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Pulmonary Sequestration Presenting with Recurrent Abdominal Pain

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Authors' contributions

This work was carried out in collaboration between all authors. Author GTO designed the study. Author GKU wrote the first draft of the manuscript. Author HSG managed the literature searches, analyses of the study performed the spectroscopy analysis. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Pulmonary sequestration is a rare congenital malformation of the pulmonary system. This non-functioning cystic or solid mass is asymptomatic unless infected. Patients primarily present with pulmonary symptoms such as cough, hemoptysis, dyspnea, chest pain, and fever due to recurrent pulmonary infections. We report herein the case of a young woman who presented with recurrent left upper abdominal pain and was diagnosed by contrast-enhanced computed tomography as intralobar pulmonary sequestration.

Keywords: Abdominal pain; pulmonary sequestration; Lung.

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1. INTRODUCTION

Pulmonary sequestration (PS) is a rare congenital malformation of the pulmonary system. This non-functioning cystic or solid mass has no communication with the tracheobronchial system and is supplied by a systemic arterial vessel rather than the pulmonary vasculature. Pulmonary sequestrations (PSs) are generally classified according to the anatomic location as intralobar or extralobar (also known as intrapulmonary and extrapulmonary sequestration, respectively). Intralobar sequestration is four times more common than the extralobar type [1,2].

PS is asymptomatic unless infected. Asymptomatic cases may be diagnosed incidentally on imaging modalities. There are several case reports on this topic with diagnosis during adulthood [3,4]. Patients primarily present with recurrent bronchitis, pneumonia, or hemoptysis. We report herein the case of a young woman who presented with recurrent left upper abdominal pain and was diagnosed as PS.

2. CASE REPORT

A 19-year-old female was admitted to the Department of Gastroenterology with complaints of recurrent epigastric and left upper quadrant pain, which radiated to the back. Her pain increased on deep inspiration. She had no significant past medical history. She had been admitted to several emergency departments with a 2- month-history of upper abdominal and chest pain. On the physical examination, the only remarkable signs were axillary temperature of 37.6°C, a few inspiratory rales at the left base, and epigastric tenderness. Laboratory findings revealed: white blood cell count: 18,100/mm³, neutrophil count: 87.7%, and C-reactive protein (CRP): 14.5 mg/L (0-5 mg/L). Her chest radiography, abdominal sonography and upper gastrointestinal endoscopy were normal. Her baseline cardiac examination and ECG are all within normal limits. The etiology of abdominal pain was not identified with these tests, and a computed tomography (CT) scan of the abdomen was performed. In the abdominal CT, abdominal organs were reported as normal, but atelectasis in the superior segment of the lower lobe of the left lung and minimal pleural effusion were detected. Thorax CT with intravenous contrast was performed to evaluate the exact pulmonary pathology, and revealed an intralobar PS in the medial aspect of the left lower lobe receiving an arterial branch from the aorta (Fig. 1). The diagnosis of PS was based on identifying this systemic arterial supply.

Ceftriaxone was administered intravenously at a dose of 2 g daily for the patient's elevated CRP level and leukocytosis (with a diagnosis of pneumonia). Her abdominal pain began to improve within three days of the antibiotic treatment. After the completion of medical treatment, the patient was referred to the Department of Thoracic Surgery for operation. The fiberoptic bronchoscope was applied to get a specific agent and to see other possible pathologies. The findings were nonspecific. The patient underwent a left posterolateral thoracotomy and left lower lobe resection. The findings at operation and the histopathological examination of operation specimens confirmed the diagnosis of intralobar PS.

3. DISCUSSION

PS comprises only 0.15-6.4% of all pulmonary malformations, making it an extremely rare disorder in adults [5]. In an adult patient, the major morbidity is recurrent pulmonary infections. PSs are generally classified according to the anatomic location as intralobar or extralobar (also known as intrapulmonary and extrapulmonary sequestration, respectively).

Intralobar sequestration is contained within the normal pulmonary tissue, usually in the posterior basal segment of the left lower lobe. It lacks separate visceral pleura, its arterial supply is mainly derived from the thoracic aorta, and venous drainage is through the pulmonary vein. Intralobar PS rarely causes problems during childhood and 15% of adult patients do not have any complaints when the sequestration is discovered. Intralobar PS, as in our case, is often identified during the second decade of life secondary to recurrent infections and almost always within the lower lobes, more commonly on the left and typically involving the posterior basal segment [6].

Extralobar PS is located outside the normal lung and wrapped by its own visceral pleura. Eighty percent of the arterial supply comes from the descending aorta, and the main venous drainage is via the systemic circulation. It is observed mostly in neonates and early childhood and is related with other congenital anomalies [7].

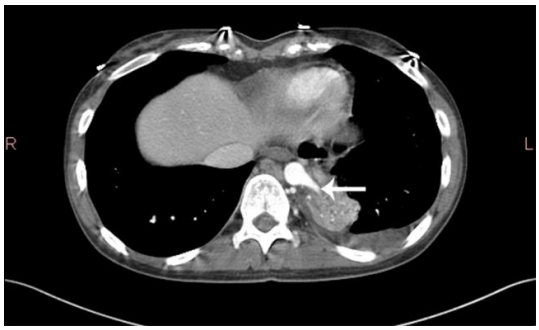

Fig. 1. Computed tomography scan of the thorax showing PS with an aberrant arterial supply originating from the descending aorta

The bronchopulmonary foregut malformation includes bronchopulmonary sequestration, congenital lobar emphysema, congenital pulmonary airway malformation, bronchogenic cysts, bronchial atresia, bronchial stenosis, and tracheal stenosis. These anomalies more frequently associated with extralobar sequestrations [7,8].

The embriyologic basis of the congenial lung malformations is unclear. Although extralobar sequestration is accepted as a congenital anomaly, the pathogenesis of intralobar sequestration is controversial, with evidence to support an acquired origin duo to chronic bronchial obstruction or pneumonia. Nevertheless, the congenital etiology of in PS has not been rejected yet.

Although numerous case reports of PS have been published in the literature, it is established that diagnosis is not easy due to the insignificant and variable clinical features. The most common complication of pulmonary sequestration is recurrent pulmonary infection, which is often the presenting sign of intralobar sequestration. The main symptoms and signs are cough, fever, chest pain, and hemoptysis, which mimic pneumonia, bronchiectasis, emphysema, and lung abscess [9,10]. PS may manifest in very different presentations, and the symptoms may vary, as illustrated in this case. The abdominal pain of our patient was a referred pain secondary to the diaphragmatic irritation from the basal pneumonia.

In recent years contrast-enhanced CT and three-dimensional reconstruction is the best diagnostic method. This noninvasive alternative diagnostic method is being used instead of traditional angiography. This imaging method confirms the presence of aberrant arterial supply and gives the definitive diagnosis. Because this was the patient's second episode of upper abdominal and chest pain, thorax CT with intravenous contrast was ordered. It revealed an intralobar PS in the medial aspect of the left lower lobe receiving an arterial branch from the aorta. The diagnosis of PS is based on identifying this systemic aberrant arterial supply [11,12].

The standard treatment is the resection of the segment or lobe that contains this non functioning sequestered tissue. The patient underwent a left posterolateral thoracotomy and left lower lobe resection. In the operation, four systemic arteries coming from aorta level and a venous drainage returning to the azygos vein at T12 were observed. Postoperative histopathological examinations excluded any other alternative diagnosis. The diagnosis of PS is approved by pathological evaluation of surgically removed specimen.

4. CONCLUSION

We aimed to demonstrate a case of PS, a rare entity, who was admitted with recurrent abdominal pain. We suggest that pulmonary pathologies should not be dismissed in the differential diagnosis of upper quadrant pain and PS in a patient with recurrent pneumonia.

CONSENT

Al authors declare that writen informed consent was obtained from the patient for publication of this case report and acompanying images.

ETHICAL APPROVAL

Ethical approval was not required.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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