EXTRALOBAR PULMONARY SEQUESTRATION : A CASE REPORT AND REVIEW OF LITERATURE OF A RARE CONGENITAL DISEASE

By
Yasser Shaban Mubarak;
Department of Cardiothoracic Surgery,
El-Minia Faculty of Medicine

ABSTRACT:

Extralobar pulmonary sequestration [ELS] is a rare congenital pulmonary malformation compared with its counterpart, intralobar pulmonary sequestration [ILS], and occurs less in females.[1]

INTRODUCTION:

Pulmonary sequestration (PS) is non-functional lung tissue , which does not connected to tracheobronchial tree, and is fed by systemic arterial circulation. It is a rare congenital anomaly. It accounts for 0.15% - 6% of all congenital pulmonary malformation. The term pulmonary sequestration was first used in 1946 by Price. Pulmonary sequestration is divided into two groups (75%-85% Intralobar and 15%-25% Extralobar) depending on the presence of pleura that covers abnormal lung tissue. There are other differentiation factors; age , localization , arterial supply , venous drainage , symptoms, and other anomalies. Both are frequently in left hemithorax and lower lobe. Males are more likely to have pulmonary sequestration than women . Extralobar sequestration occurs 4 times more in men than women. Extralobar sequestration is usually located outside the visceral pleura . The arterial supply is found to be through 95% systemic (thoracic or abdominal aorta) and 5% pulmonary artery. Venous drainage is through 75% in systemic vein (IVC, portal, azygos) and 25% in pulmonary veins. Other congenital anomalies commonly coexist with ELS (65%) are diaphragmatic hernia , pulmonary hypoplasia , congenital cystic adenamatoid malformation, and bronchogenic cyst. Majority of ELS are asymptomatic and discovered incidentally. Other symptoms are recurrent chest infection, growth retardation, and dyspnea[2].

Case Report

A 6 years old male presented with recurrent chest infection. Chest infection did not resolved clinical nor radiological by chest x-ray. Computerized tomography of the chest showed large area of destroyed lung within the medial aspect of left hemithorax (Fig.1).

Excision of left extralobar sequestration (sequestrectomy) was done through left posterolateral thoracotomy. During excision of ELS, careful attention to ligate arterial supply to avoid uncontrollable bleeding and venous drainage to avoid pulmonary infarction. Its arterial blood supply was branches from descending thoracic aorta and its venous drainage was to inferior hemiazygous vein. The sequestered lobe was separate from the normal lung and entirely covered by its own visceral pleura. There were no communications with tracheobronchial tree. After excision of ELS and securing vascular supply, the left lower lobe was healthy and inflated without residual space. Also, there were no other congenital anomalies. Routine

closure of thoracotomy and insertion of 2 intercostal chest tubes (ICT).The resected specimen was sent to pathological examination (Fig. 2). Histological analysis revealed a dilated cystic wall lined with respiratory epithelium, with mucous glands and cartilage, there was no evidence of malignancy, diagnosed an ELS.
Postoperative course passed uneventful. The patient discharged on 5th postoperative day (POD) after removal ICT.

DISCUSSION

Pulmonary sequestration is rare lung anomaly which had many theories of origin[2]. Pulmonary sequestrations are developmental anomalies defined as presence of pulmonary tissue in the thorax without bronchial communications. The blood supply to sequestrations may be from systemic or pulmonary vessels. The usual clinical presentation is recurrent localized chest infection resistant to medical treatment. Increasing the size of sequestration due to secretions causes atelectasis of the neighboring normal lung. Sometimes, in case of ELS, recurrent infection can establish a connection between sequestration and adjoining bronchus by erosion[3].

Posterolateral thoracotomy is an appropriate approach for excision of ELS with careful ligation of vascular structures[2]. Surgery to treat ELS is usually not difficult if neither inflammation nor fibrosis, from previous recurrent infections, are present. However, in some cases, no aberrant vessel can be identified preoperatively. When aberrant vessels cannot be identified preoperatively, it is necessary to pay close attention to unexpected disruption of the vessels. Preoperative confirmation of aberrant vessels by using imaging methods makes the surgery easy and safe in patients with pulmonary sequestration. Multidetector CT provides useful information without the necessity of invasive methods such as angiography in patients with congenital lung anomalies[4]. Complete excision of the sequestered lung using a video-assisted thoracoscopic approach is our hope in future.

REFERENCES:

[1] Yiping W, Liru C, Jianjun X, Dongliang Y. Intrapericardial Extralobar Pulmonary Sequestration in Adult. Asian J Cardiothorac. 2012; 3:20 -40.

[2] Atilla P, Necati C, Muzaffer M, Abdulaziz K, Alper C, Adnan S, Atilla G. The rare congenital anomaly of pulmonary sequestration experience and review of literature. Indian J Thorac Cardiovasc Surg. 2010;26: 251- 254.

[3] Birla R, Madhu A, Prabakar v, Nischal R. Intralobar pulmonary sequestration with bronchoesophageal fistula and cystic adenamatoid malformation in an adult. Indian J Thorac Cardiovasc Surg. 2012;28:31-32.

[4] Toshihiro O, Mantaro k, Tomoko T, Masako N, Junji M, Hidehiko Y. Unique extralobar sequestration with atypical location and aberrant vessels. Ann Thorac Surg. 2010;90: 1711-1712.

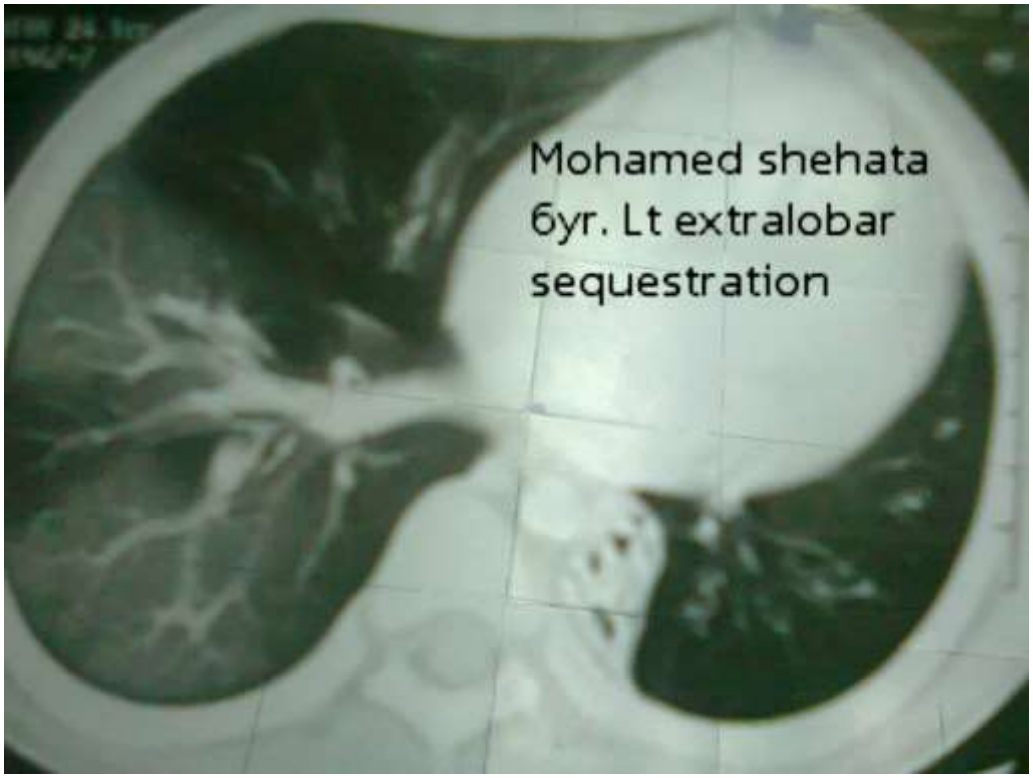

Fig.(1) Axial view, CT chest with contrast showed ELS adjacent to left lower lobe.

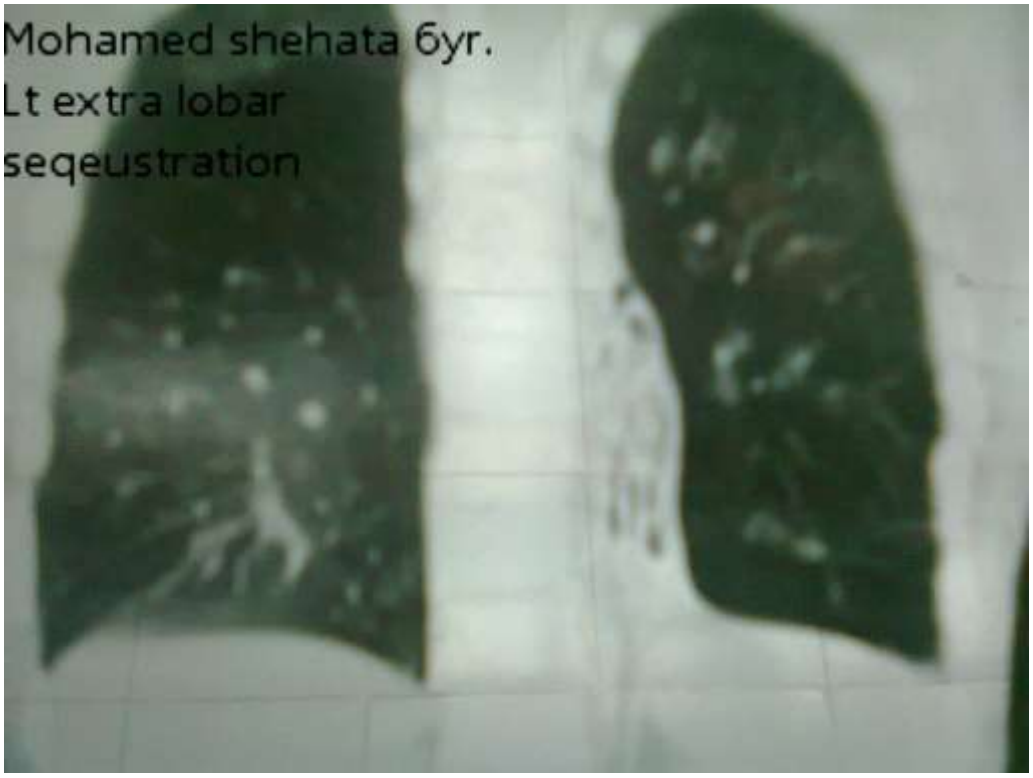

Fig. (1) Coronal view of CT chest showed ELS in left , basal , and medial hemithorax.

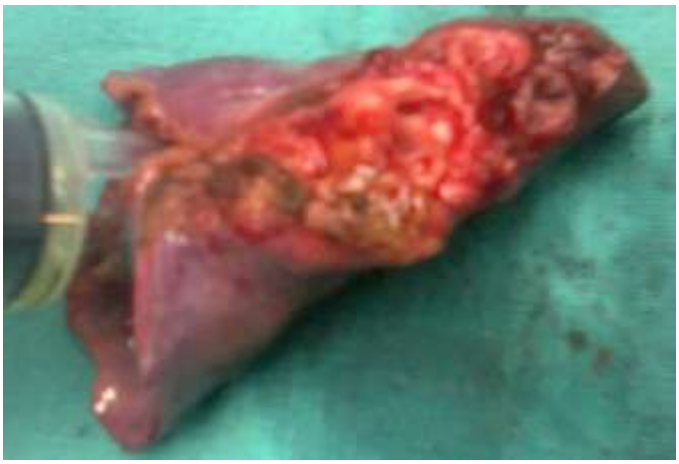

Fig.(2)The resected specimen of ELS showed that no tracheobronchial connections , aberrant blood vessels , and its own visceral pleural .