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Case Report

Extralobar pulmonary sequestration in the left superior mediastinum with an atypical pulmonary artery supply: A case report

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Abstract

Extralobar pulmonary sequestration is a rare congenital anomaly with a nonfunctional lung mass with no connection to the bronchial tree and a systemic arterial supply. Most cases are located in the lower thorax and involvement of the left upper thorax is exceedingly rare. We present a case of an incidental left superior mediastinal mass in an asymptomatic patient undergoing a screening chest radiography. Further chest CT revealed a large mass in the left superior mediastinum, with arterial supply from the left pulmonary artery. Surgical excision was performed, and histopathology demonstrated ectopic pulmonary tissue with its own pleura without bronchial connection. The case highlights the diagnostic challenges and surgical considerations in rare anatomical variations of pulmonary sequestration at the left superior mediastinum.

Keywords: Congenital lung malformation, Extralobar pulmonary sequestration, Mediastinal mass, Pulmonary sequestration.

Introduction

Pulmonary sequestration is a rare congenital malformation, accounting for approximately 0.15–6.4% of all congenital lung anomalies [1]. It consists of non-functioning lung tissue that lacks communication with the tracheobronchial tree and receives arterial supply from the systemic circulation. Pulmonary sequestration is categorized into two types: intralobar pulmonary sequestration (ILS), which is contained within the lung and has a visceral pleura covering. The other type is extralobar pulmonary sequestration (ELS), which is surrounded by its own separate pleura [1].

According to a comprehensive review of 540 cases, 133 of them are ELS, by Savic et al. [2], and over 77% of ELS cases are located between the diaphragm and the lower lobes, with a left-side predominance (78.9%). Only two cases involved the left mediastinum, without specification of whether they were in the superior or inferior mediastinum. Thus, ELS in the superior mediastinum is exceptionally rare and may present significant diagnostic and therapeutic challenges.

About 80% of the aberrant arterial supply for ELS originates from the descending aorta, including supply from other systemic arteries such as the celiac, splenic, intercostal, or subclavian arteries [3]. Its venous drainage usually occurs into systemic veins, such as the right atrium, azygos vein, or portal vein, although it can also be variable [4]. Importantly, the presence of a pulmonary arterial supply to an ELS is exceedingly uncommon [5]. Such an atypical vascular pattern can complicate the diagnosis, since a pulmonary artery feeder is more typical of intrapulmonary lesions like congenital pulmonary airway malformation (CPAM) [6].

Although a few cases of mediastinal ELS have been reported, they usually have systemic blood supply [7]. Only two cases of superior mediastinal mass with pulmonary artery supply have been reported [8,9]. Of these, only one case, which had two arterial supplies, had feeding vessels that could be identified on CT images pre-operatively [9].

Here, we present a case of left superior mediastinal ELS with a single, exclusive arterial supply from the pulmonary artery that was identifiable on preoperative CT, adding a unique example to this very limited body of literature.

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Case summary

A 58-year-old Thai woman, lifelong non-smoker residing in an urban area of southern Thailand, was referred for further evaluation after a routine screening chest radiograph revealed a well-circumscribed mass in the left superior mediastinum. She was asymptomatic, with no history of cough, dyspnea, chest pain, fever, or constitutional symptoms. Her medical and surgical histories were unremarkable.

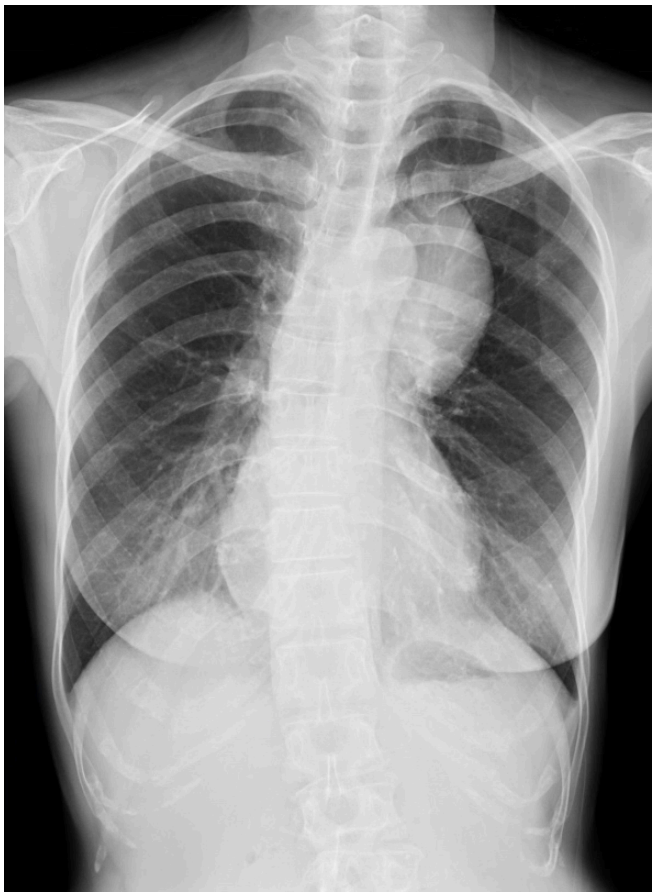

Figure 1. *Chest radiograph from the referring hospital shows a large well-defined hemispherical mass in the left upper hemithorax with preservation of the aortic knob and left hilum. No internal calcification or adjacent rib destruction is demonstrated.*

On physical examination, the patient was alert, afebrile, and hemodynamically stable. Cardiopulmonary examination was unremarkable, and there was no lymphadenopathy or peripheral edema. Contrast-enhanced chest CT demonstrated a 5.1 × 5.2 × 9.0-cm hyperdense, non-enhancing extrapulmonary mass in the superior, anterior and middle mediastinum, containing multiple internal punctate calcifications. The mass was noted to be encasing the adjacent left subclavian artery inferiorly. An arterial supply from the left main pulmonary artery was identified. No invasion of lung parenchyma or major vessels were seen. There was no mediastinal lymphadenopathy or pleural effusion.

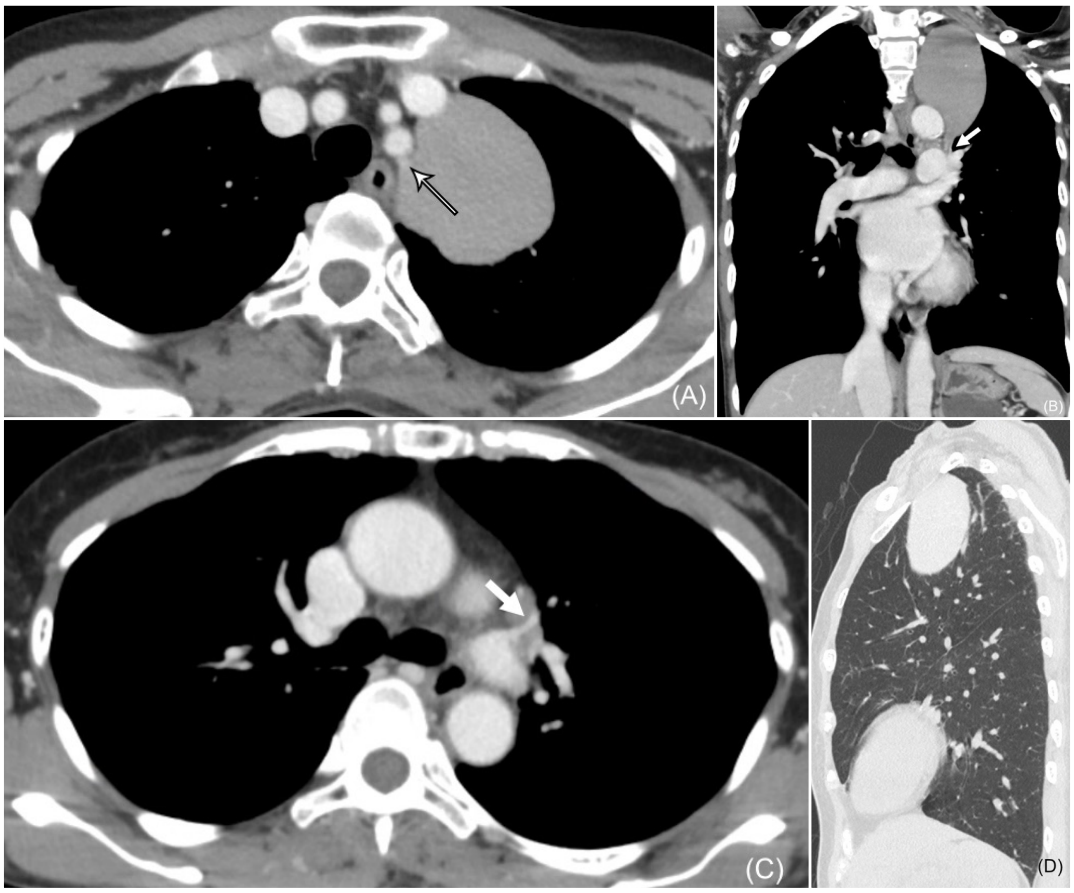

Figure 2. *Axial (A) and coronal (B) contrast-enhanced CT images demonstrate a large, non-enhancing extrapulmonary mediastinal mass containing punctate calcifications and partially abutting the proximal left subclavian artery without invasion. An additional axial image (C) shows arterial supply from the left main pulmonary artery. Lung-window sagittal imaging (D) depicts bronchial displacement without bronchial communication, confirming the extrapulmonary origin.*

The patient underwent surgical excision via an anterior mediastinal approach under general anesthesia. No gross thymic tissue was noted. A well-encapsulated cystic mass, approximately 4.0 × 8.5 cm, was identified in the left superior mediastinum. Its vascular pedicle originated from a branch of the left pulmonary artery. The feeding artery was ligated, and the mass was excised intact without rupture. Postoperative recovery was uneventful, and the patient was discharged on postoperative day 3.

Gross pathology revealed a well-circumscribed, gray-brown cystic lesion measuring 4.0 × 4.0 × 2.0 cm. Histologic sections revealed cystic spaces lined by benign bronchial-type epithelium with their own pleura. The surrounding stroma contained otherwise normal alveolar structures and pulmonary vessels without evidence of neoplasia. Foci of vascular congestion and a mild chronic inflammatory infiltrate were present in the adjacent connective tissue. No thymic tissue, granulomas, or malignant features was identified. The histological profile supports ELS with associated congestion and chronic inflammation.

At 17-month follow-up, chest CT showed no evidence of residual or recurrent disease.

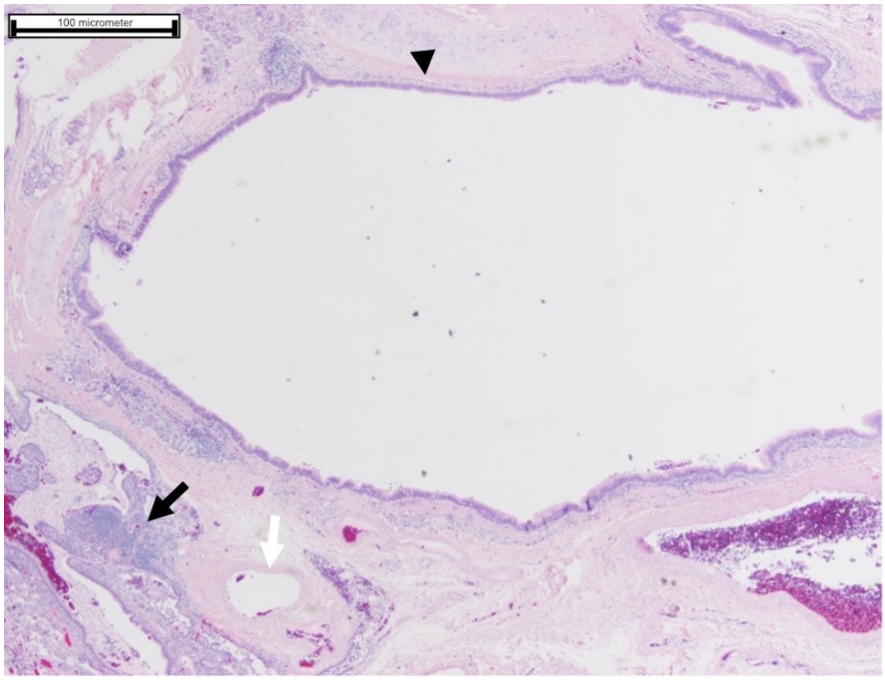

Figure 3. *H&E section (20x) shows a thick-walled vessel (white arrow) adjacent to a large cystic dilated bronchus (black arrowhead), surrounded by fibrosis involving the adjacent cyst and alveoli (black arrow).*

Discussion

Pulmonary sequestration is a rare congenital lung anomaly that arises from an abnormal development of the lung bud during embryogenesis, the extralobar form is less com mon than the intralobar type, and its location is characteristically in the lower thorax [12]. The exact etiology remains unclear, but it is believed to be from the formation of a supernumerary lung bud, which fails to integrate with the normal tracheobronchial tree [10].

A comprehensive review [2] found that the majority of ELS are located between the diaphragm and the lower lobes, with a marked predominance on the left side. A superior mediastinal location, as seen in our patient, is exceptionally rare, with only two cases in that review involving the left mediastinum without further specification. This atypical presentation creates a significant diagnostic challenge, as the lesion can mimic more common superior mediastinal masses, such as thymoma, lymphoma, or neurogenic tumors [13,14].

ELS can present with a wide range of symptoms, depending on the location, size, and associated anomalies. While some patients may be asymptomatic with the lesion being discovered incidentally, others may present with respiratory symptoms such as cough, dyspnea, or recurrent infections [15]. In rare cases, patients may present with acute symptoms such as chest pain, torsion of the sequestration [16] or even hemothorax [17]. The lesion may also be discovered during imaging studies for unrelated reasons, such as trauma or routine health screenings [18]. The diagnostic approach to ELS can be complicated due to its wide range of clinical presentations.

The most significant finding in our case, and a marked deviation from the classical definition of ELS, was the exclusive arterial supply from the left pulmonary artery. ELS is characteristically supplied by a systemic artery, with approximately 80% of vessels originating from the descending aorta or other systemic arteries [3]. An exclusive pulmonary arterial supply is exceedingly uncommon [5]. This atypical vascular pattern further complicates the diagnosis. While a few other cases of superior mediastinal masses with pulmonary artery feeders have been reported, our case remains distinct. For instance, the case reported by Osaki et al. [9] involved a complex supply from both the subclavian (systemic) and pulmonary arteries. Another report by Shadmehr et al. [8] described multiple pulmonary-artery branches but was preoperatively misdiagnosed as a thymoma, highlighting the diagnostic risk and potential for intraoperative complications. Our case is notable because it featured a single, exclusive arterial pedicle from the left main pulmonary artery that was clearly identified on preoperative CT images.

Another unusual feature of this case was the presence of multiple internal punctate calcifications on CT. Calcification in ELS is rare. In our patient, this radiologic finding correlates well with the histopathology, which demonstrated foci of a mild chronic inflammatory infiltrate and congestion. It is plausible that these calcifications were dystrophic in nature, resulting from chronic, subclinical inflammation within the sequestered tissue [19].

From an imaging standpoint, ELS should remain in the differential for well-circumscribed anterior mediastinal masses. Multiphase CTA with 3D reconstructions should document [10] an arterial inflow source, venous egress, and relationships to great vessels. These steps lower surgical hemorrhage risk and inform a limited, targeted approach. Our study identified a left main pulmonary artery pedicle preoperatively, consistent with these recommendations.

The definitive treatment is surgical resection. The goal of surgery is to completely remove the sequestrated tissue and any associated anomalies. The approach may vary depending on the location and size of the lesion, ranging from thoracoscopic resection to open thoracotomy [20]. Video-Assisted Thoracoscopic Surgery (VATS) is a minimally invasive approach that offers several advantages over open surgery, including reduced postoperative pain, shorter hospital stays, and improved cosmesis. However, it is particularly suitable for small and well-circumscribed lesions without significant associated anomalies [21,22]. In selected cases, embolization of the systemic feeding vessels may be performed to reduce blood flow to sequestration. However, this approach is typically used as an adjunct to surgery rather than a definitive treatment [10].

The prognosis for patients with ELS is generally excellent, especially when it is surgically resected [23]. Complete surgical resection is curative in most cases, and the risk of recurrence is low. However, patients with associated congenital anomalies or those who present with complications such as torsion or hemothorax may have a poorer prognosis [16].

This case highlights that ELS, despite its rarity, must be considered in the differential diagnosis of a superior mediastinal mass. The combination of an atypical location and a highly atypical vascular supply makes thorough preoperative planning essential. Our report underscores the critical role of contrast-enhanced CT with multiplanar reconstructions to precisely identify the complete vascular anatomy.

Conclusion

Extralobar pulmonary sequestration, though rare, should be considered in the differential diagnosis of superior mediastinal masses. This case demonstrates that the arterial supply can arise exclusively from the pulmonary artery, a critical variation that must be identified with preoperative imaging to ensure accurate diagnosis and safe surgical planning. With the increasing use of advanced imaging modalities, further studies are warranted to improve early diagnosis and guide surgical intervention.

Conflicts of interest: None declared.

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