CASE REPORT

Intralobar Sequestration: A Case Report

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Abstract

Introduction: Intralobar sequestration (ILS) or intrapulmonary sequestration lacks visceral pleura and is located within a normal lobe and accounts for 75% of bronchopulmonary sequestration (BPS). The majority of ILS is located in the posterior basal segment of the left lobe. Most commonly patients present with signs of infection later in life.

Objective: Chest X-rays and CT thorax is the commonest imaging done to confirm this diagnosis.

Conclusion: Surgical excision is curative extralobular sequestration, and congenital pulmonary airway malformation (CPAM) and bronchopulmonary foregut malformation (BPFM) are other anatomical classifications.

Keywords: Bronchopulmonary, Sequestered lung, Sequestration, Thoracic, Uniportal VATS.

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Introduction

Pulmonary sequestration is a rare congenital abnormality that is also known as bronchopulmonary sequestration (BPS). It is the nonfunctioning lung tissue that does not communicate with the tracheobronchial tree and derives its blood supply from systemic circulation. This condition is broadly classified as extralobar and intralobar BPS. We report a case of intralobar sequestration (ILS) left lower lobe treated surgically.

Case Report

A 38-year-old lady presented with recurrent hemoptysis and fever for 6 months. She had been seen and treated for pneumonia previously from multiple GP clinics with oral antibiotics but her symptoms persisted. In view of her persistent symptoms, she was subsequently referred to our hospital for further management. Her full blood count and baseline blood investigations were unremarkable, tuberculosis screening with the Mantoux test and sputum for acid-fast bacilli were negative and her chest X-ray did not show any abnormalities. Due to the persistent hemoptysis, a bronchoscopy was performed and there was a presence of blood clot over the left lower lobe bronchus. Subsequently, a contrast-enhanced CT thorax with supplementary angiogram was done (Fig. 1) in which a left lobe lesion suspicious of arteriovenous malformation or sequestered lung was seen.

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In view of her CT findings, an angiography was done which confirmed the diagnosis of intralobar lung sequestration in left lower lobe with an aberrant artery from the thoracic aorta (Fig. 1). The patient underwent uniportal video assisted thoracoscopic (VATS) and left lower lobectomy, as seen in Figure 2, and was discharged home day 5 postsurgery. Histopathology reported sequestered lung with chronic inflammation. On follow-up, she was well and free from her symptom.

Discussion

ILS is the most common form of BPS.[1] It comprises up to 75% of sequestrations, while the remainder 25% are extralobar

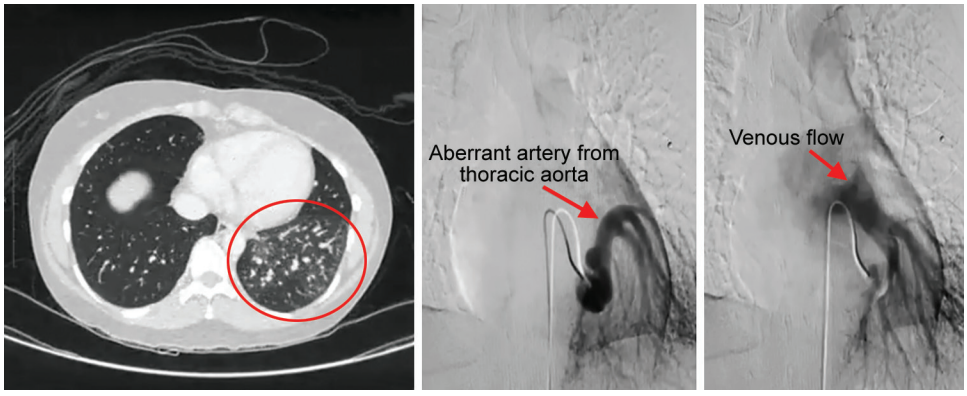

Fig. 1: Axial contrast enchanced computed tomography (CECT) thorax left lobe lesion with angiogram showing aberrant artery

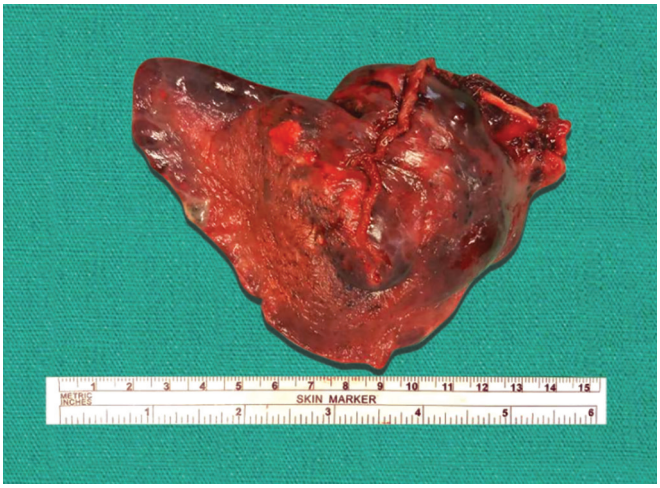

Fig. 2: Medial view of lobectomy specimen with the sequestered lung

Table 1: Differences between intralobar and extralobar pulmonary sequestration[6]

	Intralobar (75%)	*Extralobar (25%)*
Pleural covering	Within normal lung	Own covering
Etiology	Congenital	Congenital
Presentation	Infection	Incidental
Associated anomalies	Rare	Common (65%)
Arterial supply	Systemic	Systemic
Venous drainage	Systemic or pulmonary	Systemic
Macroscopic	Fibrotic with cystic areas	Solid, spongy mass

sequestration (ELS) (Table 1). Genetic predisposition for ILS is equal in both males and females; however, there is a male preponderance for ELS. It is postulated that BPS originates early in the pseudoglandular stage of lung development, during gestation, prior to the separation of the aortic and pulmonary circulations. This would explain its connection to the systemic circulation and the presence of a separate visceral pleura in ELS or the absence in ILS, the occurrence of hybrid lesions with features of BPS and congenital pulmonary airway malformation (CPAM), and the associations with bronchogenic cysts or connections to the foregut, as well as associated anomalies such as congenital diaphragmatic hernia.[2]

The presentation of bronchopulmonary sequestration depends on the type, size, and location of the lesion. Symptomatic lesions can present with respiratory distress in the neonatal period or later in adulthood with lung infection typically fever, cough, hemoptysis, and occasionally pleuritic chest pain. In ELS, diagnosis is often incidental on a chest radiograph. They may present with heart failure due to excessive flow through the aberrant artery or with bleeding.[3] Patients with ELS rarely present with infection.

Both types of BPS can be identified incidentally on a plain chest radiograph. Sequestrations typically appear as a uniformly dense mass within the thoracic cavity and recurrent infections can lead to the development of cystic areas within the mass. The left hemithorax is commonly involved for both ELS and ILS.[4] Computed tomography (CT) is best used to visualize parenchymal abnormalities associated with BPS. They most commonly appear as a solid mass which is homogeneous or heterogeneous with cystic changes. Emphysematous changes at the margin of the lesion are pathognomonic for the diagnosis.[4] However, conventional CT does not routinely demonstrate the aberrant systemic artery.[5] This is better visualized using an MRI which is well suited for the diagnosis of bronchopulmonary sequestration because of its capacity to show the sequestration which may be a well-defined, irregular, or branch-like hyperintense mass with precise anatomic localization as well as to define the size, origin, and course of both the aberrant systemic artery and the venous drainage.[5]

The approach to management depends on whether the patient is symptomatic or asymptomatic. All symptomatic patients should undergo surgical excision which will be curative and has minimal morbidity. In asymptomatic patients, we recommend surgery if the lesion occupies >20% of the lobe or if the mass has multifocal cysts or in the presence of pneumothorax. Low-risk patients are those without the above-mentioned features and can be managed conservatively and planned for elective surgical resection, because they have a moderate risk of developing infection later in life, particularly in ILS. When infection or respiratory symptoms occur, surgery becomes urgent and is associated with a higher risk of complications such as air leak, effusion, and pneumonia, as compared with elective surgery in asymptomatic patients.[1] In our center, we perform uniportal VATS and resection in patients with BPS as demonstrated in this case.

Conclusion

A case of ILS of the lung in left lower lobe in a 38-year-old lady has been presented. Diagnostic methods have been discussed. The patient underwent left lower lobectomy through uniportal VATS. The patient made an uneventful recovery. The prognosis is excellent for patients who undergo elective surgical intervention. Lung parenchyma undergoes compensatory growth and development with normal pulmonary function.

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