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Pulmonary sequestration with superadded infection in an adult

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Abstract
Pulmonary sequestration (PS) also called accessory lung is a rare congenital malformation characterized by an abnormal mass of dysplastic lung tissue supplied by an anomalous systemic artery and it is separated from normal bronchopulmonary tree.
Its incidence is around 0.1%. All cases occur sporadically It is uncommonly diagnosed in adults. It can manifest with variable presentations ranging from being asymptomatic to hemoptysis and recurrent pneumonia.
Diagnosis is confirmed with computed tomography angiogram or angiography. Treatment is embolization of the afferent artery and surgical removal of the pulmonary sequestration.
We report a case of a 55 -year-old female who presented with complaint of fever, cough breathlessness, since 1 year and was eventually diagnosed with pulmonary sequestration.

Keywords: pulmonary sequestration, Intralobar sequestration, Extralobar sequestration, Hemoptysis, aberrant artery

Introduction
Pulmonary sequestration has two distinct forms: intralobar, as in our patient, and extralobar, The diagnosis can easily missed in adults, as majority of the symptoms and the imaging findings overlap with other pulmonary diseases.
This article presents a case of intralobar pulmonary sequestration that presented to us during adulthood, and is covers a brief review of clinical features, diagnostic strategies, and management options of the PS.

Case history
A 55 year old Indian female patient prsented with a chief complaint of fever, cough and breathlessness since one year, which had worsened with hemoptysis from last one week. Her symptoms persisted despite antibiotic therapy. Vitals including oxygen saturation were within normal limits. On auscultation rough breath sounds were present in the right lower lung field.
Owing to the symptoms, patient was referred to Radiology department. Chest x-ray revealed a right basilar parenchymal opacity. Patient was then advised HRCT chest which revealed a large 7.5 x 7.4 cm sized well defined thick walled lesion in the posterior basal and medial basal segments of the right lower lobe with surrounding consolidation. The lesion showed mild peripheral enhancement on post contrast study with non-enhancing central necrotic component. Tiny foci of calcification was also noted in the periphery of the lesion probably in the surrounding area of consolidation. Few foci of air attenuation were noted in the non-dependent superior portion.
The lesion is supplied by a dilated feeding artery, arising from descending thoracic aorta at D9 vertebral level. The lesion shows venous drainage into the right inferior pulmonary vein.

Minimal right sided pleural effusion was also noted
Few conglomerate, necrotic lymph nodes were seen in the right hilar and sub-carinal regions, largest measuring 3.5x 2 cms in the subcarinal region
Given the history and imaging features characteristic of intralobar sequestration, and ongoing symptoms, the patient was referred for right lower- lobe resection. The pathological also confirmed our diagnosis of an intralobar sequestration.

Discussion
Pulmonary sequestration is a very rare representing 0.15%-6.4% of all congenital pulmonary malformations condition that is seen in adults. It is characterized by an abnormal mass of dysplastic lung tissue supplied by an anomalous systemic artery and it is separated from normal bronchopulmonary tree. Pulmonary sequestrations are divided based on their locations, as either intralobar or extralobar depending on whether it has independent pleura or not.

The 2 types are
1. Intralobar Sequestration (ILS)
a. More common (75-80%)
b. Presents later in childhood / adulthood and with recurrent infections
2. Extralobar Sequestration (ELS)
a. less common (15-25%)
b. Presents in the neonatal period with respiratory distress, cyanosis, or infection
c. M: F ratio ~4:1

Almost 97% of PS cases were located in the lower lobe, and PS in the left lower lobe is two to three times more common than that in the right lower lobe whereas in our patient, the sequestration was located in the right lower lobe.They receive blood supply from the systemic circulation, most commonly the thoracic or abdominal aorta Pulmonary sequestrations have venous drainage into either the pulmonary veins, azygous vein, hemiazygos vein, inferior vena cava, or right atrium.
The most accepted hypothesis of PS is that it results from

formation of an accessory lung bud inferior to the normal lung buds during development and gets its blood supply from foregut vascular. If the accessory lung bud develops before formation of the pleura, both the normal and sequestered lung tissue are covered by the same pleura, resulting in foremation of ILS; If the accessory lung bud develops after formation of the pleura, the sequestered lung tissue forms its own pleural covering, resulting in ELS

The investigation of choice is CT angiography as it is noninvasive compared with catheter aortography, which used to be necessary for determining the arterial supply and diagnosis. Plain chest radiograph is usually nonspecific, showsan ill-defined consolidation that mimics pneumonia, or shows a solitary soft tissue mass. Ultrasound plays an important role in the prenatal diagnosis and follow-up as it can identify the origin of the aberrant vessel feeding the lung mass. MRI can demonstrate signal voids of the aberrant feeding vessel on T2-weighted imaging, and magnetic resonance angiography can demonstrate the systemic blood supply to the sequestered lung

The treatment involves surgical resection to avoid infection and damage to the lung parenchyma Surgery is recommended even in asymptomatic patients

Conclusion

Diagnosis of pulmonary sequestration can be delayed or misdiagnosed in the adult population as the presenting symptoms are often mimicked with common pulmonary diseases, such as pneumonia and asthma. CT Angiography is preferred imaging modality for preoperative planning by identifying the lesion.

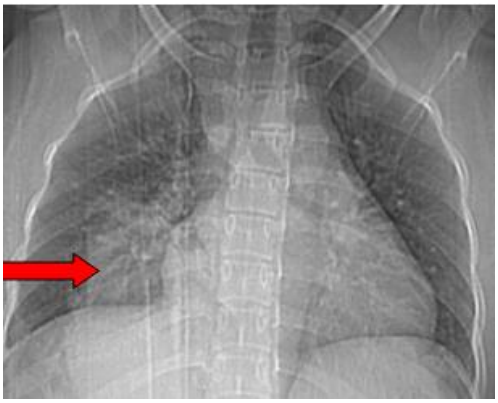

Fig 1: Radiograph of the chest (PA View) showing opacity in the right lower zone (Red arrow)

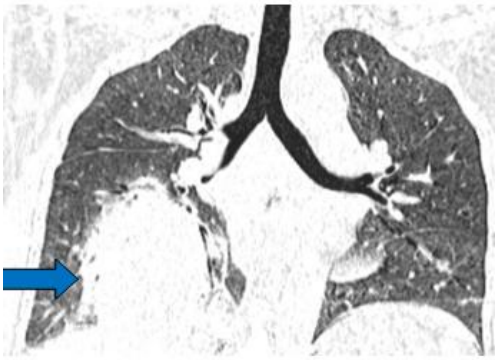

Fig 2: HRCT of the chest (lung window, coronal view) showing a large 7.5 x 7.4 cm sized well defined thick walled lesion(Blue arrow) in the posterior basal and medial basal segments of the right lower lobe with surrounding consolidation

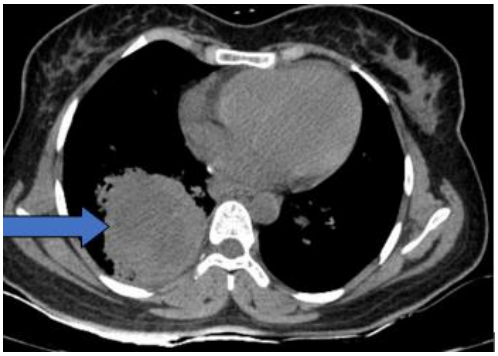

Fig 3: HRCT of the chest plain(axial view) showing a large well defined thick walled lesion(Blue arrow) in the posterior basal and medial basal segments of the right lower lobe with non enhancing central necrotic component.

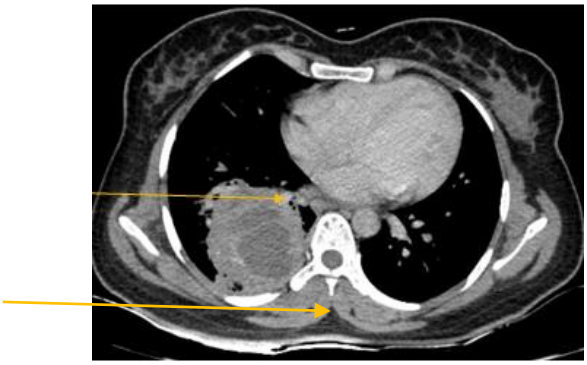

Fig 4: HRCT of the chest (axial view) showing lesion supplied by a dilated feeding artery, arising from descending thoracic aorta and venous drainage into the right inferior pulmonary vein

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