Case Report

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Intralobar bronchopulmonary sequestration in an adult: a case report

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Introduction: Bronchopulmonary sequestration (BPS) is typically a rare congenital disorder characterized by the presence of non-functioning lung tissue. There are two types of BPS: intralobar and extralobar sequestration, where extralobar sequestration can either be intrathoracic or sub-diaphragmatic.

Case presentation: In this case report, we present the case of a 70-year-old male with intralobar BPS who presented with recurrent chest infections, and a diagnosis of intralobar pulmonary sequestration was made based on a computed tomography (CT) scan.

Discussion: The diagnosis of intralobar pulmonary sequestration can be delayed as the intralobar type can present with varying imaging findings. A diagnosis can be made based on CT or MRI findings. A CT scan or MRI can show mass or consolidation with or without a cyst. Both CT and MRI can be reliable modalities to identify the arterial supply of the sequestered lung tissue, which is commonly a branch of the descending aorta.

Conclusion: Sequestration should be suspected when a posterobasal lung abnormality is supplied by an abnormal artery from the aorta or another systemic artery.

Keywords: bronchopulmonary sequestration, congenital disorder, recurrent pneumonia

Introduction

Bronchopulmonary sequestration (BPS), also called pulmonary sequestration, is a rare congenital disorder of the lower airway occurring in approximately 1 in 10 000–35 000 live births[1]. It is made up of a non-functioning mass of lung tissue that obtains its arterial blood supply from the systemic circulation and lacks normal contact with the tracheobronchial tree[2].

There are two types of BPS: intralobar and extralobar sequestration, where extralobar sequestration can either be intrathoracic or sub-diaphragmatic[2]. Among these, intralobar sequestration, which is the most common type, usually presents in late childhood or adolescence with features of recurrent chest infection[2]. Here, we present the case of a 70-year-old male with intralobar BPS. This case report has been reported in line with SCARE guidelines[3].

HIGHLIGHTS

- Bronchopulmonary sequestration (BPS) is a rare congenital disorder that typically presents with recurrent chest infections.
- Intralobar BPS can present with varying imaging findings, which can lead to a delay in diagnosis.
- The diagnosis of BPS can be made with a computed tomography (CT) or an MRI.
- A contrast-enhanced CT scan can help detect the aberrant blood supply to the congenital anomalous lung tissue.

Case presentation

A 70-year-old man presented to the emergency department of our hospital with shortness of breath and a cough that progressively increased over 2 months. He complained of two episodes of hemoptysis within 2 months. There was no chest pain, swelling of the legs, or fever. He was a known case of chronic obstructive pulmonary disease. He had received complete anti-tubercular treatment for sputum-positive pulmonary tuberculosis 45 years ago. He had recurrent episodes of pneumonia and lung abscesses. He had hernia surgery 5 years ago. He used to smoke a pack of cigarettes per day for 50 years and stopped smoking one and a half years ago. On examination, clubbing was present in bilateral hands. He had a pulse of 86 beats per minute and an oxygen saturation of 92% at 2 l/min with nasal prongs. On auscultation, he had decreased air entry on both sides, more on the right side, with wheeze present in the left inframammary area.

Laboratory tests, including a complete blood count, liver and renal function tests, and urinalysis, were all within the normal

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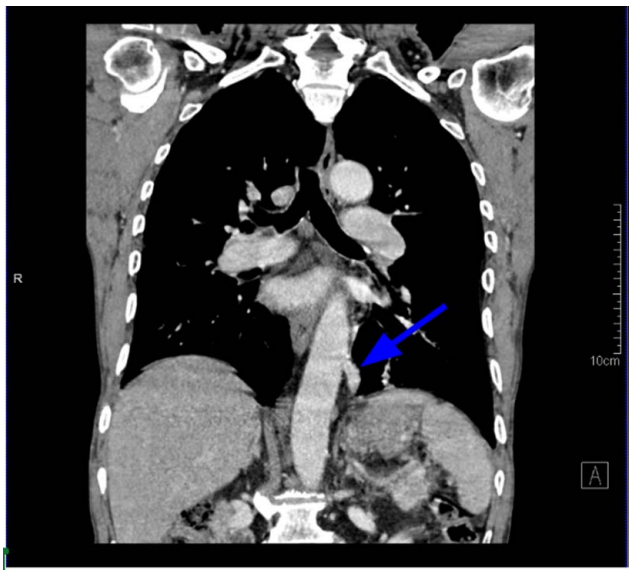

Figure 1. Coronal CT scan imaging in mediastinal algorithm showing aberrant artery arising from descending aorta and supplying the pulmonary sequestration area separately (blue arrow).

range. A chest radiograph showed a normal cardio-mediastinal contour with slightly hyperinflated bilateral lungs. Further investigation with computed tomography (CT) showed a multicystic lesion in the left lower lung lobe with a few intralesional cysts showing air-fluid level and supplied by an aberrant artery arising from the descending thoracic aorta-pulmonary sequestration (Figs 1, 2, and 3) (infected) with cystic degeneration.

He was treated with IV antibiotics. The patient was advised for surgery; however, the patient refused. The patient was referred to another center for further treatment.

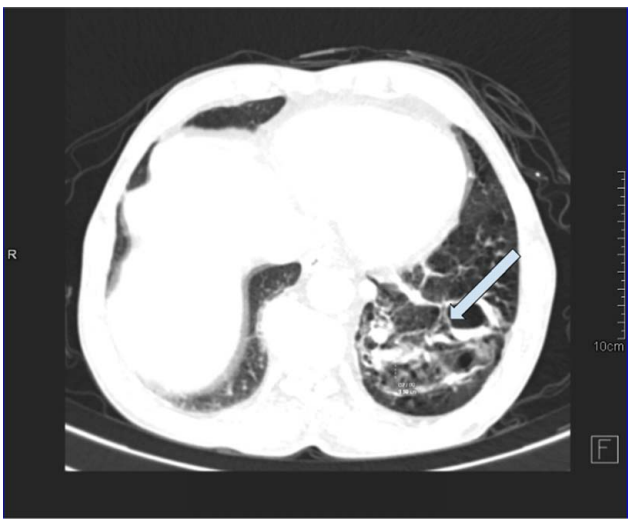

Figure 2. Axial CT scan imaging in the lung algorithm showing multicystic lesion in the left lower lung lobe with a few intralesional cysts showing an air-fluid level and supplied by an aberrant artery arising from the descending thoracic aorta-pulmonary sequestration (infected) with cystic degeneration. COPD (chronic obstructive pulmonary disease) changes with centrilobular emphysema and cardiomegaly were noted in bilateral lung fields.

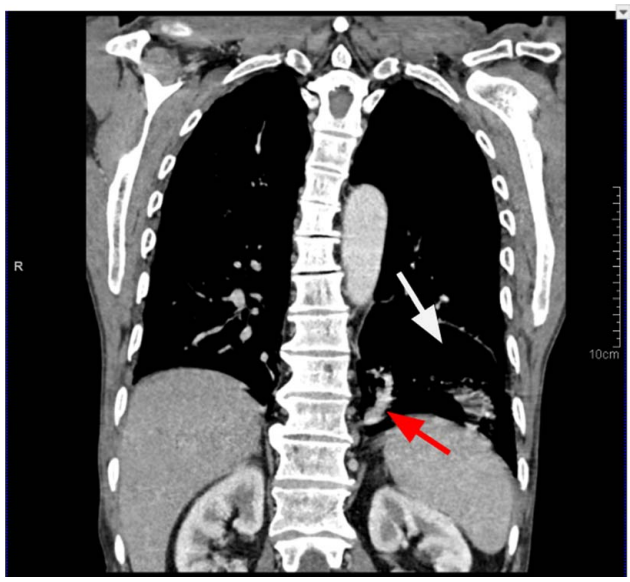

Figure 3. Chest coronal section contrast-enhanced computed tomography in mediastinal algorithm showing a section of the pulmonary sequestration as a cystic lesion (white arrow) with the systemic feeder aberrant artery from the aorta (red arrow).

Discussion

BPS is a bronchopulmonary foregut malformation that can be intralobar, extralobar, or hybrid. Intralobar type makes up 75% of BPS, which lies inside the lobe and does not have a separate visceral pleura. It is usually discovered in adolescence and early adulthood as a result of recurrent infections[2]. Patients can also present with chest pain or hemoptysis[4]. Intralobar sequestration is more common in the left lower lobe[2].

Most intralobar pulmonary sequestration occurs before the age of 20 and occurs rarely after 50 years of age. In a study by the Mayo Clinic, the median age of diagnosis of pulmonary sequestration among adults was 42 years[5]. In our case, intralobar pulmonary sequestration was diagnosed at 70 years of age. Our patient had recurrent episodes of lung abscess and two episodes of hemoptysis. The extralobar type accounts for about 25% of BPS, lies outside the normal lung, has its own visceral pleura, and presents early in life[2]. This type is usually associated with congenital anomalies like pulmonary hypoplasia, foregut duplication cysts, heart defects, or a diaphragmatic hernia or eventration. It can be diagnosed with prenatal or neonatal ultrasound, or magnetic resonance imaging (MRI)[6]. Most extralobar pulmonary sequestration also occurs in the left hemithorax.

BPS arises from an aberrant pulmonary lung bud and obtains its blood supply from foregut vessels[7]. If the accessory lung bud develops before the formation of the visceral pleura, sequestered lung tissue along with the normal pulmonary parenchyma is covered by the same visceral pleura, forming intralobar sequestration[7]. But if the accessory lung bud only forms after the formation of the visceral pleura, the sequestered lung tissue will then form its own pleura, forming extralobar pulmonary sequestration[7]. Blood supply to pulmonary sequestration is through an anomalous vessel from the systemic circulation, often from the descending thoracic aorta[4]. Many cases are incidentally diagnosed in CT chest scans. Hence, we should be aware of

the clinical features suggestive of intralobar sequestration, which include persistent cough, back pain, hemoptysis, and dyspnea, with the most common being recurrent pneumonia[8].

The diagnosis of intralobar pulmonary sequestration can be delayed as the intralobar type can present with varying imaging findings. The plain radiograph of BPS may show a solitary nodule or mass, a triangular opacity, consolidation, or a cystic or multicystic lesion if infected[4]. A CT scan or MRI can show mass or consolidation with or without a cyst, and both CT and MRI can be reliable modalities to identify the arterial supply of the sequestered lung tissue, which is commonly a branch of the descending aorta[4]. The supplying artery may also arise from the splenic artery, celiac artery, abdominal aorta, or even the coronary artery[4,9]. Sometimes, the supplying artery may not be small or thin and may not be detected[9]. The imaging finding of extralobar type differs from intralobar type in that it occurs medially to the lung and is well defined, usually airless, and homogenous[4].

The treatment for both intralobar and extralobar pulmonary sequestration is surgical excision[10]. Even when the patient is asymptomatic, intralobar type is treated with surgical excision due to the risk of future recurrent infections, abscesses, or hemoptysis[11]. For the asymptomatic extralobar type detected incidentally in adulthood, treatment has been debated, with some recommending radiological monitoring every 5–10 years[12]. Regardless of the type, when surgery has been planned, radiological evaluation of the anatomy of the blood supply of the lesion is recommended to avoid the transection of any unsuspected blood vessel[4].

Conclusion

In an adult, BPS might manifest as a consolidation, a mass, a cystic or multicystic lesion, or any combination of these. An abnormal artery from the aorta or another systemic artery supplying a posterobasal lung abnormality should raise suspicion for sequestration. It is best detected by CT or MRI. As opposed to intralobar sequestration, extralobar sequestration is medial to the lung, nearly invariably airless, clearly defined, and homogeneous.

Ethical approval

Ethical approval was not required for case reports as per our Institutional review committee of our center. Patient anonymity is maintained throughout this manuscript, and consent was obtained for publication from the patient.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of of the written consent is available for review by the Editor-in-Chief of this journal on request. We also ensured, none of the identifying characteristics are included in the case report.

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Author contribution

A.P.: manuscript preparation, editing, and review; A.P.: data collection, obtaining consent from the patient, and manuscript review; S.K.: manuscript preparation, editing, and review; A.D.: concept, manuscript preparation, editing, and review; S.A.: manuscript editing and review; A.C.: concept, manuscript editing, and review; A.C.: concept, manuscript editing, and review; P.P.: manuscript editing and review; S.B.: manuscript preparation, editing, and review.

Conflicts of interest disclosure

All authors declare that they have no conflicts of interest.

Research registration unique identifying number (UIN)

Since this is a case report, research registration has not been done.

Guarantor

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Data availability statement

Not applicable.

Provenance and peer review

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