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Case report

Unilateral proximal interruption of pulmonary artery with ipsilateral interstitial lung disease – A rare case report ☆

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ABSTRACT

Unilateral proximal interruption of pulmonary artery with ipsilateral occurrence of lung fibrosis is a very rare entity. This case report is about a 27 year old male who had complaints of progressive dysponea since 1 year. He had past history of recurrent lower respiratory tract infections. On auscultation, velcro crackles are heard on right side. Pulmonary function test showed restrictive pattern. Chest Radiography, High Resolution Computed Tomography and CT Pulmonary angiography were performed.

Introduction

Unilateral absence of pulmonary artery is a rare congenital anomaly which occurs during embryogenesis due to interruption of proximal sixth aortic arch [1,2]. It can be right or left sided. Congenital cardiac anomalies are often associated with left sided central pulmonary artery aplasia [3]. Their usual presentation is in 2nd decade. Unilateral interstitial lung disease is another rare condition.

Case report

A 27 year old male patient came with chief complaints of progressive shortness of breath and occasional hemoptysis since 1 year. Chest radiography-frontal view was performed which revealed mediastinal shift to right side with absent hilar shadow on ipsilateral side Figure 1. There is ipsilateral loss of lung volume and elevated hemidiaphragm with diminished

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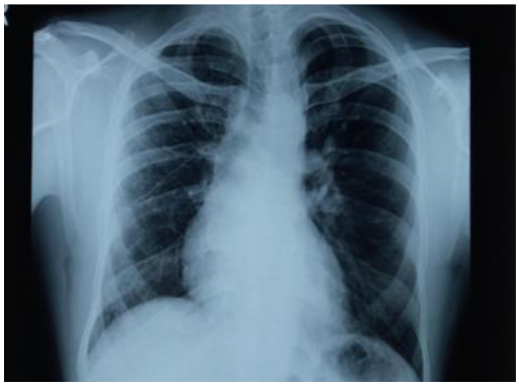

Fig. 1 – 27 year old male with right sided proximal interruption of pulmonary artery and ipsilateral interstitial lung disease who presented with progressive dysponea. Chest radiograph- frontal view shows mediastinal shift to right side with ipsilateral loss of lung volume, elevated hemidiaphragm and diminished pulmonary vascular markings. There are few scattered reticular markings seen in mid and lower zones of right lung

pulmonary vascular markings. There are few scattered reticular markings seen in mid and lower zones of right lung.

HRCT lung shows subpleural bulla with interspersed reticular thickening, interstitial fibrosis and few micronodules in upper and middle lobes of right lung. Right lower lobe shows reticular markings with interspersed fibrotic infiltrates Figure 2. There is pulmonary artery aplasia seen at its origin on right side.

CT Pulmonary angiography shows right sided proximal interruption of pulmonary artery with multiple collaterals from right intercostal, internal thoracic, sub-diaphragmatic, subclavian and coronary arteries Figure 3.

Discussion

The absence of pulmonary artery at its origin from the main pulmonary artery is known as proximal interruption of pulmonary artery [4]. The term proximal interruption of pulmonary artery is used rather than the pulmonary artery aplasia because the portion of the pulmonary artery that is in the lung is usually patent and intact. In contradiction with pulmonary agenesis where there is complete absence of lung parenchyma with associated blood vessels [5,6].

This happens when there is faulty development of sixth aortic arch in utero. System collateral branches from the aortopulmonary arteries, internal mammary artery, subclavian and intercostal arteries supplies the ipsilateral peripheral pulmonary arteries [7,8]. Left sided central pulmonary artery aplasia is often associated with cardiovascular abnormalities like tetralogy of Fallot.

Unilateral interstitial pulmonary fibrosis is a very rare lung condition associated with proximal interruption of the pulmonary artery, pulmonary vein thrombosis, ipsilateral single-lung ventilation, or radiation pneumonitis [7,9,10,11]. Agenesis of pulmonary artery can lead to unilateral interstitial lung disease [12].

The clinical presentation varies from dyspnea, exercise intolerance, recurrent chest infections, high altitude pulmonary

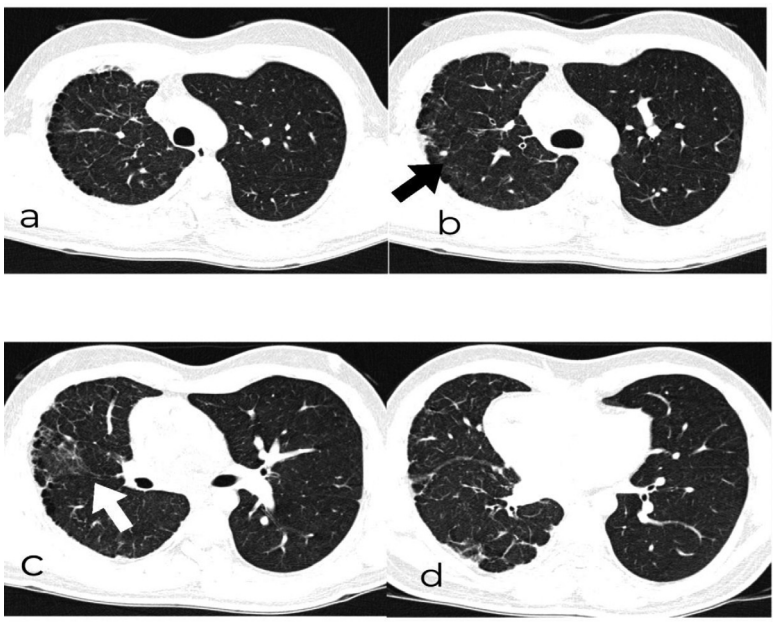

Fig. 2 – 27 year old male with right sided proximal interruption of pulmonary artery and ipsilateral interstitial lung disease who presented with progressive dysponea. (a) HRCT image shows subpleural bulla with decreased volume of right lung. (b) HRCT image shows pulmonary micronodule (black arrow) in right lung. (c) HRCT image shows area of ground glass opacity (white arrow) with interstitial fibrosis in right lung.(d) HRCT image shows interstitial fibrosis in right lower lobe.

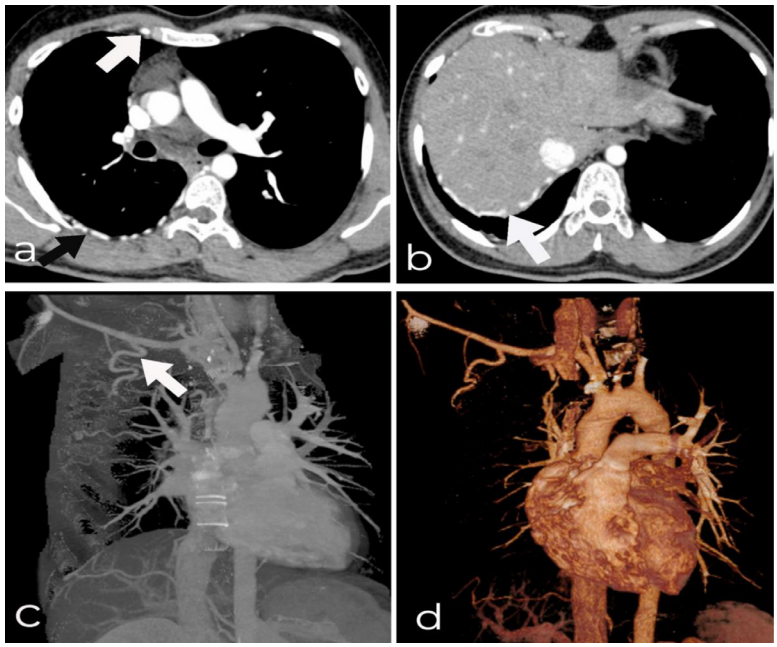

Fig. 3 – 27 year old male with right sided proximal interruption of pulmonary artery and ipsilateral interstitial lung disease who presented with progressive dysponea. (a) CT Pulmonary angiography shows proximal interruption of pulmonary artery on right side with collaterals from right internal thoracic artery (white arrow) and right intercostal artery (black arrow). (b) CT Pulmonary angiography shows right sided sub-diaphragmatic collaterals (white arrow). (c) MIP image shows proximal interruption of pulmonary artery on right side with collaterals from right sub-clavian artery (white arrow). (d) VRT image shows proximal interruption of pulmonary artery on right side with collaterals formation. (Color version of the figure is available online.)

edema, pulmonary hypertension, hemoptysis, chest pain to death. Chest radiography shows ipsilateral volume loss with diaphragmatic elevation, shift of heart and mediastinum to the affected side. The contralateral lung is herniated to the affected side. Fine non-branching linear opacities are seen at the lung periphery which indicates enlarged intercostal and transpleural pulmonary arteries [13,14].

CT mediastinal window shows absence of affected pulmonary artery at its origin or may terminate within 1cm of its origin [15]. Main differential to this is Swyer- James syndrome which shows air trapping on expiration.

Conclusion

Unilateral interstitial pulmonary fibrosis can be associated with proximal interruption of the pulmonary artery.

Patient consent

Taken priorly and documented.

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