Diagnostic peculiarities of bronchopulmonary sequestration: case and literature review

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ABSTRACT

Bronchopulmonary sequestration is an uncommon congenital anomaly in which part of the non-functioning pulmonary tissue is divided from the normal lung and has no normal connection with the tracheobronchial tree and pulmonary artery. This disease has an unknown etiology and is representing 0,1-6% of all structural lung diseases and developmental malformations. Diagnosis and management of congenital lung malformations has evolved over the past several decades. Earlier, more definitive diagnoses and, consequently, more timely intervention in utero or after birth have been enabled by advancement in imaging technology. The most commonly used imaging techniques to identify and characterize sequestration spectrum are conventional computer tomography and angiography.

Aim. To present a clinical case of bronchopulmonary sequestration diagnosed in Hospital of Lithuanian University of Health Sciences Kaunas Clinics and review the latest scientific literature on this topic.

Methods. A review of the literature using the PubMed database was performed. Publications, researching the problem of pulmonary malformations, were selected and a clinical case of bronchopulmonary sequestration was presented.

Conclusions. Pulmonary sequestration is uncommon disease, representing about 1 to 6% of all congenital lung anomalies and might be undetected during the prenatal period and early childhood years. If PS remains undiagnosed the risk of massive hemoptysis from pulmonary hypertension arises. Our purpose is to promote awareness of diagnosing this condition among various physicians. In this article we present a clinical case of bronchopulmonary sequestration with imaging characteristics.

Keywords: *pulmonary sequestration, lung malformations, anomalous systemic arterial supply.*

INTRODUCTION

Pulmonary sequestration (PS) is a congenital lung malformation, in which a segment or lobe of dysplastic lung tissue exists with no apparent communication with the tracheobronchial tree. It receives an anomalous systemic vascular supply, separate from the rest of the lung, most commonly from the descending thoracic aorta. In 1946, Pryce was the first to use the term "pulmonary sequestration" and classified this malformation into two types. Intralobar sequestration (ILS) is the more common type, which occurs within the visceral pleura of the functioning lung. In cases of extralobar sequestration (ELS), pleura separates the malformation from adjacent normal lung tissue (1–3). It is important to note that lungs with normal bronchial branching and circulation can also have an anomalous systemic arterial (ASA) supply, a phenomenon called pseudosequestration (4). This uncommon disease of still unknown definite etiology represents up to 6 percent of all structural lung diseases and developmental malformations. It usually remains undetected during the prenatal and early childhood periods raising the risk of massive hemoptysis from pulmonary hypertension (1,3,5,6). Our purpose is to raise awareness of diagnosing this condition among various physicians.

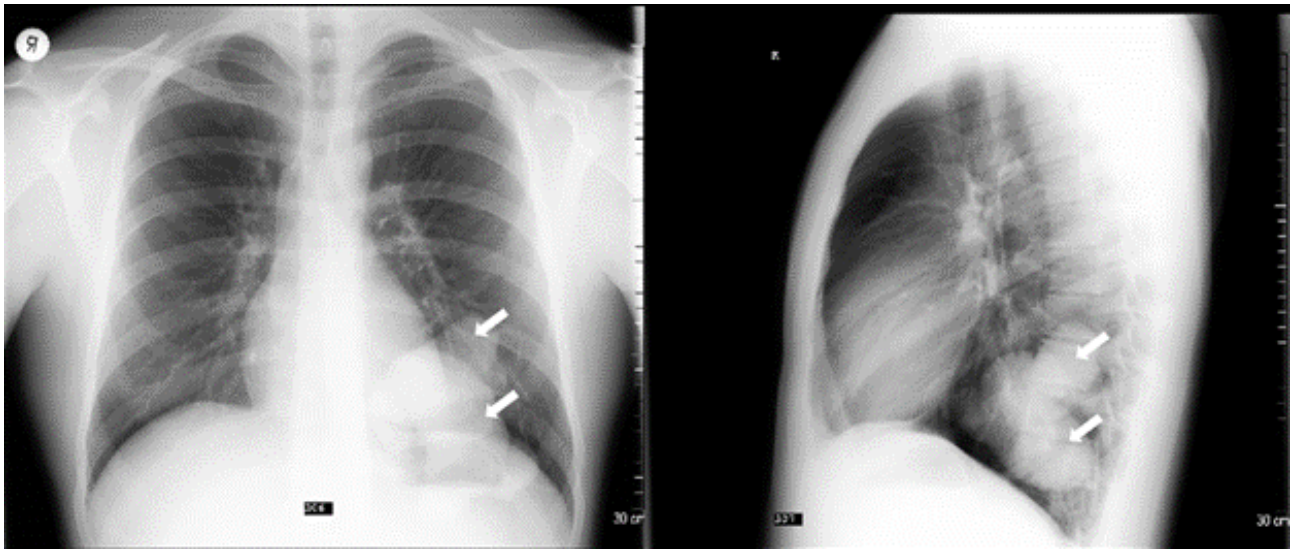
CASE REPORT

20-year-old male presented to our hospital due a persistent cough, which is exacerbated during physical activity. A year before, during a preventive health screening the patient was diagnosed with a benign cyst in the lower part of the middle lobe of the left lung. The current chest radiograph revealed an abnormal conglomeration in the basal parts of the same lung (Fig. 1).

Non-contrast chest computed tomography (CT) showed a polycyclic cystic lesion (Fig. 2). Additionally, contrast chest CT showed branches extending from the abdominal aorta although contrast enhancement was obscure. CT angiography confirmed that the affected lesion was supplied by the coeliac trunk (Fig. 3).

The patient was diagnosed with intralobar se-

Fig. 1 Chest X-ray showed an abnormal conglomeration in the left lung field (arrows).



questration as there was not enough data to confirm any arteriovenous malformation. Angiography was necessary to specify the diagnosis, but it was not performed in this case. However, physician council decided not to undergo any interventional treatment and the patient is being followed up on an outpatient basis.

DISCUSSION

In cases of PS or ASA, most patients are diagnosed coincidently (3,5–10) or present with a certain degree of hemoptysis (11–17). According to a Mayo clinic case series review, although the malformation is present from birth, the median age at diagnosis was 42 years. ILS is overall the most common form, comprising approximately 75 to 90 percent of sequestrations, while 10 to 25 percent are ELS. Males and females are equally affected with ILS, while ELS has a male predominance in most (18). Furthermore, if most probable causes of coughing, dyspnea, chest or back pain, clubbing of nails, recurrent or prolonged infections, and cardiac murmurs are excluded, PS or ASA should be considered in the differential diagnosis as well. Infections in the case of ELS are rare as the malformation is separated from the adjacent normal lung tissue (3,19). If the sequestration is of considerable size or occupies an anatomically significant location, ELS can manifest clinically in early infancy with respiratory distress, high output congestive heart failure due to right-to-left shunt, and incidental spontaneous pulmonary or pleural hemorrhage.

Fig. 2 Non-contrast chest CT showed a polycyclic cystic lesion (*). Contrast-enhanced chest CT showed branches arising from the abdominal aorta (arrow).

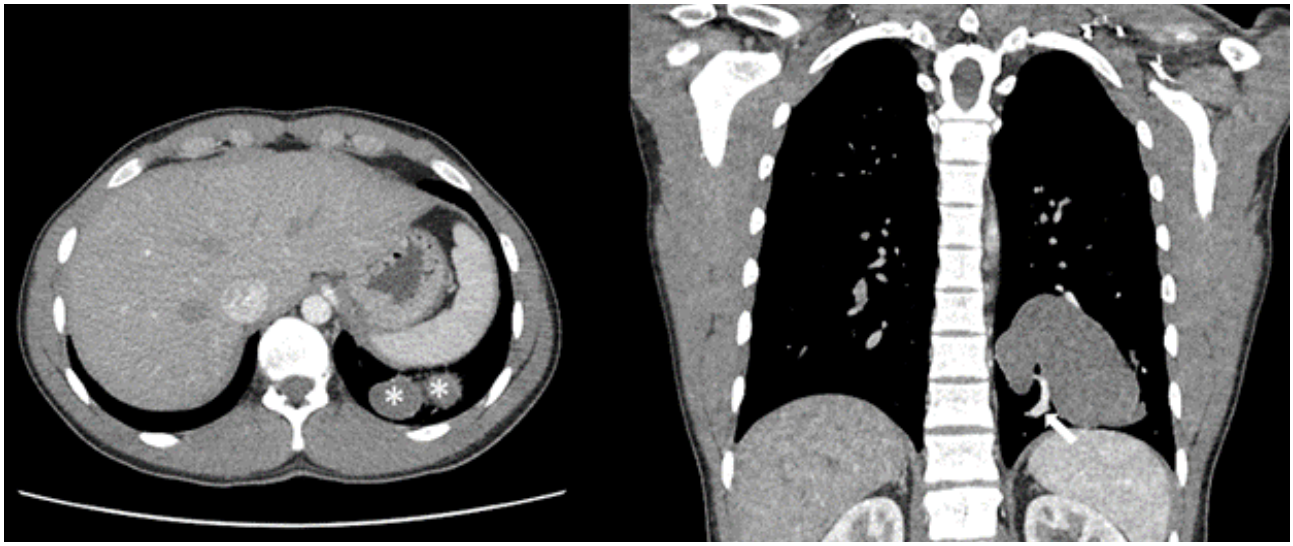
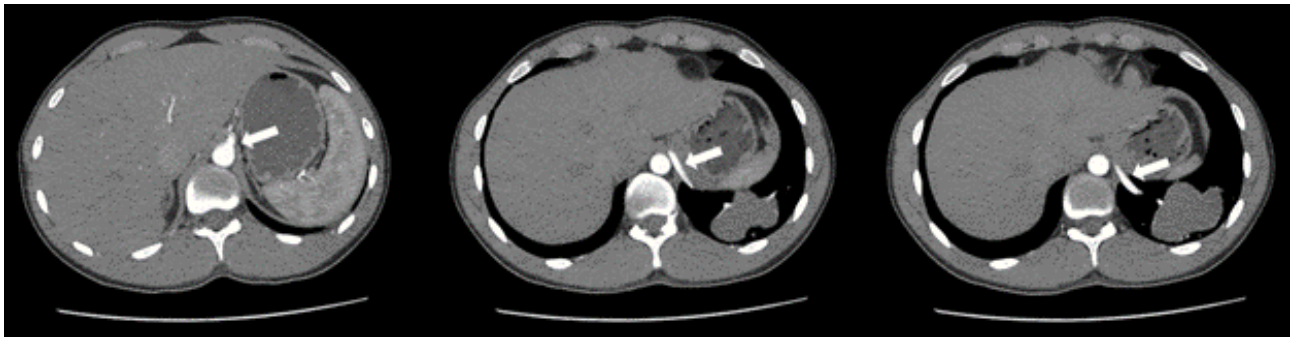


Fig. 3. CT angiography confirmed that the affected lesion was supplied by the coeliac trunk (arrows). Imaging highly suggests of intralobar sequestration.



Pseudosequestrations are generally asymptomatic. Undiagnosed cases can result in pulmonary hypertension and congestive heart failure in later years of life (4,12,20). Moreover, it is important to note that other underlying diseases can mask any of these pathologies (21).

In 1946, although being the most rare lung malformation, Pryce defined ASA supply as Pryce Type I sequestration. This malformation must be distinguished from the real bronchopulmonary sequestrations as it does not have a sequestrated lung component. Due to this fact physicians term it as pseudosequestration or anomalous systemic arterial supply to a normal lung (4,12,22).

In a chest X-ray, the area supplied by the anomalous artery is seen as an ill-defined non-specific opacity (11,14,16,21). Multidetector CT (MDCT) and CT angiography and 3-D reconstructions provide a possibility to objectively evaluate both the origin and the exact course of the anomalous artery (7,11–13,15,17). The most frequent ASA origin is the descending aorta, which in most cases course to the lower lung field and supplies it (7,10,12,14,16,17,21). Other localizations such as the abdominal aorta, inferior phrenic artery, coeliac, thyrocervical, and intercostobronchial trunks supply either the left upper field or the right lung (8–11,13,21).

Even though such new radiological technologies have emerged, conventional CT and angiography techniques remain the most frequently used methods in healthcare systems (8–10,14–17,21,23). Some authors suggest that tumor markers such as Ca 19.9 could be an effective tool in non-invasive differential diagnosis, although more in-depth research about such applications is needed (23).

After confirming the diagnosis, interventional treatment should be applied as soon as possible. Lobectomy or segmentectomy (8,9,11,12,21,23), ASA surgical ligation (8,21), and endovascular embolization (9,10,14,15) are the treatment options for the pathology. Although surgical resection via thoracotomy or video-assisted thoracic surgery (VATS) remains the most frequently applied method, embolization is a superior technique as it is both less invasive and provides satisfactory treatment results. Some researchers also note that vascular plug embolization provide better results than coil counterparts (9,15). Moreover, despite its' advantages against surgical lung tissue removal or vascular ligation, dense tortuosity and large diameter of the ASA are a few of the limiting factors of this procedure (11,16).

Procedure can be postponed only in those instances, in which the patient is asymptomatic and either too young, too old, or other patient comorbidities require more urgent physician attention (10,21,23). In rare cases, interventional treatment is optional, if ASA provides sufficient blood supply to the lung parenchyma and does not form any arteriovenous fistulas with the pulmonary veins [4]. Symptomatic treatment is also available although not recommend due to high risk to the patients' health of not undergoing interventional treatment (17).

CONCLUSION

This uncommon disease usually remains undiagnosed during the prenatal and early childhood periods raising the risk of significant complications. It is of utmost importance that physicians of different fields, especially radiology, obstetrics and gynecology, neonatology, pulmonology, and family medicine can recognize the disease to avoid negative patient treatment outcomes.

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