

Eisenmenger's syndrome and agenesis of the right pulmonary artery: A case of double diagnosis

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Abstract

We present a case of patent ductus arteriosus with severe pulmonary hypertension with the rare association of absent right pulmonary artery. (Cardiol J 2011; 18, x: xx–xx)

Key words: pulmonary hypertension, unilateral agenesis of pulmonary artery

A 42 year-old female presented with progressively increasing breathlessness of five years' duration. She was in NYHA functional class III. She had been having intermittent episodes of hemoptysis for the last three months. On clinical examination, she had a cyanosed left upper limb and both lower limbs with grade 3 clubbing and relative sparing of the right upper limb (Fig. 1). Precordial examination revealed a parasternal heave and a palpable shock in the 2nd left intercostal space. The second heart sound was single and loud with a high pitched early diastolic murmur. ECG showed right ventricular hypertrophy with P pulmonale. A clinical diagnosis of patent ductus arteriosus (PDA), Eisenmenger's was made. Chest X-ray showed prominent pulmonary arterial conus and, surprisingly, a mediastinal and tracheal shift to the right with elevation of the right diaphragm and hypoperfused right lung (Fig. 2). Three-dimensional 64-multidetector computed tomography (MDCT) for ascertaining the cause of hemoptysis confirmed the presence of a large PDA (Fig. 3) with unilateral agenesis of the right pulmonary artery. The right lung was perfused by major aorto-pulmonary collaterals arising from the posterior aspect of the arch of aorta (Figs. 4A–C).

Our patient presented as a classical case of Eisenmenger's PDA with hemoptysis. In this clinical scenario, one would suspect thrombosis in the main pulmonary arteries [1]. However, chest X-ray and MDCT revealed the rare association of Eisenmenger's PDA with unilateral pulmonary artery agenesis [2]. The hemoptysis in our patient was explained by the presence of major aorto-pulmonary collaterals (MAPCAs) which may be self-limiting or can occasionally present with massive pulmonary hemorrhage and death. The genesis of these MAPCAs is explained by the existence of small transitory branches of the dorsal aorta that supply the lung before the development of the pulmonary artery, and persist as enlarged supradiaphragmatic and infradiaphragmatic aortic branches, or as bronchial arteries in cases in which the pulmonary arteries do not develop [3].

Right-sided agenesis can occur in isolation or in association with benign cardiac conditions, and hence the diagnosis is often delayed into adulthood. Left-sided agenesis is often combined with tetralogy of Fallot or other complex congenital anomalies, and patients present earlier with symptoms of the associated conditions [4]. Pulmonary hypertension is reported in 20–25% of patients with unilateral

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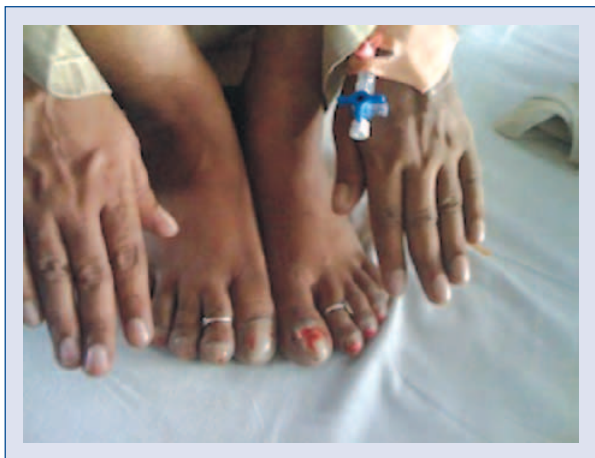


Figure 1. Toes and fingers of the lady, showing more profound cyanosis and clubbing of the left hand and the toes.



Figure 2. Chest X-ray shows prominent pulmonary artery conus as well as tracheal shift (dashed arrow) to the right with an elevated right diaphragm (solid arrow).

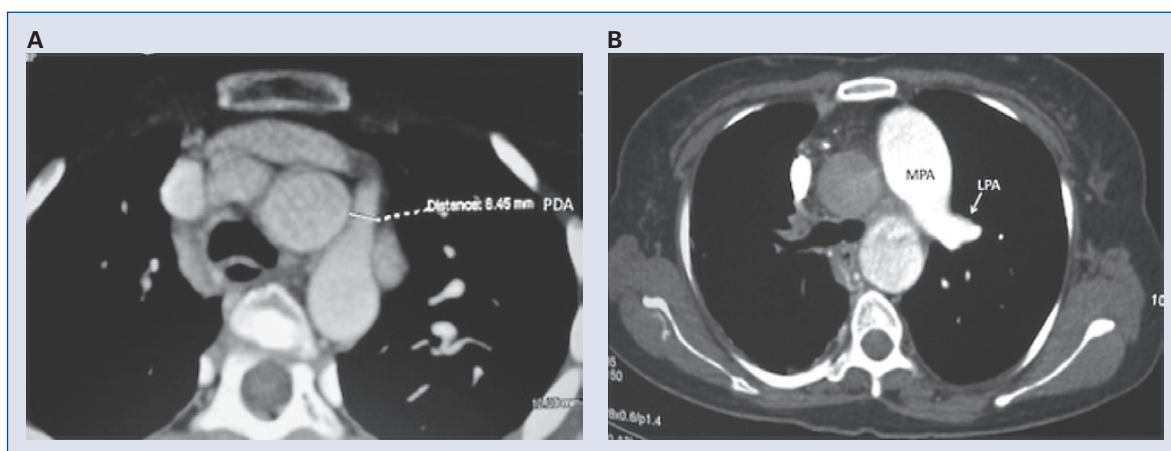


Figure 3. A. Computed tomography pulmonary angiography transaxial images showing the large patent ductus arteriosus (PDA); B. Main pulmonary artery (MPA) continuing as the left pulmonary artery (LPA). No right pulmonary artery is seen.

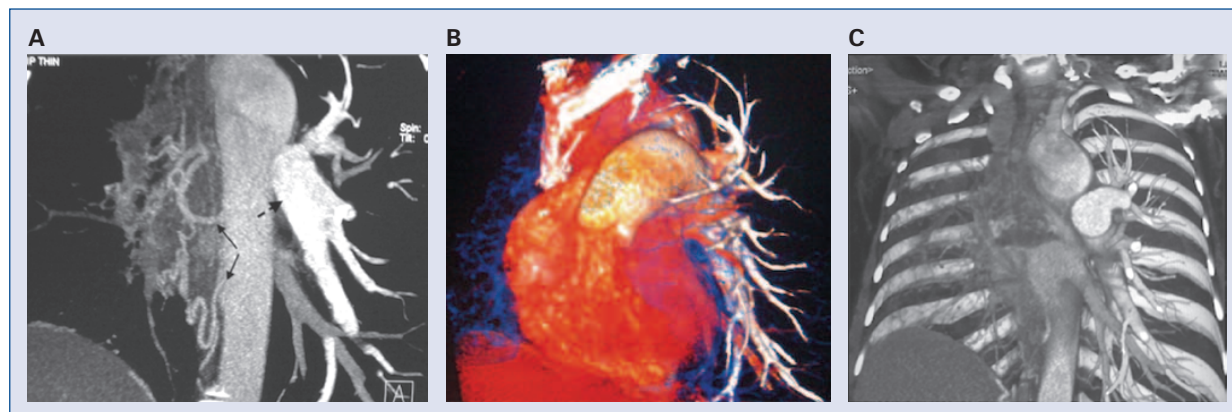


Figure 4. A. Multiplanar coronal reformation image demonstrates dilated left central pulmonary arteries (dashed arrow), absent right pulmonary artery and collateral supply to the right lung (solid arrows); B. Three-dimensional volume-rendered reformation image of the main pulmonary artery continuing as the right pulmonary artery; C. Coronal maximum intensity projection image clearly showing normal branching pattern of the left pulmonary artery and absence of pulmonary artery on the right.

pulmonary artery agenesis often unmasked by predisposing conditions such as high altitude pulmonary hypertension or pregnancy. In our patient, both the large duct as well as unilateral pulmonary artery agenesis probably contributed to the development of Eisenmenger's [5]. This case is a reminder to look for branch pulmonary arteries whenever pulmonary hypertension is present (unexplained or with other lesions).

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References

1. Wald R, Crean A. Differential clubbing and cyanosis in a patient with pulmonary hypertension. *CMAJ*, 2010; 182: E380.
2. Johnson TRC, Thieme SF, Deutsch MA et al. Unilateral pulmonary artery agenesis: Non-invasive diagnosis with dual-source computed tomography. *Circulation*, 2009; 119: 1158–1160.
3. Pfefferkorn JR, Loser H, Pech G et al. Absent pulmonary artery: A hint to its embryogenesis. *Pediatr Cardiol*, 1982; 3: 283–286.
4. Gluck MC, Moser KM. Pulmonary artery agenesis: Diagnosis with ventilation and perfusion scintigraphy. *Circulation*, 1970; 41: 859–867.
5. Harkel ADJT, Blom NA, Ottenkamp J. Isolated unilateral absence of a pulmonary artery. A case report and review of the literature. *Chest*, 2002; 122: 1471–1477.