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### Case Report

# Case report: Asymptomatic bronchopulmonary sequestration in an adult with dual celiac and aortic supply<sup>\*</sup>

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#### ABSTRACT

A rare congenital malformation of the respiratory tract, bronchopulmonary sequestration generally presents in childhood and adolescence with recurrent pneumonia or in adulthood as an incidental finding on thoracic imaging. Manifesting as intrapulmonary or extrapulmonary types, bronchopulmonary sequestration characteristically receives blood supply from the systemic rather than pulmonary circulation. We present a 45-year-old male patient who received a provisional diagnosis of bronchopulmonary sequestration following an incidental finding on routine imaging. This case describes the way in which a provisional diagnosis may be made based upon imaging as well as underscoring the importance of alleviating the burden of additional imaging studies.

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#### Introduction

Bronchopulmonary sequestration (BPS) is a rare congenital malformation, representing 0.15%-6.4% of all congenital malformations of the respiratory tract [1]. Subtypes include intralobar sequestration (ILS) and extralobar sequestration (ELS), with the ILS subtype constituting the majority of cases. ILS most commonly presents in adolescence and adulthood with either recurrent pneumonia or incidental finding on imaging studies, while the ELS subtype presents in infancy with respiratory compromise [2]. Provisional diagnosis can be made based on imaging, with identification of feeding arteries arising from systemic vasculature being pathognomic. Most commonly arising from the thoracic or abdominal aorta, the vascular supply to bronchopulmonary sequestration may originate from smaller systemic arteries such as the celiac trunk [1].

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Fig. 1 - Posteroanterior (A) and lateral (B) views of the chest show a curvilinear opacity at the left lung base.

#### **Case report**

A 45-year-old male was seen for an annual wellness exam with a chief complaint of chronic fatigue for 2 months. The patient denied a history of shortness of breath, palpitations, chest pain, tobacco consumption, or alcohol consumption in excess of 3 beers per night. He had previously been measured to have elevated liver enzymes. He had been hospitalized twice in the past 5 years for bee sting-related anaphylaxis. There was no history of recurrent respiratory infections. The remainder of his history was unremarkable. Vitals were not collected. Physical exam was unremarkable. CBC, BMP, lipid panel, liver enzyme panel, TSH and B12 were collected. A COVID-19 PCR test was negative. A plain-film radiograph of the chest was ordered to assess potential causes of chronic fatigue.

Chest radiography revealed normal cardiac and mediastinal silhouettes, although a poorly defined lobular opacity along the posterior left diaphragm was evident with associated volume loss in the left lower lobe. All other lung fields were clear. Pneumothorax and pleural effusion were not present (Fig. 1).

Follow-up thoracic CT without contrast was ordered to examine for potential atelectasis, scarring, pneumonia, or mass. The CT demonstrated a rounded, well-defined consolidation in the posteromedial aspect of the left lower lobe base with multiple associated internal air-filled cysts and mild volume loss (Fig. 2). A small, rounded nodule in the posterolateral margin of the left costophrenic angle just lateral to the consolidated lung measured  $6 \times 7$  mm. Mild scarring in the lingula of the left upper lobe was also seen. The left lower lobe findings had a chronic appearance, which at the time were attributed to sequelae of prior pneumonia or cystic bronchiectasis. Although possessing an atypical appearance for malignancy, a 4-6-month follow-up thoracic CT with IV contrast of the cystic structure was ordered for provisional diagnosis and assessment for changes.

Thoracic CT with IV contrast of the cystic pulmonary consolidation revealed 2 feeding arteries originating from the descending thoracic aorta and celiac trunk (Figs. 3 and 4), consistent with BPS.

Subsequent testing revealed a Vitamin D measurement below the reference range. The patient's fatigue responded well to Vitamin D supplementation.

#### Discussion

Although the pathogenesis of BPS is not fully understood [3], it is thought to result from the development of an accessory lung bud caudal to the main pulmonary tree. This is suspected to occur prior to the separation of pulmonary and systemic arterial circulations at week 17 of gestation [4,5]. BPS is defined as a non-functional mass of lung tissue that is not connected to the pulmonary tree and receives blood supply from systemic circulation, often the lower thoracic or upper abdominal aorta [6]. Congenital pulmonary abnormalities, including cystic pulmonary airway malformations, bronchogenic cysts, BPS, and hybrid lesions, have been estimated to occur in 1 in 10,000-35,000 live births [7], with BPS representing 0.15%-6.4% of congenital pulmonary malformations [1]. An estimated 75%-90% of sequestrations are ILS, 10%-25% of cases are ELS, and approximately 10% are associated with an aberrant connection to the gastrointestinal tract (bronchopulmonary foregut malformation), demonstrating a failure of the accessory lung bud to dissociate from the foregut during embryogenesis. A foregut malformation is associated with a higher likelihood of recurrent infections within the lesion [5].

By definition, ILS lesions are located within normal lung lobes and lack an independent visceral pleura. While these lesions can develop anywhere in the thorax, about 60% are found in the basilar posterior segment of the left lower lobe [6,8]. Generally unilateral and singular, cases of bilateral ILS lesions have been reported [9]. ILS most often receives its sys-



Fig. 2 – Coronal (A) and axial (B) thoracic CT images in lung window demonstrate the mixed cystic and consolidated opacity in the posteromedial left lung base with associated volume loss.



Fig. 3 – Coronal IV contrast enhanced thoracic CT sections show the origination (A) and supply (B) of aortic and celiac feeding arteries to the left lower lobe bronchopulmonary sequestration.



Fig. 4 – Additional coronal IV contrast enhanced thoracic CT sections show the course (A) and source (B) of the celiac feeding artery.

temic blood supply from the lower thoracic or upper abdominal aorta [10], although these feeding arteries may branch from other locations, such as the celiac trunk, as in our patient. In contrast, ELS lesions are located outside of the normal pulmonary tissue and contain their own visceral pleura. Approximately, 80% of these lesions are present inferior to the left lower lung lobe. ELS lesions less commonly manifest with recurrent infections compared to ILS, due to absence of potential communication with bronchioles. ELS lesions generally receive blood supply from the thoracic aorta and are more often detected by prenatal ultrasound, as their extrapulmonary location makes them easier to visualize [11,12]. ILS lesions are more often not discovered until adolescence or adulthood, as they are hidden within normal lung parenchyma.

Clinical manifestations of BPS can vary widely based on size, location, and presence of aberrant connections to the gut or functioning pulmonary system. Most individuals are asymptomatic at birth. Larger lesions are more likely to present symptomatically in neonates with either respiratory distress due to mass effect on healthy lung tissue or highoutput cardiac failure due to diversion of systemic blood flow to the sequestration [3,8]. Older children and adults more commonly present with pulmonary infection, demonstrating a combination of cough, fever, shortness of breath, chest pain, or hemoptysis. Recurrent localized pulmonary infections should raise suspicion for BPS in the absence of another obvious cause. Alternatively, many patients remain asymptomatic for life with a prognosis comparative to the general population. The likelihood of remaining asymptomatic is unknown, and it is suspected that many cases go undiagnosed [13].

Evaluation for suspected BPS is done by medical imaging, beginning with a chest radiograph. Typical findings include a singular, uniformly dense mass-like opacity, most often found within or below the left lower lobe. There may be cystic areas resulting from recurrent infection or air fluid levels due to connection with the main bronchial tree [14,15]. Suspicion based on either clinical or radiographic findings should prompt advanced imaging with CT or MRI to visualize the parenchymal lesion as well as its feeding artery from the systemic circulation. CT demonstrates a heterogeneous or homogenous solid mass with or without cystic changes. Cavitary lesions with air fluid levels may also be seen, but less frequently. CT and MRI are both useful for visualizing the primary lesion as well as the systemic arterial supply [2], although angiographic techniques may be required to demonstrate the feeding vessel [15]. MRI/MRA may be preferable in the pediatric population [16]. Visualization of one or multiple feeding arteries branching from the systemic circulation, often a branch of the aorta, is essential for a provisional diagnosis of BPS.

Treatment for BPS primarily consists of surgical excision of the lesion, although there is some evidence to suggest feeding artery embolization as an alternative strategy [17]. Patients with respiratory distress, recurrent infections, or imaging demonstrating a lesion occupying >20% of the hemithorax are recommended to undergo surgical removal of the lesion [18]. For patients who are asymptomatic and have BPS discovered incidentally, as in the case of our patient, the decision between elective surgery and observation is debatable, with elective surgery being generally favored due to risk of malignancy [2,19]. One study makes the argument that elective surgery is associated with a lower risk of postoperative complications, such as pneumonia or pneumothorax, when compared to emergent surgery following the onset of severe symptoms (5% and 17%, respectively) [20], but many patients will likely remain asymptomatic.

While resection is favored for all sequestration lesions, the decision to intervene is a clinical one made in accordance with the patient's values and desires. In the appropriate setting, observation with serial imaging may be considered appropriate [19]. Long-term prognosis varies considerably based on the

size, location, and communication with functional lung tissue or the gut. In general, most patients have a very good prognosis following surgery, to include pediatric patients [21]. There is limited evidence regarding long-term outcomes for asymptomatic adults with incidentally discovered BPS, but future surgical excision following the potential onset of symptoms is still associated with a very good prognosis [22]. Knowledge of the presentation, imaging, and management of patients with BPS, including those discovered incidentally, can aid in providing accurate information and efficient use of resources.

#### Conclusion

This case demonstrates one of the most common means by which bronchopulmonary sequestration is identified: incidental finding. In asymptomatic patients, CT or MR imaging provides a provisional diagnosis of bronchopulmonary sequestration if feeding arteries from the systemic circulation are identified. Longitudinal imaging showing minimal change further supports the diagnosis. In symptomatic patients, such as those with recurrent pneumonia, surgical resection and pathological examination are used to make a definitive diagnosis. Knowledge of this condition and the criteria necessary for provisional diagnosis may help to eliminate unnecessary studies.

#### Patient consent

Written, informed consent for publication of this case study was obtained from the patient.

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