

Clinical Case Seminar

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An unusual cause of chest pain

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Abstract

Case report :We present the case of a 27year-old male who went to the Emergency Room for the acute onset of chest pain irradiated to left flank and fever by 2 days. An abdominal computed tomography (CT) scan, performed for the suspicion of a kidney stone, showed a left basal pulmonary opacity. The patient was admitted to our department for suspected community-acquired pneumonia (CAP) but, a few days later, a chest CT scan with intravenous contrast revealed a thoracic congenital malformation: intralobar pulmonary sequestration. The pulmonary lesion was resected and the histopathological examination of the lesion showed the features of pulmonary sequestration.

Conclusions :Pulmonary sequestration is a rare congenital malformation that is usually diagnosed in the Pediatric age. We report here an unusual case of intralobar pulmonary sequestration in an adult mimicking a CAP and a lung cancer.

KeyWords: Pulmonary sequestration, pulmonary congenital malformation

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Introduction

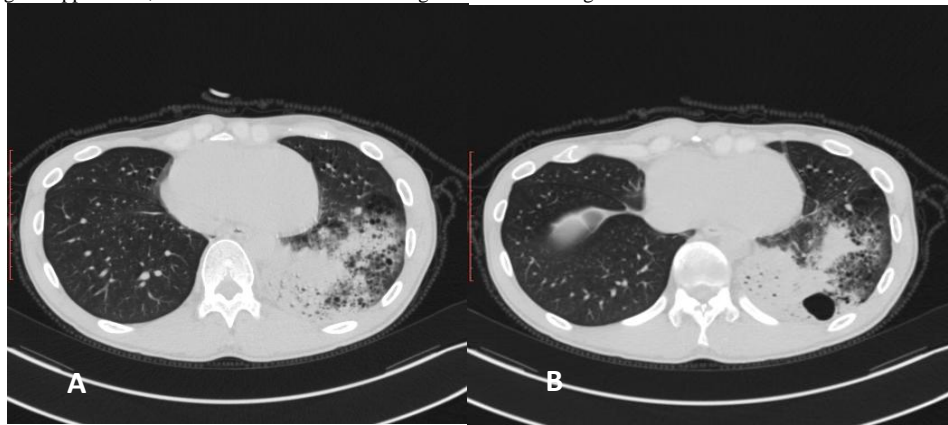
Pulmonary sequestration is an abnormal pulmonary tissue isolated from the normal bronchial tree, which generally receives its bloody supply from systemic circulation, typically from one or more anomalous systemic arteries passing through the inferior pulmonary ligament (1). In 1964 Pryce introduced the term “sequestration” (2). Pulmonary sequestration is very rare, but is considered to be the second most common congenital pulmonary anomaly (3,4).

Case Presentation

A 27year-old male, Caucasian, musician, current smoker from the age of 18 about 10 cigarettes per day (5 p-y), presented at the Emergency Room for chest subscapular pain irradiated to the left flank suddenly aroused in the evening of the day before associated with fever in the last 2 days.

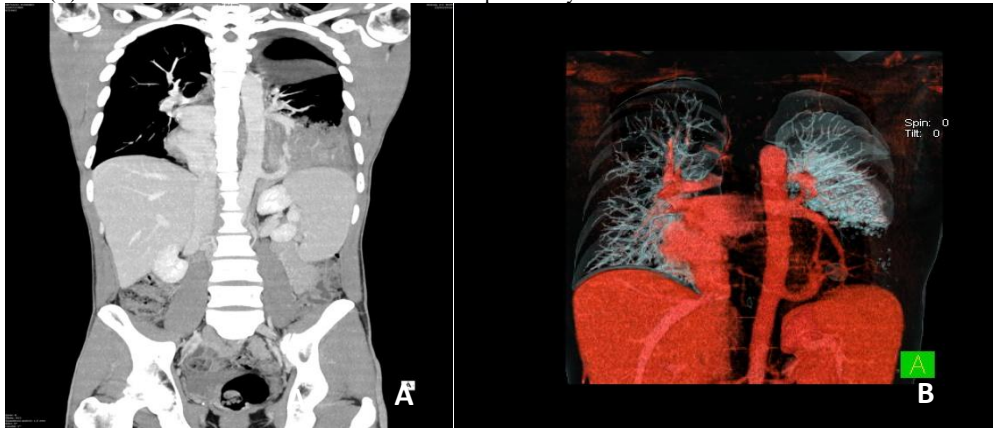
His past medical history was unremarkable and was not on regular pharmacological treatment at home. His vital signs were: systemic blood pressure 140/80 mmHg, pulse frequency 100/min rhythmic, body axillary temperature 37.1°C. Physical examination was unremarkable except for the positivity of the Giordano maneuver on the left side. An Urological consultation and an abdominal computed tomography (CT) scan were performed for the suspicion of a kidney stone, however this diagnosis was excluded whereas the CT scan revealed a pulmonary opacity in the left lower lobe (Figure 1). The patient was admitted to our Unit of Pulmonology for a suspected community-acquired pneumonia.

Figure 1. (A,B) Computed tomography (CT) scan of abdomen revealed in its most cranial scans the presence of a large parenchymal opacity of the left lower pulmonary lobe, partially solid and partially with a ground glass appearance, associated with an air bronchogram and containing an excavated area of ~ 27 mm.



At the admission the physical examination of the chest revealed on the left medium-basal lung fields decreased vocal and tactile fremitus associated with the absence of the breath sounds. Routine laboratory examination showed neutrophilic leukocytosis and increased serum levels of C reactive protein (CRP) (18.6 mg/dL, normal range <0.5 mg/dL). An arterial blood gas analysis performed with the patient breathing room air showed a mild hypoxemia (PaO₂ 74 mmHg) only. An empiric broad spectrum antibiotic treatment with ampicillin/sulbactam (1g i.v. **t.i.d.**) and clarithromycin (500 mg b.i.d. os) was started. However, during the first 5 days of hospitalization the patient had persistent chest pain and fever, despite the normalization of the blood leukocyte count and of the serum CRP level. At this point, for the suspicion of lung cancer, was performed a chest CT scan with intravenous contrast that documented an intralobar pulmonary sequestration of the left lower lobe (Figure 2).

Figure 2. (A) Contrast enhanced computed tomography scan of the chest showing an intralobar pulmonary sequestration of the left lower pulmonary lobe. Vascularization of the pulmonary lesion is supplied by a large artery originating from the descending aorta whereas venous discharge occurs by ipsilateral inferior pulmonary vein (B) 3D reconstruction of the vascularization of the pulmonary lesion better shows its vascular connections.



The pulmonary lesion was resected and the histopathological examination of the resected lesion showed the classical features of pulmonary sequestration (5) (Figure 3).

Discussion.

Pulmonary sequestrations are usually classified into two subsets (table 1). The aetiology of intralobar pulmonary sequestration is unknown. Intralobar pulmonary sequestrations may result from the formation of an accessory lung bud caudal to the normal lung buds between weeks 4 and 8 of gestation (3).

Figure 3. Hematoxylin and eosin staining of the lesion showing multiple small cysts surrounded by areas of chronic pneumonitis and aberrant small vessels.

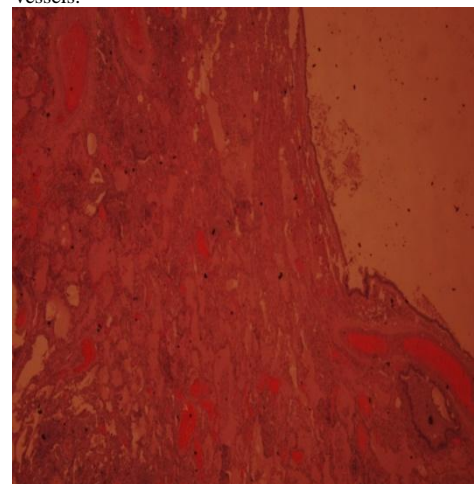


Table 1. Major differences between intralobar and extralobar pulmonary sequestration

	Intralobar pulmonary sequestration	Extralobar pulmonary sequestration
Localization and characteristics of the bronchopulmonary tissue	Confined to posterior basilar segments; no pleural separation	Found above or below diaphragm; separate pleural covering
Arterial supply	From the aorta, above or below diaphragm; usually single well developed, large artery	From pulmonary or systemic artery; usually small vessels
Venous drainage	More than 90% of the cases in the pulmonary veins	Azygos, hemiazygos or portal venous system
Side involved	~ 60% on the left	>90% on the left
Foregut communication	Unusual	More common
Association with other congenital anomalies	Rare	Frequent (often severe)

M/F ratio	1:1	3-4:1
Found in neonates	Never	Often
Clinical features	Recurrent pulmonary infection. Rarely pneumothorax, hemoptysis, haemothorax, heart failure	Respiratory distress, feeding difficulties, heart failure, antenatal fetal hydrops

Obtained with the data from the references 3 and 6

Intralobar lung sequestration can also develop postnatally following recurrent bronchial obstruction and distal infections that may lead to a fibrosis around the involved parenchyma, isolating it; in addition, recurrent bouts of inflammation may cause the development of collateral vessels in the pulmonary ligament (7). This theory is based on the observation that intralobar pulmonary sequestration, at the opposite of the extralobar, usually presents in the older child or young adult with an history of recurrent respiratory infections, occurs in equal frequency between the sexes, it is rarely associated with other congenital anomalies and there is almost always a normal pulmonary venous drainage (7).

In most cases, including the current report, the arterial supply is provided by an anomalous branch of the descending thoracic aorta. In the series of Hou and Colleagues (8) on 23 adult patients with intralobar pulmonary sequestration the aberrant arterial supply originated from the descending thoracic aorta in all cases, with the majority of the cases having a single arterial supply except three cases with multiple arterial supply.

Intralobar pulmonary sequestration is diagnosed ≤ 20 years in $\sim 50-60\%$ of cases, and it is rarely found in patients > 40 years (9).

Often the clinical presentation of intralobar pulmonary sequestration in the adulthood mimics a community-acquired pneumonia or lung cancer. In their retrospective case series of 11 adults (average age of 39.9 ± 11.3 years) with intralobar pulmonary sequestration Gompelmann and Colleagues (4) have found that the predominant symptoms were dry cough, recurrent pulmonary infections and hemoptysis. However, no patient had chest pain or flank pain as in our case.

Again in the retrospective study of Sun and Colleagues (10) of 72 adults (mean age 36.6 ± 11.8 years) patients with pulmonary sequestration (mostly intralobar) the most common clinical symptoms were cough and sputum followed by hemoptysis, whereas chest pain was present in $\sim 20\%$ of the cases.

Computed tomography (CT) of the chest usually is diagnostic. However, the CT diagnostic criteria for the diagnosis of pulmonary sequestration are not standardized. The typical appearance is that of a large pulmonary mass with or without cystic areas inside. Occasionally it appears as a mass of fluid or air-filled microcysts, or as a large cavitating lesion with an air-fluid level (3). The detection of the aberrant systemic artery, distinct from the bronchial arteries, together with the lung abnormalities described above are the keys CT features of the pulmonary sequestration in

adult patients (8). CT or MR angiography are used to demonstrate aberrant vessels (4).

In some cases, however, the diagnosis is made only during the surgical resection of the lesion as underlined in the retrospective case series of Gompelmann and Colleagues [4] where in 8 of 11 patients the intralobar pulmonary sequestration was diagnosed before the surgery by using these imaging techniques, whereas in the remaining 3 patients the diagnosis was made intraoperatively with the histological examination.

The treatment of the intralobar pulmonary sequestration is mainly represented by the surgical resection of the lesion, whenever the general clinical conditions of the patient permit this. The timing of the surgery is usually dictated by the patient's symptoms. Surgery is indicated in patients with recurrent lung infections or hemoptysis, but remains controversial in the asymptomatic cases. The surgical resection of the lesion should be performed after the resolution of the lung infection. Resection of an intralobar pulmonary sequestration almost always involves a lobectomy. The crucial part of the surgical procedure is the identification and control of the aberrant vascular supply (3,8-10).

Conclusions

We report here an unusual case of intralobar pulmonary sequestration in 27-year-old man mimicking a kidney stone, a community-acquired pneumonia and a lung cancer. CT scan with intravenous contrast has permitted the preoperative diagnosis that has been then confirmed on the surgically resected lesion.

Conflicts of Interest: There is no potential conflict of interest, and the authors have nothing to disclose. This work was not supported by any grant.

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