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Case Report

Unilateral pulmonary artery agenesis presenting with unilateral usual interstitial pneumonia in adulthood

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ABSTRACT

Unilateral interstitial lung disease secondary to unilateral pulmonary artery agenesis (UPAA) is a rare anomaly due to a malformation of the sixth aortic arch of the affected side during embryogenesis. While most of the patients present in neonatal period with either cardiac anomalies or respiratory symptoms some of them can remain asymptomatic and late diagnosis is possible when suspicious presentation is noted on chest radiography. We report a case of 32-year female with a history of recurrent respiratory tract infection, who presented with cough and expectoration and the diagnosis of unilateral interstitial lung disease secondary to ipsilateral pulmonary interruption was made.

Keywords: Pulmonary artery agenesis, Unilateral interstitial pneumonia, Unilateral usual interstitial pneumonia, Absent right pulmonary artery, Unilateral hyperlucent lung

INTRODUCTION

Unilateral interstitial lung disease secondary to unilateral pulmonary artery agenesis (UPAA) is a rare finding in adult population. UPAA is a congenital anomaly occurring due to a malformation of the sixth aortic arch of the affected side during embryogenesis.[1,2] It can occur as a single disorder or may be associated with other congenital cardiovascular malformations.[3,4] The diagnosis is usually made in adolescence, however some of the patients can remain asymptomatic and late diagnosis is possible, with patients usually presenting with one of the chronic complications, i.e., dyspnea on exertion, recurrent respiratory infection or hemoptysis.[4]

CASE REPORT

A 32-year-old female presented with cough and expectoration with right sided chest pain since 2 weeks. She was well until about two weeks before the evaluation when she experienced dyspnea on exertion which gradually aggravated and a few days later she developed cough and expectoration. She had similar past history of on and off chest pain and cough with expectoration for 1 year. However, there were no complaints of fever, weight loss, hemoptysis, night sweat or loss of appetite. No drug allergy history.

She was a homemaker with no history of prior hospitalization or any history of smoking. No family history and past history of tuberculosis, primary tumor or radiation history. On physical examination, vital signs were stable with no respiratory distress or use of accessory muscles. Lung auscultation revealed diminished respiratory sounds over the right hemithorax with crepitation over right lower lung fields. No evidence of cyanosis, clubbing or tachycardia was noted.

In the first visit, possible diagnosis such as pneumonia, pulmonary tuberculosis, bronchiectasis and pulmonary thromboembolism were considered. Which prompted for serological and radiological examination.

Her blood reports were suggestive of anemia but otherwise unremarkable, with negative reports for RA, CRP and ASO factors. ECG and 2D ECHO reports were normal. Analysis of arterial blood gases (ABGA) showed mild reduction in blood oxygen levels suggestive of hypoxemia. Pulmonary function tests revealed a mixed restrictive and obstructive pattern (FVC-68 % and FEV1-64 %). On her chest radiograph review, there were decreased vascular markings on right side with decreased volume of the right lung, mild rightward shifting of trachea and fine to coarse reticulations in right basal lung field which prompted further investigation.

HRCT was done using 16 slice CT machine which showed volume loss of right lung field, subpleural honeycombing, interlobular septal thickening suggestive of usual interstitial pneumonia (UIP) pattern, cystic and varicoid bronchiectatic changes, emphysematous changes, mild shift of mediastinum towards right side, compensatory hyperinflation of left lung and non-visualization of right pulmonary artery (Figure 1). This led to suspicion of secondary interstitial pneumonia pattern developing in absence of unilateral pulmonary circulation for which contrast study in the form of CT pulmonary angiography was carried out using nonionic contrast "iohexol". Study revealed absence of pulmonary artery from its origin on right side, reduction in diameter of right superior and inferior pulmonary veins and collaterals arising from systemic arteries- the right subclavian artery, right prominent bronchial artery and right subdiaphragmatic arteries were noted supplying right lung (Figures 2-4), the findings were compatible with unilateral congenital right pulmonary artery agenesis and dilated bronchial arteries and resultant right sided UIP changes.

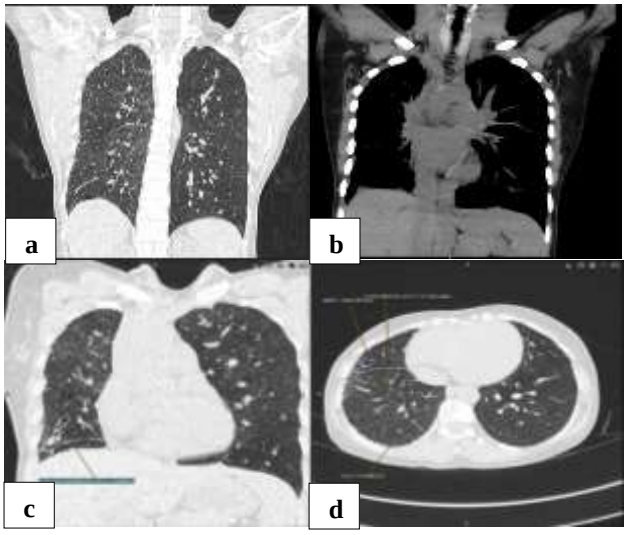

Figure 1: (a) and (c) High resolution computed tomography with coronal reformatted images, (b) soft tissue reconstructed image shows mild reduction in right lung volume with hyperinflation of left lung and subpleural honey combing and interstitial septal thickening, and (d) few varicoid bronchiectatic changes seen in right lower lobe.

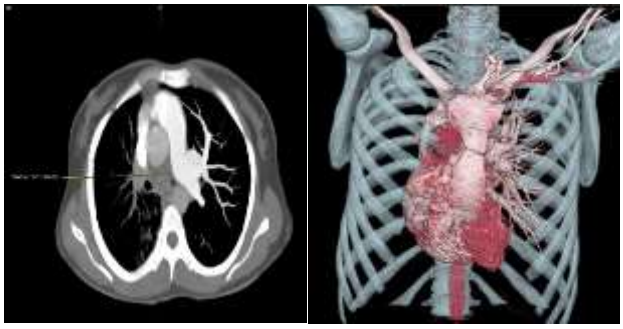

Figure 2: Axial section and reconstructed VRT images of contrast pulmonary angiography shows absent right pulmonary artery from pulmonary trunk.

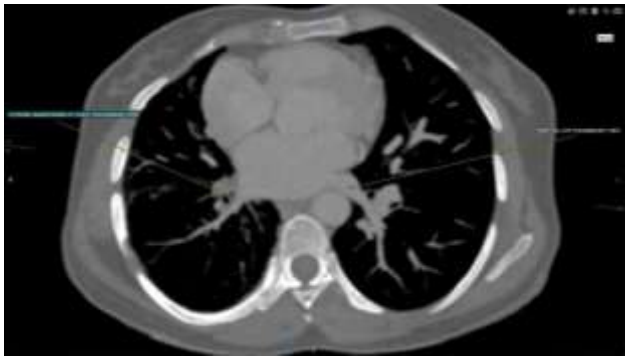

Figure 3: Axial section of venous phase of CT pulmonary angiography showed narrowed right inferior pulmonary vein compared to left.

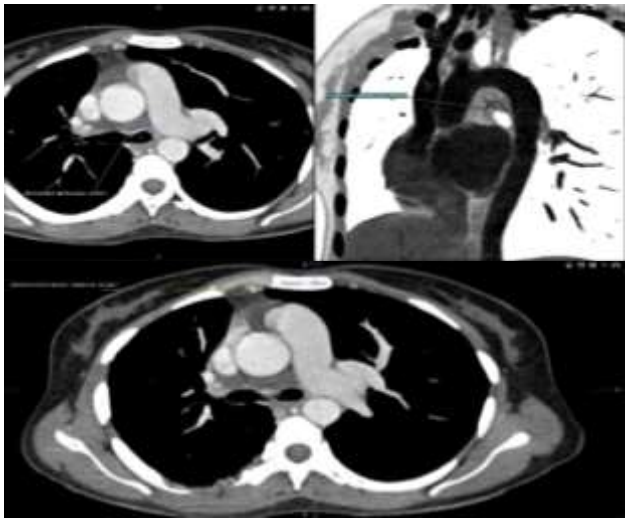

Figure 4: Axial section and reformatted images of pulmonary angiography shows Arterial supply to right lung from collaterals from right internal thoracic artery and prominent systemic supply from bronchial arteries arising from descending aorta.

After the diagnosis was made, the patient received conservative treatment with MDI budesonide and formoterol MDI nebulization and with precautionary pneumococcal and meningococcal vaccination.

In subsequent visits, she had minor respiratory symptoms with no limitations in physical activities.

DISCUSSION

Interstitial lung diseases usually present with involvement of bilateral lung fields although asymmetrical, but the differentials for unilateral involvement are far and few, being interruption of main pulmonary circulation (arterial/venous) of one lung like in UAPA, sarcoma of unilateral pulmonary artery, unilateral pulmonary venous thrombosis, and radiation-induced ILD.[2,5]

UPAA was first diagnosed in 1868 and since then, very few cases have been described in the literature. The estimated prevalence of single UPAA is 1/200,000 patients, and there is no sex predilection.[4,6] While median age of diagnosis is 14 years, in our case the diagnosis was made in adulthood.[3] In 60 percentage of cases described, the right side is affected. UPAA can be accompanied by other congenital heart abnormalities, but this was not observed in case of our patient.[4]

Pulmonary artery branches are formed from the sixth aortic arches in embryos during the fourth week of gestation. During normal development, the proximal portions of the sixth arch persist, forming the right and left main branches of the pulmonary trunk. The pulmonary vascular bed may form normally even when a main branch of the pulmonary artery is absent because it develops from ventral branches of the dorsal aorta.[7]

Blood in the affected lung is supplied by the bronchial arteries or by abnormal collaterals arising from the bronchial, subclavian, intercostal and sub-diaphragmatic arteries. In approximately 4% of the cases, a communication between coronary and bronchial arteries is present. Despite collateral circulation, the affected lung is often hypoplastic, and interstitial pulmonary pathology including bronchiectasis is revealed on CT imaging.[4]

Since the common age of presentation is the neonatal period and childhood, it is frequently misdiagnosed in the adulthood and is often not included in the list of differential diagnoses of the unilateral hyperlucent lung.[7] The most common presenting symptoms in patients with pulmonary artery agenesis include recurrent pulmonary infection, mild dyspnea and decreased exercise tolerance.[3,4] However severe symptoms including hemoptysis (in 20% of patients), pleural effusion could also occur. Pulmonary hypertension is observed in one fourth of the patients and is a determinant of long-term survival.[8]

Contrast enhanced chest CT is adequate for UPAA diagnosis, limiting the use of more invasive techniques.[9] Parenchymal findings include bronchiectasis and mosaic attenuation pattern in both lungs, possibly caused by an increased perfusion of the unaffected lung, by the development of pulmonary hypertension or by a compensatory over-inflation of the unaffected lung.[9] In addition, CT scan provides useful information regarding pulmonary hypertension, congenital heart defects and perfusion of lung parenchyma.[9] Pulmonary angiography and digital subtraction angiography are the golden standards in order to establish a definitive diagnosis and identify the collateral blood flow to the affected lung. Bronchoscopic findings include signs of chronic bronchitis, mainly in patients with recurrent pulmonary infections, bronchiectatic lesions and vascular changes (plexuses of dilated blood vessels, mesh-like vascularization) that may cause hemoptysis.

Treatment of unilateral pulmonary artery agenesis comprises surgical, pharmacological and behavioral management.[6] In cases detected early in the course where the lungs show changes of scarring from recurrent infections and changes of interstitial lung disease the treatment is pharmacological with use of steroids and vaccination against of pneumococcus and meningococcus to prevent recurrent infections. Pneumonectomy and surgical revascularization are considered in extreme cases of recurrent hemoptysis and pulmonary hypertension.[6] Selective embolization of bronchial or non-bronchial systemic arteries is a valid alternative for patients with massive hemoptysis not eligible for surgery.[10]

CONCLUSION

For patients who present with recurrent chest infections and an abnormal chest radiograph with unilateral hyperlucent lung with reticular markings, congenital anomaly like unilateral agenesis of pulmonary artery should be considered in the differential diagnosis. Pulmonary artery agenesis is a rare anomaly where the patient may remain asymptomatic till adulthood. Imaging especially contrast CT plays a major role in the diagnosis and detecting the associated findings in heart and lungs. A careful and thorough diagnosis with exclusion of all the possible differentials can avoid serious complications as pulmonary hypertension and life-threatening hemoptysis.

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