# Radiological quandary: Unilateral interstitial lung disease – Truth or dare

Sir,

Unilateral interstitial lung disease (ILD) has always intrigued clinicians. Interruption of main pulmonary circulation (arterial/venous) of one lung like in unilateral absence of pulmonary artery (UAPA), sarcoma of unilateral pulmonary artery or unilateral pulmonary venous

thrombosis, and radiation-induced ILD can potentially lead to the development of unilateral ILD.[1-3] Although asymmetrical involvement is reported in one-third of the cases of idiopathic pulmonary fibrosis, no case had pure unilateral involvement. We present a case of isolated UAPA with unilateral ILD and rheumatoid arthritis (RA). The concomitant presence of RA, which is usually a cause of bilateral disease, makes our case unique and adds to diagnostic quandary in our patient, not been discussed before in English literature.

A 32-year-old young female presented with recurrent respiratory tract infections and progressive breathlessness. She had a history of morning stiffness, which used to subside on its own within an hour, with pain in the knee joint and small (>3) joints of both hands for more than the last 10 years. She was treated intermittently with nonsteroidal anti-inflammatory drugs in the past. She was hemodynamically stable with normal $SpO_2$ (97%) at room air. There was Grade II clubbing with flexion deformity and contracture of the proximal interphalangeal joints of bilateral 5$^{th}$ fingers with no signs of inflammation/deformity in other small and large joints [Figure 1a]. Apart from basal crackles in the right lung, rest of the systemic examination was normal. Her erythrocyte sedimentation rate (61 mm at 1 h) and C-reactive protein (46.1 mg/L) raised. She had raised RA factor (956 U/mL; normal = 0–20 U/mL) with negative anticyclic citrullinated peptide antibody levels. Rest of the autoimmune profile was negative. Thus, she was diagnosed as a case of RA as per the American College of Rheumatology/European League against Rheumatism classification 2010 criteria.[4]

Her chest X-ray was suggestive of reduced lung volume with uniform haziness on the right side [Figure 1b]. High-resolution computed tomography chest revealed ground-glass opacities, perifissural and subpleural cysts, inter and intralobular septal thickening on the right side, and early honeycombing in the right paracardiac region anteriorly with the mediastinal window, showing absence of right main pulmonary artery [Figure 1c-h]. The left lung showed hyperinflation with no evidence of ILD changes. Two-dimensional echocardiography showed no other associated congenital heart anomaly.

Pulmonary function test revealed restrictive defect with reduced diffusion capacity (forced expiratory volume in one second [$FEV_1$] forced vital capacity [FVC] = 83.2%, FVC = 63% of predicted [1.95 L], $FEV_1$ = 60% of predicted [1.62 L], total lung capacity = 69% of predicted [3.09 L], and diffusion capacity for carbon monoxide = 45% of predicted [13.45 mL/min/mmHg]). She walked 406 m with no significant desaturation on 6-min walk test. Bronchoalveolar lavage [Figure 2] showed increased lymphocytes (50%). Transbronchial cryo lung biopsy showed mononuclear inflammation, temporal homogenicity, and preserved architecture with no granuloma and fibrosis, suggestive of cellular nonspecific interstitial pneumonia [Figure 2].

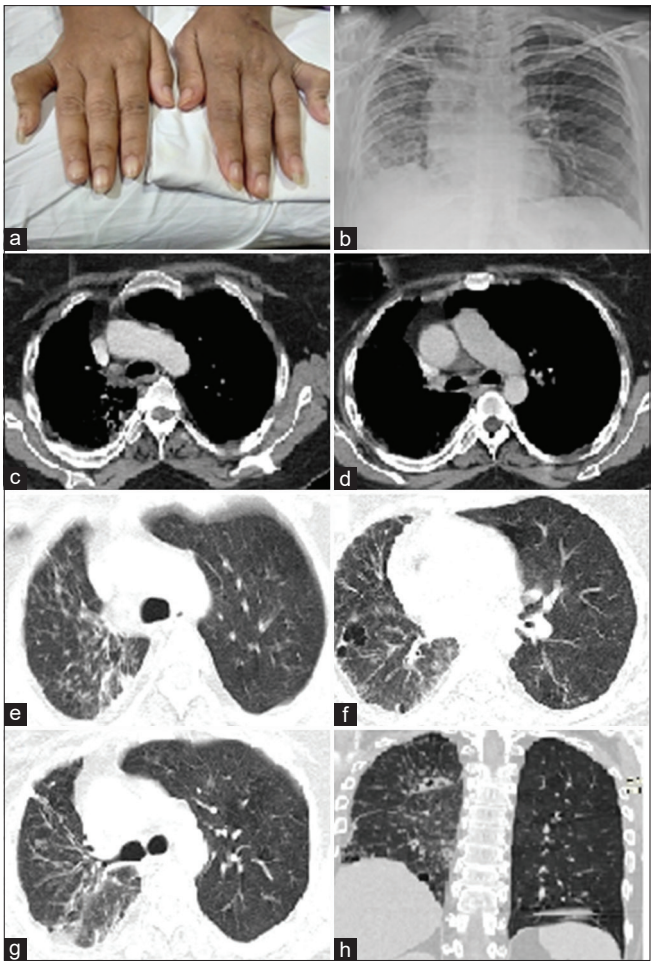


**Figure 1:** (a) Flexion deformity of the proximal interphalangeal joint of 5th fingers bilaterally. (b). Chest X-ray showing volume reduction of the right lung with mediastinal shift toward the right. (c and d) Mediastinal windows of computed tomography scan showing the absence of right main pulmonary artery. (e-h) High-resolution computed tomography scan showing foci of ground-glass opacity, cystic changes, and septal thickening in the right lower lobe with volume loss

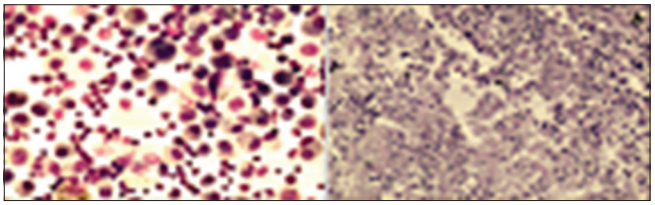

**Figure 2:** Bronchoalveolar lavage showing lymphocytic (50%) picture, and transbronchial lung biopsy demonstrating fibrotic nonspecific interstitial pneumonia

She was started on low-dose oral corticosteroids with hydroxychloroquine 200 mg twice daily. She continued to be on follow-up and is being monitored for progression of ILD and joint disease activity.

UAPA clinical presentation can vary according to the associated congenital heart abnormalities. Patients with isolated UAPA may be missed in early life and can present late, as has happened with our patient.

Absence of right-sided pulmonary artery is associated with right lung hypoplasia and can have catastrophic hemoptysis due to systemic circulation.

Connective tissue diseases such as RA are known to cause bilateral disease, although they can be asymmetrical at initial stages. We did not find any literature suggestive of the association of unilateral ILD with RA.

Thus to conclude, the association of UAPA and unilateral ILD with RA makes our case rare. This case needs to be followed to observe the effect of RA on other lung with time.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

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