

Para-aortic Extralobar Pulmonary Sequestration Mimicking a Posterior Mediastinal Tumor: A Case Report

Xiang Li

West China Hospital of Sichuan University

Shenglu Lian

West China Hospital of Sichuan University

Yixin Liu

West China Hospital of Sichuan University

Jianfeng Zhou

West China Hospital of Sichuan University

Longqi Chen

West China Hospital of Sichuan University

Yushang Yang

dyangysh@scu.edu.cn

West China Hospital of Sichuan University

Case Report

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Abstract

Background

Extralobar pulmonary sequestration is a rare congenital malformation characterized by nonfunctional lung tissue lacking communication with the tracheobronchial tree and receiving systemic arterial supply. Because of its unusual location and nonspecific imaging appearance, it may occasionally mimic a mediastinal tumor.

Case presentation

A 66-year-old woman was referred after an asymptomatic left lower thoracic mass was incidentally detected during routine screening. Contrast-enhanced computed tomography revealed a 3.3×2.5 cm paravertebral soft-tissue lesion located near the left cardiophrenic angle adjacent to the descending thoracic aorta. Three-dimensional computed tomographic angiography demonstrated an aberrant systemic feeding artery originating from the descending thoracic aorta and venous drainage into a branch of the azygos vein. Uniportal video-assisted thoracoscopic surgery was performed. Intraoperatively, the lesion was identified in the para-aortic region and was completely separated from the normal lung parenchyma. The aberrant feeding artery and draining vein were divided using an endoscopic stapling device, and the lesion was completely resected. Histopathological examination confirmed extralobar pulmonary sequestration. The postoperative course was uneventful, and the patient remained asymptomatic at 6-month follow-up.

Conclusions

Para-aortic extralobar pulmonary sequestration is rare and may mimic a posterior mediastinal tumor. Preoperative three-dimensional computed tomographic angiography is essential for identifying aberrant systemic vessels and facilitating safe thoracoscopic resection.

Background

Pulmonary sequestration is a rare congenital pulmonary malformation consisting of nonfunctional lung tissue without normal communication with the tracheobronchial tree and receiving systemic arterial blood supply[1, 2]. It is generally classified into intralobar and extralobar types according to pleural investment. Extralobar pulmonary sequestration (ELS) accounts for approximately 15–25% of all pulmonary sequestration cases and is usually identified in infancy or childhood[3, 4].

ELS most commonly occurs between the lower lobe and diaphragm and is typically supplied by branches arising from the thoracic or abdominal aorta. However, para-aortic ELS presenting as a posterior mediastinal mass in adults is uncommon and may lead to diagnostic confusion[5, 6]. We herein report a

rare case of para-aortic ELS successfully treated by uniportal video-assisted thoracoscopic surgery (VATS).

Case presentation

A 66-year-old woman was referred to our department after an abnormal shadow in the lower left thorax was incidentally identified during routine health screening. She denied cough, chest pain, hemoptysis, fever, or dyspnea. Physical examination and laboratory findings were unremarkable.

(A) Three-dimensional computed tomographic angiography demonstrating a para-aortic lesion (asterisk) supplied by an aberrant systemic artery arising from the descending thoracic aorta (yellow arrow) with venous drainage into the azygos venous system (blue arrow). (B) Thoracoscopic view showing the para-aortic sequestered lesion (asterisk) completely separated from the normal lung parenchyma. (C) Intraoperative identification and division of the anomalous draining vein (blue arrow). (D) Division of the aberrant systemic feeding artery (yellow arrow) during uniportal video-assisted thoracoscopic surgery. (E) Gross specimen of the resected sequestered lung tissue.

Contrast-enhanced chest computed tomography demonstrated a 3.3×2.5 cm well-circumscribed paravertebral soft-tissue lesion located in the left lower pleural cavity near the cardiophrenic angle adjacent to the descending thoracic aorta (Fig. 1A). Because the lesion was located in the posterior mediastinum, a neurogenic tumor was initially considered in the differential diagnosis.

Further evaluation with three-dimensional computed tomographic angiography revealed an aberrant systemic feeding artery arising directly from the descending thoracic aorta and entering the lesion, along with venous drainage into a branch of the azygos vein (Fig. 1A), findings highly suggestive of pulmonary sequestration.

The patient underwent uniportal VATS under general anesthesia in the right lateral decubitus position. A single incision was made in the fifth intercostal space along the midaxillary line. Intraoperatively, a well-circumscribed para-aortic lesion was identified near the left cardiophrenic angle and was completely separated from the normal lung parenchyma (Fig. 1B). Dense pleural adhesions surrounding the lesion were carefully dissected using electrocautery.

The aberrant systemic feeding artery arising from the descending thoracic aorta and the anomalous draining vein entering the azygos venous system were individually identified and divided using an endoscopic stapling device (Fig. 1C–D). The sequestered tissue was subsequently resected completely (Fig. 1E).

Gross examination revealed a $5.0 \times 4.1 \times 3.2$ cm sequestered lung specimen covered by a smooth pleural surface without defects. The cut surface was gray-red to gray-brown and contained multiple bronchial-like cystic structures measuring 0.2–0.6 cm in diameter (Fig. 1E).

Histopathological examination demonstrated dysplastic pulmonary tissue with cystically dilated spaces lined by ciliated columnar epithelium, focal alveolar epithelial hyperplasia, and proliferative fibrous stroma. Abnormally distributed muscular vessels were also identified within the lesion. These findings were consistent with extralobar pulmonary sequestration.

The postoperative course was uneventful. The chest tube was removed on postoperative day 2, and the patient was discharged shortly thereafter. At 6-month follow-up, she remained asymptomatic without evidence of recurrence.

Discussion

Pulmonary sequestration is an uncommon congenital anomaly characterized by dysplastic lung tissue supplied by anomalous systemic arteries. Compared with intralobar sequestration, ELS possesses a separate pleural covering and is less frequently encountered in clinical practice. ELS is often diagnosed during infancy because of associated congenital anomalies; therefore, incidental diagnosis in adulthood is relatively uncommon[1].

The present case is notable because of the unusual para-aortic location of the lesion, which closely mimicked a posterior mediastinal tumor. Posterior mediastinal masses are most commonly neurogenic tumors, and lesions located adjacent to the descending thoracic aorta are often initially interpreted as mediastinal neoplasms[7]. In this setting, identification of aberrant systemic arterial supply is critical for establishing the correct diagnosis.

Three-dimensional computed tomographic angiography played a pivotal role in the present case by clearly demonstrating the anomalous feeding artery originating from the descending thoracic aorta and the venous drainage into the azygos system. Preoperative recognition of these vascular structures is essential for surgical planning and may reduce the risk of catastrophic intraoperative hemorrhage during thoracoscopic dissection[8].

Surgical resection remains the definitive treatment for pulmonary sequestration. With advances in minimally invasive techniques, thoracoscopic resection has become increasingly feasible and safe, even in cases with aberrant systemic vessels[9, 10]. In the present case, uniportal VATS allowed successful identification and control of the anomalous vessels with favorable postoperative recovery.

Conclusions

Para-aortic extralobar pulmonary sequestration is a rare entity that may mimic a posterior mediastinal tumor. Three-dimensional computed tomographic angiography is essential for identifying aberrant systemic vessels and enabling safe thoracoscopic resection. Awareness of this uncommon presentation may help avoid diagnostic pitfalls and reduce the risk of intraoperative vascular injury.

Declarations

Ethics approval and consent to participate:

Not applicable.

Consent for publication:

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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Author Contribution

XL and SLL contributed equally to this work. XL collected the clinical data and drafted the manuscript. SLL participated in the literature review and manuscript preparation. YXL contributed to imaging analysis and interpretation. JFZ participated in the surgical management and perioperative data collection. LQC and YSY conceived and supervised the study, revised the manuscript critically for important intellectual content, and served as corresponding authors. All authors read and approved the final manuscript.

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Figures

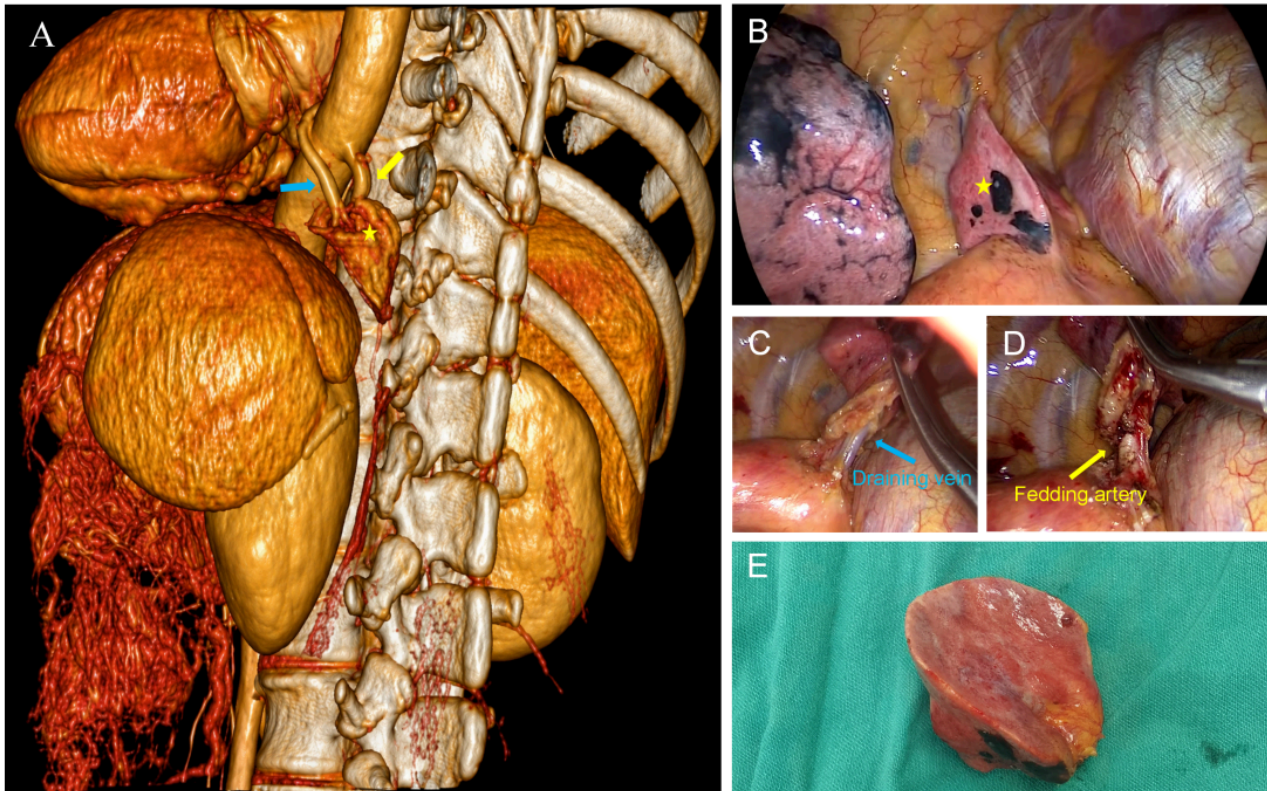


Figure 1

Imaging, intraoperative, and gross pathological findings of para-aortic extralobar pulmonary sequestration.

(A) Three-dimensional computed tomographic angiography demonstrating a para-aortic lesion (asterisk) supplied by an aberrant systemic artery arising from the descending thoracic aorta (yellow arrow) with venous drainage into the azygos venous system (blue arrow). (B) Thoracoscopic view showing the para-aortic sequestered lesion (asterisk) completely separated from the normal lung parenchyma. (C) Intraoperative identification and division of the anomalous draining vein (blue arrow). (D) Division of the aberrant systemic feeding artery (yellow arrow) during uniportal video-assisted thoracoscopic surgery. (E) Gross specimen of the resected sequestered lung tissue.