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Pulmonary Sequestration in an Adult: A Case Report

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Abstract **Case Report**

Pulmonary sequestration (PS) is a form of congenital pulmonary malformation that is uncommonly diagnosed during adulthood. Pulmonary sequestrations can manifest with variable presentations. It can remain asymptomatic or present with more severe symptoms such as hemoptysis and recurrent pneumonia. The main feature of this disease is that partial lung tissues separate from the main lung during the embryonic period, receiving blood supply from systemic circulation arteries. We report a case of a 39 year old men smoker with medical history characterized by frequent infections since childhood, chest pain and hemoptysis. CT scan showed a partial PS of left lower lobe, supplied by an abnormal artery arising from descending aorta. Surgical treatment through a lung sequestrectomy and basal lobectomy resection was performed.

Keywords: Pulmonary sequestration – adult – CT-scan**.**

INTRODUCTION

Pulmonary sequestration (PS) is a rare congenital lung malformation defined as a nonfunctioning lung tissue with no apparent communication with the tracheobronchial tree [1]. Typically, it consists of a systemic arterial supply most commonly the descending thoracic aorta, to an associated anomalous lung segment with various forms of venous drainage [2, 3]. Pulmonary sequestration is primarily considered a childhood disease, as most cases are diagnosed early in life [4]. Sequestrations are typically divided into intralobar and extralobar forms [5]. Patients with PS can be asymptomatic and the diagnosis is achieved incidentally [6]. The most important presenting symptoms are recurrent pulmonary infections or productive cough. Severe hemoptysis as the first symptomatic manifestation has been reported very rarely [7]. The gold standard method for PS diagnosis is angiography but optimal identification of parenchymal and vascular structures involved in sequestration is obtained by CT scan [8]. In confirmed cases, surgical resection is the treatment of choice [9].

OBSERVATION

We report a case of a 39-year-old men smoker (22 pack years) with medical history characterized by frequent infections since childhood, recently complaining of right chest pain, and recurrent hemoptysis of mild-to-moderate abundance. Laboratory results including complete blood count, complete metabolic panel, and coagulation studies were unremarkable.

CT scan showed a large mass in the right lower lobe measuring 84 x 70 cm in axial dimensions with cysts and excavation areas. PS was supplied by an abnormal artery arising from descending aorta. Surgical treatment through a lung sequestrectomy and basal lobectomy resection was performed.

Pathological examination revealed that the mass did not contain atypical or malignant cells, but bronchial epithelial cells, macrophages, and other inflammatory cells.

After surgery, the patient was stabilized and discharged home without complications.

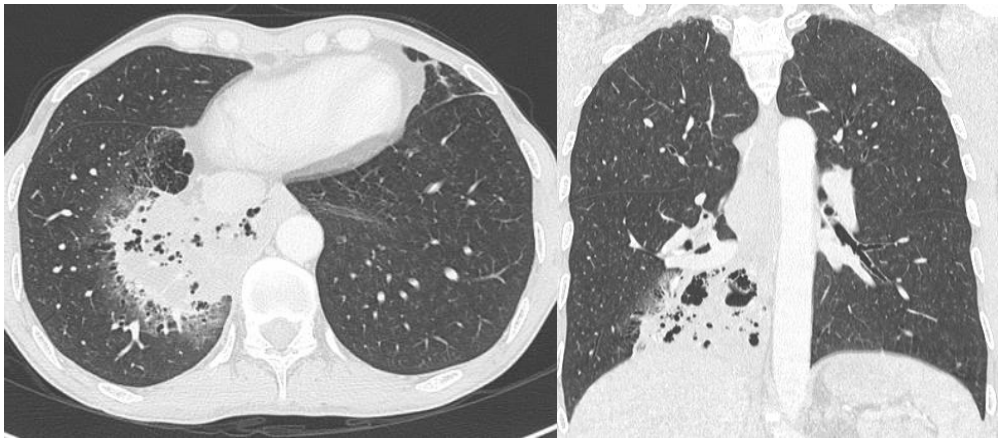

Figure 1: Chest CT scan lung windows in axial and coronal cross sections show a large mass in the right lower lobe with cysts and excavation areas

DISCUSSION

Firstly described in 1861 by Rokitanski and Rektorzik, Pulmonary Sequestration (PS) encompasses a spectrum of congenital lung anomalies exhibiting anomalous systemic arterial supply to the lungs and/or abnormal connection between one or more of the major components of lung tissue [10]. In 1946, Pryce introduced the term 'sequestration' to describe congenital abnormalities characterized by an anomalous systemic arterial supply to the lung and atresia or hypoplasia of the pulmonary artery [11]. In detail, sequestration was defined by Pryce as a “disconnected bronchus-pulmonary mass or cyst with an anomalous systemic blood supply” [11].

The incidence of congenital pulmonary airway malformations is 1 per 8300 to 35,000 accounting for 0.15% to 6.4% of all congenital lung malformations [5]. Sixty percent of lung sequestrations are diagnosed within the first decade of life [12]

In agreement with Pryce's nomenclature, PS is an area of nonfunctioning lung tissue with no apparent communication with the tracheobronchial tree [1]. Typically, it consists of a systemic arterial supply most commonly derived in 75% of cases from the thoracic aorta, 20% from the abdominal aorta, and in 15% from two different origins [5]. Venous drainage is accomplished by pulmonary veins [9]. Lung sequestration must be distinguished from Scimitar syndrome defined by partial anomalous pulmonary venous connection [13].

There are two forms of lung sequestration: intra-lobar pulmonary sequestration (IPS) or extra-lobar pulmonary sequestration (EPS) [5, 14]

Intrapulmonary sequestration, representing 85% of all lung sequestrations, is incorporated within normal lung tissue and the venous drainage is usually to the pulmonary circulation, and the posterior basal segment is most often affected [9,14].

EPS is separated from normal pulmonary tissue by its own visceral pleura and the lesion is located outside the lung; in EPS the venous drainage is usually to the systemic circulation [15]. 90% of EPS affects the left lower and are more frequently associated with other congenital pulmonary or cardiac malformations and are 4 times more common in males than in females [5, 9].

Patients with PS can be asymptomatic and the diagnosis is achieved incidentally [3]. Other presenting symptoms may include fever, cough, chest pain, wheezing, regurgitation, cyanosis, hemoptysis, and dyspnea [15, 16]. ELS rarely become infected because it is separated from the tracheobronchial tree by its own pleural investment. Therefore, misdiagnosis with other pathologies such as tuberculosis, pneumonia, pulmonary cyst, lung abscess, pulmonary tumor, pectus excavatum as well as asthma is reported [5].

Traditionally, the diagnosis of lung sequestration has been made by arterial angiography. Today, noninvasive imaging techniques, like CT or MR angiography, are used to demonstrate aberrant vessels and parenchymal disorders such as in our case report [9]. There are multiple radiologic manifestations of PS on computed tomography (CT) which include mass, consolidation with or without cysts, bronchiectasis, and cavitary lesions [15,16]. The arterial supply to PS most arises from the thoracic aorta, abdominal aorta, celiac artery, splenic artery, or even a coronary artery [3]. Most ILS drains to pulmonary veins while venous drainage for most ELS is to the azygos or hemiazygos vein or the inferior vena cava [12].

The optimal treatment for pulmonary sequestration involves surgical resection to avoid infection and damage to the lung parenchyma. Surgery is recommended even in asymptomatic patients [17]. In cases of pulmonary infection or destruction of normal lung tissue, a major resection such as lobectomy or pneumonectomy may be considered. Two alternative approaches should be noted. One is the exclusion of the

aberrant arterial supply via an endovascular approach, utilizing various occlusion devices. This carries the downside of retaining the un-aerated pulmonary parenchymal tissue that is still subject to recurrent infection [18]. The other option is resection via minimal-access procedures such as VATS lobectomy [19]. The advantages of this approach must be weighed against potential difficulty controlling the systemic arterial branches [14].

CONCLUSION

Pulmonary sequestration is a rare and difficult-to-diagnose lung disease, especially in the adult population. Due to not specific clinical presentation, PS recognition may be delayed and differential diagnosis may result challenging. Crucial for diagnosis is the contrast-enhanced lung CT scan which reproduces a detailed assessment of the vascular tree structure allowing the exclusion of other diseases like neoplasms and smoking-related diseases. The decision regarding surgical resection needs to weigh various factors including clinical manifestations related to PS, risk of surgical complications, comorbidities, and individual patient preferences.

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