

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

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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 jednorôčnú anamnézu námahovej dušnosti, únavy, opuchu tváre a palpitácií. Echokardiografické vyšetrenie zaznamenalo útvar veľkosti 60 × 70 mm v pravej komore, spontánny echokонтast v pravej predsieni, maximálnu rýchlosť prietoku krvi v pľúcnom kmeni 3 m/s, zväčšenie pravej predsieni 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 predsieni a pravej komory.

Diskusia a záver: Nádory pravých oddielov srdca môžu viesť k obštrukcii výtokového traktu pravej komory, s príznakmi zlyhania pravej komory. Napriek dobrým perioperačným výsledkom a nízkemu riziku recidívy je skorá diferenciálna diagnostika a včasný chirurgický zákrok kľúčový pre optimálnu prognózu pacientov.

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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 re-

ports 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

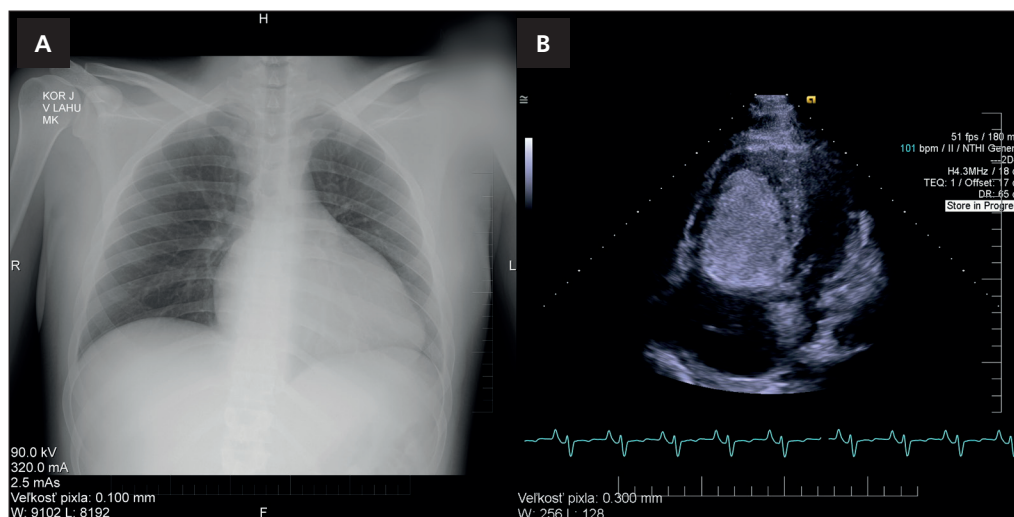


Fig. 1 – (A) X-ray image of the patient's thorax prior to surgery; (B) echocardiographic imaging of the tumor in the right ventricle.

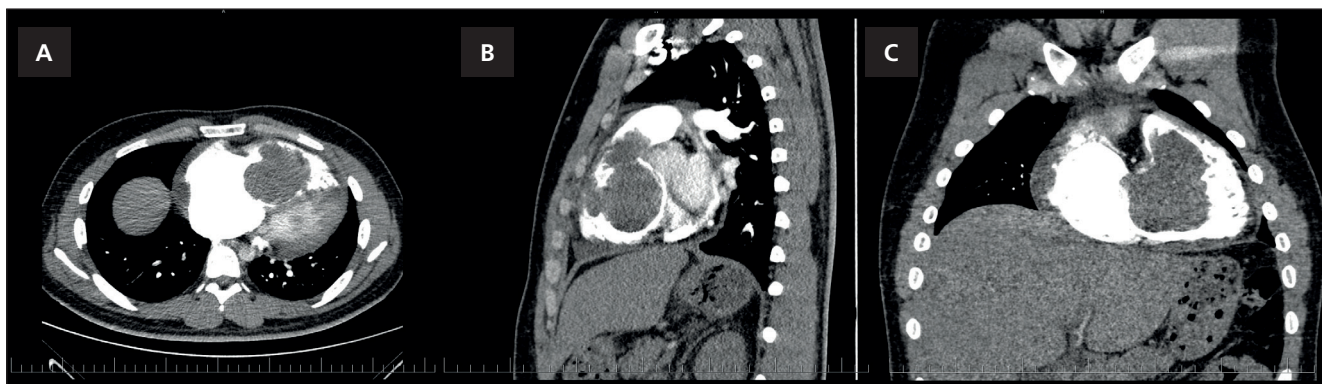


Fig. 2 – Computed tomography of the cardiac tumor. tumor. (A) Axial plane; (B) sagittal plane; (C) coronal plane.

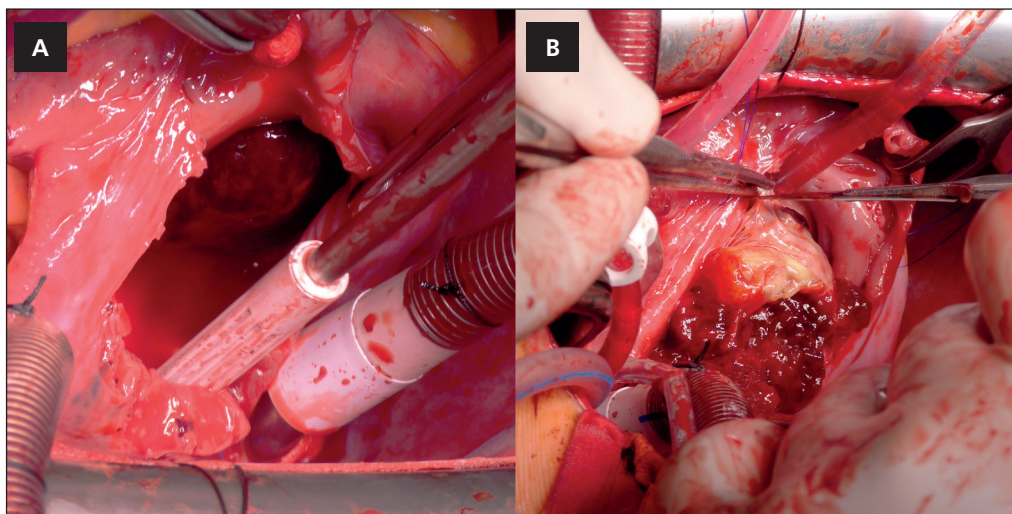


Fig. 3 – Intraoperative photographs: (A) view of the right atrium with a tumor obstructing nearly the entire annulus of the tricuspid valve; (B) dissection of the tumor's attachment from the leaflets of the tricuspid valve.

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-

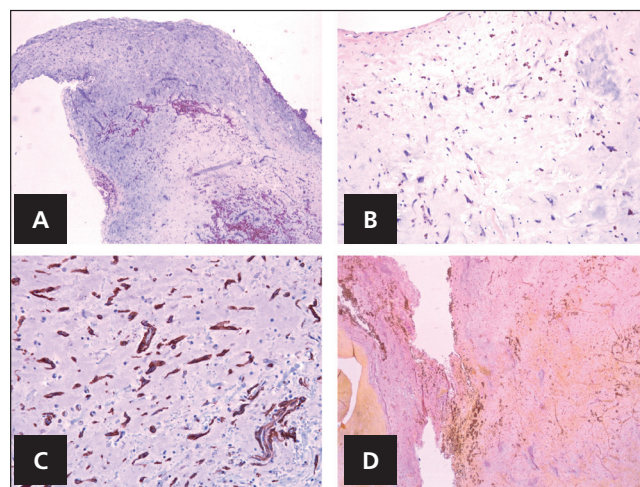


Fig. 4 – Histological appearance of the resected right ventricular myxoma. (A, B) Hematoxylin & eosin staining; (C) CD34 antibody; (D) mucicarmine staging; (A, D – 25×; B, C – 100×).

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

None.

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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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