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

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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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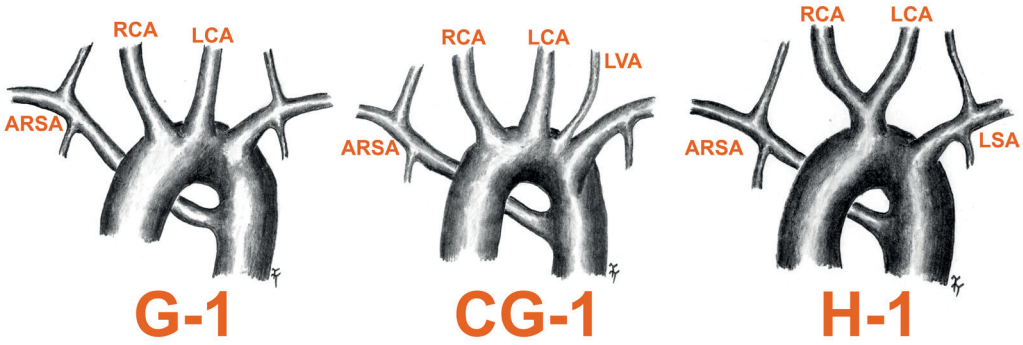

Fig. 1 – Adachi–Williams classification of arteria lusoria subtypes. ARSA – aberrant right subclavian artery; LCA – left carotid artery; LSA – left subclavian artery; LVA – left vertebral artery; RCA – right carotid artery.

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]

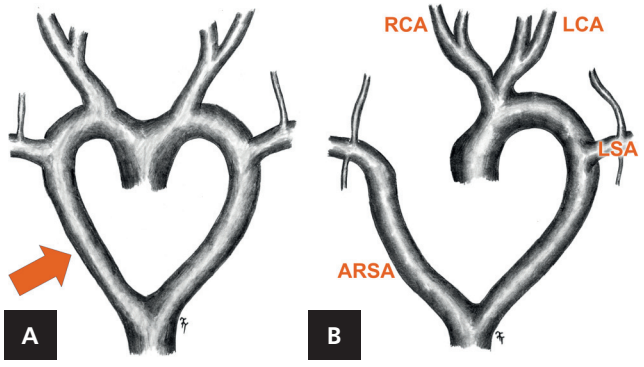

Fig. 2 – Depiction of embryonic development of ARSA. (A) Normal development of the aortic arch according to the double aortic arch hypothesis by Edwards and (B) degeneration of the right 4th aortic arch and proximal right dorsal aorta leading to ARSA. ARSA – aberrant right subclavian artery; LCA – left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

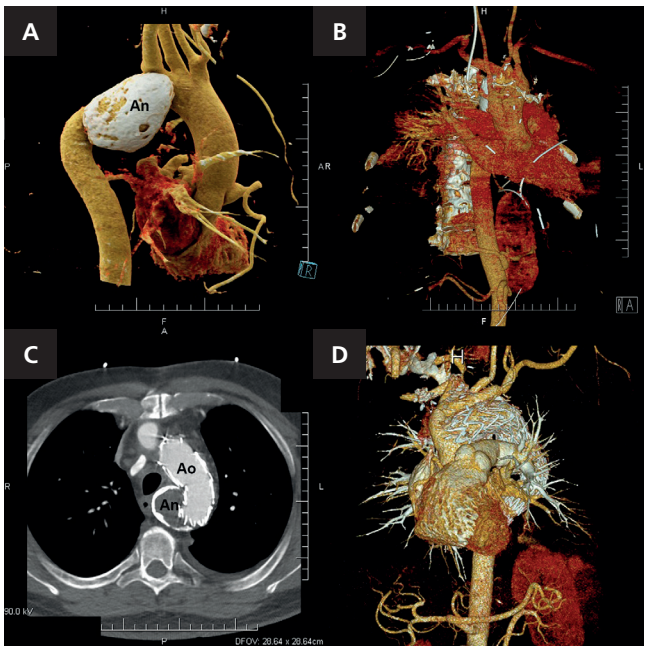

Fig. 3 – CT angiography images depicting the course of treatment. (a) Pre-operative 3D-reconstructed image, dorsal view, (B) 3D-reconstructed image after aortic debranching and before TEVAR implantation, ventral view, (C) image after TEVAR implantation, axial view and (D) 3D-reconstructed image after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

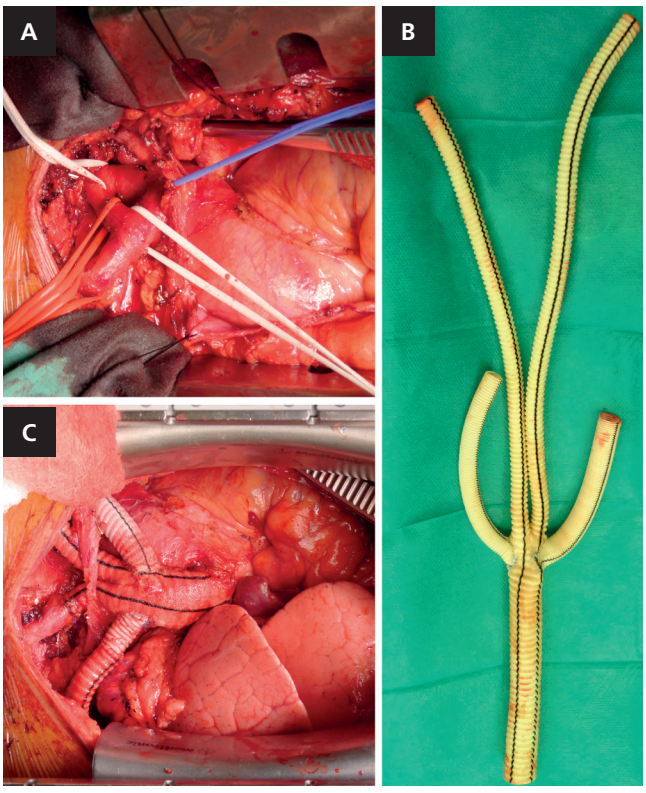

Fig. 4 – Images depicting surgical course of the first stage of treatment. (A) Intra-operative division of the supra-aortic branches using vessel loops, (B) 4-limbed Dacron graft used to construct the anastomoses and (C) post-anastomotic result of aortic debranching plus trans-anatomical bypass.

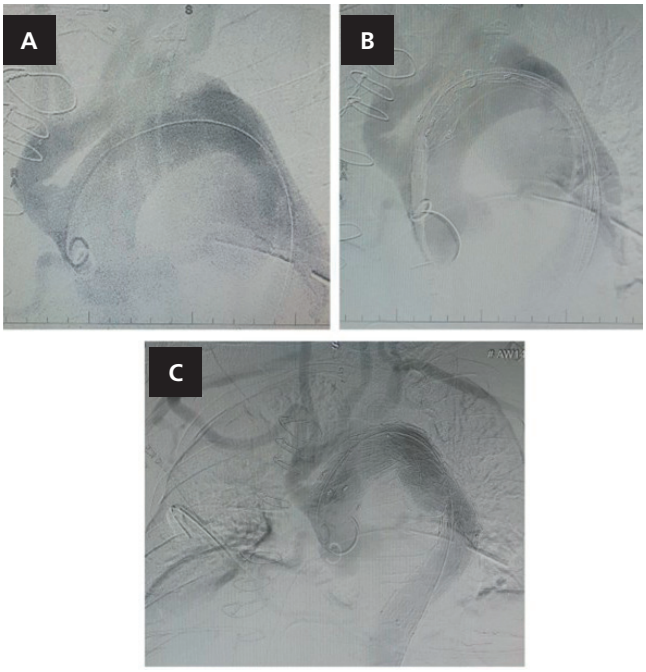

Fig. 5 – Images depicting course of the second stage of treatment under DSA guidance. (A) Pigtail catheter advancement into the ascending aorta, (B) stent graft advancement into the aortic arch, (C) final TEVAR result after stent graft deployment and ARSA occlusion.

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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