Unilateral interstitial lung disease in a woman aged 35 years

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DESCRIPTION

A woman aged 35 years presented to the respiratory services with a history of asthma and recurrent lower respiratory tract infections. Chest X-ray (figure 1) showed mediastinal shift, volume reduction and increased interstitial markings throughout the right lung. Pulmonary function testing revealed a mixed obstructive and restrictive pattern.

CT scan demonstrated the absence of the right main pulmonary artery (figure 2). Collateral supply to the right lung was provided from the enlarged right internal mammary and inferior phrenic arteries (figure 3). Fine reticular peripheral interstitial markings, focal areas of ground glass and bronchiolitis were seen in the right lung (figure 4).

Radiographical findings were consistent with a diagnosis of interruption of the right pulmonary artery. This is a rare congenital condition



Figure 3 CT showing enlarged inferior phrenic artery.



Figure1 Chest X-ray.

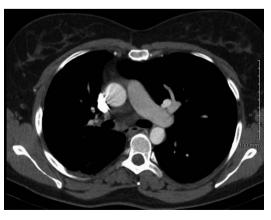


Figure 2 CT thorax showing the absence of the right pulmonary artery.



Figure 4 CT showing interstitial markings.

caused by a development anomaly of the sixth branchial arch.¹

The condition may present with recurrent respiratory infections. Despite collateral circulation, the affected lung is often hypoplastic, and interstitial pulmonary pathology including bronchestasis is revealed on CT imaging.²

Complications include haemoptysis due to the rupture of collateral vessels. Management is primarily conservative. In the presence of large volume haemoptysis, systemic collateral vessels may be embolised.³

Learning points

- Agenesis of a pulmonary artery can lead to unilateral interstitial lung disease.
- CT allows evaluation of pulmonary vasculature and lung parenchyma when investigating an abnormal chest X-ray.



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Contributors DJOH is the primary author of the paper. CTOC aided critical appraisal of the submitted article and provided literature review help. TS is the consultant radiologist who reported the patient's scans and reached the correct diagnosis. AOB is the consultant respiratory physician who managed the care of the patient, and provided critical appraisal of the final article.

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