Case Report

Intralobar pulmonary sequestration: incidental finding in an asymptomatic patient

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ABSTRACT

We describe a case of a 58-year-old male who presented to the emergency room with symptoms related to an appendicitis. A computed tomography scan with contrast confirmed the diagnosis of acute appendicitis but also revealed a mass medially in the base of the inferior lobe of the right lung. The mass measured 6.7 cm $\times$ 3.7 cm transverse. It had multiple lobulations and the anterior aspect was of very low density, possibly representing accumulated mucoid material. The mass had an arterial connection from the descending thoracic aorta and a venous drainage into the right pulmonary vein, classical features of intralobar pulmonary sequestration. The physical exam was unremarkable, and the patient had no history of pulmonary symptoms. This case helps increase awareness of intralobar pulmonary sequestration, a rare condition that may be asymptomatic.

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Introduction

Pulmonary sequestration (PS) is a congenital malformation of the respiratory tract in which an ectopic, nonfunctioning mass of lung tissue develops without a connection to the tracheobronchial tree [1,2]. The ectopic mass derives its blood supply from the systemic circulation rather than the pulmonary circulation, commonly via an aberrant artery arising from the thoracic aorta [3,4]. PS is rare, accounting for only 0.15%-6.4% of congenital pulmonary malformations [3].

PS is classified as extralobar or intralobar. In extralobar PS, the abnormal lung tissue has its own pleural covering that completely separates it from the adjacent normal lung whereas in intralobar PS, the abnormal lung parenchyma is contained within the visceral pleura adjacent to the normal lung [2,5]. The relative frequency of extralobar PS and intralobar PS is reported to be approximately 25% and 75%, respectively [3].

Extralobar PS typically manifests early in infancy, with symptoms including respiratory distress, cyanosis, and infection. Intralobar PS, by contrast, usually presents later in child-
hood or in adulthood. Clinical symptoms of intralobar PS reflect pneumonia or other recurrent pulmonary infection, and patients typically present with recurrent cough, fever, hemoptysis, and chest pain [4–8]. Infrequently, patients with intralobar PS may be asymptomatic and the sequestration is identified incidentally. Here, we describe the case of a man who presents with acute appendicitis and was diagnosed with an intralobar PS found incidentally on computed topography (CT).

**Clinical case**

A 58-year-old male presented to the emergency room complaining of 5-hour history of generalized abdominal pain. The dull pain localized to the patient’s right lower quadrant with a severity of 9/10 without radiation. The patient experienced nausea but denied cough, shortness of breath, chest pain, or palpitations. Apart from a distant smoking history prior to 1990, the patient’s medical history was unremarkable. Physical exam revealed tenderness to palpation in the right lower quadrant without rebound or guarding. Lungs were clear to auscultation and percussion, and the remainder of the physical exam was unremarkable. Labs showed mild leukocytosis with increased granulocytes in addition to a random glucose level consistent with diabetes (131 mg/dL).

A CT scan with contrast was ordered and performed at the local hospital. It showed an appendix with a diameter of 11 mm and mild para-appendiceal inflammation, but without perforation or gangrene, suggestive of acute appendicitis. CT also revealed a mass in the medial aspect of the base of the right lung involving the right lower lobe (Fig. 1). The mass measured 6.7 cm AP × 3.7 cm transverse. It had multiple lobulations, and the anterior aspect was of very low density, possibly representing accumulated mucoid material. The mass had an arterial connection from the descending thoracic aorta (Figs. 2 and 3) and a venous drainage into the right pulmonary vein.

The patient was given Zosyn 3.375 g IV. His case was discussed with the on-call surgeon, and the he was admitted for treatment of his appendicitis. The patient opted out of surgical resection of the PS. To date, 18 months after diagnosis, he has not returned with pulmonary symptoms and at this point has been lost to follow-up.

**Discussion**

The patient presented in this report is a 58-year-old male who was diagnosed with an intralobar PS, found incidentally when he presented to the emergency department with acute appendicitis. Intralobar PS is a rare condition that usually presents with recurrent infection [6] and symptoms that include chronic or recurrent cough, fever, hemoptysis, chest pain, pneumonia, chills, and back pain [4,5,7–9]. Infrequently, patients with intralobar PS may be asymptomatic, in which case intralobar PS is typically diagnosed incidentally during chest CT. Case series reviews have reported the percentage of asymptomatic patients to be between 9.7% [7] and 15% [3].

Unlike extralobar PS, which is generally viewed as a developmental anomaly [5], the etiology of intralobar PS is less clear. There is evidence to support the hypothesis that intralobar PS is an acquired pathology following infection but there
is also evidence to support that it may have a congenital origin [5,10]. Sade et al [1] suggested a “sequestration spectrum” exits, which accounts for the origins of both intralobar PS, extralobar PS, and the variations observed in the vascular supply to the sequestered segment.

Diagnosis of intralobar PS is made through imaging, with the goal of identifying the aberrant artery to guide surgical resection [5]. Arterial supply most commonly originates from the descending thoracic aorta (73%) but it may arise from the abdominal aorta, celiac, or splenic arteries (21%), or less commonly from other sources [2,5]. In our patient, blood supply to the sequestration was from an anomalous artery from the descending thoracic aorta, and this helped confirm the diagnosis of intralobar PS. Venous drainage was into the right pulmonary vein, which is consistent with the pattern reported in 95% of cases [5]. Physiologically, this pattern of venous drainage creates an unusual “left-to-left” shunt as the venous blood enters the left atrium [5].

Screening chest radiographs taken before CT angiography (CTA) typically reveal a homogenous opacity, cystic spaces, or a prominent vascular shadow [5]. While angiography is the gold standard of diagnosing a PS, CTA is the modern imaging modality of choice because it is much less invasive and involves lower radiation doses [5,11]. Intralobar PS presents on CTA as a homogeneous or heterogeneous opacity, with possible areas of cavitation, cystic spaces, emphysematous changes, and calcifications. CTA also provides better spatial resolution over other imaging techniques for the visualization of the aberrant arterial supply and lung parenchyma [5]. In a retrospective study analyzing the CTA of patients who had been diagnosed with pathologically proven PS, Long et al [12] found CTA correctly diagnosed 100% of the aberrant supplying systemic arteries and draining veins. These authors also found that 70% of intralobar PS were located in the left lower lobe of the lung and 30% were located in the right lower lobe. In the case presented here, intralobar PS was diagnosed with CT with contrast and the sequestration was located in the right lower lobe. The low density of the sequestration in our patient could be due to an accumulation of mucoid material, which may be seen in cystic spaces of intralobar PS [5].

In cases of symptomatic intralobar PS, surgical resection is the recommended treatment [2,4,6]. Cases of asymptomatic intralobar sequestration have been more controversial, and practitioners have generally let the patient choose between close observation and preventative surgical resection of the PS [10]. However, a recent study by Li et al [13] provides evidence that preventative surgical resection may be the desired approach in many cases. The team retrospectively reviewed

Fig. 3 – (a and b) Serial IV contrast-enhanced CT images, axial projections, displaying aberrant systemic artery supplying the sequestered segment in the right lower lobe. (a) The aberrant artery (arrow) is seen arising from the descending thoracic aorta. (b) Adjacent slice, demonstrating the aberrant artery (arrow) supplying the sequestered segment. Small specks of calcification are visible within the aberrant artery. (c and d) Reference CT images at lung windows. (c) coronal projection, and (d) axial projection.
data on 37 patients diagnosed with asymptomatic intralobar PS, where 17 patients received video-assisted thoracic surgery (VATS) immediately after diagnosis while the other 20 patients chose to wait and undergo observation. The study revealed that 16 of the 20 patients who had chosen to undergo observation eventually returned with symptoms and underwent VATS, but these patients had worse values of blood loss, median duration of chest tube insertion, postoperative hospital stay, and lung function compared to the patients who received surgery immediately after diagnosis [13]. These data suggest that both symptomatic and asymptomatic patients with intralobar PS have improved outcomes if they undergo resection immediately after diagnosis. Our patient, who presented asymptomatic for intralobar PS and without any history of pulmonary symptoms, opted to not receive surgical or medical management. To date, 18 months after diagnosis, he has not returned with pulmonary symptoms.

Traditionally, a lobectomy performed during an open thoracotomy was gold standard surgical treatment of intralobar PS [2]. More recently, VATS has become the treatment of choice due to its less invasive nature [13,14]. In comparison with an open thoracotomy, VATS is associated with less intraoperative bleeding and shorter postoperative hospital stays [15]. Alternative management options include endovascular embolization and coiling in cases where open thoracotomy is likely to cause high levels of morbidity and mortality [16,17] or hybrid thoracic endovascular aortic repair-lobectomy treatment for cases of intralobar PS involving an aneurismal feeding vessel [18,19].

This case helps increase awareness of intralobar PS and its diagnosis by CT. As this case highlights, intralobar PS may be asymptomatic and the sequestration identified incidentally. Given the potential for intralobar PS to manifest clinically as recurrent pulmonary infections and other long-term morbidities, treatment options should be considered, even in the case of asymptomatic patients. Management options will vary, depending on the patient’s history of pulmonary symptoms, current health status, and consideration of the patient’s wishes. In the asymptomatic patient, these may range from preventative surgical resection, VATS, or coil embolization, to a more conservative approach of observation.

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