

Pulmonary Sequestration: An Unusual Finding

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ABSTRACT

Pulmonary sequestration is a rare congenital malformation that receives its blood supply from a systemic artery. We report a case of pulmonary sequestration initially diagnosed as a hydatid cyst of the lung, surgical findings revealed an abscessed collection related to a pulmonary sequestration of the left lower lobe. Pulmonary sequestration remains a diagnostic and therapeutic challenge.

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Introduction

Pulmonary sequestration is a congenital anomaly defined as a non-functioning lung tissue, separated from the rest of the lung and supplied with blood from an unusual source, often an artery from systemic circulation. The thoracic computed tomography (CT) scan and aortography allow the diagnosis. Surgery is the treatment of choice for this malformation.

Case report

A 42-year-old rural patient with a history of recurrent left basal pneumonia and a history of contact with dogs was hospitalized with left basal chest pain and chronic cough with no evidence of hydatid vomiting.

Chest X-ray (Fig. 1) showed a poorly limited left basal opacity. Bronchial endoscopy noted an inflammatory aspect of the left lower lobar bronchus, with negative hydatid serology. Thoracic CT scan (Fig. 2) showed the presence of a left basal rounded fluid density formation not enhanced by the contrast product and without the visualization of aberrant vascularization.



Fig 1. Front chest X-ray showing a poorly limited left basal pulmonary opacity

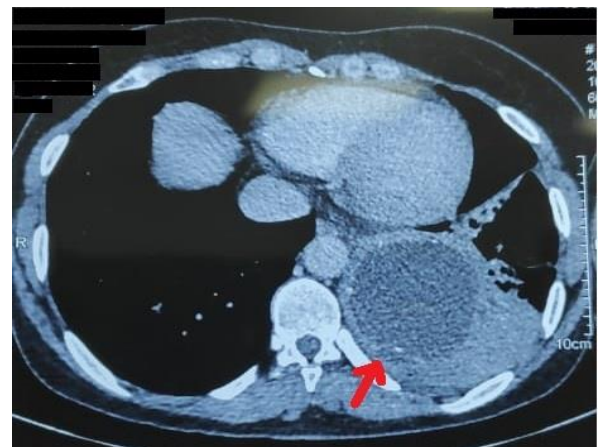


Fig 2. axial thoracic CT image showing a rounded basal fluid density cyst with thick wall of the left lower lobe (arrow) not enhanced by the contrast product.

The patient was treated by antibiotics for three weeks with clinical improvement and a clear regression of the radiological opacity. (Fig. 3).

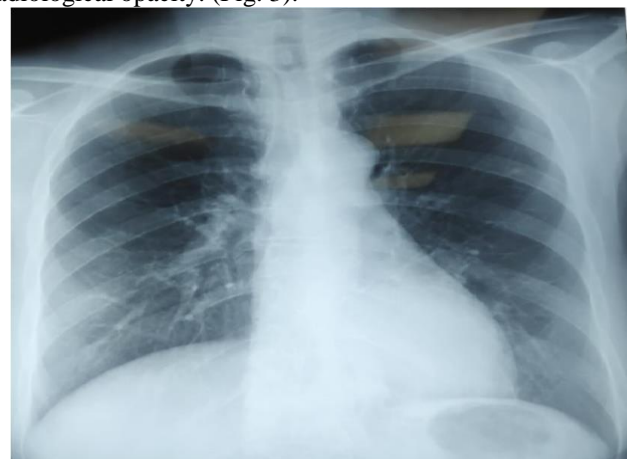


Fig 3. Front chest X-ray showing the radiological improvement

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Posterolateral thoracotomy in the sixth left intercostal space showed a collapsed postero-basal segment of the lower lung lobe. Dissection of the triangular ligament revealed the two aberrant systemic arteries leading from the aorta, each had a diameter of approximately 0.5 mm. A ligation-section of both arteries was performed. Venous return from the inferior lobe occurred at the inferior pulmonary vein. Re-expansion of the inferior lobe, including the postero-basal segment, was not good and a left inferior lobectomy was performed. The operative data led to the conclusion that it was a Pryce's Type II pulmonary sequestration.

The postoperative outcome was simple. Venous return from the inferior lobe occurred at the inferior pulmonary vein. Re-expansion of the inferior lobe, including the postero-basal segment, was not good and a left inferior lobectomy was performed. The operative data led to the conclusion that it was a Pryce's Type II pulmonary sequestration.

The postoperative sequelae were simple. Clinical and radiological check-ups at one, three and six months did not reveal any abnormalities, the chest CT scan at one year was satisfying. With an 18-month follow-up, the patient remained asymptomatic and the radiological checks revealed no abnormalities.

Discussion

Pulmonary sequestration is a non-hereditary congenital malformation secondary to an anomaly occurring during pulmonary organogenesis (22nd to 24th day). It accounts for 0.15 to 6.4% of all congenital lung defects and can be seen at any age with a slight prevalence in the first two decades. Described for the first time by Rokitanski and Rektorzic in 1861, it was Pryce [1] who, in 1946, gave a precise definition and distinguished two types: intra- and extralobar.

In intralobar sequestrations (ILS) the abnormal parenchyma is included in the normal lung: it is enveloped by the same visceral pleura; the usual seat is the postero-basal region of the lower lobe without predominance on the side.

Extralobar sequestrations (ELS) have no connection with the normal lung due to their development from a supernumerary bronchial bud. There is therefore a complete anatomical and physiological separation between the two. ELS have their own pleural envelope. They are found in 75% of the cases between the diaphragm and the lower lobe (80% on the left). Other localization in the middle and upper lobes are rare [2,3].

The sequestered areas constitute a non-functional structure excluded from its bronchoarterial connections. The main abnormality is the systemic origin of the vascular supply [1,4,5]. The vasculature of the sequestered territory most often comes from the thoracic aorta (ILS: 75%, ELS: 46%) or the abdominal aorta (ILS: 19%, ELS: 32%). But the origin of the systemic artery can be a branch of the aorta (diaphragmatic, intercostal artery, celiac trunk, renal arteries, 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 return varies according to the type of sequestration. In extralobar forms, the venous return is dependent on the azygos system, the inferior vena cava or, exceptionally, the portal vein. In intralobar forms the venous return is often ensured by a pulmonary vein in normal

position, sometimes by a venous trunk of the cellular system with sub- or transdiaphragmatic path.

ILS can manifest as episodes of recurrent pneumonitis with fever and sometimes hemoptysis, chest pain. Sometimes, the discovery is made during a complication: hemothorax or heart failure by shunt effect.

The macroscopic appearance of the sequestered parenchyma is often atelectatic or dystrophic. Histological examination shows air cavities with a bronchial structure with a collagen wall coated with a cylindrical epithelium.

The chest X-ray can evoke the diagnosis in front of an opacity of posterior base seat in particular on the left. CT with injection is an excellent examination for the diagnosis and preoperative assessment of pulmonary sequestration [6,7].

It recognizes the nature of the mass and shows the systemic artery [2,6-8]. But magnetic resonance imaging (MRI) and in particular angio-MRI is undoubtedly the ideal method for diagnosing pulmonary sequestration [9,10]. Aortography has no diagnostic indication at present and its practice is now reserved for embolization techniques [2,7].

Treatment of pulmonary sequestration is necessary due to the risk of repeated respiratory infection and spontaneous hemothorax [3,11]. It is essentially surgical. Control of the systemic nourishing artery is difficult given the fragility of its embryonic wall and its elasticity with the risk of retraction in the mediastinum or through the diaphragm. The surgical procedure often consists of a lobectomy, rarely a segmentectomy. A simple ligation of the afferent arterial pedicle can be performed each time the pulmonary parenchyma is considered normal and there are no dystrophic lesions: Campbell et al. [12] as well as Ernst et al. [13] reported cases. Another observation is our observation. Furthermore, Yamanaka et al. [4] report a case where the aberrant artery was severed at its origin and anastomosed laterally with the lower edge of the left pulmonary artery without pulmonary parenchymal resection.

Recently, surgical treatment by videothoracoscopy has been performed [14]. It seems technically more easily achievable in extralobar sequestrations and in children. Currently, endovascular techniques are proposed for the treatment of pulmonary sequestrations and mainly in patients whose general condition is altered or in patients with heart failure. Izzillo et al. [15] reported a case of type I intralobar sequestration of Pryce treated by percutaneous vaso-occlusion using metal coils with the occurrence of a rapidly resolving pulmonary infarction.

This technique may be insufficient and must then be supplemented by a surgical procedure. In addition to the occurrence of pulmonary infarction, embolization involves other risks, notably that of paraplegia [16].

Conclusion

Pulmonary sequestration is a rare malformation, the diagnosis of which must be evoked in the face of repeated infections in the same pulmonary territory, in particular the postero-basal territory (especially on the left). CT or angio-MRI are the exams of choice for diagnosis and assessment. Surgical treatment retains a prominent place.

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