Double aortic arch without vascular ring: an unusual variation

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A 3-day-old female newborn was transferred to our hospital due to cyanosis after delivery. The echocardiogram confirmed the diagnosis of tetralogy of Fallot (TOF), consisting of a subaortic ventricular septal defect (VSD), overriding aorta, and right ventricular outflow tract stenosis with a pressure gradient of 67 mmHg. Moreover, the aortic valve was noted to be bicuspid, and on suprasternal images, a double lumen was noted in the descending aorta distal to the left subclavian artery. In addition to the confirmed TOF diagnosis, a suspected double aortic arch (DAA) was diagnosed. Cardiac multidetector computed tomography (MDCT) was ordered for further workup. MDCT was performed using two different 256-detector-row CT scanners (SOMATOM® Definition Flash; Siemens Healthcare, Forchheim, Germany). With the scanner, the following parameters were used: 24 mA, 80 kV, 16 × 0.75 mm collimation, 1 mm slice thickness, and 0.6 mm reconstruction interval. Seventy millilitres of iodinated contrast medium was injected intravenously at 4.5 mL/s, followed by 40 mL of saline at 2.5 mL/s. Electrocardiogram synchronization and sedation were not used. On MDCT, the VSD, overriding aorta, and infundibular pulmonary stenosis were visualized, and TOF was confirmed (Panel A). Additionally, a right-sided aortic arch and pulmonary hypoplasia were observed (Panel B). Surprisingly, sagittal and axial maximum intensity projection (MIP) and three-dimensional volume rendering (3D VR) images revealed a DAA without a vascular ring around the trachea and oesophagus. Two stacked aortic arches were observed at the sagittal (Panels C and D) and axial planes (Panels E and F) and designated as inferior and superior arches. The bilateral subclavian and common carotid arteries originated from the superior aortic arch (Panels G and H). There was no vessel originating from the inferior arch. The branches originating from the superior aorta arch were determined to be the left carotid artery, right carotid artery, right subclavian artery, and left subclavian artery.

The DAA occurs due to the persistence of both left and right fourth aortic arches. The axial images are a good tool to the diagnosis of anterior and posterior arches of DAA. In our case, the aortic arches presented as superior and inferior arches on sagittal plane on MDCT imaging. No vascular ring or narrowing of the trachea was found. A persistent fifth aortic arch (PFAA) is an extremely rare anomaly and haemodynamically, and it is usually inconsequential and the diagnosis is often coincidental. The PFAA was first described as a DAA lumen, which has two well-separated parallel, complete aortic arches as in our case. The origins of the main vascular structures arising from the aortic arch were different than from reported PFAA cases. Moreover, the present case showed that DAA may present with superior and inferior arches instead of posterior and anterior arches. Therefore, sagittal and 3D VR imaging should be added to routine axial imaging for differential diagnosis of DAA.

MDCT and magnetic resonance imaging (MRI) are important diagnostic modalities for identifying anomalies of the aortic arch and its branches. However, MRI is time-consuming imaging modality, which may require prolonged sedation in paediatric patients. Because of its comparatively high spatial resolution, MDCT is preferred to MRI for the evaluation of complex congenital anatomy. Recent advances in CT technology indicated that radiation protection techniques deliver not only a low radiation dose of around or below 1 mSv for a cardiac study, but also showed that this mode provides diagnostic image quality.

Panel A. MDCT imaging demonstrating a VSD (arrow). Ao indicates the overriding aorta; LV, left ventricle; RV, right ventricle.

Panel B. Axial view of MDCT revealing hypoplasia of the main pulmonary artery and its branches. Ao indicates the right aortic arch; MPA, main pulmonary artery; RPA, right pulmonary artery; LPA, left pulmonary artery.

Panel C. Sagittal MIP images demonstrating the DAA. Ao indicates the overriding aorta; asterisk, superior aortic arch; arrow, inferior aortic arch, T, trachea.

Panel D. Posterior views from 3D VR imaging demonstrating the DAA. Asterisk, superior aortic arch; arrow, inferior aortic arch.

Panel E. Axial view at the level of the superior aortic arch revealing no vascular ring around the trachea. SA, superior aortic arch; T, trachea.

Panel F. Axial view at the level of the inferior aortic arch revealing no vascular ring around the trachea. IA, superior aortic arch; T, trachea.
Panel G. Posterolateral of 3D VR imaging demonstrating that the branches of the aorta arch originate from the superior aortic arch. Ao indicates the descending aorta; asterisk, superior aortic arch; LSA, left subclavian artery; RSA, right subclavian artery; RCCA, right common carotid artery; LCCA, left common carotid artery.

Panel H. Posterior views of 3D VR imaging demonstrating that the branches of the aorta arch originate from the superior aortic arch. Ao indicates the descending aorta; asterisk, superior aortic arch; LSA, left subclavian artery; RSA, right subclavian artery; RCCA, right common carotid artery; LCCA, left common carotid artery.

Ruptured unknown Stanford Type A aortic dissection with huge mediastinic emathoma mimicking pulmonary embolism

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We report the case of a 84-year-old patient, with a history of coronary artery bypass graft (CABG) and mitral valve replacement 11 years ago and a recent access to an emergency department for acute chest pain. The patient was quickly discharged because of negative troponin without any radiological examination.

Six days later, the patient referred to our emergency department for chest pain and dyspnoea. Owing to an elevated D-dimer, patient underwent chest computed tomography (CT) for suspected pulmonary embolism (PE).

CT pulmonary angiography did not show PE but a significant pulmonary artery compression (Panels A and B). A subsequent arterial phase demonstrated a Type A aortic dissection originating from the sinotubular junction and a 2-mm wall rupture with huge mediastinal haematoma (diameters 5.7 × 12.3 cm), causing significant compression of pulmonary artery trunk, left atrium and superior left pulmonary vein (Panel C).

Owing to the very high risk of mortality, the surgical intervention was refused by the patient’s relatives and an appropriate anti-hypertensive therapy was administered.

A 7-day follow-up thoracic CT angiography showed no significant differences in the size of haematoma but the onset of bilateral pleural bleeding (Panel D). The patient died 3 days after CT examination.

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