Triple thoracic anomalies: right-sided aortic arch, aberrant left subclavian artery and Kommerell's diverticulum

Subodh Kumar,[1] Mayank Mishra [2]

[1]Department of Emergency Medicine, All India Institute of Medical Sciences Rishikesh, Rishikesh, India
[2]Department of Pulmonary Medicine, All India Institute of Medical Sciences Rishikesh, Rishikesh, India

Correspondence to
Dr Mayank Mishra;
virgodrmayank@gmail.com

Accepted 3 August 2021

DESCRIPTION

A 30-year-old diesel-engine-repair worker presented with cough and nasal regurgitation while swallowing liquids for last 10 years, impaired balance while walking and hand tremors for last 5 years, and mild exertional dyspnoea for last 1 year. General examination was normal and respiratory examination revealed mild tachypnoea and occasional scattered crepitations, while neurological assessment disclosed a drunken gait and positive cerebellar signs. Detailed neurosurgical evaluation subsequently identified the patient's symptoms to be due to atlantoaxial dislocation with type 1 Chiari malformation, and he improved following neurosurgical correction.

However, during work-up, chest imaging revealed three rare anomalous findings: right-sided aortic arch (RAA), aberrant left subclavian artery (ALSA) and an aortic aneurysmal dilatation (Kommerell's diverticulum, KD) at the base of ALSA (figure 1). A clinical diagnosis of incidentally detected triple thoracic aortic anomalies (RAA +ALSA+KD) was arrived at and communicated to the patient. The need for regular, close follow-up was emphasised given a possibility of future symptom emergence due to this condition, in which case surgical correction was kept as the back-up treatment plan.

It is important for the clinician to be aware of these entities not only because of their rarity but also because of their potential clinical significance. Although they are often asymptomatic and discovered as an incidental radiographic finding, they can produce symptoms due to compression of adjacent mediastinal structures.

Vascular abnormalities of the aorta account for 15%–20% of all congenital cardiovascular diseases.[1] These malformations arise from errors in embryologic development or abnormal persistence of various vascular structures. Organogenesis involves formation of six pairs of branchial arches between week 2 and week 7.[2] The normal adult aortic arch is formed from the fourth primitive left aortic arch, while the right fourth arch disappears. A congenital aortic arch anomaly may arise as a consequence of faulty development of these branchial arches. Based on abnormal anatomical development, aortic arch anomalies are essentially of four types—left-sided aortic arch (LAA), RAA, double aortic arch and cervical aortic arch. Details of these have been described elsewhere,[2] except the RAA which is pertinent to our case and, hence, has been elaborated here.

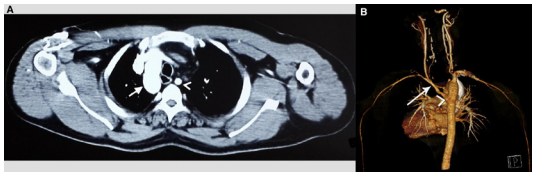

Figure 1 (A) CT angiogram (axial section) showing right-sided aorta (arrow) and aberrant left subclavian artery (ALSA) (arrowhead). (B) CT angiogram with three-dimensional-reconstructed image (posterior view) showing ALSA (arrow) and Kommerell's diverticulum (arrowhead).

RAA arises because of dissolution of the left primitive fourth arch and persistence of the right. It has an estimated prevalence of 0.04%–0.1% based on a necropsy series.[3] It may be asymptomatic, only to be discovered incidentally as widened mediastinum on imaging done for other reasons, and may mimic a mediastinal mass. There are three types of RAA—type I (mirror image RAA), type II (with ALSA) and type III (with isolation of left subclavian artery).[4] Type I RAA represents a mirror image of the normal LAA, with its vascular branching pattern comprising of the left brachiocephalic artery as the first branch, and the right common carotid and right subclavian arteries as the second and third branches, respectively. Congenital cyanotic cardiac defects may occur in up to 75% of these patients, which require surgical correction. Type II RAA is the most common. Herein, the ALSA may originate either directly from the RAA as its fourth branch or from an aneurysmal dilatation called KD (as in the present case),[5] to course retroesophageally and supply the left upper extremity. Compression of the adjacent trachea or oesophagus by the dilated aneurysm or by formation of vascular rings may

Learning points

- Vascular anomalies of the aorta are rare conditions that may or may not be symptomatic.
- Their incidental discovery deserves mention given the unusual clinical entities that are associated with them.
- Clinicians must be aware of these anomalies and symptoms which they may produce by compression of adjacent mediastinal structures or vascular rupture.

To cite: Kumar S, Mishra M. *BMJ Case Rep* 2021;**14**:e245539. doi:10.1136/bcr-2021-245539

cause respiratory distress or dysphagia lusoria, respectively. Occasionally, catastrophic clinical consequences may arise due to aneurysmal rupture. Treatment involves surgical resection of KD and correction of any cardiac anomalies if present. Type III RAA is associated with isolation of the left subclavian artery and produces ischaemic symptoms and reduced blood pressure in the left upper limb. It is the least common variety seen in only 0.8% of patients.[6]

Contributors Both the authors in this paper were equally involved in conception, design, literature review, preparation of first draft, critical revision for important intellectual content and final approval of the submitted version.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

ORCID iD
Mayank Mishra http://orcid.org/0000-0001-7982-6267

REFERENCES

1 Goldmuntz E. The epidemiology and genetics of congenital heart disease. *Clin Perinatol* 2001;28:1–10.
2 Stojanovska J, Cascade PN, Chong S, *et al*. Embryology and imaging review of aortic arch anomalies. *J Thorac Imaging* 2012;27:73–84.
3 Hastreiter AR, D'Cruz IA, Cantez T, *et al*. Right-sided aorta. I. Occurrence of right aortic arch in various types of congenital heart disease. II. right aortic arch, right descending aorta, and associated anomalies. *Br Heart J* 1966;28:722–39.
4 Edwards JE. Anomalies of the derivatives of the aortic arch system. *Med Clin North Am* 1948;32:925–49.
5 van Son JA, Konstantinov IE. Burckhard F. Kommerell and Kommerell's diverticulum. *Tex Heart Inst J* 2002;29:109–12.
6 Hara M, Kitase M, Satake M, *et al*. A case of right-sided aortic arch with isolation of the left subclavian artery: CT findings. *Radiat Med* 2001;19:33–6.