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IMAGE

Unusual Late Presentation of a Rare Complex Aorto-Pulmonary Malformation Diagnosed on Computed Tomography Angiography

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A 5-year-old girl presented with cyanosis, dyspnoea on moderate exertion and palpitations. Transthoracic echocardiography revealed a large type III aorto-pulmonary window (APW) with a suspicion of associated aortic arch interruption. She was further advised a computed tomography angiography (CTA) for accurate assessment of the aortic arch along with evaluation of the primary defect and any other cardiac or extra-cardiac anomaly.

Computed tomography angiography confirmed the presence of two separate semilunar valves and a large communication between the ascending aorta and the main pulmonary artery, extending from just above the level of sinuses of Valsalva to the distal ascending aorta. The right pulmonary artery (RPA) appeared to be arising from the ascending aorta. There was associated type A aortic arch interruption (interrupted aortic arch distal to left subclavian artery) and the descending thoracic aorta was seen to be reformed by the ductus (Figure 1, panels A, B and C).

Berry syndrome is an extremely rare and complex aorto-pulmonary malformation comprising a distal aorto-pulmonary window, aortic arch interruption and right pulmonary artery originating from the ascending aorta in presence of an intact ventricular septum. It almost always presents in early infancy (usually before 6 months of life) and neonates with this condition have been reported to have undergone successful single-stage repair [1]. However, in rare cases, especially in the presence of a large, non-restrictive aorto-pulmonary window, as seen in our case, the pulmonary vascular resistance may not fall significantly. This subset of patients may remain relatively asymptomatic initially. However, despite the patient being free of symptoms, irreversible pulmonary vascular obstructive disease characteristically develops with time. It is at this point, that symptoms like easy fatigue, exertional dyspnoea, and cyanosis start to appear.

Our patient presented at the age of 5 years and is most likely to have developed irreversible pulmonary hypertension. However, a right heart catheterisation, being the gold standard for measuring haemodynamic parameters in patients with questionable operability, may be performed and cardiac catheterisation criteria of operability such as the Lopes and O'Leary criteria may be considered to decide on operability [2]. To our knowledge, this is the first ever reported case of Berry syndrome presenting this late in childhood.

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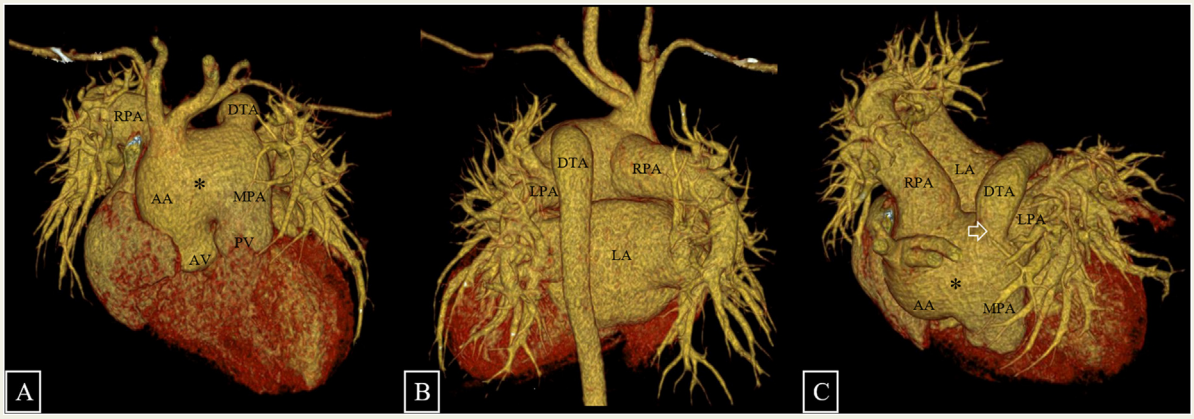

Figure 1 Volume rendered images of CT angiography (A, B and C; anterior oblique view, posterior view and cranio-caudal view respectively) reveal presence of a large communication (*) between the AA and the MPA, extending from just above the level of sinuses of Valsalva (AV, PV) to the distal AA. There is interruption of arch distal to the common trunk and the DTA is reformed by patent ductus arteriosus (block arrow). The RPA appears to be arising from the ascending aorta.
Abbreviations: CT, computed tomography; AA, ascending aorta; MPA, main pulmonary artery; AV aortic valve; PV pulmonary valve; DTA descending thoracic aorta; RPA, right pulmonary artery; LPA, left pulmonary artery; LA, left atrium.

Conflicts of Interest

The authors declare no conflicts of interest.

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