Sri Lanka Journal of Medicine **SLJM**

Case Report

Citation: Arudchelvam J, Jazeel MN. 2024. Right aortic arch and aberrant left subclavian artery. Sri Lanka Journal of Medicine, pp 40-44. DOI: https://doi.org/10.4038/sljm.v33i4.547

Right Aortic Arch and Aberrant Left Subclavian Artery Presenting with Dysphagia Lusoria: A Case Report and Review of the Literature

J Arudchelvam [1,2], MN Jazeel [2]

[1]Department of Surgery, Faculty of Medicine, University of Colombo
[2]National Hospital Colombo, Sri Lanka

Correspondence:
J Arudchelvam
Email: joelaru@srg.cmb.ac.lk
: https://orcid.org/0000-0002-4371-4527

ABSTRACT

Right aortic arch (RAA) occurs in 0.05% to 0.1% of the individuals. Dysphagia lusoria is the difficulty in swallowing due to the compression of the oesophagus due to aberrant vessels. We report a case of aberrant left subclavian artery (ALSA) arising from the RAA resulting in dysphagia in a 59-year-old male. However, he was able to take meals with minimal difficulties. A computerised tomographic scan (CT) of the chest revealed a RAA and an ALSA running posterior to the oesophagus compressing it. Considering that the patient can take meals and maintain his weight, he was managed conservatively.

Keywords: Aberrant left subclavian artery, development of the aorta, right aortic arch

INTRODUCTION

Dysphagia lusoria (Latin - "lusus naturae" - freaks of nature) is difficulty in swallowing due to the compression of the oesophagus by an aberrant vessel. This was first described in a patient with an aberrant right subclavian artery (1). The common vascular anomalies causing dysphagia lusoria are the aberrant right subclavian artery (ARSA), the right aortic arch (RAA) with aberrant left subclavian artery (ALSA) and the double aortic arch (DAA) (2) (3). We report a case of ALSA arising from the RAA resulting in dysphagia in a 59-year-old male.

CASE REPORT

A 59-year-old male presented with a history of slowly progressive dysphagia for 18 months. He did not have any respiratory symptoms. The upper gastrointestinal endoscopy did not reveal any mural pathology. A pulsatile bulge was noted on the posterior wall of the oesophagus which is about 23 cm to 24 cm from the incisor teeth. The endoscope passed through this area without any difficulty. A computerised tomographic scan (CT) of the chest revealed a right aortic arch (RAA). An aberrant left subclavian artery (ALSA) was arising from the distal RAA and was running towards the left side, posterior to the oesophagus was noted. The ALSA was compressing the oesophagus. Kommerell diverticulum; an outpouching at the origin of the ALSA was observed (Figure 1) (Figure 2). However, the Kommerell diverticulum was not compressing the oesophagus. In the CT no other pathologies were revealed. Even though the

Received: 2024-09-12 **Accepted revised version: 2024-12-13** **Published: .....**

patient had dysphagia, he was able to have meals with minimal difficulty. It was not associated with weight loss.

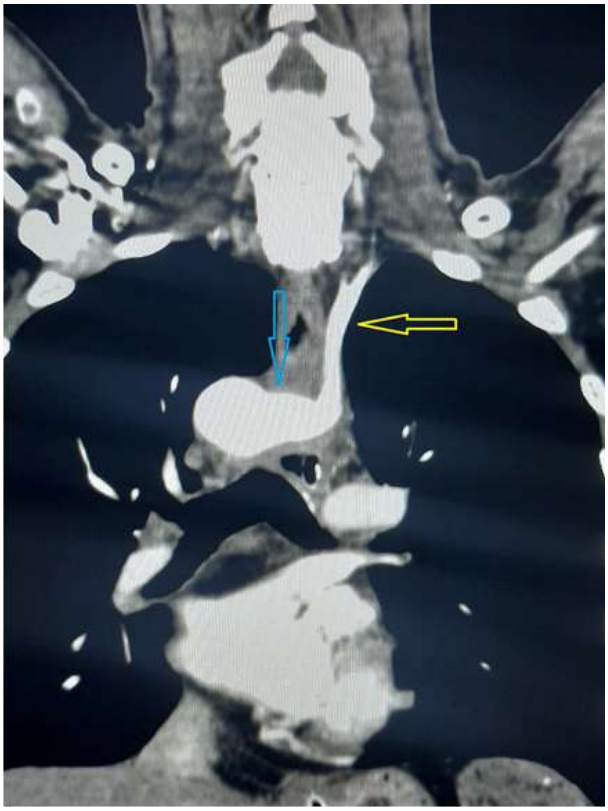

Figure 1: Computerised tomographic scan (CT) showing Kommerell diverticulum (blue arrow) and the aberrant left subclavian artery (ALSA) (yellow arrow)

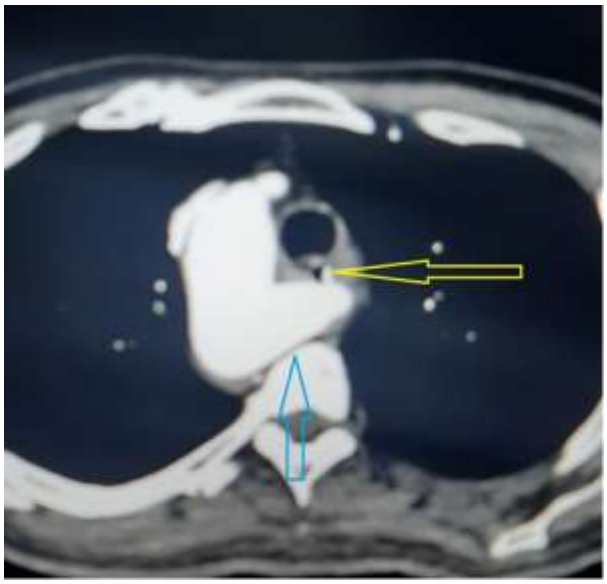

Figure 2: Computerised tomographic scan (CT) (axial view) showing Kommerell diverticulum (blue arrow) and the oesophagus (yellow arrow)

On further evaluation, it was found that the patient had poor cardiac and respiratory functions. Considering the ability of the patient to have meals, maintain his weight and the high risk for surgical intervention, it was decided to manage him conservatively.

DISCUSSION

Aortic arch abnormalities occur in 1% to 3% of the population (4). When the aortic arch crosses over the right bronchus it is called the RAA. In the case of the RAA, the descending thoracic aorta runs inferiorly on the right side of the spinal column. Most patients with aortic arch anomalies with oesophageal compression remain asymptomatic (60%-80%) (5). The symptomatic patients with oesophageal compression present with dysphagia in 91% of the cases. Others present with chest pain (20%), Horner syndrome and symptoms due to rupture of the Kommerell diverticulum (5). Patients can also develop symptoms of tracheal and oesophageal compression soon after birth.

Embryonic development

During embryonic development, two primitive aortas form on the ventral and dorsal sides of the foregut (ventral and dorsal aortas). The two ventral aortas join at the proximal end to form the aortic sac, while the rest of the ventral aorta remains as right and left aortic horns (Figure 3).

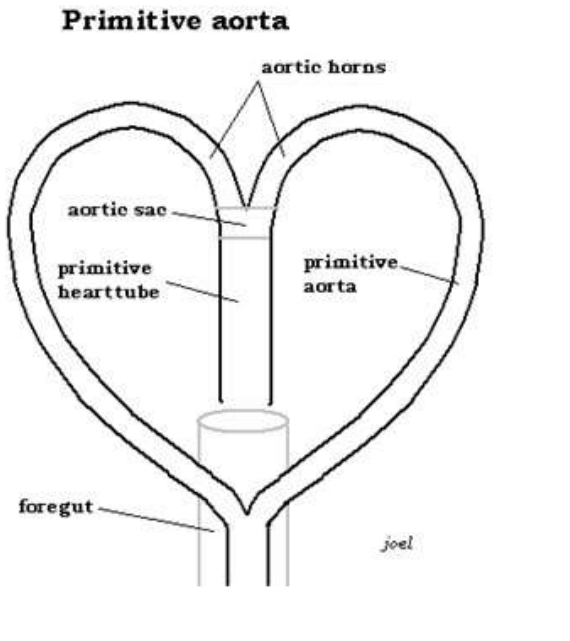

Figure 3: Primitive aorta

During further development, a series of pharyngeal arches connect the aortic sac with the dorsal aorta (Figure 4).

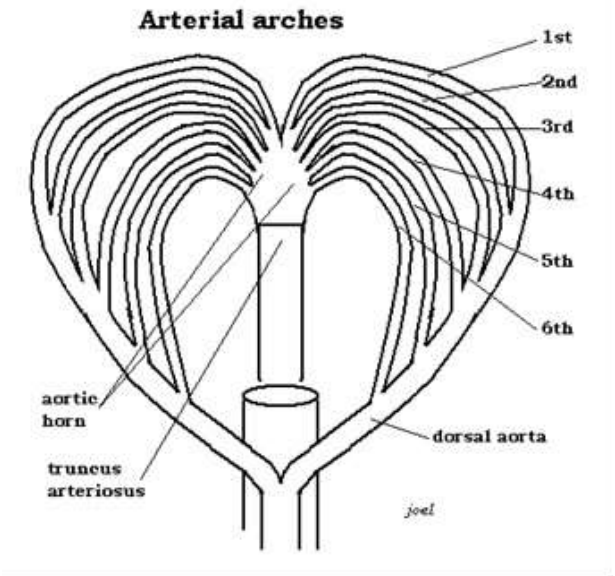

Figure 4: Pharyngeal arterial arches

During further development, some pharyngeal arch arteries disappear and the remaining parts unite to form the adult pattern of the aorta (Figure 3, Figure 4).

The adult aorta is formed by the fusion of (Figure 5) the part of the aortic sac, left horn, left 4th pharyngeal arch and left dorsal aorta.

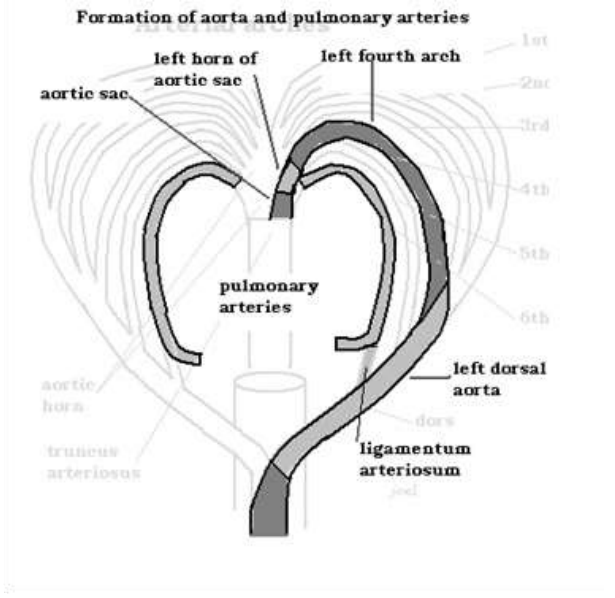

Figure 5: Development of adult pattern aorta

Variations in the fusion and regression of the arterial components result in variations of the aortic arch.

Right-sided aortic arch (RAA)

The RAA arises as a result of the persistence of the right dorsal aorta and the disappearance of the left dorsal aorta (Figure 6). It occurs in 0.05% to 0.1% of the individuals (6) (Figure 6). RAA is associated with ALSA. This ALSA runs posterior to the oesophagus resulting in compression of the oesophagus, resulting in dysphagia. This is aggravated if Kommerell diverticulum (abnormal dilatation at the site of the origin of the ALSA) is present.

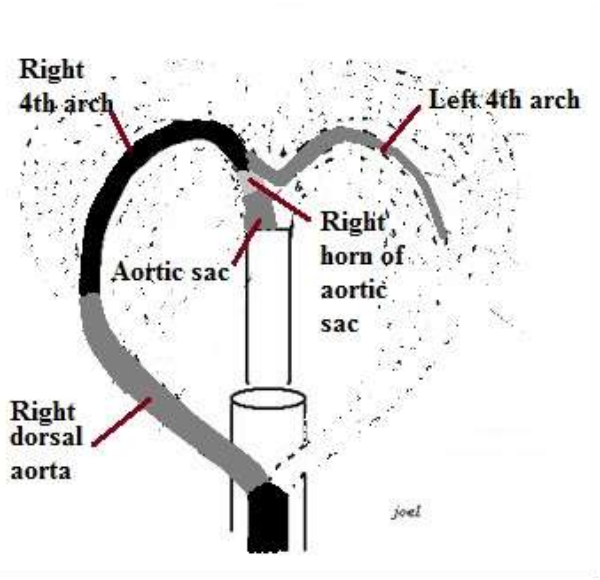

Figure 6: Right-sided aortic arch (RAA)

Patients with RAA can be asymptomatic or present due to the compression of the airway or the oesophagus. Other patients present due to the associated congenital heart diseases (e.g. tetralogy of Fallot, truncus arteriosus) or with features of chromosomal abnormalities (e.g. Di George syndrome - 22q11) (7) (8). patient mentioned in this case did not have any evidence of congenital heart disease or chromosomal abnormalities.

Compression symptoms often appear later in life due to the rigidity of the arteries caused by the age-related changes and atherosclerosis (9), the aneurysmal dilatation of the Kommerell diverticulum and motility abnormalities of the oesophageal wall with ageing. Even though the above-mentioned patient had hypertension, the arteries were not significantly thickened on imaging.

The investigations done to diagnose RAA and dysphagia lusoria are contrasted with swallow imaging, upper gastrointestinal endoscopy (UGIE)

and CT scan. The contrast swallow is a useful study to show the functional (dynamic) and the anatomical site of the obstruction. The site of compression is shown as an indentation at the level of the aortic arch. However, the site of compression may be missed if the lateral or oblique views are not obtained.

During the UGIE, the area of vascular compression appears as a pulsatile mass on the posterior wall of the oesophagus. Endoscopic ultrasound can be used to visualise the pulsatile vessel.

The gold standard for the diagnosis of the oesophageal compression and the vascular anomaly is the CT or magnetic resonance (MR) scan. The CT scan will also facilitate the planning of the intervention.

Management

Indications for interventions include compression symptoms, development of arch aneurysm or aneurysm of the Kommerell diverticulum (3 cm) and the dissection of the descending thoracic aorta (10).

The management options include conservative and surgical. Non-complicated or complicated cases with mild symptoms can be managed conservatively. The conservative management includes dietary modifications, reassurance and follow-up.

There are many surgical interventions described. The first such intervention was ligation and disconnection of the subclavian artery (SCA) through a cervical incision (11). However, ligation of the left SCA proximal to the origin of the vertebral artery can result in stroke in 9.1% of patients. Therefore, the current approaches to reduce the risk of stroke are to do a common carotid to SCA bypass followed by ligation and disconnection of the SCA. The other option includes the transposition of the SCA to the common carotid artery (12).

Therefore, vascular anomalies as a cause of compression of the oesophagus should be carefully evaluated with CT or MR scans. The associated congenital heart diseases should be excluded from further evaluation. Patients with severe symptoms and associated complications are offered surgical management after careful planning.

Author declaration

Acknowledgement
None.

Authors' contributions:
Conceptualization and Design: JA, MNJ; Literature Survey: JA; Manuscript Writing: JA; Critical Review and Editing: JA, MNJ; Images: JA.

Conflicts of interest:
The authors declare that there is no financial or non-financial conflict of interest.

Funding statement:
Self-funded.

Ethics statement:
Written informed consent was obtained from the patient before they participated in the case, ensuring their understanding of the purpose, procedures, and potential implications involved.

Statement on data availability:
All data generated during this study are available upon request from the corresponding author.

REFERENCES

1. Panduranga P, Al-Delamie T, Ratnam L, Al-Mukhaini M, Zachariah S. Repair of Kommerell's diverticulum with aberrant left subclavian artery in an elderly patient with right aortic arch and dysphagia lusoria. J Card Surg. 2011; 26: 637–640. https://doi.org/10.1111/j.1540-8191.2011.01344.x
2. McNally PR, Rak KM. Dysphagia lusoria caused by persistent right aortic arch with aberrant left subclavian artery and diverticulum of Kommerell. Dig Dis Sci. 1992; 37: 144–149. https://doi.org/10.1007/bf01308358
3. Zhao J, Liao Y, Gao S. Right aortic arch with retroesophageal left ligamentum arteriosum. Tex Heart Inst J. 2006; 33: 218-221. https://pubmed.ncbi.nlm.nih.gov/16878631/
4. Levitt.B, Richter J. E. Dysphagia lusoria: a comprehensive review. Diseases of the Esophagus. 2007; 20: 455–460. https://doi.org/10.1111/j.1442-2050.2007.00787.x
5. Cina CS, Althani H, Pasenau J, Abouzahr L. Kommerell ' s diverticulum and right-sided aortic arch : a cohort study and review of the literature. J Vasc Surg. 2004; 39: 131–139. https://doi.org/10.1016/j.jvs.2003.07.021
6. Asherson N. Bayford David. His syndrome and sign of dysphagia lusoria. Ann R Coll Surg Engl. 1979; 61: 63-67. https://pubmed.ncbi.nlm.nih.gov/369446/
7. McElhinney DB, Hoydu AK, Gaynor JW, Spray TL,

Goldmuntz E, Weinberg PM. Patterns of right aortic arch and mirror-image branching of the brachiocephalic vessels without associated anomalies. Pediatr Cardiol. 2001; 22: 285-291. https://doi.org/10.1007/s002460010231

8. Evans WN, Acherman RJ, Berthoty D, Mayman GA, Ciccolo ML, Carrillo SA, Restrepo H. Right aortic arch with situs solitus. Congenit Heart Dis. 2018; 13: 624-627. https://doi.org/10.1111/chd.12623
9. Myers PO, Fasel JHD, Kalangos A, et al. Arteria lusoria: Developmental anatomy, clinical, radiological and surgical aspects. Ann Cardiol Angeiol (Paris). 2010; 59: 147-154. https://doi.org/10.1016/j.ancard.2009.07.008
10. Albacker TB, Roselli EE, Pettersson GB et al. Surgical management of right aortic arch with tailored surgical approach. J Card Surg. 2012; 27: 511–517. https://doi.org/10.1111/j.1540-8191.2012.01488.x
11. Carrizo GJ, Marjani MA. Dysphagia lusoria caused by an aberrant right subclavian artery. Tex Heart Inst J. 2004; 31: 168-171. https://pubmed.ncbi.nlm.nih.gov/15212130/
12. Benjamin O Patterson, Peter JH, Christoph N, Ronald MF, et al. Management of the left subclavian artery and neurologic complications after thoracic endovascular aortic repair. Journal of Vascular Surgery. 2014; 60: 1491 - 1498. https://doi.org/10.1016/j.jvs.2014.08.114