

Double aortic arch with right positioned descending thoracic aorta and coexistent aortic kinking

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DESCRIPTION

An 82-year-old woman presented with fever, severe dyspnoea and pneumonia, which required non-invasive ventilation and intravenous antibiotics. She had a history of recurrent airway infections with inspiratory stridor and dysphagia. During the pneumonia work up, in order to unveil a predisposing condition for pneumonia, and rule out alternative diagnoses, a high resolution CT scan of the chest revealed the presence of a double aortic arch. Following this finding, a contrast-enhanced three-dimensional CT scan confirmed the diagnosis of incomplete double aortic arch (IDAA) (figure 1) with right positioned descending thoracic aorta and coexistent aortic kinking (figure 2). IDAA is a rare congenital abnormality resulting from atresia rather than complete involution of the distal left arch in a fibrous cord between the left arch and descending

thoracic aorta.¹ This anatomic anomaly is due to the persistence of the fourth right and left arches and dorsal aortas, resulting in the abnormal formation of vascular rings encircling the trachea and oesophagus. IDAA is generally diagnosed in childhood because of the early onset of symptoms related to tracheal and oesophageal obstruction. Anecdotal cases have been reported in adult patients, who usually became symptomatic because of asthma-like symptoms or swallowing difficulties as a consequence of the compression of the trachea and oesophagus by the abnormal aortic arches.¹⁻³

Our patient was evaluated at the cardiac surgery unit, but because of age, comorbidities and the mildness of her symptoms, medical management of potential IDAA-related complications and short-term follow-up were chosen.

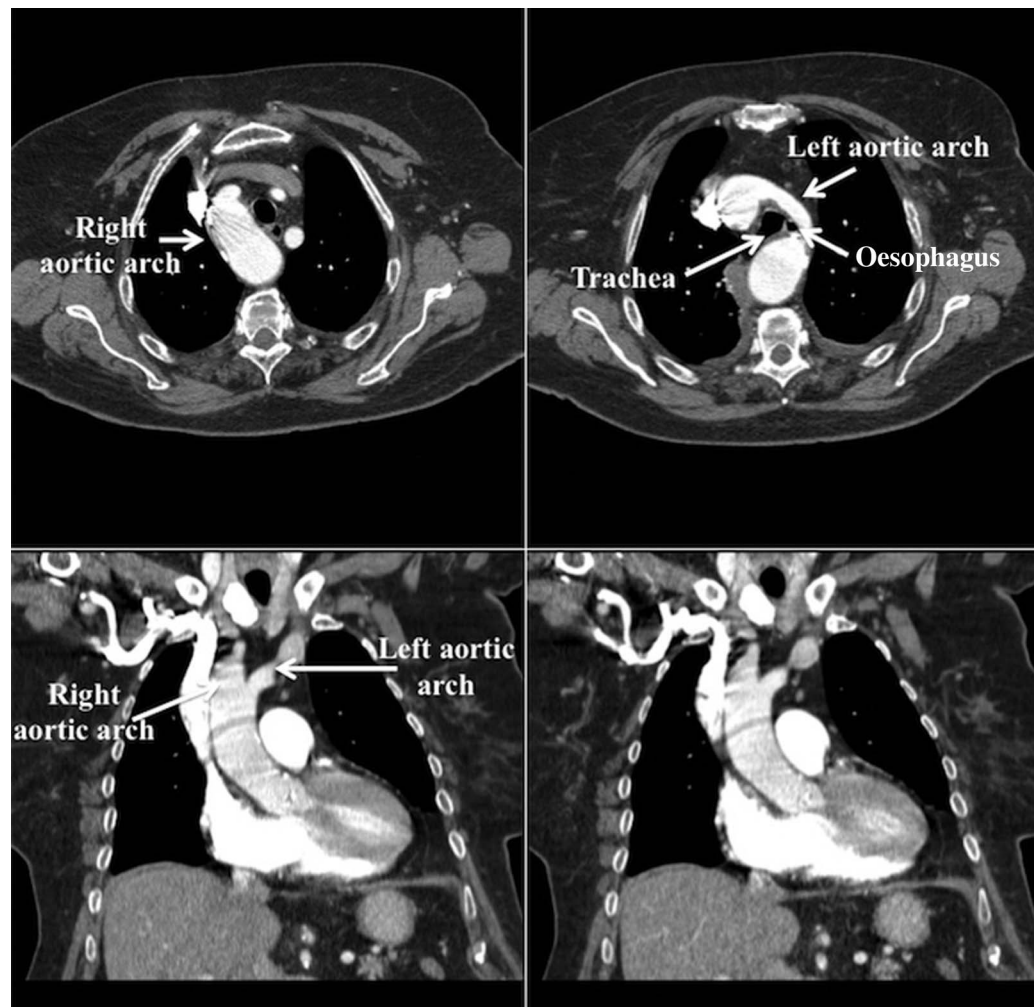


Figure 1 Contrast-enhanced CT scan of the chest showing right-sided dominant aortic arch and left aortic arch encircling the trachea and oesophagus.



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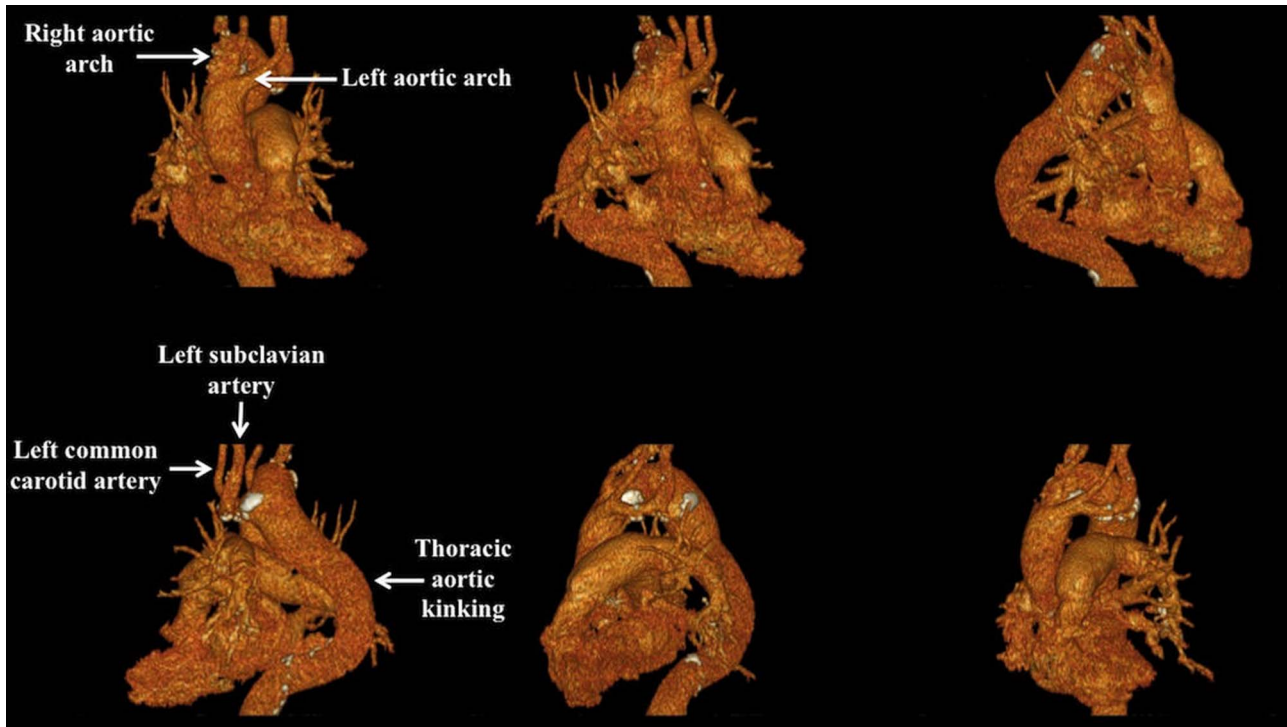


Figure 2 Contrast-enhanced three-dimensional CT scan showing incomplete double aortic arch with right positioned descending thoracic aorta and coexistent aortic kinking.

Learning points

- ▶ Vascular rings represent 1–2% of all congenital diseases of the heart and aorta.
- ▶ In most cases, this abnormality is diagnosed in childhood because of symptoms related to oesophageal or tracheal obstruction.
- ▶ Symptomatic adult patients usually experience asthma-like symptoms and swallowing difficulties because of the compression of the trachea or oesophagus by the abnormal aortic arches.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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