Isolated absence of right pulmonary artery: Radiographic and multi-detector CT demonstration

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CASE SUMMARY

A 24-year-old male presented to clinic for a routine pre-employment chest radiograph. The clinical history was uneventful. There was no significant medical illness or operative history in the past. Clinical examination findings were also reported to be within normal limits. Subsequent, pulmonary function tests showed mild restrictive defect. The oxygen saturation level was approximately 85% in the lower limbs while breathing room air. Radiograph chest was performed followed by contrast-enhanced computed tomography of the chest.

Radiograph chest findings

The chest radiograph, however, showed smaller right lung fields on the right side with rightward shift of the mediastinum and upward shift of the right hemi-diaphragm [Figure 1]. There is relative paucity of the vascular markings on the right side. The left lung shows relative hyperinflation with increased translucency and prominent vascular markings. The lower lobe pulmonary artery was visualized on the left side and non-visualized on the right side. The tracheal and bronchial structures showed rightward shift and were otherwise optimally visualized. No definite focal parenchymal opacity or pleural effusion is identified in this study. The findings raise suspicion of a developmental abnormality involving the right sided pulmonary arterial system and CT study of chest was advised for further evaluation.

CT findings

Contrast-enhanced Multi-detector CT (MDCT) chest was performed, which showed complete absence of the right pulmonary artery [Figure 2a] with dilatation of the right-sided bronchial artery. There was paucity of the vascular markings in the right lung field, as compared to left side. The visualized vessels in the right lung were also small in calibre [Figure 2b and c]. Small volume of the right lung is also noted along with pleural thickening and bronchial dilatation on the right side. Based on this constellation of radiological findings, the diagnosis of right-sided unilateral agenesis of pulmonary artery (UAPA) was made. No other definite cardiovascular or pleuro-parenchymal abnormality was identified in this study. Subsequent pulmonary functions tests however, showed mild restrictive changes. The oxygen saturation level was approximately 85% in the lower limbs while breathing room air. In absence of any symptoms, the patient refused further evaluation or any form of treatment and is presently under follow-up.

Figure 1: Radiograph chest - frontal projection showing smaller right lung field with rightward shift of the mediastinum and upward shift of the right hemi-diaphragm. There is relative paucity of the vascular markings on the right side. The left lung shows relative hyperinflation with increased translucency and prominent vascular markings. The lower lobe pulmonary artery was visualized on the left side and non-visualized on the right side.
DISCUSSION

UAPA is a rare congenital anomaly which is often accompanied with cardiovascular anomalies, most commonly tetralogy of Fallot\cite{1} or septal defects.\cite{2} UAPA also may occur as an isolated finding and as much as 108 such cases were described in the database of National Library of Medicine between 1978 and 2000.\cite{3} There are case reports which described isolated UAPA in completely asymptomatic patients,\cite{4} while other reports mentioned of severe pulmonary hypertension and congestive heart failure,\cite{5} as the clinical presentation. In a retrospective analysis of 108 cases of UAPA,\cite{3} most patients presented with recurrent pulmonary infections (37%), dyspnea or limited exercise tolerance (40%), or hemoptysis (20%). Only 14 out of 108 patients were found to be asymptomatic in this review.

Several etiologic hypotheses to explain this abnormality include chromosomal origin, vitamin A deficiency, intrauterine infections and environmental factors. The reduction of pulmonary arterial blood flow prevents normal lung development and causes pulmonary hypoplasia.\cite{6} The frequent respiratory infections may be explained by underlying bronchiectasis or impaired muco‑ciliary clearance. Though right‑sided bronchial dilatation and pleural thickening were also seen in the present case, the patient denied any previous history of chest infection. Hemoptysis in these patients may be caused by excessive collateral circulation through the bronchial, intercostal, subclavian or subdiaphragmatic arteries.

Even in absence of clinical symptoms, pulmonary hypertension is an important complication mainly related to exacerbating factors like high altitude or pregnancy. This emphasizes the role and relevance of early imaging‑based diagnosis of UAPA to prevent pulmonary hypertension. Chest radiograph provides a crucial opportunity to suspect this abnormality. CT, magnetic resonance imaging (MRI), echocardiography or ventilation‑perfusion scintigraphy is useful for confirmation of the diagnosis. Digital subtraction angiography is only required for evaluation of finer vascular details prior to a revascularization surgery. If reparative surgery is not feasible, oral administration of calcium channel blockers or or continuous infusion of prostaglandins have been used for pulmonary hypertension. In patients with massive hemoptysis, selective embolization of the collaterals and even pneumonectomy procedures are described in literature.\cite{3}

The present article thus highlights the role of chest radiograph and CT in the diagnosis of isolated unilateral absence of right pulmonary artery in an asymptomatic individual who had his routine pre‑employment chest radiograph.

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