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Pulmonary Sequestration Supplied by an Aberrant Infra-diapharagmatic Artery

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Abstract

Sequestered lung is a discrete mass of lung tissue, where airways do not connect with the rest of the trachea-bronchial tree, and that receives its blood supply from a systemic artery rather than a pulmonary one, while sometimes maintaining normal venous drainage to the heart. Abnormal feeding arteries have been reported from the thoracic and abdominal aorta, celiac trunk, coronaries among others. In this article we describe a patient with left lower lobe sequestration supplied by a peculiar infra-diaphragmatic artery.

Keywords: Pulmonary embryology; Pulmonary sequestration; Intralobar sequestration; Rare lung diseases

Introduction

Bronchopulmunary sequestration (BPS) is a rare congenital anomaly that probably results from faulty embryogenesis of the trachea-bronchial tree. There are two types of sequestrations: intralobar and extralobar, depending on whether they share their pleural with the rest of the lung or not [1,2]. Sequestered parts of the lung tend to occur in the lower lobes. Detection of an abnormal feeding vessel originating from the systemic circulation is essential both for the diagnosis of pulmonary sequestration and for safe surgical resection of the lesion [3,4].

Case Presentation

A 22 year old non-smoker man presented to our department with two year history of persistent dry cough more at night. His past medical history was significant for frequent admissions to other hospitals because of recurrent pneumonias in the left lung. He had a persistent consolidation in the left lower lobe that failed to resolve on multiple courses of antibiotics. Otherwise, he had free past medical and surgical history. Previous evaluations included bronchoscopy with BAL and Brush which were unremarkable.

The patient was well built, vital signs were normal, physical exam was positive for decreased breath sounds and dullness on percussion over the left lung lower zone. Chest CT scan revealed consolidation involving the posterior segments of the left lower lobe (L9, L10) very suggestive of sequestration (Figure 1). CT aortogram was done and showed an anomalous artery arising from the abdominal aorta that supplies the consolidated LLL. (Figures 2 and 3). A diagnosis of

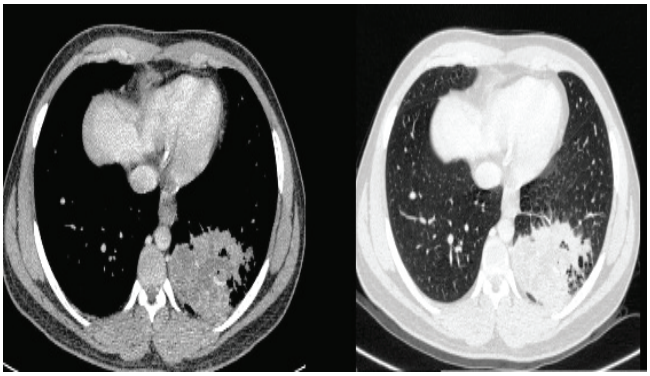

Figure 1: Chest CT scan showing consolidation in the left lower lobe (L9,L10) very suggestive of sequestration.

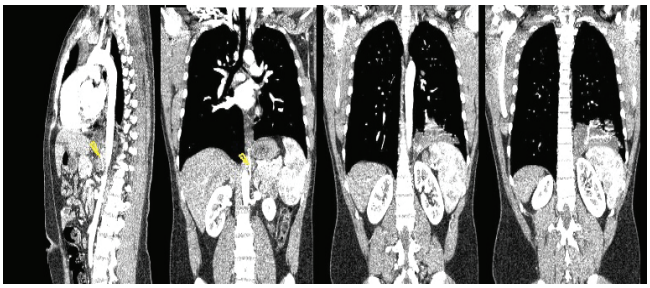

Figure 2: Sagittal and coronal CT scans with IV contrast showing the course of an aberrant artery arising from abdominal aorta and supplying the LLL. The artery penetrates the left hemidiaphragm.

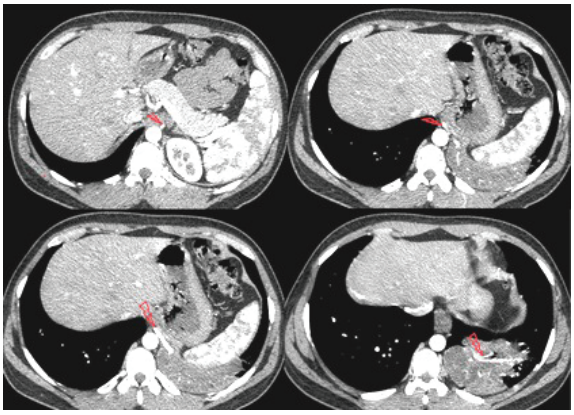

Figure 3: Axial CT scan with IV contrast: Aberrant artery supplies the sequestered lobe.

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Pulmonary intralobar LLL sequestration was made, and patient was referred for surgery in view of his symptoms. Informed consent was taken from the patient.

Discussion

Sequestration is a common pulmonary lesion that can be detected in childhood when extralobar, or it can evade diagnosis until late adulthood especially with intralobar lesions. The most common presentation in adulthood is with recurrent pulmonary infections [5].

Both types of pulmonary sequestration have systemic arterial supply, but differ in their pleura and venous drainage. Whereas the intralobar form shares the same pleura with rest of the ipsilateral lung and drains into left atrium via pulmonary veins, the extralobar form has a separate pleural and systemic venous drainage. Other congenital lung abnormalities include congenital pulmonary artery malformations (CPAM) that have an abnormal connection to the airways, but maintain pulmonary arterial supply and pulmonary venous drainage, Hybrid PBS/CPAM lesions that have histologic features of CPAM and a systemic artery similar to sequestration, and Bronchopulmonary-foregut malformation (BPFM) where airways connect to the gastrointestinal tract [6].

The etiology of pulmonary sequestration and several other bronchopulmonary anomalies is not fully understood. It is thought to occur when a second lung bud arises from the ventral aspect of primitive foregut, just below the normal lung bud. If the accessory bud arises before the development of pleura, it shares the same pleura with the rest of the ipsilateral lung and becomes an intralobar sequestration, where as if it arises after pleural formation, it grows separately and acquires its own pleural becoming an extralobar sequestration.

Once sequestration is discovered, it is recommended that patients undergo surgical resection of the sequestered portion-even in the absence of symptoms- to avoid future complications, such as infection, bleeding and potential malignant transformation [6,7].

Conclusion

Pulmonary sequestration refers to lung tissue that receives aberrant arterial supply from the systemic vessels, mainly the aorta and it`s major branches. In this case, the sequestered left lower lobe received blood supply from an abnormal vessel, branching off the abdominal aorta, and ascending through the esophageal hiatus.

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