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

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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 emphasized 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. 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. 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, except the RAA which is pertinent to our case and, hence, has been elaborated here.

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.

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).
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.  

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.

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REFERENCES  