



Asymptomatic Giant Aneurysm of the Arteria Lusoria Treated by Debranching and Aneurysmal Resection

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The aberrant right subclavian artery (ARSA, arteria lusoria) is the most common intrathoracic vascular anomaly, affecting up to 2% of the population. However, aneurysms of congenital anomalies are extremely unusual and often present with dysphagia, dysphonia, or dyspnea due to compression of the surrounding structures. We report a case of an asymptomatic 57-year-old male with chronic kidney disease who was incidentally found to have a large aneurysm of the ARSA on preoperative computed tomography for laparoscopic nephrectomy. Surgery is unequivocally warranted as these aneurysms are associated with a high risk of complications, including thrombosis, embolism, and rupture. We debranched the ARSA, followed by anastomosis to the right carotid artery through a right neck incision. Subsequently, aneurysmal resection was performed through left thoracotomy. The patient had an uneventful postoperative recovery and was asymptomatic during the follow-up.

Key Words: Aberrant subclavian artery, Aneurysm, Computed tomography angiography, Thoracotomy, Left heart bypass

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INTRODUCTION

The aberrant right subclavian artery (ARSA) or arteria lusoria is formed by the involution of the right fourth vascular arch and proximal right dorsal aorta, coupled with persistence of the seventh intersegmental artery from the proximal descending thoracic aorta [1]. Although ARSA is the most common intrathoracic vascular anomaly with 0.2% to 2% of the population affected, aneurysms of this congenital anatomical variant are extremely rare [2]. As it conventionally passes behind the esophagus, a retroesophageal ARSA aneurysm (ARSAA) usually manifests as dysphagia lusoria, owing to esophageal compression. We describe the case of a 57-year-old male who was incidentally diagnosed with a large asymptomatic ARSAA during preoperative imaging for laparoscopic nephrectomy for chronic kidney

disease (CKD). Ethical approval (no. 441/2021) was obtained from Kasturba Medical College and Kasturba Hospital Institutional Ethics Committee on June 29, 2021. A written informed consent was obtained from the patient for publication and use of images.

CASE

A 57-year-old male with CKD and hypertension presented to the hospital with generalized weakness and urinary incontinence for one month. Ultrasonography revealed bilateral retrograde hydronephrosis due to staghorn calculus impaction. The patient was scheduled for right-sided percutaneous nephrolithotomy and laparoscopic nephrectomy of the non-functioning left kidney. During routine preoperative imaging, chest radiography showed asymmetrical

haziness and widening of the right upper mediastinum (Fig. 1). Computed tomography (CT) revealed an ARSA with fusiform aneurysmal dilatation of the proximal segment, measuring 5.6 cm×9.6 cm×7.2 cm (anteroposterior×transverse×superoinferior) with an eccentric mural thrombus of 4.5 cm in size (Fig. 2, 3). The patent aneurysmal luminal diameter was 3.8 cm×5.7 cm (anteroposterior×superoinferior). The esophagus and trachea were displaced anteriorly, with no signs of luminal compromise. The ARSA orifice was spared from the aneurysmal process, and there was no



Fig. 1. Chest X-ray scan showed mediastinal haziness in the upper right mediastinum.



Fig. 2. A coronal section of chest computed tomography revealed a fusiform aneurysmal dilatation of the proximal segment of an aberrant right subclavian artery, with the transverse (9.6 cm) and superior-inferior (7.2 cm) diameters marked.

evidence of Kommerell diverticulum (KD). The left subclavian artery originated proximal to the aneurysm. The aorta proximal and distal to the ARSA were measured 3.2 cm×3.6 cm and 2.6 cm×2.3 cm, respectively. Diffuse atherosclerotic changes with calcifications were noted in the aortic arch and descending thoracic aorta. The patient was diagnosed with non-KD-associated ARSAA and planned to undergo cervical debranching and open aneurysmal resection, deferring to laparoscopic nephrectomy.

Under general anesthesia, a double-lumen endotracheal tube was inserted using a bronchoscope to prevent potential vascular injuries. A right neck incision was made, and the right common carotid artery (RCCA) and right subclavian artery (RSCA) were looped. The RCCA was found to be diffusely atheromatous. The RSCA was ligated proximal to the origin of the right vertebral artery. An 8-mm polytetrafluoroethylene graft was used to create an anastomosis between the RCCA and RSCA. Then, the aorta was approached via left posterolateral thoracotomy at the 4th intercostal space in the mid-axillary line. The ribs were not cut to explore the mediastinum. After appropriate cannulation of the left inferior pulmonary vein and descending thoracic aorta, the patient underwent left heart bypass. The aneurysm was visualized, and the surrounding tissue was dissected (Fig. 4, 5). Proximally, the aortic arch was clamped distal to the origin of the left subclavian artery, and distally, the descending thoracic aorta was clamped. The aneurysm was excised along with a portion of the adjacent aorta, and a 28-mm Dacron graft was used to anastomose the aortic arch to the descending thoracic aorta (Fig. 6). The patient was placed on a centrifugal bypass pump for 47 minutes. During left heart bypass, normothermia was maintained with a mean arterial pressure of 80 mmHg and activated clotting time of 246 seconds. The postoperative course was

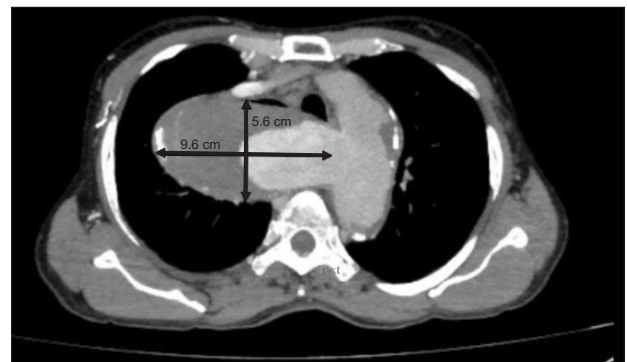


Fig. 3. An axial image of chest computed tomography revealed a fusiform aneurysmal dilatation of the proximal segment of an aberrant right subclavian artery, with the transverse (9.6 cm) and anteroposterior (5.6 cm) diameters marked.

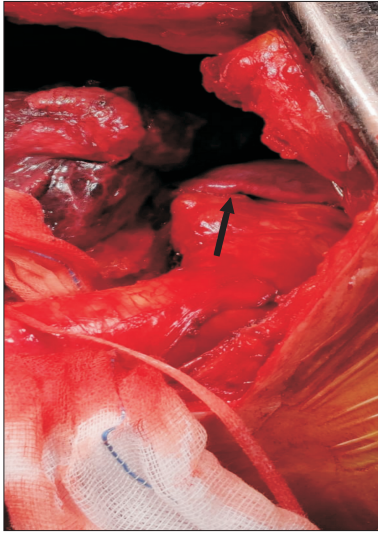


Fig. 4. An intraoperative photograph showed the aneurysmal portion of the aorta (arrow).

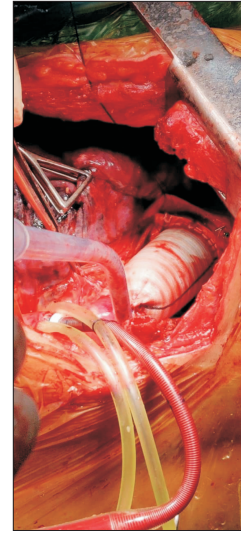


Fig. 6. An intraoperative photograph showed the Dacron interposition graft placed after excising the aneurysmal portion of the aorta.

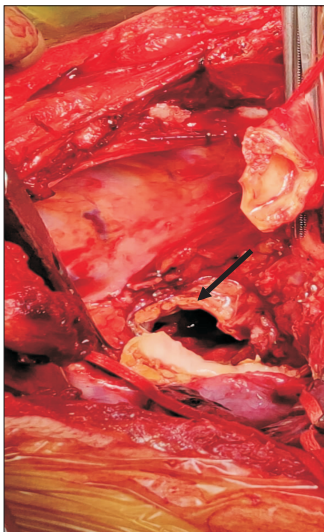


Fig. 5. An intraoperative photograph showed the opening of the blood vessel of the aneurysm (arrow) between the transected descending thoracic aorta.



Fig. 7. Postoperative computed tomography aortography showed the patent aortic graft and right carotid-subclavian graft with no residual aneurysm.

uneventful, and follow-up CT after one month revealed patent arteries without complications (Fig. 7).

DISCUSSION

After Hunauld initially described ARSA in 1735 [3], Bayford first labeled the clinical entity of “dysphagia lusoria” in 1787 in a female with a long-standing history of dysphagia who was found to have an ARSA at autopsy [4]. The word *lusoria* originates from the Latin phrase *lusus naturae*, which means “freak of nature.” ARSA may be classified into

three Neuhauser types based on the course of the artery [5]: retroesophageal (80%), between the trachea and esophagus (15%), and anterotracheal (5%). In contrast, Adachi and Williams classified ARSA into four basic morphological types [5]: Type I/G, the ARSA arises from the aortic arch as the final branch; Type II/CG-Type I/G with the left vertebral artery arising from the aortic arch; Type III/H, three branches arising from the aortic arch, including a common trunk of the CCAs (truncus bicaroticus), left subclavian artery, and ARSA; and Type IV/N, an aberrant left subclavian artery

arises from the right-sided aortic arch as the final branch, essentially a mirror image of Type I/G. In our case, the ARSA was retroesophageal and Type I/G.

ARSA is occasionally accompanied by vascular anomalies, such as KD (15%-60%), truncus bicaroticus (19%-29%), aneurysm of the anomaly (13%), and right-sided aortic arch (9%) [5]. The clinical presentation of ARSAs depends on their size and proximity to adjacent vital structures, potentially manifesting as dyspnea, dysphonia, or dysphagia. Surprisingly, in this case, the patient remained asymptomatic despite the aneurysm being 9.6 cm (transverse) in maximum dimension. After examining common symptoms using barium studies and chest radiography, the diagnosis of an ARSAA is usually confirmed using contrast-enhanced CT angiography [6-8]. CT allows for accurate localization and assessment of the integrity of the surrounding vital structures, enabling the surgeon to plan the best surgical approach and anticipate potential perioperative difficulties [9].

Surgery is the ideal treatment modality, as ARSAs have a high risk of thrombosis, embolization, and rupture [6]. If an aneurysm ruptures, massive blood loss can lead to systemic shock and death. Choosing the appropriate surgical approach is the most crucial step in preoperative planning because of the proximity of the vital structures. We agree that the repair technique should be built on the foundation of relevant concepts, as outlined by Harrison et al. [8]. Atherosclerosis is a potentially significant factor in the genesis of ARSAA, similar to traditional subclavian aneurysms [10]. In our case, extensive atherosclerosis in the aorta and carotid arteries in the background of CKD suggested a similar etiology. Atherosclerotic debris or a mural clot in the aneurysm may embolize via the vertebral and distal subclavian vasculature, if additional care is not taken during surgery. Additionally, improper revascularization of the subclavian vessels can result in ischemia and subclavian steal syndrome. To avoid these circumstances, management of the subclavian circulation should be prioritized before manipulating the aneurysm [8]. A right cervical incision should be employed to disconnect the RSCA proximal to the vertebral artery and transpose it to the RCCA by an end-to-side anastomosis [8]. Although a separate second incision may be required, exploration of the aneurysm should be performed via a left thoracotomy because proximal control of the aneurysm may be challenging from the right side [8]. Alternatively, proximal occlusion of the ARSA has been performed using midline sternotomy, or in the case of a right aortic arch, right-sided thoracotomy [7]. Particularly, if a single incision is desired, midline sternotomy can be extended into the neck [7]. With associated low mortality rates, aneurysmal excision and repair by grafting is the rec-

ommended approach for aneurysm management in adults [2]. Although the incidence of complications with the repair and reconstruction of an ARSAA is unknown, the rate in subclavian aneurysms generally tends to be as high as 26% [10]. Complications include recurrent laryngeal nerve palsy, upper limb ischemia, graft occlusion, brachial plexus injuries, chylothorax, esophagopleural fistula, and cardiac complications [10]. In our case, there were no complications, and the recovery was satisfactory.

Recently, hybrid procedures that combine traditional operative and endovascular modalities have been reported for the definitive treatment of ARSAA [11,12]. Thoracic endovascular aneurysm repair (TEVAR) in conjunction with extrathoracic revascularization for ARSAA is an attractive option because it mitigates the risk of mortality associated with thoracotomy, extracorporeal circulation, and major open surgery [12]. TEVAR is associated with lower in-hospital and perioperative mortality rates and pulmonary complication rates [13,14]. However, long-term mortality rates were similar [13,14]. Although several studies assessing the advantage of TEVAR over open repair reported mixed results in thoracic aortic aneurysms [13-15], very little research has been conducted on ARSAA in this context. While surgeons have advocated a hybrid approach in the management of isolated ARSAA, open repair is usually recommended, as the origin of the ARSA can only be ligated using thoracotomy [12,16]. In addition, long-term studies have shown that open repair has better long-term durability than the hybrid approach [13,15,17], making it a more reasonable option for younger populations and those from a lower socioeconomic background. Endovascular repair is associated with complications such as proximal site leakage, stent migration, and increased incidence of aortic dissection, requiring long-term follow-up [14,15]. However, we believe that the utility of the endovascular approach would be beneficial in high-risk patients with ARSAA who would not tolerate major surgery. Particularly, total endovascular repair may be performed as demonstrated by Mazzaccaro et al. [18] using the periscope technique in patients who may not tolerate extrathoracic anastomosis. Overall, there is insufficient evidence to highlight the potential advantages of endovascular or hybrid repair of ARSAs over open repair.

In conclusion, comprehensive preoperative surgical planning with CT is recommended to analyze the ARSAA anatomy and proximity of vital structures to reduce the risk of perioperative complications. Especially in young patients with a lower socioeconomic status and poor healthcare-seeking behavior, open repair may be preferred over hybrid or endovascular repair, owing to better durability and less requirement for long-term specialist follow-up. Although

initial studies are promising for hybrid repairs [16], more long-term studies are necessary to support the role of endovascular intervention in the definitive management of ARSAA.

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CONFLICTS OF INTEREST

The authors have nothing to disclose.

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