

Case Report

Pulmonary sequestration: a case report and literature review

Xin Qian^{1,2*}, Yuhui Sun^{3*}, Dan Liu^{1,2}, Xiaofeng Wu^{1,2}, Zhenxiong Wang⁴, Yijun Tang^{1,2}

Departments of ¹Respiratory Medicine, ³Emergency Medicine, ⁴Medical Image Center, Taihe Hospital Affiliated to Hubei University of Medicine, Shiyan, China; ²Institute of Respiratory Medicine, Taihe Hospital Affiliated to Hubei University of Medicine, Shiyan, China. *Equal contributors.

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Abstract: Purpose: Pulmonary sequestration (PS) is a rare congenital lung malformation. It is characterized by an abnormal mass of dysplastic lung tissue supplied by an anomalous systemic artery and separated from normal bronchopulmonary tree. Misdiagnosis and inadequate treatment can lead to recurrent pneumonia and fatal hemoptysis. Methods: We report a 45 years female was diagnosed PS, and performed a brief review about the clinical features, diagnostic strategies, and management options of the PS. Results: Her remarkable symptoms were cough and hemoptysis, the contrast-enhanced computed tomography of the chest revealed a multiloculated cystic solid mass filled with low density lesions and a feeding artery from the descending abdominal aorta to the cystic solid mass was visualized, then the patient suffered a right lower-lobe resection, and the surgery and pathological examination all supported the diagnosis of intralobar sequestration. Conclusions: Symptomatic patients of the pulmonary sequestration should be treated by surgery to avoid the risk of death due to massive hemoptysis.

Keywords: Pulmonary sequestration, intralobar sequestration, extralobar sequestration, hemoptysis, aberrant artery, surgery

Introduction

Pulmonary sequestration (PS) is a rare congenital lung malformation characterized by an abnormal mass of dysplastic lung tissue supplied by an anomalous systemic artery and did not have a connection with the tracheobronchial tree [1]. There are two distinct forms: intralobar, as in our patient, and extralobar, depending on whether it has independent pleura [1]. The diagnosis may be easily missed in adults, as many of the symptoms and the computed tomogram (CT) manifestation overlap with other pulmonary processes especially lung cancer. This article presents a case of intralobar pulmonary sequestration that presented during adulthood, and a brief review of the clinical features, diagnostic strategies, and management options of the PS was performed.

Case report

A 45 years female patient was admitted to Taihe hospital (Shiyan, China) with a chief com-

plaint of a recurrent cough about one year, worsened and accompanied hemoptysis and right-sided chest pain about one week. Her symptoms persisted despite antibiotic therapy. Physical examination revealed a healthy appearing, well nutrition. Vital signs, including oxygen saturation on ambient air, were all within normal limits. Chest auscultation was remarkable for rough breath sound in the right lower lung field but without rhonchi or rales. A CT scan showed a clear round high density mass (5.8 cm × 3.6 cm). Laboratory tests including complete blood count, liver and renal function tests, electrolytes, urinalysis, erythrocyte sedimentation rate, tuberculosis antibody tests, carcinoembryonic antigen and neuron specific enolase were negative. Flexible bronchoscope showed a normal tracheal lumen without visible tracheal tumor. Bronchoalveolar lavage fluid revealed no tumor cells and acid-fast bacillus. Subsequent contrast-enhanced computed tomography of the chest revealed a multiloculated cystic solid mass filled with low density lesions and a feed-

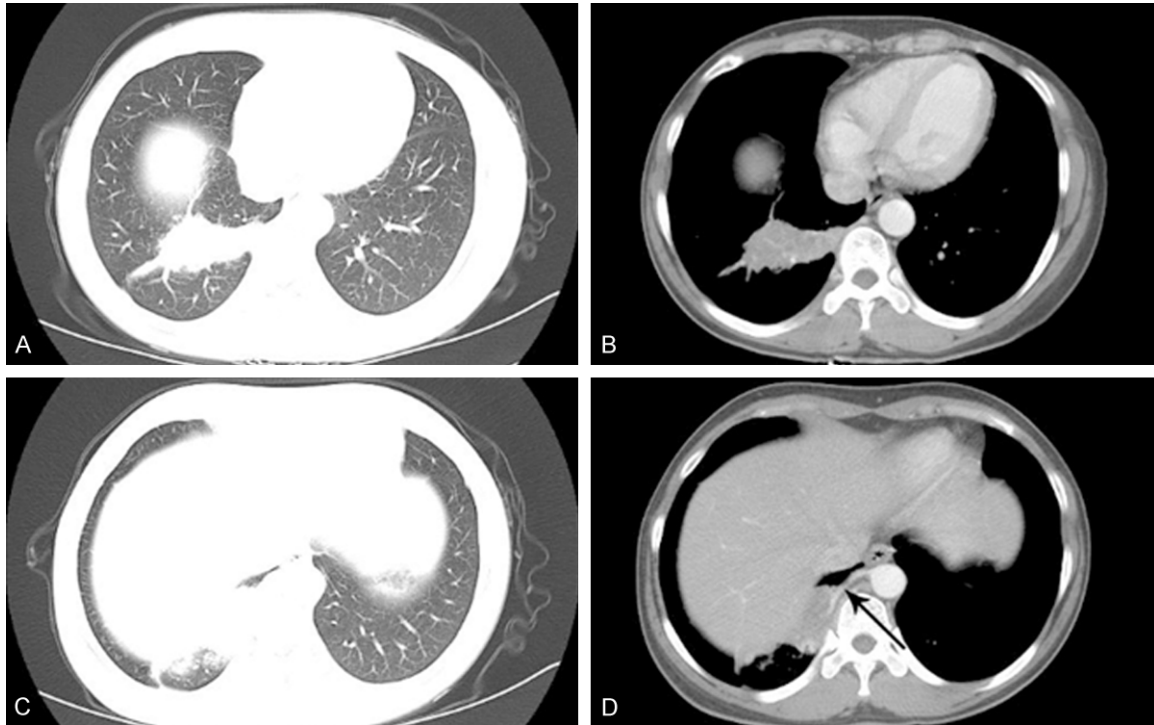


Figure 1. The typical contrast enhanced thoracic computed tomography images of a 45-year-old female with intralobar sequestration. A, C. A round high density mass was seen in lung window; B. A multiloculated cystic solid mass filled with low density lesions were visualized in mediastinal window; D. A feeding vessel coursed from the abdominal aorta directly into the cystic solid mass (arrow).

ing artery from the descending abdominal aorta to this cystic solid mass was visualized (**Figure 1**). Given the history, imaging features characteristic of intralobar sequestration, and ongoing symptoms, the patient was referred to right lower-lobe resection. Pathology of the resected specimen showed inflammatory lung parenchyma, with multiple cystic spaces filled with purulent secretions and with the cystic bronchioles contained neutrophils. The pathological examination revealed an intralobar sequestration.

Discussion

Pulmonary sequestration is a rare congenital abnormality characterized by an abnormal mass of dysplastic lung tissue supplied by an anomalous systemic artery and did not have a connection with the tracheobronchial tree, representing 0.15%-6.4% of all congenital pulmonary malformations [1, 2]. There are two subtypes: intralobar and extralobar, depending on whether it has independent pleura. An intralobar sequestration shares the visceral pleura of the adjacent lung, while the extralobar type is

contained within its own visceral pleura, separate from the involved lung [1]. Intralobar sequestration (ILS) is more common (75% of cases) than the extralobar sequestration (ELS), roughly in a 3:1 ratio [2]. In China, ILS is constituted approximately 83.95%-92.5% of the PS cases [3, 4]. Multiple theories of the pathogenesis of pulmonary sequestration have been proposed, but all remain controversial. The most widely accepted hypothesis is that it results from formation of an accessory lung bud inferior to the normal lung buds during development and gets its blood supply from foregut vascular [5]. If the accessory lung bud develops before formation of the pleura, both the normal and sequestered lung tissue are covered by the same pleura, resulting in ILS; If the accessory lung bud develops after formation of the pleura, the sequestered lung tissue forms its own pleural covering, resulting in ELS [5].

Localization of pulmonary sequestration

Almost 97% of PS cases were located in the lower lobe, and PS in the left lower lobe is two

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to three times more common than that in the right lower lobe [2-4]. As in our patient, the sequestration was located in the right lower lobe (**Figure 1**).

Aberrant arterial supply of pulmonary sequestration

In most cases, PS has a single feeding artery, occasionally there are multiple systemic arteries supplying the PS. Arterial supply of PS mainly originated from thoracic aorta (46.1-86.1%) and abdominal aorta (6.9%-31.6%), the other feeding aorta include intercostal artery, diaphragmatic artery, aortic arch, subclavian artery, anastomotic artery, pulmonary artery, left gastric artery, coronary artery, arteria lienalis, celiac trunk and renal artery [2-4, 6]. The key to establishing the diagnosis of the PS lies in identifying the aberrant arterial supply. In our patient, the enhanced CT scan demonstrated that the feeding vessel coursing from the abdominal aorta directly into the cystic solid mass (**Figure 1D**).

Symptoms of pulmonary sequestration

The two types of sequestration are associated with very different clinical features. ELS, often discovered on prenatal or neonatal ultrasound or magnetic resonance imaging (MRI), is frequently associated with other congenital anomalies, including heart defects, pulmonary hypoplasia, foregut duplication cysts, vertebral anomalies, or diaphragmatic abnormalities [5]. Infants with ELS may be asymptomatic or may suffer from respiratory distress owing to lung hypoplasia or mass effect. ELS rarely becomes infected, as its pleural investment prevents contact with inhaled air [5]. Rarely, adult patients become symptomatic because of hemothorax, pulmonary infarction, infection, or hemoptysis [5]. In contrast, ILS usually present during childhood, but up to half present after the age of 20. The most patients are asymptomatic and carry the abnormality for years, only to be diagnosed during a routine physical examination or with recurrent bacterial pneumonia in the affected lower lobe [1]. Symptoms are often nonspecific, the most common symptom of PS was cough or expectoration, fever, hemoptysis and chest pain [2-4]. As in our patient, the remarkable symptoms were cough and hemoptysis.

Imaging of PS

Ultrasound plays an important role in the prenatal diagnosis and follow-up of PS as it can identify the origin of the aberrant vessel feeding the lung mass [7, 8]. However, ultrasonographic findings are nonspecific and has limited value in the antenatal, neonatal periods and in adults, and the differential diagnosis is wide in these findings [4]. MRI can demonstrate signal voids of the aberrant feeding vessel on T2-weighted imaging, and magnetic resonance angiography can demonstrate the systemic blood supply to the sequestered lung [7]. ELS most commonly manifests as a well-defined pyramidal, oval, or round mass in the pleural space near the posteromedial aspect of the ipsilateral hemidiaphragm and can be seen on fetal sonography as early as 16 weeks gestation [5, 8]. Because of its separate pleural investment from aerated lung, ELS almost never contains air. On CT or MRI, ELS appears as a well-defined mass of uniform soft-tissue attenuation, there is often a single anomalous artery arising from the thoracic or abdominal aorta [5].

ILS has diverse imaging features. Plain chest radiograph is usually nonspecific, showing an ill-defined consolidation that mimics pneumonia, or shows a solitary soft tissue mass or nodule, or a cystic or multicystic lesion [4, 5]. On CT, the most and common manifests of ILS as a mass lesion, cystic lesion that may be filled with fluid, air, or both and cavitary lesion [3-5]. CT scan with intravenous contrast and preferably CT angiography (CTA) is the method of choice for identifying the arterial supply.

Current treatment

Surgical treatment should be considered for most patients, especially for symptomatic patients or when cancer cannot be excluded [4, 5]. Asymptomatic ILS is often treated by surgery to avoid the risk of death in adulthood due to massive hemoptysis [1, 5]. Surgery usually involves lobar resection via standard thoracotomy or video assisted thoracic surgery (VATS). Since Wan et al. first described VATS lobectomy for treating pulmonary sequestration in 2002 [9], the use of VATS for pulmonary sequestration resection has been used widely. Percutaneous endovascular embolization of the feeding systemic vessel has been per-

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formed for definitive treatment [10, 11]. However, the major concerns of the embolization include possible incomplete occlusion of vascular supply, subsequent evolution of the sequestered tissue and possible recurrence of symptoms [11].

The management of ELS is more controversial, it is known that these lesions can remain asymptomatic throughout the patient's life but the complications may develop [12]. So strict observation is required. When symptoms develop, however, patients should be referred for definitive surgery.

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Disclosure of conflict of interest

None.

Address correspondence to: Dr. Yijun Tang, Department of Respiratory Medicine, Institute of Respiratory Medicine, Taihe Hospital Affiliated to Hubei University of Medicine, 32 Renmin South Road, Shiyan 442000, Hubei Province, China. Tel: (+86) 13886827301; Fax: (+86) 7198801821; E-mail: tangyijun_799@163.com

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