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Case Report

Abundant hemoptysis revealing an aneurysm of an aberrant systemic artery to the left lower pulmonary lobe: A case report ☆

Karim Haddar[a,b,*], *Leila Haddar*[a,b], *Hajar Siouri*[a,b], *Nadia El Mahi*[a,b], *Mohamed Lemine Abdesselam*[a,b], *Siham Nasri*[a,b], *Imane Kamaoui*[a,b], *Imane Skiker*[a,b]

[a] *Faculty of Medicine and Pharmacy, Mohammed I University, Oujda, Morocco*
[b] *Department of Radiology, Mohammed VI University Hospital Mohammed I University Oujda Morocco*

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ABSTRACT

Pulmonary sequestration (PS) is a congenital anomaly characterized by a lung region that is isolated from its normal bronchial and vascular connections. It typically receives blood supply from an aberrant systemic artery. An aneurysm of that aberrant artery is extremely rare. We report the case of a 55-year-old female patient who was incidentally diagnosed with intralobar PS after experiencing severe hemoptysis. The diagnosis was set by a chest contrast-enhanced computed tomography (CT), which also revealed an aneurysm of the aberrant systemic artery. A left lower lobectomy was successfully performed which confirmed the diagnosis. PS is characterized by a mass of lung tissue that lacks connection to the normal bronchial tree and is supplied with blood from the systemic circulation. There are 2 primary types: intralobar (ILS), located within a lung lobe, and extralobar (ELS), situated outside the lobes with abnormal vascular connections typically originating from the thoracic or abdominal aorta. In ILS, drainage usually occurs through pulmonary veins, whereas ELS often drains via the azygos or portal venous system. CT and magnetic resonance imaging (MRI) are preferred for diagnosing PS. They can also reveal complications such as an aneurysm of the aberrant artery, as in our case. PS is a rare yet significant anomaly to consider in cases of recurrent pneumonia or persistent consolidative opacities, especially when involving the lower lobe of the left lung. CT and MRI are highly valuable for pinpointing the lesion and identifying the feeder blood vessel associated with it.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

* Corresponding author.
E-mail address: karimhaddar@yahoo.fr (K. Haddar).

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Introduction

Pulmonary sequestration (PS) is an uncommon congenital anomaly where an abnormal lung lacks connections to the bronchial tree or pulmonary arteries. It is classified into intralobar (ILS) (75%) and extralobar (ELS) (25%) types:

- ILS shares the same visceral pleura as the normal lung and drains through the pulmonary veins.
- ELS is enveloped by its own separate pleura distinct from the normal lung and drains through systemic veins.

Case presentation

Our 55-year-old female patient, a nonsmoker with no significant medical history, arrived at the emergency department with a significant episode of hemoptysis along with isolated dyspnea. She did not report fever or chest pain. Upon clinical examination, no abnormalities were noted.

A contrast enhanced CT scan showed a mass in the left lower lobe characterized by soft tissue density, well-defined borders, and peripheral calcification. The mass lacks visceral pleura around it. During the arterial phase, there is no enhancement observed. The mass is supplied by a single arterial branch originating from the descending aorta with a partially thrombosed aneurysm, and venous drainage occurs via the left pulmonary veins (Figs. 1-4).

During a posterolateral thoracotomy at the fifth intercostal space on the right side, surgeons identified a vascularized intrapleural structure on the left side (Fig. 5). This structure was fed by a pedicle originating from the descending thoracic aorta and had venous drainage into the inferior pulmonary vein of the lower lobe. Subsequently, a left lobectomy was performed, with no complications observed during or after the procedure.

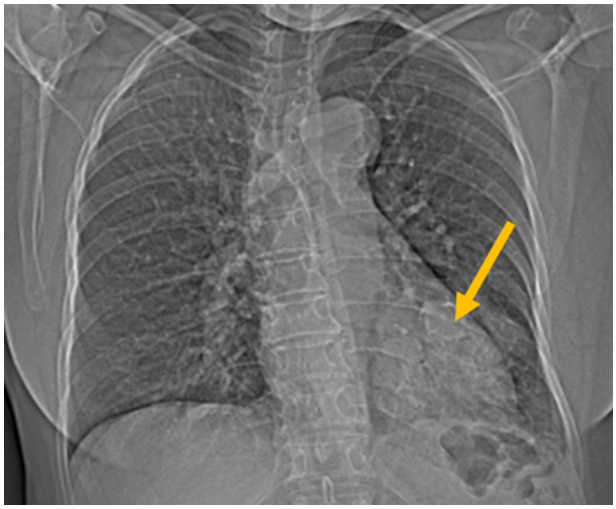

Fig. 1 – CT scout view showing a basal opacity located in the left lower lobe. The opacity appeared heterogeneous and poorly demarcated.

Discussion

Pulmonary sequestration (PS) is a rare condition where part of the lung tissue becomes nonfunctional and receives abnormal blood supply from the general circulatory system [1]. It predominantly occurs in the left lower lobe in 95% of cases [2] and is categorized into 2 types based on its pleural covering: intralobar or extralobar. The exact causes and characteristics of various pulmonary anomalies, including PS, remain unclear, leading to variability in prevalence, classification, diagnostic methods, and surgical indications.

Diagnosis of PS relies heavily on identifying systemic arterial blood supply to the affected lung tissue. Intralobar sequestration (ILS), more prevalent at 75%, shares the same visceral pleura as the normal lung and drains venously into the pulmonary veins. In contrast, extralobar sequestration (ELS) (25% of cases) possesses its own distinct visceral pleura separate from the normal lung, drains venously into a systemic vein, and functions akin to an accessory lobe, also referred to as “Rokitanski’s lobe”. Roughly 10-15% of ELS are located within or below the diaphragm [3]. ELS typically manifests early in childhood as a congenital malformation associated with foregut development, often accompanied by other anomalies. In contrast, ILS usually presents later in childhood or young adulthood and is localized within the normal lung structure [4].

Imaging studies are crucial for diagnosing PS, focusing on identifying abnormal vascular anatomy which aids in precise diagnosis and surgical planning to minimize vascular complications. Chest radiography plays a pivotal role in detecting ILS, particularly noting lesions in the posterior basal segment of the left lower lobe in many cases, often showing signs of chronic inflammation such as cavitations or cystic spaces. Diagnostic evaluations for PS include assessing vascular supply, detecting abnormal vessels, and searching for associated congenital anomalies and signs of infection. According to various sources, ILS primarily receives arterial blood supply from the thoracic aorta in about 75% of cases, while 15%-20% receive it from the abdominal aorta. Occasionally, aberrant arteries from smaller branches of these vessels can also be observed. Venous drainage for ILS usually returns to the left atrium via the pulmonary veins [6–8]. ILS cases often correlate with various infections. In ELS, the sequestered lung tissue is typically vascularized by an abnormal artery arising directly from the aorta in approximately 80% of cases. Venous drainage is usually directed to the right atrium through the azygos venous system or superior vena cava [7,9,10]. It’s important to consider the potential presence of accompanying congenital anomalies, such as those affecting the diaphragm, as well as signs of infection during the diagnostic assessment of patients with ELS. Aneurysmal dilation of the aberrant artery is extremely rare with only few cases having been reported in the literature.

Management strategies vary depending on the type and location of sequestration, guided by detailed preoperative imaging and clinical evaluation. Surgical resection involving isolation and division of abnormal systemic feeding arteries is considered the primary treatment for

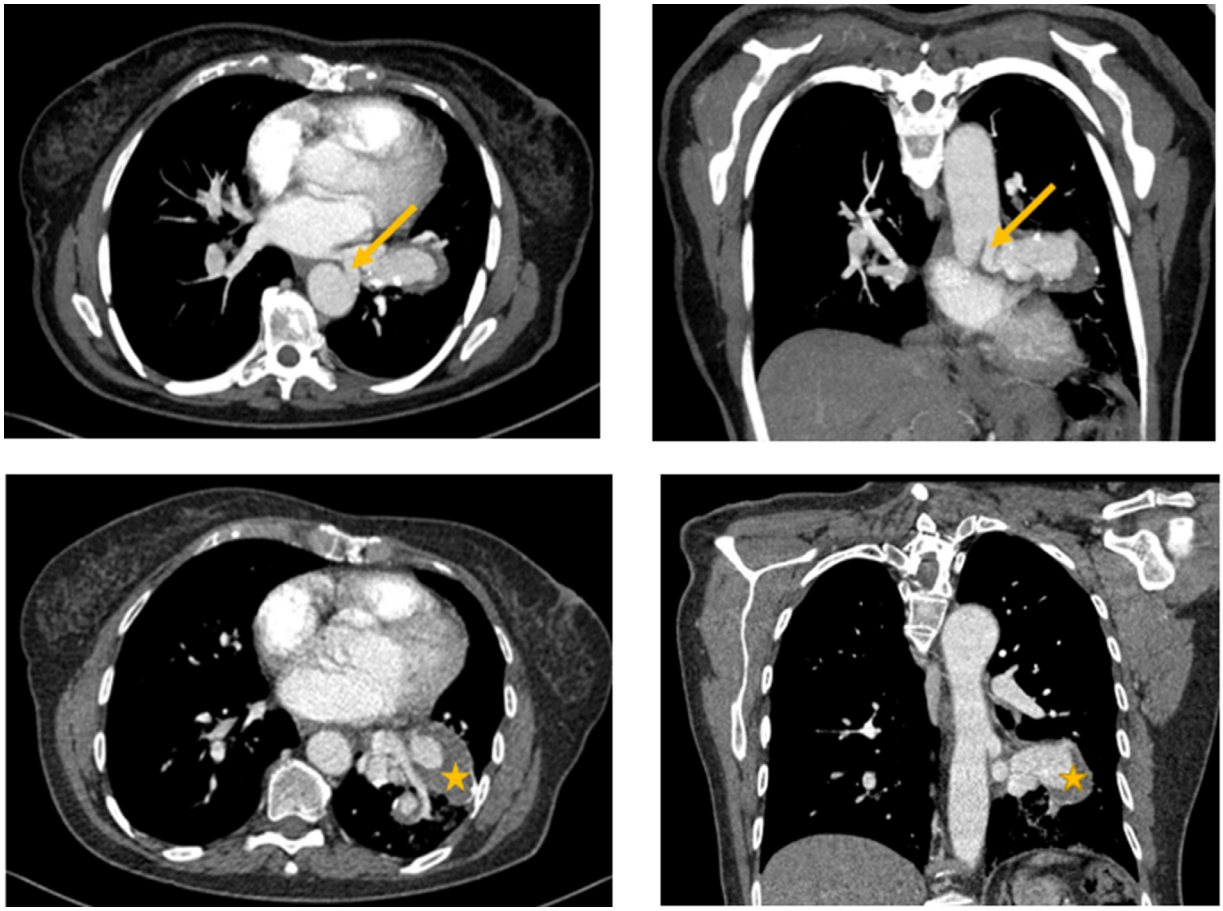

Fig. 2 – Axial and coronal contrast-enhanced computed tomography (mediastinal window) shows a well-defined left lower lobe heterogeneous mass supplied by a systemic artery (arrow) that arises from the distal descending thoracic aorta with a partially thrombosed aneurysm (star).

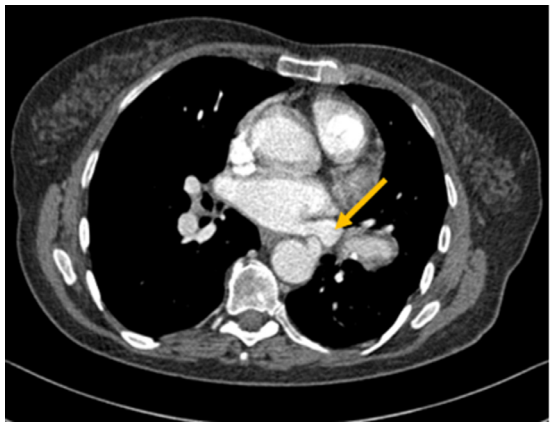

Fig. 3 – Axial contrast-enhanced computed tomography (mediastinal window) shows the venous return to the left atrium in relation to the pulmonary veins.

pulmonary sequestration. This approach is recommended due to the high risk of recurrent infections, potential need for extensive resection in cases of chronic infectious states, risk of bleeding, and potential for malignant tumor development. Even in asymptomatic patients, surgical intervention is advised to prevent complications that may arise from potential infections [4,5,11–13].Typically, a lobectomy is the preferred surgical procedure, although for smaller lesions located away from the pulmonary hilum and surrounded by substantial normal lung tissue, wedge-shaped resection via thoracotomy or video-assisted thoracoscopy may be preferred [14]. Endovascular occlusion of the arterial blood supply can also be employed to reduce blood flow to the sequestered tissue, promoting necrosis, fibrosis, and gradual resolution. This approach can be beneficial as a preoperative measure to minimize intraoperative bleeding risk and, in some cases, may serve as an alternative to surgery, particularly for small PS lesions (less than 3 cm) or in cases complicated by acute hemoptysis [5,15,16]. In instances where a large and tortuous aberrant feeding artery is present, exclusion of the arterial supply using a thoracic endograft has been reported as an effective method. This technique involves covering the orifice of the feeding artery to halt blood flow, which can be simpler and more effective than direct embolization [16].

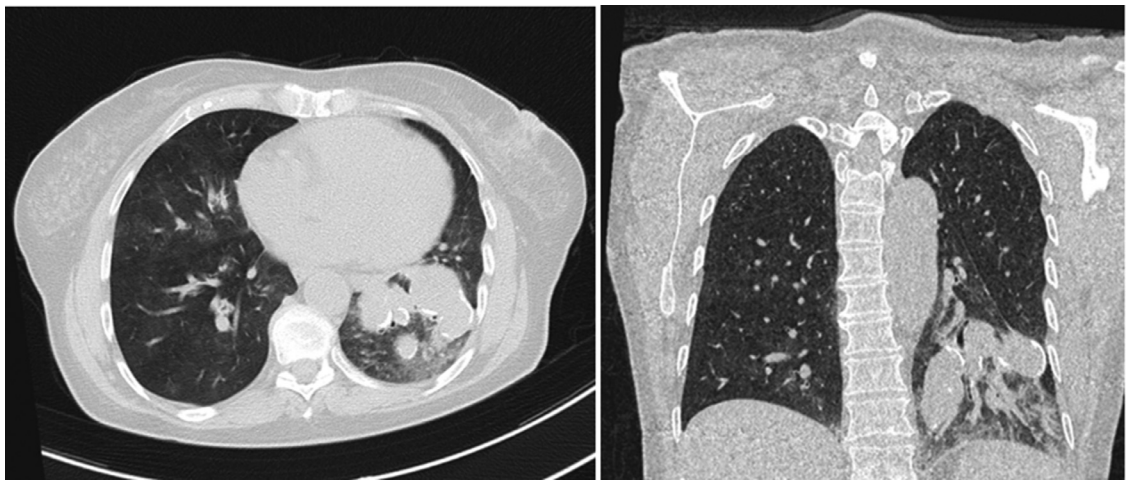

Fig. 4 – Axial and coronal computed tomography (lung window) shows a left lower lobe mass with peripheral calcifications, associated with adjacent ground-glass opacities, suggesting alveolar hemorrhage.

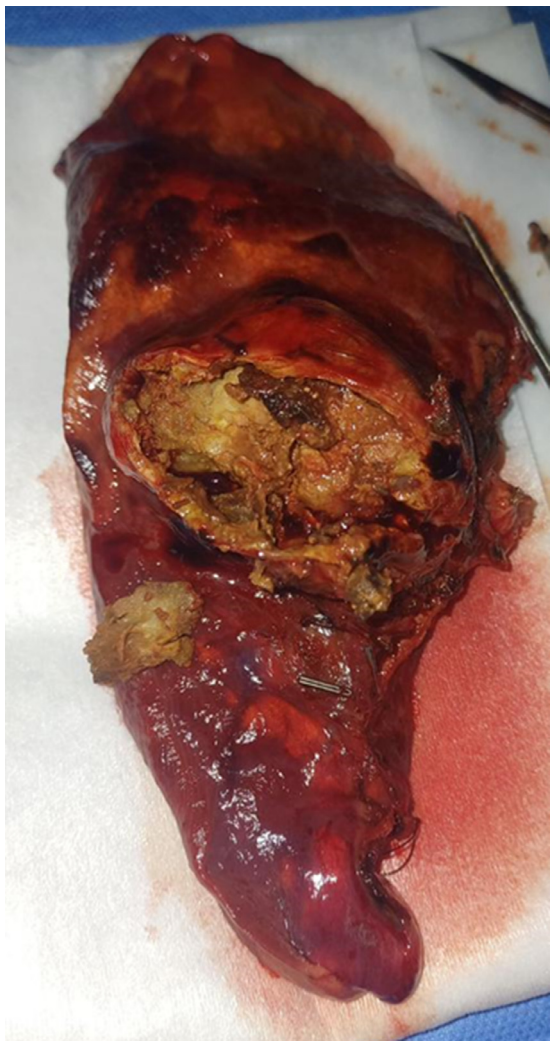

Fig. 5 – Peroperative picture of the inferior left lobe during lobectomy.

Conclusion

Pulmonary sequestration poses a diagnostic dilemma. When recurrent pneumonia, episodes of hemoptysis, and a lower lobe lung mass coexist, suspicion of this uncommon condition should be heightened. Diagnosis confirmation typically involves visualizing an abnormal systemic artery using angio-CT or angio-MRI. The preferred treatment is surgical resection, typically involving the ligation of the feeding vessel and occasionally requiring lobectomy.

Patient consent

Informed consent was obtained from the patient.

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