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ABSTRACT

Introduction
Pulmonary sequestration represents a rare congenital anomaly of the lower respiratory tract. Interlobar sequestration is the most common form and usually presents in the left hemithorax during the second decade of life or earlier. Its blood supply usually arises from systemic circulation. Surgical intervention is the treatment of choice in patients with PS.

Case Report
We describe a late presentation of an intralobar sequestration in a 59 years old male patient. It presented with recurrent attack of hemoptysis. Radiological investigations revealed pulmonary sequestration. Two unusual features were found in this case: being in the right side and having double blood supply from both systemic and pulmonary circulation.

Conclusion
In this case report, we described a case of unusual presentation of ILS in old patients, right sided with double blood supply from both pulmonary artery and abdominal aorta.

Keywords: Intralobar pulmonary sequestration, Lung anomalies, Pulmonary angiography
TITLE: Intralobar pulmonary sequestration: Rare presentation in adult

INTRODUCTION
Pulmonary sequestration (PS) is a rare congenital malformation of the lower respiratory tract. It defined as a nonfunctioning primitive solid or cystic aberrant lung mass which has no connection with tracheobronchial tree and has systemic arterial supply [1].

Intralobar sequestration (ILS) is overall the most common form. In 50–60% of cases the diagnosis of ILS is made during the second decade or younger. Later presentation is rare [2].

Surgical excision is the treatment of choice of PS. The preoperative assessment includes besides confirmation of the diagnosis the proper identification of the vascular supply of the sequestration [3].

We describe intralobar pulmonary sequestration in a male patient 59 years old, who presented with recurrent attacks of hemoptysis. The discovered ILS was abnormally supplied with a double blood supply.

CASE REPORT
Male patient aged 59 years old, current smoker presented with recurrent attacks of hemoptysis. The first attack started 12 years ago, after that he experienced recurrent attacks of blood-tinged sputum (average 3 attacks per year). The patient was first investigated in the primary health care facility where chest X-ray (Figure 1) was done and was interpreted as a case of bronchitis despite the obvious right Para cardiac opacity. The patient was treated with supportive measures. Then the patient was admitted to a general hospital due to another attack of hemoptysis with syncope. Thorax CT scan with contrast revealed right lower lobe opacity with calcification (Figure 2). Due to the suspicious of malignancy, ultrasonographic-guided aspiration was done twice and the pathological examination revealed fibrous tissue with dilated vascular spaces. Finally, the patient was referred to our chest medicine department for further evaluation and bronchoscopy. After revision of his history and chest radiology, the PS was suspected besides other benign lung lesions and CT angiography was ordered before any further interventions. It revealed
evidence of right intralobular pulmonary sequestration. It has dual arterial supply from both right pulmonary artery and multiple branches from aorta arise, the venous drainage into right pulmonary vein (Figure 2 and 3). Finally, the patient was transferred to the surgical department for surgical intervention, but the patient refused to perform any interventions.

DISCUSSION

Pulmonary sequestration was first described by Pryce in 1946. Its name was derived from the Latin verb ‘sequestrate’ which means to set apart. It can be defined as a developmental lung disease with non-functioning pulmonary tissue, which has no communication with the bronchial tree and receive a systemic blood flow [4].

PS is classified into 3 subtypes: 1) Intralobar pulmonary sequestration (located within normal lung lobe and has its own visceral pleura), 2) Extra lobar pulmonary (located outside the lung lobe and has its visceral pleura), and 3) Bronchopulmonary- foregut malformation which is a rare variant of sequestration and is connected to the gastrointestinal tract [5].

The incidence of the PS is rare and represented about 0.15 to 6.4% of all congenital pulmonary malformations [6]. Generally, ILS is the commonest form of pulmonary sequestration. It represents about 75 to 90% with no sex differentiation [5].

The embryologic basis of the pulmonary sequestration remained unclear. Many possibilities had been suggested. The first referred to very early abnormality in the development during lung bud formation. Another theory suggested mechanical separation of a portion of the developing lung due to compression or traction by aberrant vascular structure or inadequate pulmonary blood flow. However, the mechanical theory cannot fully explain all types of the pulmonary sequestration especially BPFM [7]. The third theory suggested that ILS may be an acquired rather than developmental lesion [8]. Recently, researchers found that the abnormal vascular development in the pulmonary arterial blood supply can lead to retention and proliferation of the nascent systemic capillary network [1].

ILS usually presents in patients less than 20 years of age in 50% of the patients and rarely found in patients older than 50. Lower lobe above the diaphragm is typically the mostly affected area and left side is common in 55-60% of the patients [2, 4].
Here, we describe presentation of ILS in male patient 59 years old which is not only atypical due to the old age but also due to the atypical site in the right lower lobe. The time of ILS presentation is variable. It usually present in late childhood or adolescence. It presented with recurrent lower respiratory tract infection. Hemoptysis and chest pain were also reported. It may be also asymptomatic and discovered in routine chest radiology in 15.5% of the patients with ILS [6]. In rare cases, heart failure occurs due to high flow through the anomalous artery [1].

The radiological assessment aimed not only to confirm the sequestration diagnosis, but also to evaluate its vascular supply for further management. Besides the chest radiography, ultrasonography, computer tomography and magnetic resonance imaging, the relatively recent introduced angiography represent the diagnostic tool of choice to define the vascular supply prior to any surgical intervention [3]. Single feeding blood supply was detected in 71% of the patients. It arises from the thoracic aorta in 36% [6].

The intralobar sequestration can be classified according to Pryce et al into: type (1) presence of aberrant artery without sequestration, type (2) the aberrant artery supply the sequestration as well as the adjacent normal lung and type (3) the aberrant supply only the sequestration [2].

The main management of the pulmonary sequestration in symptomatic patient is surgical resection for curative purposes. Proper identification and ligation of the feeding vessels is crucial. While in asymptomatic patients with ILS the surgery is also recommended to prevent recurrent infections and the unfavorable cardiac influence caused by the existing aortopulmonary shunt. Recently published data has introduced the emerging successful role of video-assisted thoracoscopic surgery (VATS) for pulmonary sequestration resection despite the difficulties in surgical dissection due to recurrent inflammation and fibrosis owing to the recurrent infections [9]. Arterial embolization of the feeding vessels has been also reported [10].

**CONCLUSION**

The interesting points in our case include the late presentation of the patient (59 years old), unusual right-sided ILS and the double blood supply from both pulmonary artery and abdominal aorta. Finally, we should emphasize the importance of proper
interpretation of the chest radiology by the primary medical care providers as a standard tool of chest examination for early and adequate diagnosis of catastrophic hidden chest diseases.

**CONFLICT OF INTEREST**

The authors have any financial interest or any conflict of interest.

**AUTHOR'S CONTRIBUTIONS**

Ahmed Ehab
- Group 1 - Substantial contributions to conception and design, acquisition and analysis of data
- Group 2 - Drafting the article, revising it critical for important intellectual content
- Group 3 - Final approval of the version to be published

Marwa Ghanem
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- Group 2 - Drafting the article, Critical revision of the article
- Group 3 - Final approval of the version to be published
REFERENCES


FIGURE LEGENDS

Figure 1: Chest X ray: right paracardiac shadow.

Figure 2: (A) - CT chest with contrast reveled right lower opacity (B) - Chest angiography with appearance of the aberrant blood supply of PS.

Figure 3: CT angiography (Volume rendering) show the arterial blood supply of PS from both right pulmonary artery and multiple branches from aorta.

FIGURES

Figure 1: Chest X ray: right paracardiac shadow.
Figure 2: (A) - CT chest with contrast revealed right lower opacity (B) - Chest angiography with appearance of the aberrant blood supply of PS.

Figure 3: CT angiography (Volume rendering) show the arterial blood supply of PS from both right pulmonary artery and multiple branches from aorta.