

**PULMONARY SEQUESTRATION – RARE FINDINGS****DR. SUBRAMANIAN S.*¹, DR. APAR JINDAL², DR. ABU BUCKER³
AND DR. N. MEENAKSHI⁴**¹*Associate Professor – Respiratory Medicine, Chettinad Hospital and Research Institute, Chennai*²*M.D. Post Graduate, Respiratory Medicine, Chettinad Hospital & Research Institute.*³*Associate Professor – Radiodiagnosis, Chettinad Hospital and Research Institute, Chennai*⁴*Professor and HOD – Respiratory Medicine, Chettinad Hospital and Research Institute, Chennai***ABSTRACT**

Pulmonary sequestration is a developmental disorder characterized by nonfunctioning abnormal pulmonary parenchyma that has no connection with the tracheobronchial tree and receiving blood supply from a systemic artery. Two forms have been described, Intralobar (within the normal lung visceral pleura), and Extralobar (totally separate with its own pleural covering). We present a case of a 48 year old female with recurrent hemoptysis for one year. Chest radiograph demonstrated a well defined opacity in the right lower zone. CECT thorax showed a round heterogeneously enhancing mass in Right Lower Lobe Lateral Basal Segment. 3D reconstruction imaging revealed an anomalous artery arising from the Right Renal Artery supplying the mass and venous drainage into the Inferior Pulmonary Vein establishing a diagnosis of Right Lower Lobe Intralobar Sequestration. The rare findings in our case were the anomalous arterial supply from a branch of right renal artery and sequestration in Right Lower Lobe.

KEYWORDS: Pulmonary sequestration, Recurrent haemoptysis, Foregut malformation, Contrast Enhanced CT scan of thorax with 3D reconstruction



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INTRODUCTION

Pulmonary sequestration is an uncommon disease of nonfunctioning abnormal pulmonary parenchyma having no connection with the tracheobronchial tree and receiving blood supply from a systemic artery. Recurrent hemoptysis is a recognized but uncommon complication of intralobar sequestration. A definitive diagnosis requires the demonstration of the systemic arterial supply to the sequestered lung.

Case Report

A 48 years old female presented with complaints of recurrent hemoptysis for one year. Routine blood investigations were normal, sputum was negative for Acid Fast Bacilli, bacteria and fungus. The chest

radiograph demonstrated a well defined opacity in the right lower zone (Fig 1). A CECT scan of the thorax revealed a round heterogeneously enhancing mass with abundant peripheral vascularity of size 5.3 X 3.5 X 4.6cms in right lower lobe lateral basal segment. Punctate calcification noted within the mass with surrounding consolidation and displacement and compression of the adjacent bronchial tree (Fig.2,3,4). 3D Reconstruction images revealed an anomalous artery arising from the Right Renal Artery and passing upwards into the thoracic cavity to supply the sequestered lung tissue (Fig 5,7,8). Venous drainage noted into the inferior pulmonary vein (Fig 6). Hence a diagnosis of right lower lobe intralobar sequestration was made.

Figure 1
CT Scout film showing large non homogeneous opacity
In the Right Lower Zone



Figure 2

CECT Thorax showing a Round heterogeneously enhancing mass of size 5.3X3.5X4.6cms in right lower lobe lateral basal segment. Punctate calcification noted within the mass.

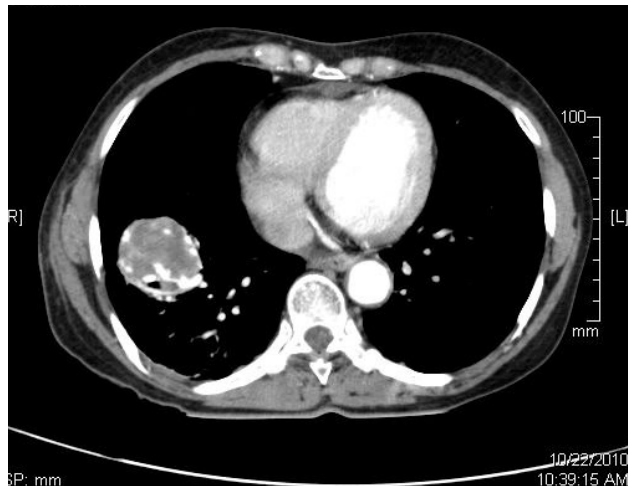


Figure 3

Adjacent bronchial tree displaced by the mass and mass noted compressing the distal bronchi and causing adjacent Bronchiectasis. No air bronchogram noted within the mass.



Figure 4

Anomalous tortuous artery noted supplying the mass.

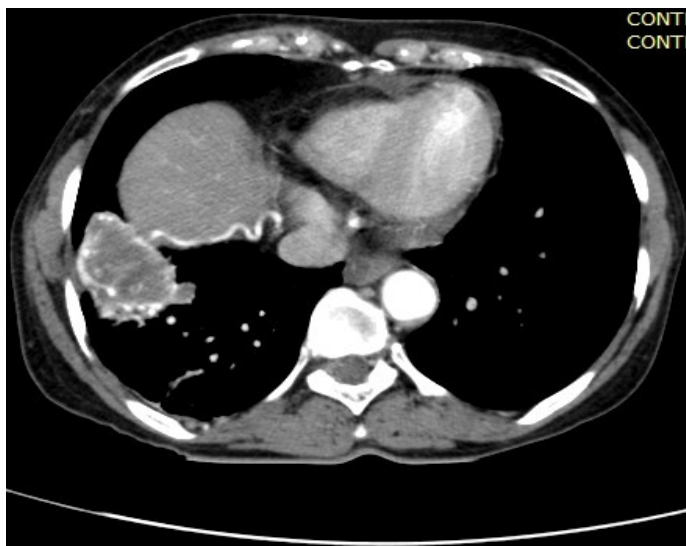


Figure 5

Venous drainage into the Right Inferior Pulmonary Vein.

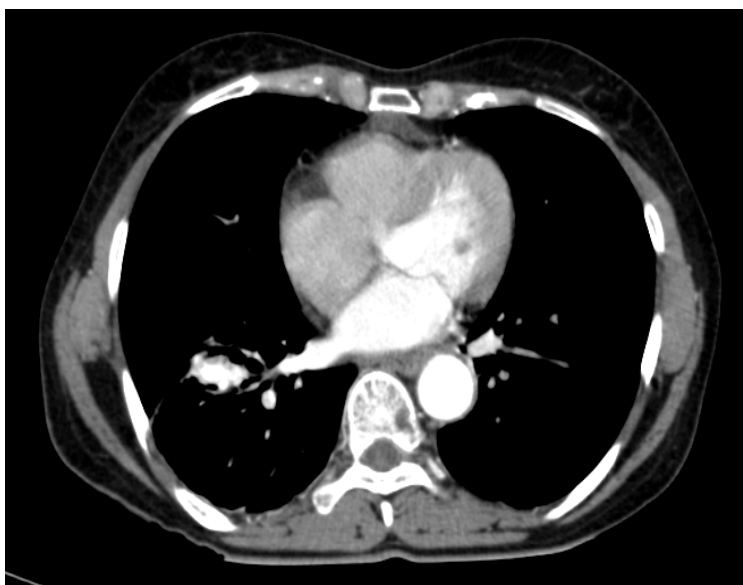
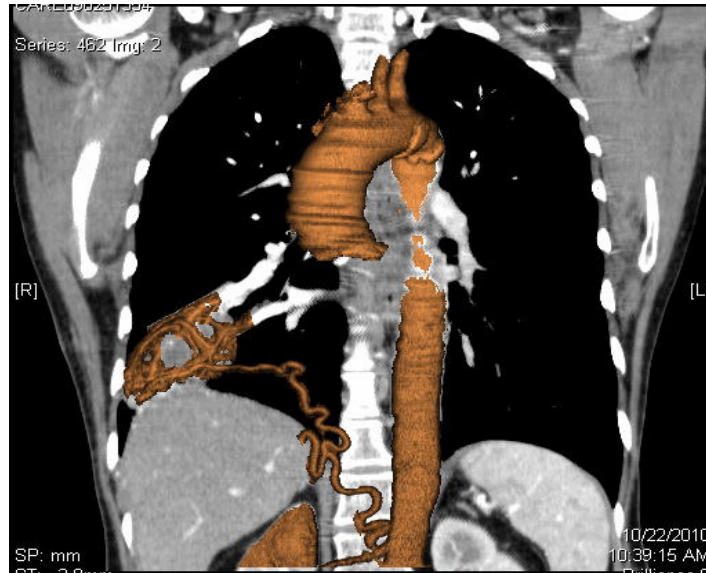


Figure 6

3D Reconstruction image demonstrating anomalous artery originating from the Right Renal Artery and passing upwards into the thoracic cavity, supplying the sequestered lung tissue.



DISCUSSION

Bronchopulmonary sequestration is a developmental malformation of the lung which involves a portion of the lung parenchyma detaching itself from the normal tracheobronchial tree and receiving blood supply from a systemic artery. It is classified into Intralobar sequestration where the anomaly is located within the same visceral pleura and Extralobar sequestration where it is enclosed in its own pleural covering. They are thought to occur due to development of a separated branch fragment and retention of its embryonic vascular supply. Early separation from the primitive lung bud or from a separated foregut diverticulum with a separated pleural covering and development in the mediastinum or outside the thorax forms the extralobar sequestration. By contrast the intralobar sequestration is thought to arise from separated bronchial divisions of the developing lung bud, continuing to develop within the lung itself. Intralobar sequestration is usually found in the paravertebral gutter within the posterior segment of the left lower lobe and less commonly in the posterior segment of right lower lobe. Infrequently it is associated with oesophageal diverticula, diaphragmatic hernia and a variety of skeletal

and cardiac defects¹. The abnormal tissue derives its arterial supply from the aorta or its branches, most commonly the descending thoracic aorta. Venous drainage is almost always via the pulmonary venous system. Extralobar sequestration is usually situated between the inferior surface of the lower lobe and the diaphragm, or within the diaphragm or less commonly in the mediastinum or retroperitoneum. The arterial supply is usually derived from the abdominal aorta and venous drainage is into the systemic veins inferior vena cava, azygous or hemi-azygous veins or the portal system. It is most commonly seen in neonates and is associated with other congenital abnormalities like paralysis of ipsilateral hemidiaphragm and diaphragmatic hernia. Clinically most patients are asymptomatic. Intralobar sequestration usually presents in adulthood¹ as recurrent respiratory infections. Signs and symptoms are usually of acute lower lobe pneumonia. Extralobar sequestration is most commonly identified incidentally as a homogeneous soft tissue mass in an asymptomatic patient. Since it is covered in its own separate pleura chances of infection are less². Radiographically intralobar sequestration presents as a homogeneous

opacity in the posterior basal segment of the left lower lobe, usually along the hemidiaphragm.^{3,4,5} In contrast extralobar sequestration appears as a sharply defined triangular opacity in the posterior costophrenic angle, usually adjacent to the left hemidiaphragm. Definitive diagnosis is based on demonstration of anomalous arterial supply. Aortography usually shows the feeding arteries with multiples feeders identified in about 20% cases⁶. Review of literature revealed similar cases of intralobar sequestration being supplied by branches from the right renal artery^{7,8}. Other similar rare presentations found in the literature search were from the coronary artery and right bronchial artery^{9, 10}. Identification of the systemic arterial supply can also be accomplished by CT^{3,5}. Demonstration of the course of the aberrant systemic vessel has improved considerably with the use of multiplanar reformations including 3D

reconstruction imaging. With the introduction of Magnetic Resonance Imaging and Magnetic Resonance Angiography more patients can be diagnosed with less invasive methods^{11,12,13}

CONCLUSION

High degree of suspicion towards congenital lung anomalies is needed in cases of unexplained hemoptysis, as in our case. The rare findings here were the presence of the sequestered lung tissue in the Right Lower Lateral Basal Segment with arterial supply from the Right Renal Artery. The advent of multidetector CT with 3D reconstruction imaging and Magnetic Resonance Angiography has greatly revolutionized the diagnosis of sequestration by demonstrating the systemic arterial supply by less invasive means.

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