

Unilateral interstitial lung disease: A unique manifestation of asymptomatic pulmonary artery atresia

Akriti Gujral¹, Namita Singh², Giriraj Singh³, Ankush Bansal⁴

From ¹Medical Student, Clinical Rotation, Richmond University Centre, Staten Island, New York, ²Consultant Teleradiologist, Department of Radiodiagnosis, Telerad Solutions, Bengaluru, Karnataka, ³Professor, Department of Radiodiagnosis, Military Hospital, Agra, Uttar Pradesh, ⁴Consultant Radiologist, Department of Radiodiagnosis, Bansal Diagnostic Centre, Panipat, Haryana, India

ABSTRACT

Interstitial pulmonary fibrosis is idiopathic, bilateral, or secondary to exposure to organic antigens, or collagen disorders. Unilateral pulmonary fibrosis is uncommon and intriguing as it has different pathogenetic mechanisms. We present a case of a patient with a short 5-day history of dyspnea with no ailment in childhood. His radiograph revealed increased reticular markings in the right lung with a small right hilum. Contrast-enhanced computed tomography showed unilateral pulmonary fibrosis and confirmed the presence of an atretic right pulmonary artery contributing to the small hilum, with collateral supply from the right internal mammary artery. This asymptomatic patient had unilateral pulmonary fibrosis which had developed as a consequence of impaired blood supply to the right lung due to atresia of the right pulmonary artery with collateral from systemic circulation unable to keep abreast with normal pulmonary development

Key words: Pulmonary artery atresia, Interstitial lung disease, Unilateral pulmonary fibrosis


Unilateral presentation of interstitial lung disease (ILD) is an uncommon condition with very few case reports mentioning this condition [1]. Detection of such a condition in an asymptomatic patient is very intriguing. Unilateral pulmonary artery atresia (UPAA) is an uncommon condition with only a total of 350 cases reported in the literature [2]. It occurs when the 6th aortic arch fails to fuse with the pulmonary trunk during fetal development [3]. In the pediatric age group, it is seen associated with congenital cardiac anomalies. Rarely when not associated with congenital heart disease, it is detected as an asymptomatic condition in adults, where the ventilation of the contralateral normal lung provides the oxygen supply to meet the body's demands. Although the main pulmonary artery is atretic, the intrapulmonary vascular network is formed and is recruited by collaterals from systemic arteries to supply blood to the affected lung [4]. The alternative arrangement of collateral circulation is, however, not as efficient as the presence of the main pulmonary artery in supplying blood to the lung. Thus, the affected lung develops bronchiectasis, interstitial fibrosis, and recurrent lung infections [5].

CASE REPORT

A 50-year-old male presented with a short history of dyspnea for 5 days. There was no history of fever or weight loss. He had an asymptomatic childhood with no history of recurrent lung infections or dyspnea in the past.

No abnormality was detected in the cardiovascular or respiratory systems on auscultation. His vitals were within the normal range. There was no finger clubbing or cyanosis.

His blood counts were within normal limits. The clinician ordered a chest radiograph which showed increased interstitial markings in the right lung with a small right hilum (Fig. 1). Non-contrast computed tomography was then done, which showed volume loss in the right hemithorax with mediastinal shift to the right. There was interlobular septal thickening in a peripheral and subpleural distribution in the right lung with an apicobasal gradient (Fig. 2a and b). There was subpleural honeycombing in the right lung, more prominent in the basal segments of the right lower lobe and the right middle lobe. There was an associated right pleural effusion. The right pulmonary artery was not visualized (Fig. 3a). The left lung did not show any significant abnormality. Contrast-enhanced computed tomography (CECT) showed an absence of the right pulmonary artery (Fig. 3b). No cardiac anomaly was detected on the CECT. There was subpleural

Access this article online	
Received - 23 November 2023 Initial Review - 05 December 2023 Accepted - 20 January 2024	Quick Response code 
DOI: 10.32677/ijcr.v10i2.4364	

Correspondence to: Giriraj Singh, Military Hospital, Agra, Uttar Pradesh, India.
E-mail: girigujral@gmail.com

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Figure 1: X-ray chest shows increased reticular markings in the right lung with blunted right CP angle due to a small pleural effusion and a small right hilum

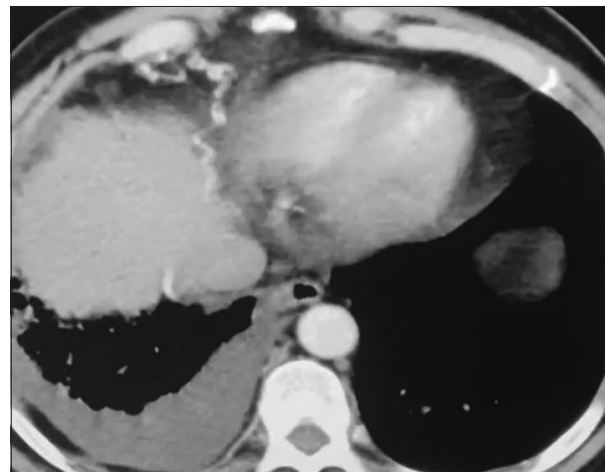


Figure 4: Contrast-enhanced computed tomography shows collateral circulation to the right lung from the right internal mammary artery

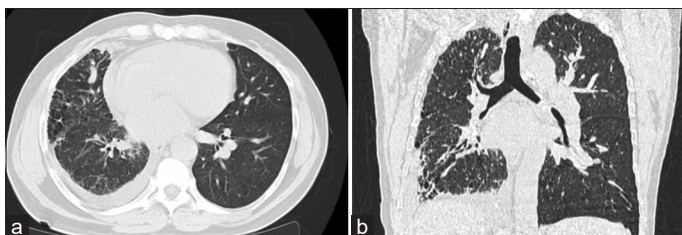


Figure 2: (a and b) Axial and coronal CT shows interlobular septal thickening in a peripheral and subpleural distribution in the right lung with an apico-basal gradient

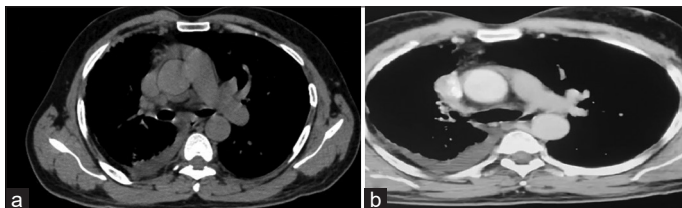


Figure 3: (a) The main pulmonary artery is leading into the left pulmonary artery. The right pulmonary artery is not visualised on the non-contrast computed tomography; (b) Contrast-enhanced computed tomography shows an atretic right pulmonary artery with conical tapering to a narrow point

honeycombing in the right lung more prominent in the basal segments of the right lower lobe and right middle lobe associated with right pleural effusion (Fig. 4). The left lung was normal in appearance. A diagnosis of unilateral ILD with right pulmonary arterial atresia was made.

DISCUSSION

ILD is usually bilateral but can have an asymmetrical distribution. Unilateral lung involvement by ILD is rarely encountered in clinical practice. This uncommon condition can be associated with disorders of pulmonary circulation such as proximal interruption of the pulmonary artery, pulmonary vein thrombosis, post-radiation lung changes, lung injury caused by ventilatory support, bronchiectasis, and gastroesophageal reflux [6].

UPAA can be associated with congenital heart disease or can occur in isolation. When associated with congenital heart diseases such as ventricular septal defect, Tetralogy of Fallot, coarctation of the aorta, and right-sided aortic arch, it is usually detected in childhood [7,8]. A second subset is seen in adulthood when there is no associated congenital heart disease [9]. Such patients are usually asymptomatic and are detected incidentally, or present with dyspnea, hemoptysis, and recurrent lung infections [5]. Our patient had an uneventful childhood and had no history of any lung ailment. His only presenting symptom was a short history of dyspnea for 5 days.

The pulmonary artery develops from the sixth aortic arch during fetal development. Any defect in the sixth arch will lead to developmental abnormality in the pulmonary artery. Impaired blood supply to the lung results in the development of unilateral pulmonary abnormality on the affected side with increased blood flow to the contralateral normal side resulting in the development of pulmonary artery hypertension [9]. Collaterals develop between systemic and pulmonary circulation to supply the affected lung [10,11]. In our case, collaterals were seen to arise from the right internal mammary artery (Fig 4). In UPAA, the main pulmonary trunk is normal or slightly enlarged, but the affected arterial segment is either atretic or incompletely developed. This leads to lung hypoplasia of the affected lung with the development of bronchiectasis, interstitial fibrosis, cyst formation, and recurrent infections [8]. No bronchiectasis was seen in our case. There was no history of recurrent lung infection in our patient.

In patients with UPAA, collaterals develop between the systemic and pulmonary circulation to supply blood to the hypoplastic lung enabling it to develop and grow [10,11]. Collaterals have been reported from bronchial, intercostal, internal mammary, and subdiaphragmatic arteries. Hypertrophied collaterals are at risk of rupture and can present with pulmonary hemorrhage or unilateral rib notching if collaterals develop from intercostal arteries [12]. In our case, we found hypertrophied collaterals from the internal mammary arteries.

Despite compensatory blood supply from systemic circulation, the affected lung fails to develop normally and pathological

conditions such as bronchiectasis, cysts, and interstitial fibrosis develop in the affected lung [13]. A study that reviewed CT findings in eight cases with UPAA found cystic bronchiectasis and honeycombing in two of the eight cases [14]. In our subject, despite collateral blood supply from systemic arteries, the right lung developed interlobular septal thickening and honeycombing, while the left lung which had a normal blood supply showed no evidence of ILD.

CONCLUSION

Unilateral interstitial fibrosis is a rare entity and should alert the radiologist to look for disorders of pulmonary circulation as a possible cause, especially when there is no history of unilateral lung injury in the form of radiation or gastroesophageal reflux. UPAA has a bimodal presentation, occurring with congenital heart disease in the paediatric age group and without congenital heart disease in the adult population where it is usually detected incidentally.

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Funding: Nil; Conflicts of interest: Nil.

How to cite this article: Gujral A, Singh N, Singh G, Bansal A. Unilateral interstitial lung disease: A unique manifestation of asymptomatic pulmonary artery atresia. *Indian J Case Reports*. 2024;10(2):41-43.