

A rare configuration of a complete vascular ring with the ductus arteriosus arising from the aberrant subclavian artery

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DESCRIPTION

A 2-year-old child, who presented to the emergency room with breathing difficulty, dysphagia and failure to thrive, underwent a cardiac CT angiography (CTA) to look for presence of any vascular ring. Review of CTA images showed situs solitus, levocardia and a left-sided aortic arch. A Kommerell's diverticulum gave origin to the aberrant right subclavian artery (SCA) and a patent right arterial duct which connected to the ostial segment of the right pulmonary artery (figure 1). A vascular ring, completed by segment of aortic arch on the left side along with the retroesophageal Kommerell's diverticulum, ductus on the right, and pulmonary artery on the anterior aspect, was thus formed encircling and compressing the trachea (figure 2) and oesophagus.

Complete vascular rings are congenital anomalies of the aortic arch system, which causes airway and esophageal compression. The described case is a rare pattern of vascular ring with a patent arterial duct arising contralateral to the side of the aortic

arch. This case also demonstrates the importance of cross sectional imaging by cardiac CT, which helps in accurate identification of the morphology.

A patent ductus arteriosus (PDA) which represents 5%–10% of all congenital heart disease, is generally caused by persistence of the left sixth aortic arch between a left pulmonary artery (PA) close to the pulmonary artery bifurcation and the aorta just distal to the left SCA.¹ A PDA is generally ipsilateral to the side of the aortic arch with origin and embryology as described above. An arterial duct contralateral to the side of aortic arch is rare, with the possible origins of the duct from brachiocephalic artery, SCA (with/without isolation), aberrant SCA (with/without Kommerell's diverticulum) or the common carotid artery in the presence of aberrant SCA.

An aberrant right SCA is caused by disappearance of the right fourth arch between the common carotid and seventh cervical intersegmental artery and persistence of the distal right dorsal aorta. A Kommerell's diverticulum which represents persistence of the right sixth arch component that

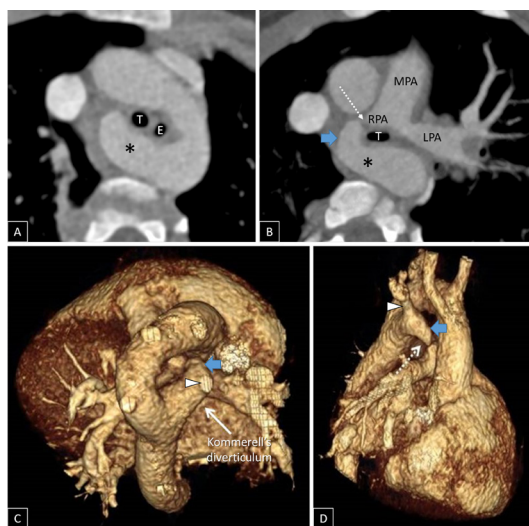


Figure 1 Oblique axial images (A,B) and volume rendered images (C,D) reveal a left-sided aortic arch with a Kommerell's diverticulum (*) giving origin to the aberrant right subclavian artery (arrowhead) and a patent right arterial duct (blue arrow) which was seen joining the ostial segment of the right pulmonary artery (RPA). Significant stenosis (dotted arrow) was noted at the site of insertion of right arterial duct into the right pulmonary artery. E, esophagus; LPA, left pulmonary artery; MPA, main pulmonary artery; T, trachea.

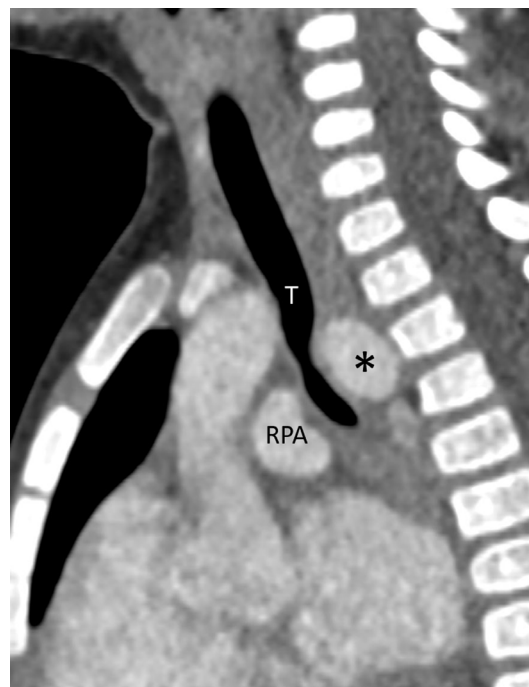


Figure 2 Sagittal image reveals compression of the trachea (T) between the Kommerell's diverticulum (*) posteriorly and the right pulmonary artery (RPA) anteriorly.

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forms a right-sided ductus arteriosus is seen in 15%–30% of cases of aberrant right SCA.² In the setting of such a diverticulum, the PDA arises from the junction between the diverticulum and the normal-sized SCA, and forms a complete vascular ring. If there

is no retroesophageal diverticulum, the PDA is usually present on the side of the aortic arch. A complete vascular ring with the described configuration is also more commonly observed in a right aortic arch with aberrant left SCA as a retroesophageal diverticulum is seen in 60% of the cases.²

Learning points

- ▶ A patent ductus arteriosus contralateral to the side of aortic arch is rare, with the possible origins of the ductus from brachiocephalic artery, subclavian artery (SCA), aberrant SCA with or without Kommerell's diverticulum or even the common carotid artery in the presence of an aberrant SCA.
- ▶ A complete vascular ring with a retroesophageal Kommerell's diverticulum and a ductus contralateral to the side of aortic arch is exceedingly rare, especially in a left-sided aortic arch.
- ▶ Cross-sectional imaging by cardiac CT plays an important role in identification of the accurate morphology in cases of vascular rings.

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