Circumflex retroesophageal right aortic arch: rare differential of mediastinal widening

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DESCRIPTION
A 48-year-old man presented to the outpatient department with complaints of inspiratory stridor and occasional dysphagia. Chest radiograph revealed bilateral symmetric widening of the superior mediastinum at the level of aortic arch with descending aorta seen to the left of spine. No lateral displacement of tracheal air column was noted (figure 1A). The patient was referred to us for a CT angiography (CTA) with a provisional diagnosis of an aneurysm or a mediastinal mass. Review of CTA images revealed a right-sided aortic arch crossing the midline posterior to the trachea and the oesophagus in the upper mediastinum and further descending on the left side of spine (figure 1B–E). A Kommerell’s diverticulum arising from the anterosuperior aspect of aorta, on the left side, gave rise to an aberrant left subclavian artery (LSCA). The circumflex arch was ectatic and measured 37 mm in its maximum diameter. Moderate compression on the posterior aspect of trachea by the ectatic circumflex arch was noted (figure 1E–F).

A circumflex retroesophageal right aortic arch is an extremely uncommon anomaly. Embryologically, a circumflex right aorta with an aberrant LSCA results from regression of the left dorsal aorta between the left common carotid artery and the LSCA, with the right dorsal aorta forming the distal portion of the arch. It has also been postulated that the third aortic arch gives rise to the proximal aortic arch while the distal arch is formed by the fourth arch, which can explain the elongated transverse arch as well as the relatively higher position of the proximal aortic arch. Even in the absence of a ligamentum arteriosum, the circumflex aorta itself, especially if ectatic, can cause symptomatic...
tracheal compression, as in this case, which makes identification of this anomaly pertinent; as such, patients may require aortic uncrossing procedure, where the distal aortic arch is transected, the retrooesophageal segment mobilised and brought anterior to trachea followed by its anastomosis with the ascending aorta.²

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REFERENCES

Learning points
► Circumflex retrooesophageal right aortic arch can mimic an aneurysm or a superior mediastinal mass on a chest radiograph.
► Identification of this anomaly is important as, even in the absence of a ligamentum arteriosum, the circumflex aorta itself, especially if ectatic, can cause symptomatic tracheal compression and such patients may require aortic uncrossing procedure.