Pulmonary Sequestration: A Case Report

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Received: 10 Dec. 2018; Accepted: 18 Jun. 2019

Abstract- Pulmonary sequestration is a relatively rare congenital malformation characterized by an abnormal mass of dysplastic lung tissue. This mass is separated from the bronchopulmonary tree and vascularized by an aberrant systemic artery. Pulmonary sequestration common symptoms are chest pain, persistent dry cough, shortness of breath, hemoptysis, and recurrent attractions of pneumonia. We report a case of a 45-year-old man who suffered from recurrent cough for one year and bleeding with pain in the left chest. He was eventually diagnosed with intralobar pulmonary sequestration. Surgical removal of the sequestration generally has good outcomes.

Acta Med Iran 2019;57(6):398-401.

Keywords: Lung; Bronchopulmonary sequestration; Congenital abnormalities; Chest pain

Introduction

Pulmonary sequestration is a relatively rare congenital abnormality where non-functioning lung tissue is separated from the bronchopulmonary tree and supplied with blood from an unusual source (1). Rokitansky and Rektorizic reported the first case in 1861, with their "Fraction Theory" which hypothesized that the isolated entity is a separation of naturally developed lung, and after that, several theories were presented to describe the emergence of this abnormality (2). In 1946, Pryce *et al*., used the term Sequestration for the first time to define this abnormality (3,4). There are two types of pulmonary sequestration:

1) Intralobar pulmonary sequestration (ILS)
2) Extralobar pulmonary sequestration (ELS)

In intralobar pulmonary sequestration, abnormalities are part of the normal parenchyma without lining (Figure 1A), but in the extralobar pulmonary sequestration, the abnormality is separated from the lung tissue with pleural effusion (Figure 1B) (4,5).

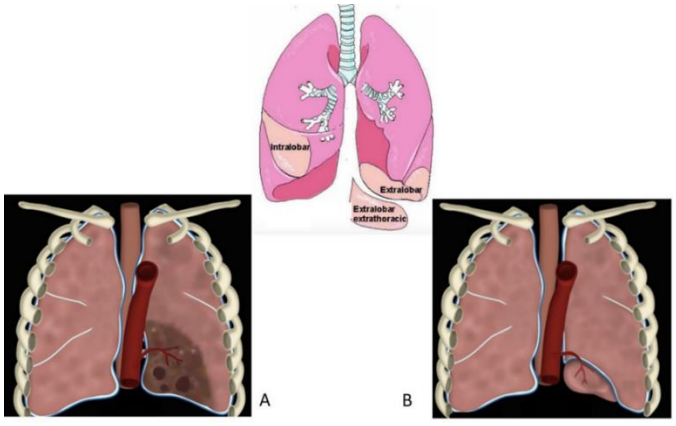

Figure 1. Schematic illustration of four different types of pulmonary sequestration anomalies. (A) Type (ILS), (B) Type (ELS)

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The ILS is more common, and they are approximately six times more prevalent than the ELS. Among the ELS cases, about 10-15% of cases are seen in the abdomen. Extralobar sequestration in the abdomen shows up in two forms:

(a) Infra diaphragmatic extralobar pulmonary sequestrations are commonly reported and (b) intradiaphragmatic extralobar pulmonary sequestration with less reported cases.

In general, pulmonary sequestration is mainly seen on the left side of the body, and more precisely, intralobar sequestration is commonly seen in the lower lobes of the lung (6,7).

Pulmonary sequestration is usually asymptomatic. However, common symptoms are chest pain, cough, shortness of breath, hemoptysis, as well as recurrent attractions of pneumonia (8). We now demonstrate a case of a 45-year-old man with recurrent cough and left-side chest pain, which eventually was diagnosed as ILS detected by Computed Tomography Angiography (CTA).

Case Report

A 45-year-old male patient suffered from a recurrent cough for one year, the situation worsened with bleeding and pain in the left chest, and finally he was wheeled into an emergency room in the hospital. His physical exam was normal; he had some persistent coughs and was febrile with a temperature of 38° C and blood pressure 120/80. Chest examination showed diminished breath sounds in the left lower lung area and wheezing in the upper lung area with no rhonchi. The patient had no history of smoking and alcohol, and his Purified Protein Derivative (PPD) and Human Immunodeficiency Virus (HIV) tests were negative.

His laboratory tests, including liver and renal function tests, complete blood count, erythrocyte sedimentation rate, electrolytes, urinalysis, and tuberculosis antibody tests were all negative.

CTA scan showed a pleura-based lesion (47 x 6 mm) in the back, medial, and inferior of the left lung. This mass was initially concerned with malignancy, but the biopsy showed a negative result with no malignant and abnormal cells in this mass. Interventional radiology showed that the feeding of this lung mass is a systemic artery that originates from the distal part of the descending thoracic aorta, and the venous drainage was through a pulmonary vein.

These symptoms were consistent with the pulmonary sequestration symptoms. Considering the history of the disease and the symptoms of the patient, including imaging features and pathological tests, it was found that the observed mass is ILS (Figure 2).

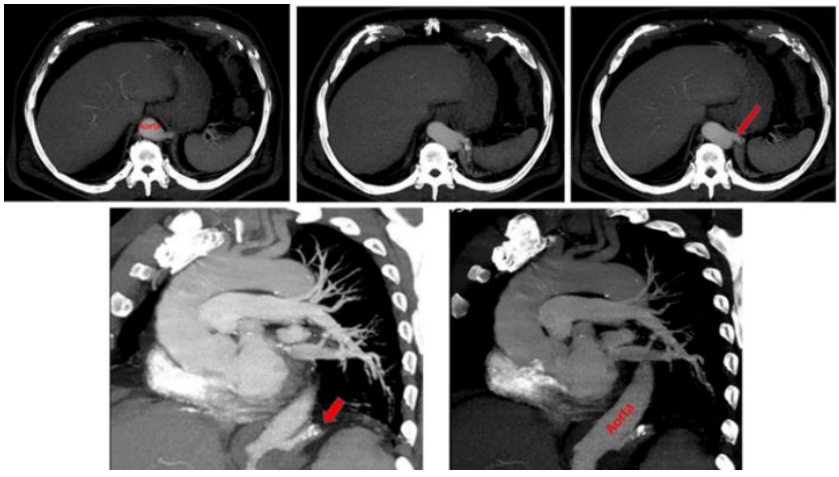

Figure 2. Axial and sagittal contrast-enhanced CT scan, Spiral is showing aberrant vessels arising from descending thoracic aorta. Red arrows show the lung mass feeding vessels

Discussion

Pulmonary sequestration is a rare malformation that includes a small fraction of the total pulmonary abnormality (about 6.4-0.15%) that is caused by a non-functional mass into the lung and is not associated with a bronchial tree.

ILS and ELS are differentiated based on the formation of an accessory lung bud to the normal lung buds during development and venous drainage. Such that if the

accessory lung bud develops before formation of the pleura, both the normal lung tissue and sequestered tissue covered with pleura and drains into the pulmonary veins, resulting in ILS; if the accessory lung bud develops after formation of the pleura, the sequestered tissue forms its own pleural covering and drains into the systemic veins, resulting in ELS (1,9).

The blood supply of ELS is typically from systemic arteries. This arises directly from the thoracic or abdominal aorta in approximately 80% of cases. The feeding vessel is typically single and measures between 0.5cm and 2cm in diameter. In approximately 15% of cases, ELS is supplied by smaller arteries from splenic, gastric, subclavian, and intercostal branches. Approximately 20% of ELS are supplied by multiple arteries.

In 5% of cases, the sequestrated mass is supplied by branches of the pulmonary artery or by both the pulmonary and systemic circulations. The venous drainage of ELS is usually systemic (80%), through the azygos system, the emiazygos system, or the vena cava to the right atrium. In approximately 25% of ELS, the venous drainage is partly through the pulmonary veins. Less common routes of drainage include the portal, intercostal, suprarenal, and other abdominal veins. An ELS supplied by pulmonary arteries is more likely to have pulmonary venous drainage (10,11).

ILS, which is typically located in the medial or posterior basal segments of the left lower lobe supplied by the anomalous aortic located within the inferior pulmonary ligament. In ILS, anomalous systemic arterial supply is via the descending thoracic aorta (72%) via the abdominal aorta, celiac axis, or splenic artery (21%), via intercostal artery (3%), and rarely via the subclavian, internal thoracic, and pericardiacophrenic arteries. Venous drainage is usually via the pulmonary veins, but it can also go through the azygos vein/hemiazygos system, portal vein, right atrium, or IVC (11,12).

Diagnosis of pulmonary sequestration depends upon the type of anomaly and presenting symptoms. Computed Tomography (CT) is normally sufficient in most adult cases and in some cases there is a need for CT angiography (12). In this case report, the chest CT angiography scan was clearly efficient in making the diagnosis and delineate the anatomic features notable for operation planning.

To treat pulmonary sequestration, the age of the patient should be taken into consideration, and appropriate treatment should be chosen based on the patient's condition to avoid any threatening issue to the patient. Some authors recommend embolization of the systemic artery as a treatment option. However, most authors recommend surgical removal, especially for extrathoracic lesions due to concern of malignant transformation (6).

Surgical resection is the primary treatment of pulmonary sequestrations, which frequently used for younger patients, while elderly or asymptomatic patients may be treated with antibiotics and corticosteroids. Two other treatments can be performed for patients with pulmonary sequestration: one consists of occluding the aberrant supply by endovascular approach; the other, which is widely used in the pediatric population is resection via minimal-access procedures such as video-assisted thoracoscopic surgery (VATS) (13).

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