CASE REPORT

Intralobar pulmonary sequestration: diagnostic expertise

Joana Sotto Mayor,[1] Diogo Rocha,[2] Sofia Esperança,[1] Antonio Oliveira e Silva[1]

[1]Department of Internal Medicine, Hospital de Braga, Braga, Portugal
[2]Department of Radiology, Hospital de Braga, Braga, Portugal

Correspondence to
Dr Joana Sotto Mayor,
s.mayor.joana@gmail.com

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SUMMARY

An obese 22-year-old man with a history of recurrent respiratory infections presented to the emergency room with left pleuritic chest pain, productive cough with mucupurulent sputum and an axillary temperature of 37.7°C. Blood work showed elevated inflammatory parameters and chest X-ray was relevant for heterogeneous infiltration in the left base and opacity of the left costophrenic angle. An angio-CT scan revealed areas of bilateral consolidation with presence of an arterial branch originating from the aorta to the collected area of the left lower lobe, consistent with a respiratory infection grafted on a intralobar pulmonary sequestration of the left lung base. The infectious process was treated and the patient was planned for a lower left lobectomy.

BACKGROUND

In 1777, Hubber described an anomalous artery that originates in the aorta and supplies a normal lower lung lobe.[1] In 1861, Rokitansky and Rektorzik described a series of cases that resembled what is now known as extralobar pulmonary sequestration (EPS).[1] Pryce, in 1946, introduced the term, PS, and defined all its anatomy.[1 2]

PS is a rare congenital malformation of the lower respiratory tract, responsible for 0.15–6.4% of all congenital lung malformations. It consists of an area of pulmonary tissue that does not usually communicate with the tracheobronchial tree, and receives its blood supply from the systemic circulation, typically from an accessory vessel originating on the thoracic or abdominal aorta.[1]

From an anatomical point of view, the PS is classified into two types: intralobar PS (IPS), where the abnormal lung mass is located within a normal lung lobe and is not coated with a proper visceral pleura; and EPS, where the mass of abnormal lung is separated from the rest of the normal parenchyma by proper visceral pleural coating. IPS is more common than EPS, corresponding to about 75–90% of all PS, probably due to the pathophysiological mechanisms involved in the development of each.

We present a case of a young man admitted to the hospital because of community-acquired pneumonia (CAP) and whose clinical research path led to a diagnosis of IPS with institution of appropriate therapy.

CASE PRESENTATION

A 22-year-old man with a history of pulmonary infections, a non-smoker taking no chronic medication, was admitted to our emergency department for the second time in 2 weeks, for a pleuritic pain of the left hemithorax and progressively worsening productive cough and asthenia. He denied dyspnoea and had no other symptoms.

On pulmonary auscultation, a vesicular murmur was present and symmetric, crackles were present on the left pulmonary base. He was tachycardic (100 bpm). Blood tests showed a C reactive protein of 270 mg/L and a leucocyte count of 179 000/µL. Blood gas analysis breathing room air was normal. Chest X-ray showed heterogeneous infiltrates in the left pulmonary base, with opacity of the left costophrenic angle (figure 1).

Given the clinical presentation and laboratory tests results, the patient was admitted for CAP to monitor therapy response in a twice presenting young patient.

Blood and sputum cultures were made and empirical antibiotic therapy with ceftriaxone and clarithromycin was started.

On the second day of hospitalisation, the patient's status worsened, with new-onset dyspnoea and hypoxaemia on breathing room air.

INVESTIGATIONS

An angio-CT scan was ordered to exclude pulmonary thromboembolism (PTE), which was considered given the sudden appearance of the symptoms. The CT revealed: 'Consolidation of the lower lobes, especially on the left. In the lower left lobe, the consolidation area had associated cystic bronchiectasis, and adjacent to this, particularly in the basal segments, there was a fluid density area with multiple dispersed gas bubbles not communicating,

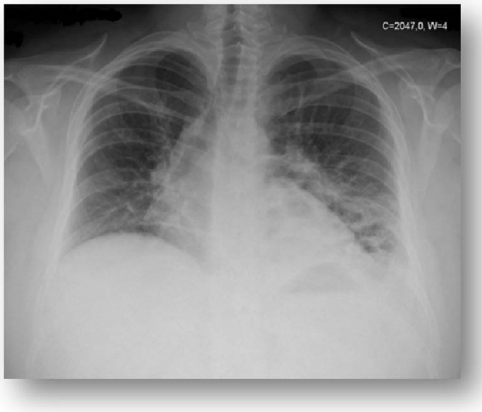

Figure 1 Motley infiltrate in the left base, with obliteration of the costophrenic left angle.

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suggesting thick liquid/collected. An arterial branch originating from the aorta (thoracic-abdominal transition) coexisted, and was directed to the epicentre of the aforementioned net area, suggesting a probable respiratory infection grafted on a pulmonary left lung base sequestration, without clear images of PTE' (figure 2).

TREATMENT

Antibiotics were switched to meropenem and vancomycin due to the clinical deterioration.

On the third day after admission the patient, already on broad-spectrum antibiotics, develops an episode of severe dyspnoea and hypoxaemia, requiring a high-concentration oxygen mask, and was transferred to an intermediate care unit for increased surveillance and monitoring. A day later, after being clinically stabilised, the patient returned to the internal medicine department.

OUTCOME AND FOLLOW-UP

Progressive clinical and analytical improvement was achieved, and on the eighth day of hospitalisation, oxygen therapy was no longer needed. The patient completed 14 days of antibiotic therapy with meropenem and vancomycin, and a control angio-CT scan revealed 'significant improvement in imaging, with resolution of almost all the consolidated parenchyma in the left lower lobe. Within the left lower lobe, cystic bronchiectasis was identified' (figure 3).

Plethysmography and bronchoscopy were requested and revealed no changes, with normal bronchoalveolar lavage liquid analysis.

The patient was discharged on the 18th day, completely asymptomatic, and was directed to a cardiothoracic surgery outpatient appointment to undergo lower lobectomy and maintenance of respiratory kinesiotherapy.

DISCUSSION

PS is a rare lung disease that can occur as one of two possible variants: extralobar and intralobar, with the latter being three times more frequent.[1] The intralobar form has a similar distribution between genders and is usually diagnosed after 20 years; it is a result of acute pneumonia, most often in the third decade of life.[2–5]

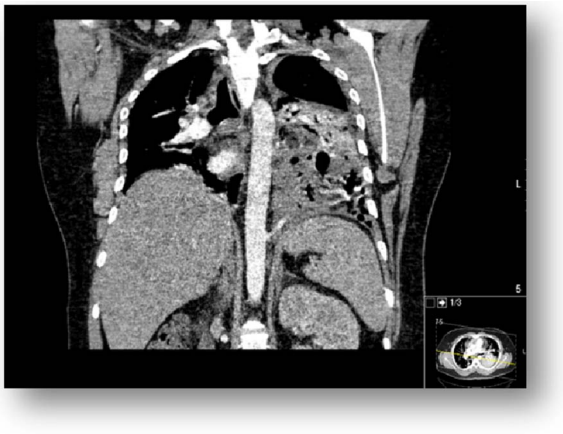

Figure 2 Pneumonic bilateral process, with areas of water density and non-communicating gas bubbles. Arterial branch originating from the aorta that leads to the area of collection, highlighting the intralobar pulmonary sequestration.

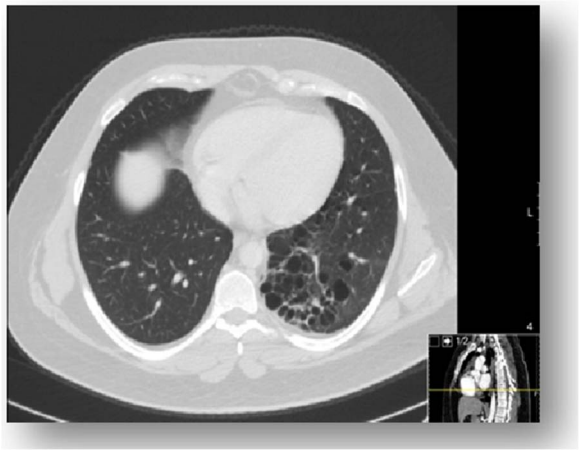

Figure 3 Almost complete resolution of the left consolidated parenchyma. Left lower lobe cystic bronchiectasis.

Regarding its physiopathology, the embryological basis for the development of PS and other congenital malformations of the lower respiratory tract is not yet fully understood.[6 7] It is known, however, that changes can occur at a very early stage of embryonic development, even before the separation of aortic and pulmonary circulations.[8] One possibility, supported by several studies, is the fact that all defects involving embryonic lung buttons represent a spectrum of the same embryological defect.[8–10]

Typically, PS is revealed after an acute respiratory infection or after cardiac symptoms.[1] Dyspnoea, cyanosis, haemoptysis and respiratory failure are often found in these patients,[1] and the vast majority have a history of repeated respiratory infections in childhood, which is not required to establish the diagnosis.[11] If present, cardiac symptoms are due to a left–right shunt and are directly related to the volume of blood flow, which in turn is dependent on the vessel's calibre anomaly.[12]

Chest roentgenogram is one of the fulcral examinations for the diagnosis of PS, requiring confirmation by arteriography and CT scan.[2 11] In the chest X-ray, PS usually appears as a radiopaque image or parenchymal area with diffuse density. The safest diagnostic preoperative approach is arteriography, which has the ability to show the anomalous systemic artery that supplies the lesion.[13] CT scan has been used to demonstrate abnormal parenchymal characteristics, but it may also identify aberrant blood supply sequestration in the lung.[11] Preoperative angiography not only confirms the diagnosis, but also shows the origin of the blood supply, crucial to the success of the intervention.

Treatment of PS consists of surgical resection of the sequestered lung parenchyma. This intervention is usually a simpler variant in extralobar type, since the malformation is well demarcated from normal lung by the visceral pleura.[14] In the case of an intralobar embodiment, the inflammatory changes resulting from previous infections may complicate the procedure and, in some cases, destroy the intersegment plane. Therefore, lobectomy resection of the sequestered segment is often the treatment of choice.[14–16] There are few postoperative complications of the procedure, which substantially improves the quality of life of these patients.[11]

The histopathological examination of the lung tissue with PS typically reveals a cystic or solid mass containing normal lung constituents, such as smooth muscle, bronchial epithelium and cartilage, but with the absence of bronchial structure and, in

most cases, no communication with the lung tissue. As noted by Halkic *et al*[3] and Louie *et al*,[12] the long-term evolution of surgical patients is excellent, confirming the highly positive results of surgical treatment of PS.[13]

In the case presented, the fact that the first therapeutic treatment was not accompanied by clinical improvement, motivated admittance to the emergency room (ER), and may be justified due to the structural changes resulting from the congenital malformation, which favoured occurrence of infections by antibiotic-resistant microorganisms.

So, when the patient re-presented to the ER, with continuing symptoms and the absence of clinical improvement, an antibiotic regimen recommended for CAP, namely, a combination of amoxicillin/clavulanic acid with clarithromycin, was instituted.

The second episode of respiratory decompensation was probably due to the fact that, once again, the predisposition to infections associated with sequestration area favoured the growth of microorganisms with increased resistance, so the current antibiotic therapy would be insufficient for the aetiological agent responsible for the symptoms. For this reason, and with a worsening clinical condition in a young patient with no other risk factors to justify decompensation, broad-spectrum antibiotic therapy with meropenem and vancomycin was immediately initiated.

At the time of clinical decompensation that motivated transfer to the intermediate care unit, and with the angio-CT allowing better characterisation of vascular and lung parenchyma, exclusion of a PTE was also achieved, since the patient's young age did not favour thrombosis prophylaxis; however, since the patient was overweight and was mostly immobilised in bed during hospitalisation, this was also a possibility that had to be considered as a potential cause of decompensation.

It is important to highlight that a bronchoscopy was performed at an advanced stage of admission, given the consensus of the medical and pneumology team not to perform invasive tests (such as a bronchoscopy) while there was no sustained clinical stability. Therefore, its realisation coincided with the final phase of the broad-spectrum antibiotic treatment cycle, with the infection being virtually resolved. That is possibly the reason why the bronchoalveolar lavage revealed no changes.

Learning points

Intralobar pulmonary sequestration is a malformation that predisposes to recurrent respiratory infections. It is difficult to diagnose unless a more comprehensive directed investigation (to the vasculature and pulmonary parenchyma) is undertaken. Early diagnosis is of paramount importance to prevent recurrent respiratory infections, which are often refractory to treatment with antibiotics. CT scan and angiography are the preferred tests for a safe and definitive diagnosis. The classical treatment involves surgical intervention, with full quality of life recovery only a short time after surgery.

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Competing interests None declared.

Patient consent Obtained.

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