Case Report

Bilateral pulmonary sequestration complicated with a bibasal pneumonia in an adult patient

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A B S T R A C T

Pulmonary sequestrations (PS) are rare congenital pulmonary malformations, characterized by non-functioning and dysplastic pulmonary tissue, without a normal connection to the tracheobronchial tree and supplied by the systemic arterial circulation. PS typically occur in the lower lobes and the radiologist should consider the possibility of a PS in a patient with persistent or recurrent pneumonia in the lower lobes, especially in children. We hereby present a rare case of bilateral intralobar PS complicated with bilateral pneumonia, in a previously healthy 37-year-old male patient, who was referred to the emergency department by his general practitioner because of persisting dyspnea and fever. The hospital stay was complicated with central nervous aspergillosis due to septic emboli.

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Introduction

Pulmonary sequestrations (PS) are rare congenital pulmonary malformations, characterized by non-functioning and dysplastic pulmonary tissue, without a normal connection to the tracheobronchial tree. They occur predominantly in the lower lobes and are typically supplied by the systemic arterial circulation, hence contrast administration is useful to highlight this aberrant systemic blood supply [1,2]. The first description of this rare entity dates from 1777 [2].

Case report

A previously healthy 37-year-old male patient was referred to the emergency department by his general practitioner because of persisting dyspnea and fever. Ten days before he developed headaches, general malaise and fever. Empiric antibiotic treatment was prescribed but symptoms persisted. Lung auscultation revealed bilateral diminished breathing sounds, most noticeable in the basal lung fields. A chest radiograph was requested to rule out pneumonia.

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The chest radiograph showed bilateral lung opacities in the lower lobes (Fig. 1).

Due to the persistent symptoms and high fever, a subsequent computed tomography (CT) with intravenous contrast administration was requested to rule out empyema. This CT demonstrated bibasal lung consolidations containing air bronchograms and also hypodense regions. Four aberrant arteries on the right and two on the left, originating from the distal thoracic aorta, ran towards the consolidations (Figs. 2-4). The diagnosis of bilateral intralobar PS was made. The hypodense regions were compatible with the PS.

**Fig. 1** – Frontal chest radiograph. Bilateral lung opacities in the lower lobes.

**Fig. 2** – Chest CT with intravenous contrast administration. Arterial phase. Axial slice demonstrates bilateral pneumonic consolidations with aberrant arteries originating from the distal thoracic aorta (white arrows) entering the bilateral PS.

**Fig. 3** – Chest CT with intravenous contrast administration. Coronal image in the lung window demonstrates bilateral pneumonic consolidations with air bronchograms.

**Fig. 4** – 3D-angiographic reconstruction showing 3 out of 4 aberrant arteries to the right PS and 1 out of 2 aberrant arteries to the left PS, originating bilaterally from the distal thoracic aorta (white arrows).
A nasopharyngeal swab for detection of viral infection using polymerase chain reaction returned positive for influenza type A. Note that this patient presented to our emergency department during the coronavirus pandemic. Empirical treatment with antiviral and antibiotic agents was initiated and the patient was hospitalized. Over the course of the following days however, the patient’s condition worsened. A bronchoalveolar lavage was performed, which showed influenza type A and also revealed high levels of Aspergillus antigen. Microbiological culture was also positive for Aspergillus fumigatus. Antimycotic treatment was initiated. On day 5 of hospitalization, the patient’s condition quickly deteriorated with impending respiratory failure. Control CT of the chest revealed an important amount of bilateral pleural fluid and progression of the bilateral consolidations. Non-invasive ventilation was started, pleural fluid drainage was performed, and antibiotic treatment was adjusted. On day 7, respiratory parameters were stable. Nevertheless, the neurological condition of the patient further declined. CT scan of the brain showed multiple hypodense areas bilaterally, possibly indicating septic emboli. The following day magnetic resonance imaging (MRI) of the brain with contrast administration took place. This MRI investigation revealed multiple cerebral and cerebellar ring-enhancing lesions with diffusion-restriction (Figs. 5 and 6). The diagnosis of central nervous aspergillosis due to septic emboli was made. Antifungal therapy was continued. Inflammatory parameters declined over the course of the following days. A month later the neurological condition was improved sufficiently to transfer the patient to a specialized revalidation hospital. Surgical treatment with resection of the bilateral PS was recommended, but the patient refused surgical treatment. Further clinical follow-up is ongoing.

![Fig. 5 – MRI of the brain. Contrast-enhanced T1 sequence. Axial image. Multiple ring-enhancing lesions.](image)

**Discussion**

PS are the second most common congenital lung anomaly, accounting for 1%-6% of all congenital lung abnormalities. Some studies propose an incidence of 0.15%-1.8% [2,3]. Bilateral PS are extremely rare, with only a few cases known in the literature. As shown in a large retrospective analysis by
Wei et al. in 2011, only 3 of their 2625 patients with PS had bilateral PS [2,4–6].

There are two types of PS: intralobar PS are the most common form, accounting for 75% of the cases, while the remaining 25% is extralobar [3].

The difference between intra- and extralobar pulmonary sequestration is based on their pleural covering. While extralobar PS are covered by their own pleura, intralobar PS are located within a normal anatomic lobe of the lung and are contiguous with the adjacent normal lung parenchyma [1,2].

Intralobar PS are located in 98% of cases in the lower lung lobes, with a preference for the left hemithorax (60%). Similarly to intralobar PS, extralobar PS are preferentially located in the left thoracic cavity (65%) and are located in 63% between the lower lung lobes and the diaphragm [2].

Finding a systemic arterial blood supply to an aberrant lung region supports the diagnosis of pulmonary sequestration [7]. However, the inability to identify an aberrant artery does not exclude the diagnosis of PS [8]. Multiple aberrant arteries are present in 16% of PS [7].

While both intralobar and extralobar PS mainly receive their arterial supply by the thoracic and abdominal aorta, venous drainage is different between them. Venous drainage from the intralobar PS is predominantly performed by the pulmonary venous system, while extralobar PS show predominantly a systemic outflow [2].

Different imaging methods can suggest the possibility of a pulmonary sequestration. On chest radiograph and CT, PS can exhibit a wide spectrum of imaging appearances. The most challenging are intralobar PS, because they are invested in the normal visceral pleura [1,2]. Intralobar PS can appear as a pneumonia, as in our patient. Other presentations of intralobar PS are as a mass, a cavitary lesion, a cystic lesion or a combination of previous presentations [3,5,7]. Extralobar PS almost never contain air, since they are separated from the normally aerated lung lobes by their own pleura and generally appear as a well-defined mass on CT [7,9].

One should always consider the possibility of a pulmonary sequestration in a patient with persistent or recurrent pneumonia in the lower lobes, especially in children [2]. Not only in the diagnosis, but also in the therapeutic workup of PS, CT plays an important role, more specifically CT angiography, because it can detect the aberrant systemic artery in most cases [1]. MRI and conventional angiography are other imaging techniques to identify this aberrant arterial blood supply [7].

Extralobar PS are often discovered early, within the first year of life, probably because of the association with other congenital anomalies. In contrast, intralobar PS are diagnosed during late childhood or early adulthood, as with our patient. Common clinical symptoms are cough, hemoptysis, fever, and chest pain. A small percentage of patients are asymptomatic [1,5,7].

Late diagnosis is common because of the misinterpretation of recurrent pulmonary infections as standalone incidents. Especially when someone suffers from recurrent pneumonia in a single lung lobe, the clinician and radiologist should include a possible pulmonary sequestration in the differential diagnosis [1,3,7].

Treatment of intralobar PS preferably involves surgery with either segmentectomy or lobectomy. The correct ligation of the aberrant vessels is important during surgery, emphasizing the value of preoperative workup with imaging [3,7].

In conclusion, we want to stress the importance of contrast administration for making the diagnosis on CT, without which, the diagnosis would be challenging in the case presented above. Furthermore, we want to emphasize the importance to include the possibility of PS in the differential diagnosis in patients with recurrent or persistent pneumonia especially in late childhood or early adulthood.

REFERENCES

[1] Lee EY, Boiselle PM, Cleveland RH. Multidetector CT evaluation of congenital lung anomalies. Radiology 2008;247(3):632–48.
[2] Corbett HJ, Humphrey GM. Pulmonary sequestration. Paediatr Respir Rev 2004;5(1):59–68.
[3] Walker CM, Wu CC, Gilman MD, Godwin JD, 2nd, Shepard JA, Abbott GF. The imaging spectrum of bronchopulmonary sequestration. Curr Probl Diagn Radiol 2014;43(3):100–14.
[4] Enfield KB, White WC, Jones D, Truitt JD. Bilateral pulmonary sequestration. Am J Respir Crit Care Med 2011;184(1):141.
[5] Wei Y, Li F. Pulmonary sequestration: a retrospective analysis of 2625 cases in China. Eur J Cardiothorac Surg 2011;40(1):e39–42.
[6] Oliveri Aruete F, Candelario Cáceres A, Alonso Mallo E. Bilateral Intralobar pulmonary sequestration in a young adult. A Case Report. Arch Bronconeumol 2017;53(5):281–2.
[7] Frazier AA, Rosado de Christenson ML, Stocker JT, Templeton PA. Intralobar sequestration: radiologic-pathologic correlation. Radiographics 1997;17(3):725–45.
[8] Ahmed M, Jacoby V, Vogl TJ. Multislice CT and CT angiography for non-invasive evaluation of bronchopulmonary sequestration. Eur Radiol 2004;14(11):2141–3.
[9] Zylak CJ, Eyler WR, Spizarny DL, Stone CH. Developmental lung anomalies in the adult: radiologic-pathologic correlation. Radiographics 2002;22 Spec No:S25–43.