A rare case of anomalous origin of the left pulmonary artery from the ascending aorta with ventricular septal defect and pulmonary atresia

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A 14-year-old female patient was evaluated for episodes of cyanosis and fatigue. Initial physical examination revealed signs such as central cyanosis, digital clubbing, a single-second heart sound, and a 2/6 systolic ejection murmur audible at the upper left parasternal border. Radiographic examination of the chest revealed a cardiothoracic index of 60%, a left-sided aortic arch, and a typical "boot-shaped" cardiac silhouette suggestive of a congenital cardiac malformation. Additionally, chest X-rays showed an absence of a pulmonary artery segment, reduced right pulmonary vasculature, and prominent left pulmonary vasculature (Figure 1A).

A further diagnostic evaluation with transthoracic echocardiography demonstrated a large misaligned subaortic ventricular septal defect with aortic overriding and bi-directional shunting (Figure 1B). The ventricles were balanced in size, with the left pulmonary artery originating from the ascending aorta. Neither a separate pulmonary valve nor the main and right pulmonary arteries were discernible. Right ventricular ejection fraction (EF) using Simpson's rule was 50%, and tricuspid annular plane systolic excursion (TAPSE) was 26 mm. During cardiac catheterization, the systolic aortic pressure and the systolic pressure of the anomalously originating left pulmonary artery were both recorded at 90 mm Hg, consistent with expectations. Angiographic findings indicated a complete absence of native pulmonary arteries supplying the right lung (Figure 1C). Computed tomography provided definitive evidence supporting the aberrant origin of the left pulmonary artery from the ascending aorta (Figure 1D), the absence of an extraparenchymal right pulmonary artery, and the existence of rudimentary intraparenchymal branches of the right pulmonary artery (Figure 1E). Given that no collateral arteries associated with these pulmonary arteries were identified, it was postulated that these intraparenchymal pulmonary arteries were likely nourished by the bronchial arteries. The patient was subsequently initiated on a therapeutic regimen to manage pulmonary hypertension. We assumed that the right lung might have initially received blood supply from the right arterial duct, which could have ceased functioning postnatally, leaving the right lung undersupplied; an assumption that necessitates further investigation to thoroughly comprehend the complex physiological adaptations accompanying such rare cardiac anomalies.

Anomalous origin of the pulmonary artery from the aorta (AOPA) is a cardiac anomaly observed infrequently. It may manifest as an isolated malformation or in conjunction with other cardiac abnormalities, such as ventricular septal defect [1] or tetralogy of Fallot [2]. Among these, the anomaly involving the left pulmonary artery (AOLPA) originating from the aorta is observed less frequently than its right-sided counterpart [1]. Although AOLPA is predominantly associated with a right aortic arch, this association was not observed in the presented case. A configuration featuring AOLPA concomitant with right pulmonary artery atresia presents as a rare cardiac anomaly [1, 2]. As argued by Pepeta et al. [1] and Nagulakonda et al. [2], such abnormalities are seldom documented in the literature, underlining the unique nature of our case.

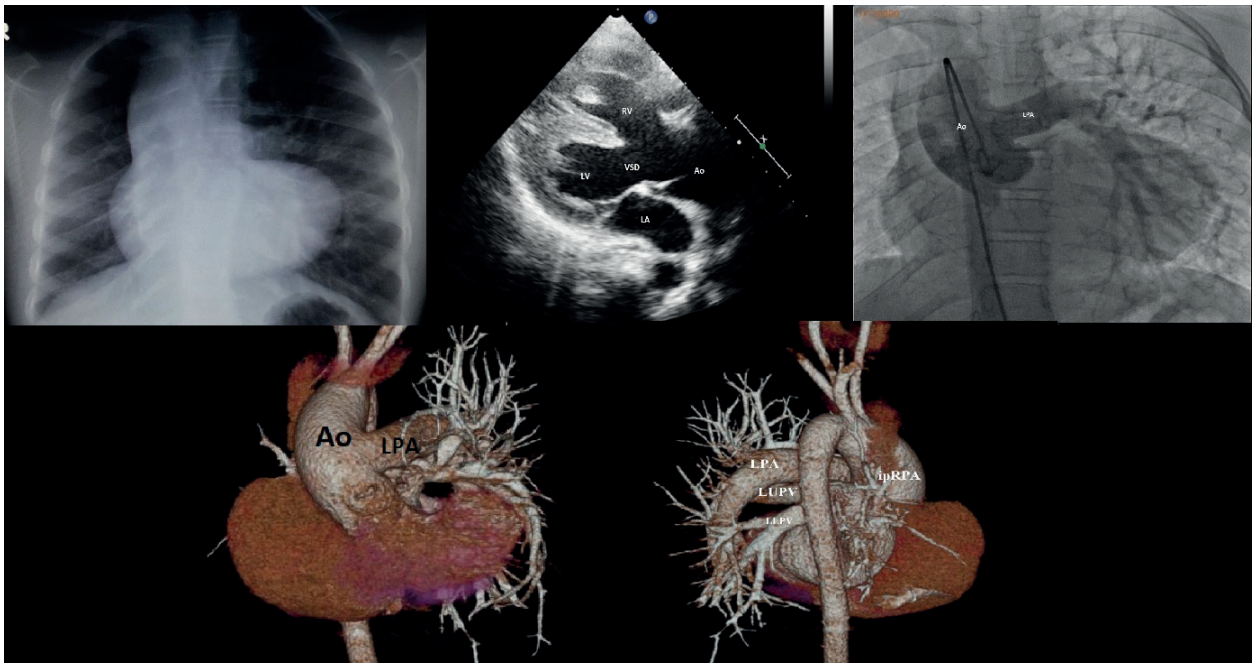

Figure 1. A. Chest X-ray showing levocardia, a boot-shaped heart, and an absent pulmonary artery segment. **B.** Two-dimensional echocardiographic study showing a large misaligned subaortic ventricular septal defect with aortic overriding. **C.** Ascending aortogram in antero-posterior view showing the left aortic arch, the origin of the left pulmonary artery from the aorta, and the absence of the right pulmonary artery. **D.** Computed tomography angiography image anterior view showing an anomalous left pulmonary artery arising from the proximal ascending aorta. **E.** Computed tomography angiography image posterior view showing the intraparenchymal right pulmonary artery branches

Abbreviations: Ao, aorta; ipRPA, intraparenchymal right pulmonary artery; LA, left atrium; LLPV, left lower pulmonary vein; LPA, left pulmonary artery; LPA, left pulmonary artery; LUPV, left upper pulmonary vein; LV, left ventricle; RV, right ventricle; VSD, ventricular septal defect

Modern imaging techniques can correctly identify such aberrations. The importance of preoperative computed tomography angiography as a specific diagnostic tool for complex cardiac anomalies is underscored in a study by Nagulakonda et al. [2], in which a case of tetralogy of Fallot, pulmonary atresia with AOLPA, and collateral arteries supplying the right lung was described in detail. Considering the substantial risk of pulmonary hypertension development, prompt detection and intervention are indispensable to prevent irreversible pulmonary damage and improve long-term patient prognosis.

Article information

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