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Intralobar Pulmonary Sequestration: A Case Report

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Abstract **Case Report**

Pulmonary sequestration is a rare malformation. It is characterised by a territory of non-functional bronchopulmonary tissue vascularised by one or more aberrant systemic arteries. We report the case of a 51-year-old man admitted to hospital with recurrent haemoptysis, a thoracic angioscan revealed left lower lobar sequestration by two systemic vessels. The patient was treated with a left lower lobectomy. Pulmonary sequestration remains a diagnostic and therapeutic challenge.

Keywords: Pulmonary sequestration, arteries, haemoptysis, bronchopulmonary tissue.

INTRODUCTION

Many theories and speculations have been put forth as to the etiology of this condition, but it is primarily accepted to be a rare congenital malformation of the lower respiratory tract occurring between the 5th and 7th weeks of gestation, in a critical period of lung development that leads to a variety of developmental malformations [1]. In other words, pulmonary sequestration is an embryological malformation characteristically involving broncho-pulmonary foregut connection because the sequestrated lung segment develops with its blood supply arising from the aorta or the intercostal vessels instead of the pulmonary artery [2].

A pulmonary sequestration (PS) is a rare congenital condition with aberrant proliferation of the primitive foregut. It is manifested either in its complex form - continuous with the rest of the lung through the trachea, creating a broncho-pulmonary foregut connection, or in its simple or isolated form, a mass of non-functioning pulmonary tissue that has lost its connection with the tracheo-bronchial tree [3]. Extrapulmonary sequestration (EPS) is the term applied when this non-functioning lung tissue is located outside the thorax, most commonly in the vicinity of the diaphragm, in other words, with or without abnormal vascular connections [1].

CASE REPORT

We report the case of a 51-year-old patient who smoked 25 packs per year. He had a history of recurrent bronchopulmonary infections. He was hospitalised for moderate haemoptysis associated with left basal chest pain. Clinical examination was unremarkable.

The chest X-ray showed a basal opacity in the left lower lobe (Fig 1). A blood investigation was performed, including d-dimer, Genexpert on sputum, and aspergillosis serology, all of which were negative. Biological investigations showed hyperleukocytosis with PNN at 17,000/mm3 and CRP at 130 mg/l. Bronchial endoscopy revealed inflammation of the right lower lobar bronchus.

A chest computed tomographic angiography revealed a mass measuring 30 x 22 x 42 mm in the left lower lobe. This formation was systematised and well limited, and was vascularised by two systemic arteries from the descending thoracic aorta (Fig 2).

The diagnosis of intralobar pulmonary sequestration by two arteries was accepted. A probabilistic antibiotic treatment was started, and radical treatment by inferior lobectomy was proposed. The patient was operated on using video-assisted thoracic surgery (VATS), which was intraoperatively converted to a left posterolateral thoracotomy. The post-operative care is uneventful.

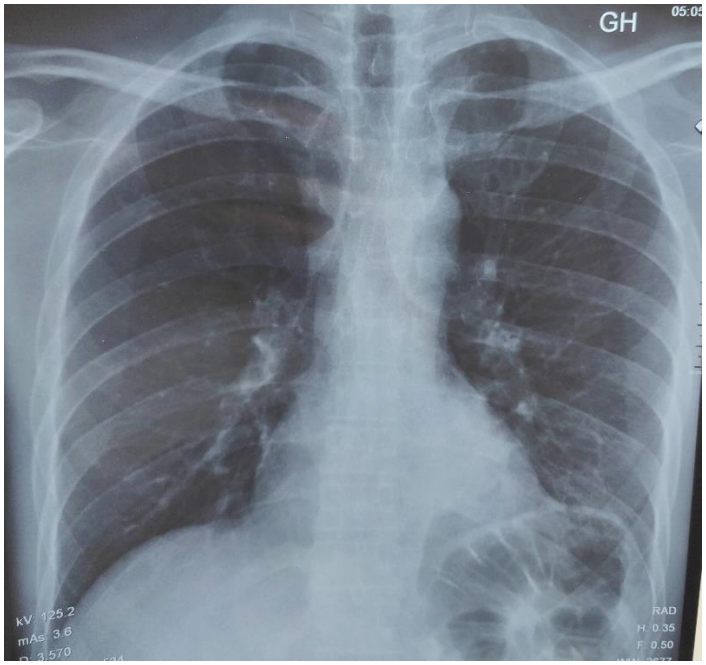

Figure 1: Front chest X-ray showing a left basal pulmonary opacity

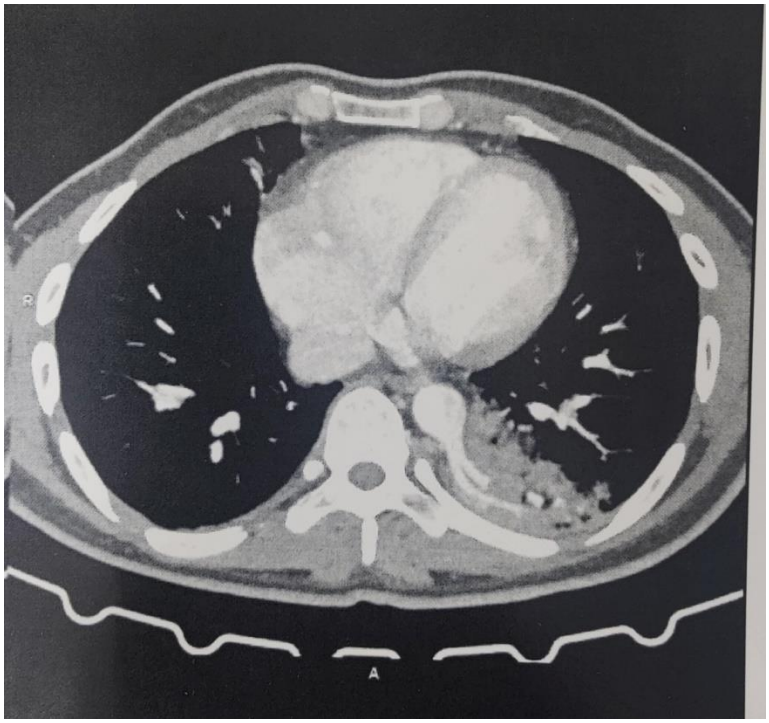

Figure 2: Chest computed tomographic angiography a dense and homogeneous mass in the left lower lobe irrigated by two systemic arteries

DISCUSSION

The cause and risk factors of pulmonary sequestration-associated malformations are not well known. Embryologically, pulmonary sequestration can be categorized into two types: intralobar and extralobar sequestration. Intralobar sequestration is not separated from the normal lung and is covered with the visceral pleura of the normal lung. The blood supply from the aorta passes through the normal lung. Extralobar sequestration is separated from the normal lung with its own visceral pleura, and the blood supply from the aorta does not pass through the normal lung [1, 4].

The sequestered regions represent non-functional entities devoid of their bronchoarterial connections. The primary anomaly lies in the systemic sourcing of vascular supply. Typically, vascularization of the sequestered region predominantly stems from the thoracic aorta (intralobar sequestration: 75%, extralobar sequestration: 46%) or the abdominal aorta (intralobar sequestration: 19%, extralobar sequestration: 32%).

However, the systemic artery's origin may also manifest as a branch of the aorta, including the diaphragmatic, intercostal artery, celiac trunk, renal arteries, or subclavian artery.

According to Pryce, there are three pulmonary sequestration types:

Type I: Normal lung with anomalous systemic arterial supply;
Type II: Anomalous artery supplying disconnected lung and adjacent normal lung
Type III: Anomalous artery to disconnected lung

The venous drainage pattern varies based on the classification of sequestration. In extralobar variants, venous return relies on the azygos system, the inferior vena cava, or, in rare instances, the portal vein. Conversely, intralobar types commonly exhibit venous drainage facilitated by a normally positioned pulmonary vein, occasionally supplemented by a venous trunk of the mediastinal system with a sub- or transdiaphragmatic trajectory.

Pulmonary sequestration commonly involves patients who are younger than 30 years of age, and its incidence rate is approximately 1.1 [5]. Pulmonary sequestration is usually asymptomatic in the newborn period and may occur during infancy or later in 5–10% of cases. Many symptoms of the intralobar type are recurrent or prolonged pneumonia, coughing, wheezing, and hemoptysis. The most common symptoms of the extralobar type are respiratory distress, coughing, recurrent pneumonia, and sepsis [6].

The examination of choice is currently nuclear magnetic resonance angiography (MRI angiography) and computed tomography (CT) with contrast injection and image reconstruction, as is now possible with helical angioscanner [7]. Chest computed tomographic angiography is the best diagnostic test to not only make the diagnosis, but also outline the appropriate therapeutic intervention that is necessary to treat the patient's symptoms.various radiological aspects are possible: peripheral nodules, a single nodule, or parenchymal consolidations in the left lower lung [8]. Other related findings are the enlarged left hilum, separation, and enlarged pulmonary blood vessels. The differential diagnosis is lung cancer, aspergilloma, invasive aspergillosis, and active tuberculosis. As seen in our patient's case, the diagnosis of lung sequestration can be made in the presence of lower lobe opacity on chest X-ray.

Most patients with pulmonary sequestrations have a benign course and do not develop serious complications. The mortality rate is approximately 0.5-2%; even without treatment, most patients suffer chronically with respiratory symptoms and recurrent infection [6]. Reported late complications include hemoptysis, pneumonia, and lung abscess formation; both right- and left-sided pulmonary sequestrations have been associated with these complications. Another less common late complication is the development of malignancy. In a case series of 150 patients with pulmonary sequestrations, 14 (9.8%) patients developed one or more complications. Eight patients developed recurrent pneumonia, two developed abscess formation, two had hemoptysis requiring hospitalization, and one patient developed a malignancy. An additional patient had an early death from the pulmonary sequestration [9].

Currently, the standard treatment of pulmonary sequestration, especially in adults, is surgical excision, and more difficult in the intralobar form than in the extralobar, given the absence of a clean pleura in the sequestrated parenchyma. Surgical treatment consists of ligation-section of the aberrant artery(s) and resection of the affected lung parenchyma, usually by lobectomy [10]. Surgery under video-thoracoscopy is a controversial approach, as the absence of a cleavage plane between normal and malformed parenchyma makes it difficult to perform.

The mortality and the morbidity after surgical excision are reported to be less than those of conservative management in such cases. However, perioperative morbidity and the risks inherent to surgical treatment are considerable when patients have the risk factors for progression of arterial aneurysms, especially arteriovenous fistulas, that would cause heart failure. Pre-embolization or preoperative transcatheter embolization of the feeding artery reduces the possibility of cardiovascular dysfunction during surgery for pulmonary sequestration [11]. A prophylactic approach with embolization in a poor surgical candidate is reasonable when symptoms exist, particularly in the presence of recurrent episodes.

CONCLUSION

Pulmonary sequestration represents a rare anatomical anomaly, necessitating consideration in cases of recurrent infections localized to specific pulmonary territories, notably the postero-basal region, particularly on the left side. Imaging modalities such as CT or angio-MRI serve as the primary diagnostic tools for accurate assessment. Surgical intervention remains a cornerstone in the management of this condition.

REFERENCES

1. Cachão, J., Raminhos, I., D Martins, J., & Bravio, I. (2020). Congenital pulmonary airway malformation and bronchopulmonary sequestration in infants–a rare diagnosis. *Nascer e Crescer-Birth and Growth Medical Journal*, *29*(4), 200-203.
2. Wu, Y., Xi, L., Pan, Z., Wu, C., & Li, H. (2023). Torsed extralobar pulmonary sequestration associated with congenital lobar emphysema: a rare entity. *Therapeutic Advances in Respiratory Disease*, *17*, 17534666231164535.

3. Yamanaka, A., Hirai, T., Fujimoto, T., Hase, M., Noguchi, M., & Konishi, F. (1999). Anomalous systemic arterial supply to normal basal segments of the left lower lobe. *The Annals of thoracic surgery*, *68*(2), 332-338.
4. Cong, C. V., Ly, T. T., & Duc, N. M. (2022). Intralobar pulmonary sequestration supplied by vessel from the inferior vena cava: Literature overview and case report. *Radiology Case Reports*, *17*(4), 1345-1353.
5. Arjun, P., Palangadan, S., Haque, A., & Ramachandran, R. (2017). Intralobar sequestration. *Lung India*, *34*(6), 559-561.
6. Muri, J., Makovicky, P., Kamarad, V., Durcova, B., Vecanova, J., & Hvizdosova, N. (2024). Pulmonary sequestration in adulthood: clinical-morphological study. *Clinical Study*, *153*, 158.
7. Ikezoe, J., Murayama, S., Godwin, J. D., Done, S. L., & Verschakelen, J. A. (1990). Bronchopulmonary sequestration: CT assessment. *Radiology*, *176*(2), 375-379.
8. Gabelloni, M., Faggioni, L., Accogli, S., Aringhieri, G., & Neri, E. (2021). Pulmonary sequestration: what the radiologist should know. *Clinical imaging*, *73*, 61-72.
9. Transcatheter Management of Pulmonary Sequestrations in Children—A Single-Center Experience - PMC [Internet]. [cité 8 mai 2024]. Disponible sur: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC10378210/
10. Nakagawa, T., Wada, A., Aruga, N., Watanabe, H., Masuda, R., Yamada, S., & Iwazaki, M. (2022). Surgical strategy for pulmonary sequestration: Focus on precautions for aberrant vessels under minimally invasive surgery. *Proceedings of Singapore Healthcare*, *31*, 20101058211058551.
11. Zhang, Y., Qiu, Y., & Li, Y. (2022). Debating the embolization of a large aberrant systemic artery for pulmonary sequestration using an Amplatzer duct occluder: a case report and literature review. *Cardiology in the Young*, *32*(2), 331-336.