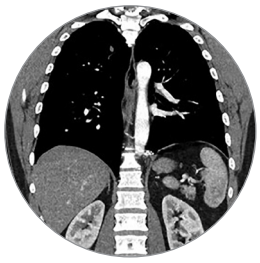

Recurrent Pneumonia as A Manifestation of Pulmonary Sequestration. Case Report

Neumonía recurrente como manifestación de secuestro pulmonar. Presentación de caso

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Palabras clave (DeCS)

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Tomografía computarizada multidetector

Summary

Pulmonary sequestration is a congenital respiratory disease that involves the pulmonary parenchyma and its vasculature. It can be divided into intralobar or extralobar depending on its relationship with normal visceral pleura. The extralobar subtype is usually diagnosed prenatally or in the first months of life, while the intralobar occurs in young adults. It represents approximately 0.15-6.4% of all congenital lung malformations and does not show differences with respect to sex with a 1:1 ratio. We present the case of an average adult male patient with recurrent pneumonia in whom multiples imaging studies are performed, with tomographic findings compatible with intralobar pulmonary sequestration in the left posterior basal segment, with the aim of performing a literature review to raise a discussion on the importance of the proper and early diagnosis of this pathology. It is necessary to have a high clinical suspicion -it is a disease of variable presentation-, to request the most appropriate imaging study, since the images are those that give the definitive diagnosis of pulmonary sequestration. In some cases, management can be performed in a minimally invasive manner by interventional radiology, as in this case.

Resumen

El secuestro pulmonar es una enfermedad congénita del tracto respiratorio que compromete el parénquima pulmonar y su vasculatura, puede dividirse en intralobar o extralobar dependiendo de su relación con la pleura visceral normal. El subtipo extralobar usualmente se diagnostica prenatalmente o en los primeros meses de vida, mientras que el intralobar se presenta en adultos jóvenes. Representa aproximadamente el 0,15-6,4 % de todas las malformaciones pulmonares congénitas y no tiene diferencias respecto al sexo con una relación 1:1. Se presenta el caso de un paciente masculino adulto medio con neumonías a repetición en quien se realizan estudios imagenológicos con hallazgos tomográficos compatibles con secuestro pulmonar intralobar en el segmento basal posterior izquierdo, con el objetivo de realizar una revisión de la literatura para plantear una discusión sobre la importancia del diagnóstico adecuado y a tiempo de esta patología. Es necesario tener una alta sospecha clínica —se trata de una enfermedad de presentación variable—, para solicitar el estudio imagenológico más adecuado, puesto que las imágenes son las que dan el diagnóstico definitivo del secuestro pulmonar. En algunos casos, el manejo se puede realizar de manera mínimamente invasiva a cargo de radiología intervencionista, como en este caso.

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Introduction

Pulmonary sequestration (PS) is a congenital disease of the respiratory tract characterized by an area of dysplastic and non-functional lung tissue, which is not in continuity with the tracheobronchial tree and is also irrigated by aberrant systemic vessels, usually coming from the aorta (1). However, these vessels can also originate from the celiac trunk, the splenic artery, the intercostal arteries and even the coronary arteries (2). It represents approximately 0.15-6.4% of all congenital pulmonary malformations and shows no difference with respect to sex, with a ratio of 1:1 (3, 4).

Two types of pulmonary sequestration have been described: intralobar (IPS) and extralobar (EPS). The reference diagnostic test is arteriography. This is the clinical case of an average adult patient with a diagnosis of intralobar pulmonary sequestration and who received minimally invasive management with satisfactory results. An analysis of the case and a review of the literature are carried out.

Clinical case

Average adult male patient with a history of recurrent pneumonia, who goes to the emergency department for fever and respiratory symptoms. On admission, temperature of 38.3 ºC, tachycardia and decreased vesicular murmur at the left lung base were recorded. Paraclinics are performed, which report leukocytosis of 16,880 mm3, with neutrophils of 85% and PCR of 101.48 mg/L. PA and lateral chest radiography is performed with evidence of alveolar type opacity at the height of the lower left lobe in the lower and posterior segments (Figure 1). Antibiotic therapy is initiated for the management of the acute picture; However, because of the history of recurrent pneumonia, a chest CT scan with contrast medium is performed, in which alveolar infiltrations are observed in the posterior basal segment of the left lower lobe, with arterial vascular structure originating from the distal thoracic aorta, with venous drainage to the pulmonary veins, findings compatible with intralobar pulmonary sequestration in the left posterior basal segment (Figure 2). In view of the findings described by imaging and the reiterative clinic of pulmonary infectious processes, embolization and selective occlusion of aberrant arterial vessel by means of coils and microparticles are performed, a procedure performed without complications, with satisfactory evolution and adequate postoperative control (figures 3 and 4).

Discussion

Intralobar lung sequestration accounts for 75% of all lung abductions. This consists of an abnormal lung segment that shares visceral pleura with a normal lung lobe and lacks of a normal communication with the tracheobronchial tree, irrigated mainly by anomalous branches coming from the aorta, which are usually located through the lower pulmonary ligament and draining through the pulmonary veins into the left atrium. Your location mainly occurs in the lower lobe with a predominance of slightly larger in the left lung than in the right one (3). Findings consistent with the case presented in which it was evident clinical and imaging features in the left lung field. Both types of pulmonary sequestration present a pathogenesis different. EPS is a congenital anomaly whose etiopathogenesis is still a bit controversial, but is believed to develop from an outbreak supernumerary lung that obtains its blood supply from of the primitive splenic vessels surrounding the anterior bowel (5).

On the other hand, IPS is not related to congenital malformations, so it is thought to be an acquired condition secondary to multiple infections, chronic inflammatory processes in the lungs, or bronchial obstructive conditions (6). This is consistent with the diagnosis of the patient, in whom the only significant history was lower respiratory tract infection of the recurrent pneumonia type, which is one of the most common symptoms of patients with intralobar sequestration. The diagnosis of IPS can be made at any age in more than half of the patients. It is usually diagnosed after the second decade of life. In contrast, EPS is diagnosed primarily during the fetal or neonatal period associated with diaphragmatic hernias, heart abnormalities and even pulmonary hypoplasia (3, 7).

The clinical features of patients with IPS typically present with a chronic history of coughing, mucopurulent expectoration, and recurrent pneumonias, usually caused by pyogenic bacteria (8). When IPS is suspected, diagnostic tests should focus on identifying aberrant circulation and looking for communications with the gastrointestinal tract or associated congenital abnormalities (6). The initial examination should be a simple chest x-ray, in which parenchymal abnormalities or vascular shadows suggesting the diagnosis can be identified. In this case, the radiographic findings were consistent with left pneumonias on multiple occasions; however, they are non-specific findings for the diagnosis of pulmonary sequestration.

Angiography is considered the standard test for identifying the nutrient circulation of the PS; however, although this study is capable of identifying the aberrant arterial supply of a sequestered lung, it is invasive, requires hospitalization, and does not provide sufficient information about lung structure. Currently, noninvasive imaging techniques including computed tomography (CT), magnetic resonance imaging (MRI), and contrast Doppler ultrasound have displaced diagnostic angiography as the first option.

In particular, multi-detector CT with intravenous contrast has been shown to be equally effective as angiography and if these two are performed simultaneously they serve to confirm an abnormal arterial supply and to distinguish pulmonary sequestration from other pulmonary opacities (6).

CT is very useful in identifying parenchymal changes and the extent of lung involvement. Typical radiological findings of intralobar lung sequestration are a homogeneous or heterogeneous mass of soft tissue in the lower lobe, usually accompanied by heterogeneous consolidation zones and emphysemic changes at the edges of the lesion. Generally, the rest of the non-sequestered lung parenchyma is healthy, findings consistent with those described in the case, where parenchymal alterations were found at the level of the left posterior basal segment associated with an arterial vessel from systemic circulation (aorta) in the single chest CT and with contrast medium.

The treatment is usually surgical, even some authors consider it even in asymptomatic cases, mainly due to the malignant transformation of the aberrant parenchyma and other possible complications (9). Total resection of the abducted lung parenchyma by videothoracoscopy is considered the surgical management of choice (10). However, some authors consider arterial embolization as an alternative that allows the reduction of the size of the lesion, for an eventual later removal. An abnormal systemic arterial supply not only confirms the diagnosis, but also provides a preoperative vascular roadmap for the surgeon, thus minimizing the chances of involuntary vascular injury (7). In the case presented, an endovascular embolization of the aberrant artery was performed by interventional radiology. By means of post-procedure arteriography, the total devascularization of the arterial tree corresponding to the pulmonary sequestration was confirmed. The procedure was carried out without complications and the patient has not presented new acute low respiratory episodes, which confirms the PS as predisposing for the development of repeat pneumonia. An imaging control was performed two months after the interventional procedure, which showed a satisfactory evolution.

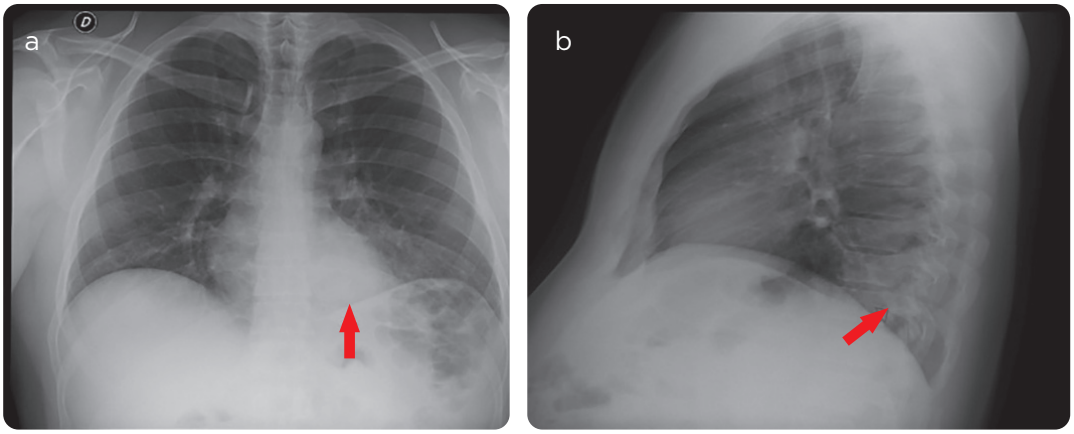

Figure 1. Chest X-ray PA and lateral, which shows alveolar opacity at the level of the lower left lobe (arrows).

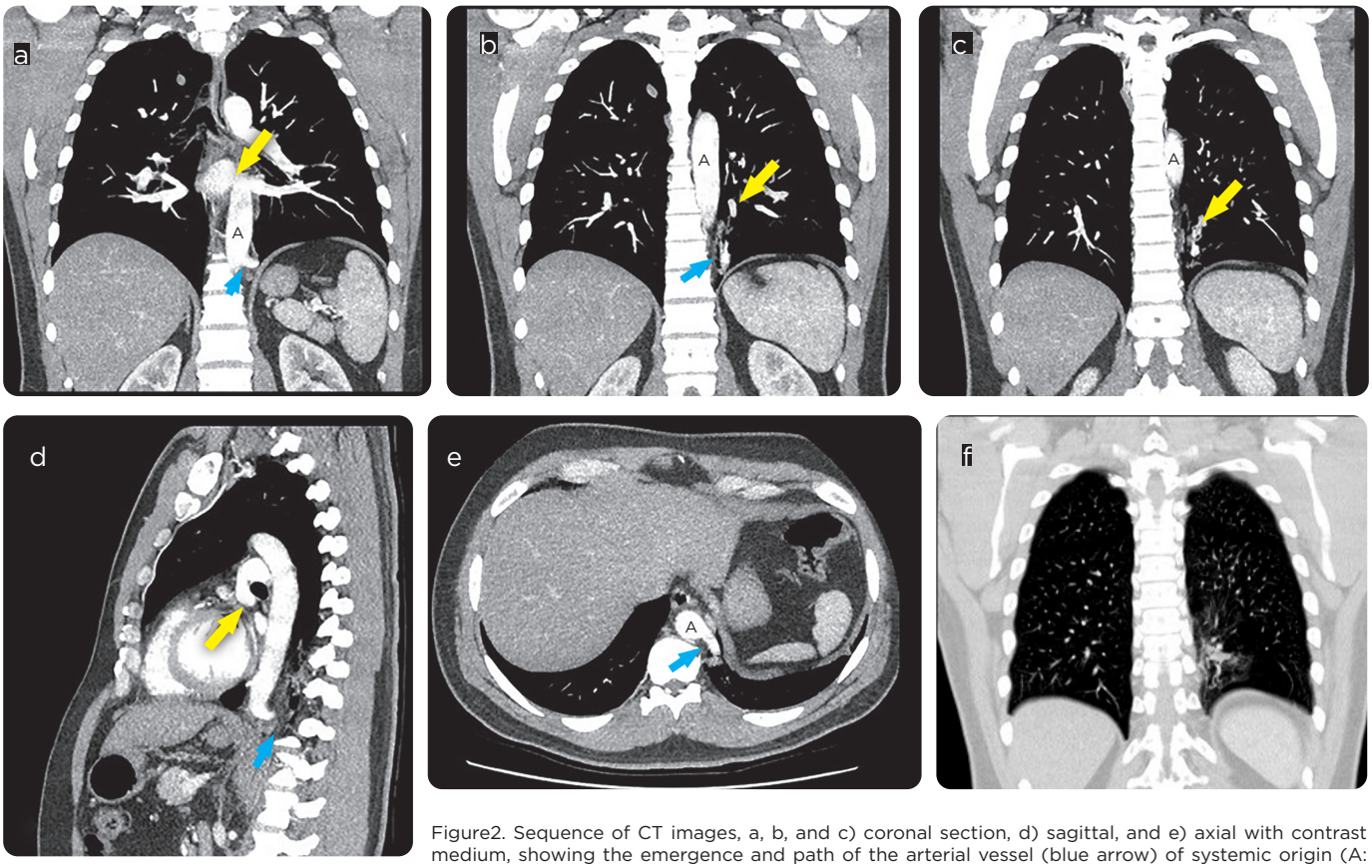

Figure2. Sequence of CT images, a, b, and c) coronal section, d) sagittal, and e) axial with contrast medium, showing the emergence and path of the arterial vessel (blue arrow) of systemic origin (A: thoracic aorta) and the venous drainage a (yellow arrow) pulmonary veins. f) Image in coronal section window for lung showing alveolar type infiltrates in the parenchyma.

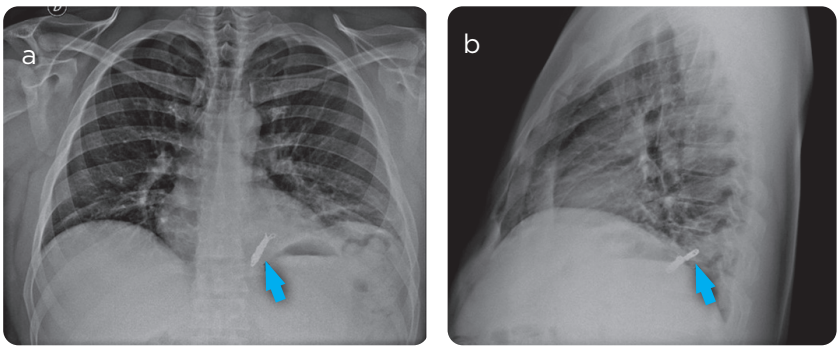

Figure 3. Chest X-ray with PA and lateral view showing embolization material in the left lung base, without significant alterations in lung fields.

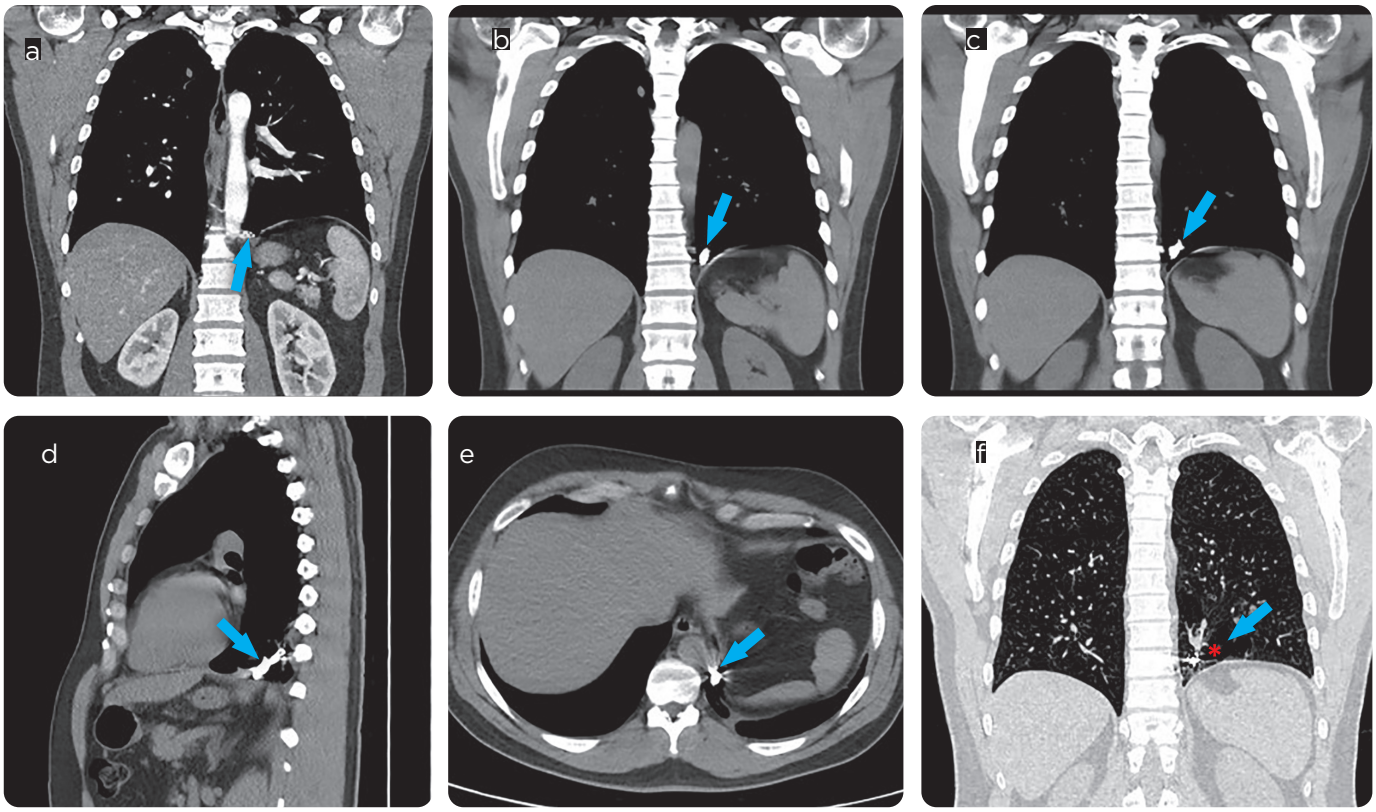

Figure 4. Sequence of CT images of the chest in a) coronal, b) and c) coronal cuts, d) sagittal and e) axial, in cuts similar to figure 2 (comparatively pre-treatment and post-treatment), showing surgical embolization material (blue arrow) in aberrant artery vessel and f) coronal cut in lung window, showing fibrous tracts and volume loss of the compromised segment (asterisk). Normal changes expected for the procedure.

Conclusions

Sequestration of the lung is a rare congenital pathology that occurs prenatally and/or in the neonatal period (extralobar subtype) and in young adults (intralobar subtype), usually with recurrent lower respiratory tract infections without other characteristic findings. Arteriography is considered the standard method for diagnosis, but currently, thorax angiotomography is accepted as the method of choice to make evident the aberrant arterial vessel associated with the parenchymal changes described, which suggests that the diagnosis of this pathology is purely radiological and hence the importance of radiologists being familiar with this type of disease, in order to avoid diagnostic errors and delays in treatment. As it became evident, in selected cases it can be performed by means of embolization, which offers a less invasive therapeutic alternative than conventional resection of dysplastic lung tissue.

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