-sided ventriculotomy when the anatomical relationship between the tumor, right ventricle, tumor stalk, and tricuspid orifice does not allow for its safe extirpation via an atriotomy approach.[3,8] Atriotomy was also chosen in the presented case. Augmenting the surgical approach with an incision in the pulmonary trunk helps prevent the risk of periprocedural embolism. In the case of a right ventricular myxoma without direct anatomical continuity between the tumor and the tricuspid valve, myxoma resection can be performed via right atriotomy while preserving the functionality of the tricuspid valve.[2,4,10] In the presented case, valve preservation was not possible due to the direct attachment of the myxoma to the tissue of the valve leaflets. The literature also presents resection as a therapeutic option for secondary cardiac tumors. The goal of this therapy is not only in the treatment of the obstruction of cardiac chambers but, in the case of solitary metastasis, such resection can also be curative for the patient.[6] In the context of right ventricular outflow tract obstruction and the presence of tricuspid valve regurgitation without annular dilation, tumor resection that effectively eliminates the RVOTO can also resolve the tricuspid valve regurgitation without the need for surgical intervention on the valve itself.[17] In rare cases, the literature describes the resection of right ventricular tumors using cardiopulmonary bypass on a beating heart.

The results of surgical resection are optimistic. In this case report, the resection was complicated by the development of third-degree atrioventricular block. Such a complication has also been described in other case reports, particularly when the extent of the tumor necessitated replacement of the tricuspid valve.[6] Perioperative mortality for cardiac myxoma is less than 5%.[2] Recurrence occurs in 5% of cases.[8,17] The development of recurrence can be diagnosed using transthoracic echocardiography (TTE), typically within months but also years after the surgery.[8,9]

Conclusion

Cardiac myxoma, along with thrombus and infective endocarditis, is among the three primary diagnoses that are most commonly presented as a tumorous mass in the ventricular chambers. Their manifestation involves constitutional symptoms, embolization, and intracardiac obstruction. The last one, when located in the rare right-sided heart chambers, may primarily present as right-sided heart failure. Despite good perioperative outcomes and a low risk of recurrence, early differential diagnosis

and early surgical intervention are critical for an optimistic prognosis for the patient.

Author contributions
Štefan Lukačín: conceptualization, formal analysis, investigation, methodology, validation, funding; Tomáš Toporcer: conceptualization, formal analysis, investigation, methodology, project administration, resources, supervision, validation, visualization, writing – original draft, writing – review & editing; Vilém Rohn: conceptualization, formal analysis, supervision, validation; Pavel Kočan: data curation, validation; Anton Bereš: data curation; Marián Homola: data curation; Michal Trebišovský: data curation; Adrián Kolesár: conceptualization, formal analysis, supervision, validation. All authors have read and agreed to the published version of the manuscript.

Conflict of interest
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None.

Ethical statement
The work was carried out in compliance with the Declaration of Helsinki.

Informed consent
The patient gave an informed consent with publishing the case report.

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Late-onset acute limb ischemia after transcatheter patent foramen ovale closure with Occlutech Figulla Flex II PFO Occluder

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Perkutánny uzáver
Trombus

SÚHRN

Kontext*:* Trombogenéza je veľmi zriedkavá komplikácia po katétrovej oklúzii foramen ovale patens (PFO). Kazuistika neskorej symptomatickej trombózy oklúzora Occlutech Figulla Flex II PFO nebola podľa vedomostí autorov dosiaľ publikovaná.
Kazuistika: Ide o prípad 45-ročného muža s anamnézou periférnej artériovej embolizácie s následným uzáverom PFO oklúzorom Occlutech Figulla Flex II PFO. Pacient absolvoval 6-mesačnú duálnu protidoštičkovú liečbu klopidogrelom (75 mg denne) a kyselinou acetylsalicylovou (100 mg denne) a následne pokračoval v monoterapii klopidogrelom ďalších 8 mesiacov. Päť mesiacov od ukončenia protidoštičkovej monoterapie (t.j. 19 mesiacov od uzáveru PFO) bol rehospitalizovaný s klinickým a ultrasonografickým obrazom akútnej končatinovej ischémie ľavej dolnej končatiny. Transtorakálna a transezofagová echokardiografia verifikovali rozsiahlu masu v ľavej predsieni, fixovanú na ľavopredsieňový disk oklúzora. Reziduálny skrat nebol prítomný. Pacient absolvoval katétrovú embolektómiu z ľavej femorálnej artérie. Nález na počítačovej tomografii po dvoch mesiacoch antikoagulácie warfarínom bol identický ako na echokardiografii. Zistila sa rezistencia na aktivovaný proteín C. Realizovali sme chirurgickú extirpáciu masy a uzáver defektu predsieňového septa perikardiálnou záplatou. Histopatologický nález extirpovanej masy svedčil pre trombus.
Záver: Tromboembolizmus po uzávere PFO pomocou Occlutech Figulla Flex II PFO oklúzora je výnimočná komplikácia, ktorá môže vzniknúť neskoro po výkone napriek antitrombotickej profylaxii.

Keywords:
Acute limb ischemia
Occlutech Figulla Flex II PFO occluder
Patent foramen ovale
Percutaneous closure
Thrombus

ABSTRACT

Background*:* Thrombus formation is an exceedingly rare complication after transcatheter patent foramen ovale (PFO) occlusion. To the best of the authors' knowledge, a case of late symptomatic thrombosis of Occlutech Figulla Flex II PFO occluder has not yet been published.
Case presentation: A 45-year-old man with a history of peripheral artery embolism, and subsequent PFO closure with Occlutech Figulla Flex II PFO occluder. The patient completed dual antiplatelet therapy with clopidogrel (75 mg/day) and aspirin (100 mg/day) after 6 months and subsequently continued single antiplatelet therapy (SAPT) with clopidogrel for other eight months. Five months after the discontinuation of SAPT (i.e. 19 months after PFO closure), he was readmitted with a clinical and ultrasound presentation of acute limb ischemia of the left lower extremity. Transtoracic and transesophageal echocardiography revealed a huge left atrial mass, attached to the left atrial disc of the occluder. The residual shunt was not present. A catheter embolectomy from the left femoral artery was performed. Computed tomography after two months of warfarin anticoagulation showed identical findings to echocardiography. Activated protein C resistance was found. The mass was surgically extirpated and the atrial septal defect was corrected with a pericardial patch. Pathology confirmed the mass to be a thrombus.
Conclusion: Thromboembolism after PFO closure with an Occlutech Figulla Flex II PFO occluder is an exceptional complication that can occur late after the procedure despite antithrombotic prophylaxis.

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Introduction

Percutaneous occlusion of patent foramen ovale (PFO) is indicated in precisely selected patients with cryptogenic central or peripheral thromboembolism and a high probability of a causal relationship with PFO after excluding other identifiable causes.[1]

Numerous devices have been used for the PFO closure. Figulla Flex II occluders are among the most commonly used devices. The observed incidence of adverse effects after implantation of this device during long-term follow-up is low. Based on the published studies, thrombus formation on the Figulla Flex II occluder was not observed during the mid-term follow-up of 6 months to 1.1 years.[2–4]

We present a case of symptomatic late thrombus formation 19 months after percutaneous closure of PFO with an Occlutech Figulla Flex II PFO occluder.

Case presentation

A 45-year-old man with a history of right femoral and popliteal artery embolism treated with thrombolysis. The patient was subsequently diagnosed with a PFO with a massive right-to-left shunt. The channel length and diameter were 9 and 4 mm, respectively. Other causes of peripheral thromboembolism were excluded. Paradoxical embolism through a PFO was believed to be a cause of acute limb ischemia. So, he underwent a PFO closure with Occlutech Figulla Flex II PFO Occluder 23/25 mm, resulting in no residual shunt. Dual antiplatelet therapy (DAPT) with long-term clopidogrel (75 mg/day) and aspirin (100 mg/day) during the first six months following the procedure was recommended. The patient completed DAPT as recommended after 6 months and subsequently continued single antiplatelet therapy (SAPT) with clopidogrel.

At transthoracic echocardiography (TTE) study 12 months after PFO occlusion, the occluder was free of leak and thrombus. The patient discontinued clopidogrel monotherapy 14 months after PFO closure.

Five months after the discontinuation of SAPT (i.e. 19 months after PFO closure), he was admitted repeatedly with a clinical and ultrasound presentation of acute limb ischemia of the left lower extremity. On presentation, his vital signs were normal: blood pressure 130/84 mmHg, heart rate 84 beats per minute, temperature 36.5°C, and respiratory rate 18 breaths per minute. The left calf was cool, pale, without palpable pulsations on the popliteal artery and, more distally, with a movement disorder of the fingers. The rest of the physical examination was unremarkable. Initial basic routine laboratory tests (blood count, biochemistry, INR, aPTT, fibrinogen) did not reveal significant abnormalities. The electrocardiogram recording and the chest radiograph were normal.

TTE found a large homogeneous hypoechogenic 34 × 15 × 12 mm mass in the left atrium below the anterior mitral leaflet. The rest of the TTE examination was unremarkable.

Two- and three-dimensional transesophageal echocardiography (TEE) confirmed a huge left atrial ovoid 30 × 24 xx 12 mm mass, which was attached by a 7 mm wide stalk to the left atrial disc of the Figulla Flex occluder (**Fig. 1**). This partially mobile mass was not prolapsing into the left ventricle. There was no mitral valve obstruction or regurgitation. Both discs of the occluder were in the correct position. The residual shunt was not present. The mass displayed on echocardiography was considered as likely to be an extensive thrombus. Myxoma was unlikely.

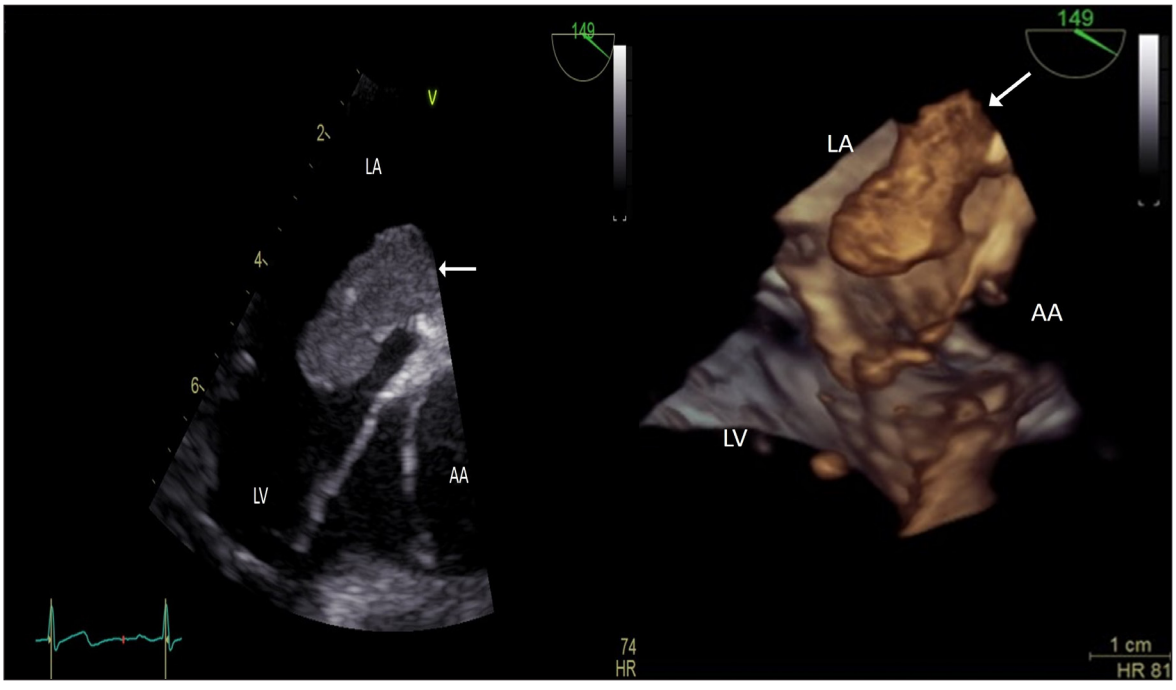

Fig. 1 – Two-dimensional (left) and three-dimensional (right) transesophageal echocardiography. Large hypoechogenic left atrial mass (←) attached to a PFO occluder. AA – ascending aorta; LA – left atrium; LV – left ventricle.

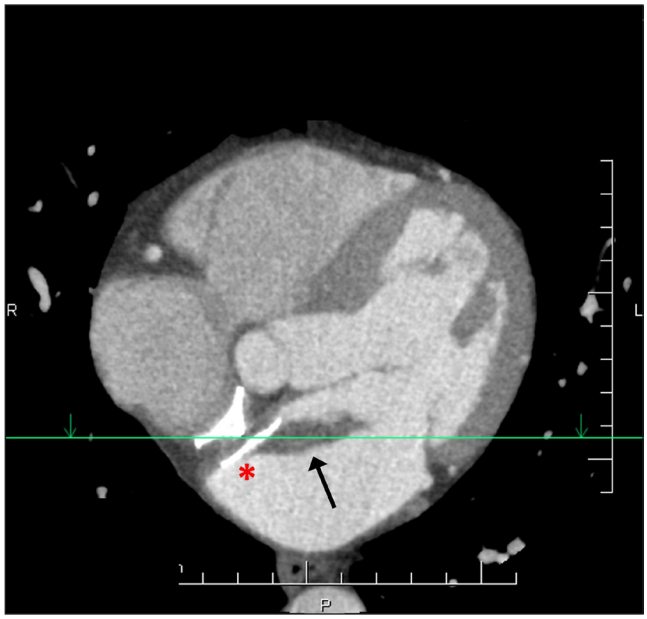

Fig. 2 – CT scan showing a mass (→) on the left atrial disk (*) of the PFO occluder.

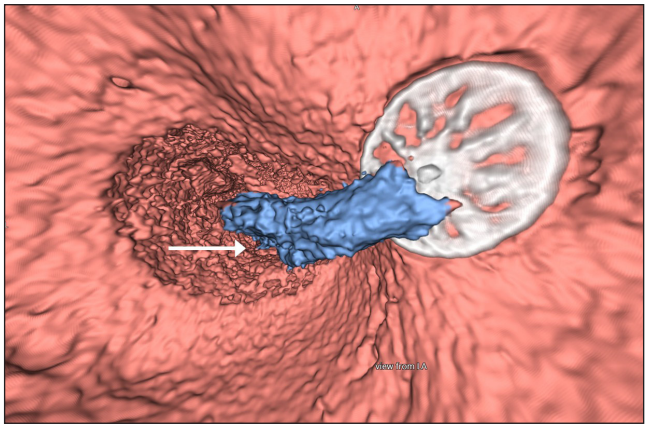

Fig. 3 – Three-dimensional CT scan reconstruction (view from the left atrium) demonstrating a mass (→) on the left atrial disk of the PFO occluder.

Successful arterial embolectomy from the left common, superficial, and deep femoral artery was performed using the Fogarty embolectomy catheter. A warfarin therapy was started.

Computed tomography (CT) after two months of oral anticoagulation showed an analogous finding as in the previous TEE – a hypodense 34 × 11 × 10 mm mass with a stalk attached to the left atrial disk of the PFO occluder (**Figs 2 and 3**). The mass was thought to be a thrombus rather than a myxoma. CT coronary angiography demonstrated the coronary artery disease without significant stenoses. Surgical management was advised and unfractionated heparin drip was started. The mass and the occluder were extirpated (**Fig. 4**). The atrial septal defect was corrected with a pericardial patch. The histopathological examination of the removed mass was consistent with a thrombus. Coagulation assays were performed to

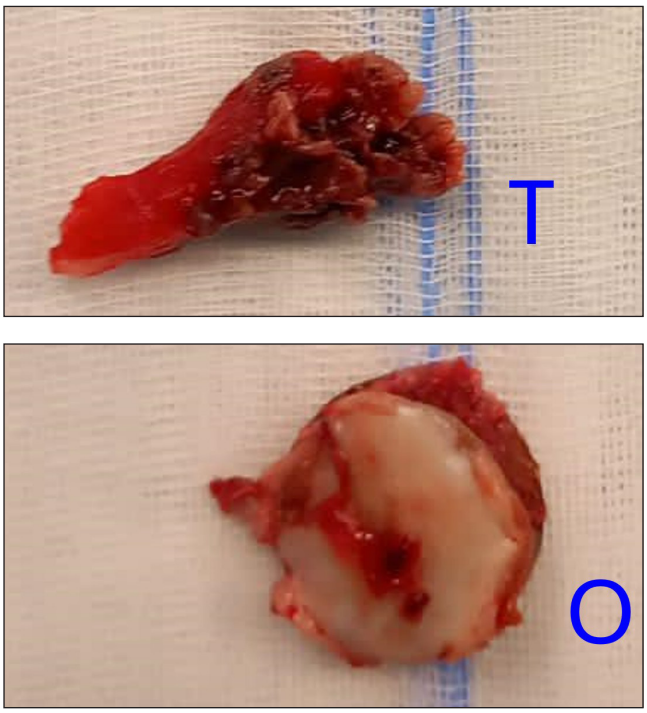

Fig. 4 – Explanted Figulla-Flex II Occluder (O) and thrombus (T).

detect hereditary or acquired thrombophilic disorders. Activated protein C resistance was found. Other thrombophilic conditions were not found. Rivaroxaban was recommended for three months to prevent recurrent thromboembolism. The postoperative status was complicated by hemodynamic instability and sepsis caused by erysipelas bullosum of the right calf. After appropriate treatment, the patient was stabilized and later discharged on the tenth postoperative day.

Discussion

Thromboembolism is an exceedingly rare complication after transcatheter PFO closure. However, its consequences can be serious and even life-threatening.[5] In our case, we reported acute limb ischemia caused by thrombosis on the Occlutech Figulla Flex II PFO Occluder 19 months after the procedure. Thromboembolism occurred despite antithrombotic prophylaxis. To the best of the authors' knowledge, a case of late symptomatic thrombosis of this type of PFO occluder has not yet been published.

Our case report raises the question of how long antiplatelet prophylaxis should last after PFO occlusion. According to the European position paper, it is reasonable to propose DAPT for 1 to 6 months after PFO closure. Single antiplatelet therapy should be continued for at least 5 years. However, the strength of both statements is only conditional.[1] The extension of the single antiplatelet therapy beyond 5 years should be based on the balance between the patient's overall risk of stroke for other causes and hemorrhagic risk. However, real-world practice is heterogeneous and the duration of thromboprophylaxis is often shorter than 5 years.

The thromboembolic complication in our patient is in contrast to the results of published studies. Trabattoni et al. reported 406 consecutive patients after PFO occlusion with either the Occlutech Figulla (n = 227) occluder or the Amplatzer PFO occluder (n = 179) after cerebral ischemic attack ascribed to the PFO. Aspirin (100 mg/day) was administered at least 24 hours before the procedure and continued for six months after the closure. Clopidogrel was started immediately after the closure and continued for three months. No thrombi were detected on TTE within 24 hours after the procedure. The occurrence of thrombi on the occluders in further follow-up is not reported.[6]

In a study by Toggweiler et al., 193 consecutive patients who underwent PFO closure with the Amplatzer occluder or the Occlutech Figulla-Flex-II occluder were included. DAPT with aspirin (100 mg/day) and clopidogrel (75 mg/day) was recommended during the first 3 months after the procedure, followed by 3 months of aspirin monotherapy. No thrombus on the PFO device was found during the follow-up of 6-month.[4]

In both studies, DAPT with aspirin (100 mg/day) and clopidogrel (75 mg/day) was recommended during the first 3 months after the procedure, followed by 3 months of aspirin monotherapy.[4,6] No thrombus on the PFO occluders was noted during echocardiographic follow-up in these studies. Our patient had more intensive antiplatelet therapy than in both studies (DAPT for 6 months, followed by 8 months of clopidogrel monotherapy). Despite this, he developed a thrombus on the PFO occluder 19 months after PFO closure.

Neuser et al. occluded PFO with Figulla Flex II Occluder in 57 patients with cryptogenic thromboembolism. DAPT with aspirin (100 mg/day) and clopidogrel (75 mg/day) was recommended during at least the first 6 months following the procedure. Before discontinuation of DAPT thrombus formation on the occluder was ruled out by TEE. Examinations did not reveal any thrombus formation.[2]

The largest published retrospective multi-center registry evaluating the safety and efficacy of PFO closure with Figulla Flex II PFO and UNI occluders was the RISE study. Pioch et al. included 527 patients. The median follow-up was 1.1 years. Thrombus formation on both devices was 0%.[3] Postprocedural thromboprophylaxis was not specified.

Belgrave et al. reported several risk factors for thrombus formation on occluders – coagulopathies, female gender, age, the relative safety of the individual device, use of anticoagulant therapy, arrhythmias, atherosclerosis of the aorta, and the time since implantation. Most studies found events within the first year after the procedure.[5] Our patient developed a thromboembolism 19 months after PFO closure, and five months after the discontinuation of antiplatelet therapy. Of the known risk factors, he presented only with thrombophilia (activated protein C resistance).

The risk of thrombus formation and thromboembolic events raises the question at what intervals should repeat echocardiography be performed after device implantation. The Amplatzer device manufacturer recommends intervals of one week, six months, and one year after the closure.[5] Thrombus can rarely occur up to 5 years after Amplatzer device implantation.[7]

Myxoma should also be considered for masses on the occluders. Gupta et al. presented a TEE finding of a large hypermobile mass arising from the left atrial disc two years after PFO closure with an Amplatzer occluder. Histopathological analysis confirmed myxoma.[8]

Conclusion

Thromboembolism after the closure of PFO with an Occlutech Figulla Flex II occluder is an exceptional but potentially serious complication that can occur late after the procedure despite antithrombotic prophylaxis.

Conflict of interest
Nothing to declare.

Funding
Nothing to declare.

Ethical statement
This case report was conducted in accordance with the principles of the Declaration of Helsinki.

Informed consent
The authors confirm that written consent for the publication of this case report has been obtained from the patient.

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