Intralobar Bronchopulmonary Sequestration with Large Aberrant Vessel Presenting as Recurrent Pneumonias

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ABSTRACT

Bronchopulmonary sequestration is a rare congenital malformation of the lower respiratory tract comprising of a non-functioning lung tissue mass that lacks normal communication with the tracheobronchial tree. The diagnosis may be easily missed as many of the symptoms of bronchopulmonary sequestration overlap with that of other pulmonary diseases. Bronchopulmonary sequestration can be complicated by recurrent infections, hemorrhage and malignant transformation and, therefore, needs to be timely diagnosed and resected to decrease both morbidity and mortality. A high degree of suspicion in the differential diagnosis helps diagnose the positive cases. The parenchymal abnormalities associated with bronchopulmonary sequestration are best visualized using computed tomography, although their appearance is variable. We report the case of a 14 years old boy with intralobar bronchopulmonary sequestration with the sole manifestation of recurrent pneumonias.

Key Words: Bronchopulmonary sequestration. Aberrant vessel. Intralobar. Pneumonia.

INTRODUCTION

Bronchopulmonary sequestration is a rare congenital lung malformation characterized by an abnormal segment of lung tissue supplied by an anomalous systemic artery. The diagnosis may be easily missed as many of the symptoms of bronchopulmonary sequestration overlap with that of other pulmonary diseases. Two forms of pulmonary sequestration are described depending on whether or not the abnormal lung tissue possesses its own pleural covering i.e. intralobar and extralobar sequestration. Extralobar sequestration has its own sac that is anatomically separated from the rest of the lung. Surgical resection provides definitive cure and is usually reserved for patients with symptoms.

The present report describes this rare condition with recurrent pneumonias.

CASE REPORT

A 14 years old young boy of South-East Asian ethnicity presented with excessive cough productive of purulent sputum and high grade fever for the last one week. He described having intermittent episodes of productive cough for last one year and received multiple courses of antibiotics with presumed diagnosis of recurrent pneumonias. He had no other significant past, personal, drug, family or occupational history.

On examination, he was febrile (101°F) with respiratory rate of 19/minute and oxygen saturation of 95% at room air. He had of lean physique with no evidence of cyanosis, pallor, clubbing or lymphadenopathy. His respiratory examination revealed dull percussion note with decreased air entry and decreased vocal resonance in left lower half of chest. Rest of the systemic examination was unremarkable.

His laboratory tests revealed leukocytosis of 17,700/mm³ with 87% neutrophils and markedly raised erythrocyte sedimentation rate of 100 mm in the first hour. His liver function tests, renal function tests, serum electrolytes and electrocardiogram were all normal. His chest radiograph showed a non-homogeneous opacity occupying left middle and lower lung fields (Figure 1A).

A wide spectrum of diagnosis related to radiologically evident lung mass was considered (Table I). A preliminary diagnosis of left sided pneumonia was made. His sputum was sent for Gram staining and acid
Contrast enhanced CT chest; transverse section showing intralobar bronchopulmonary sequestration with air fluid level (white arrow head). C: Contrast enhanced CT chest; coronal section showing intralobar bronchopulmonary sequestration with aberrant vascular supply (white arrow head). D: CT chest; coronal section showing sequestration with lack of communication to tracheobronchial tree (white arrow head); the patent airway (tracheobronchial tree) is indicated by black arrow heads. E: Gross view of the successfully removed lung mass (sequestration). F: Postoperative chest X-ray (PA view) showing clearance of the opacity in the left lung field.

fast bacillus. Sputum specimen was also taken for culture and sensitivity testing. The patient was started on broad spectrum antibiotics. His sputum testing did not reveal any abnormality and his antibiotics were continued for two weeks.

There was a significant symptomatic improvement with the above treatment but the repeat chest X-ray remained unchanged. At this point, the diagnosis was reviewed. A contrast enhanced computed tomography of the chest was organized which revealed a large, well defined, left lower lobe multi-loculated cystic cum solid mass with multiple enhancing internal septae and an air fluid level (Figure 1B). The mass had an anomalous arterial supply from the descending thoracic aorta and venous drainage into the left pulmonary vein (Figure 1C). There was no communication of the mass with the tracheobronchial system (Figure 1D). All of these findings suggested the diagnosis of an intralobar bronchopulmonary sequestration with superimposed infection.

In view of the above radiological findings and repeated episodes of lower respiratory tract infections, the patient was referred to the thoracic surgeon for resection of the left lower lobe bronchopulmonary sequestration.

At thoracotomy, the large anomalous artery originating from the lateral wall of descending thoracic aorta was identified, carefully isolated and ligated. The venous drainage into the left pulmonary vein was also identified and transected. The mass was removed successfully and sent for histopathological examination (Figure 1E).

Gross histopathological examination revealed a 390 gram lung mass measuring 18 x 9 x 6 cm. There were multiple necrotic and hemorrhagic areas with the solid part being 6 x 5 x 3 cm and the cystic cavity measuring 4 x 4 x 3 cm, all filled with blood. The microscopic examination showed infiltration of the tissues with lymphocytes, plasma cells, hemosiderin laden macrophages and eosinophils. There were microabscesses formation and destruction of normal lung parenchyma along with thick and tortuous vessels and dilated bronchioles. The above confirmed the diagnosis of intralobar sequestration with acute and chronic inflammatory changes.

Patient’s subsequent postoperative course was uneventful. His chest radiograph showed clearance of the left sided lung opacity and he made good recovery without no subsequent chest infections on follow-up (Figure 1F).

**DISCUSSION**

The term sequestration, first used in the medical literature by Pryce in 1946, originates from the Latin word *sequestare*, which means to separate. Hence bronchopulmonary sequestration is the term given to a region of lung parenchyma that is partially or completely separated from the bronchopulmonary tree of the lung proper. Its blood supply is usually from an aberrant artery arising from the aorta or one of its branches. The venous drainage is into the left atrium, although abnormal connections to vena cava,azygous vein or right atrium may occur as well. Bronchopulmonary sequestration is estimated to comprise 0.15 - 6.4% of all congenital pulmonary malformations, making it an extremely rare disorder. In several reports, even tertiary care hospitals diagnose less than one case per year.

Sequestrations are classified anatomically. Intralobar pulmonary sequestration (also known as intrapulmonary sequestration), in which the lesion is located within the parenchyma of a normal lung lobe and lacks its own visceral pleura, Extralobar pulmonary sequestration (also known as extra-pulmonary sequestration), in which the mass is located outside the normal lung tissue and has its own visceral pleura.

The aim of roentgenological imaging for sequestration is to define and characterize the lesion, rule out alternative pathology, and clearly identify the source of the aberrant arterial supply of the sequestration to facilitate operative management. The parenchymal abnormalities associated with bronchopulmonary sequestration are best visualized using computed tomography, although their appearance is variable. The most common appearance is a solid mass that may be homogeneous or heterogeneous, sometimes with cystic changes, as in our patient. Less frequent findings include a large cavitory lesion with an air-fluid level, a collection of many small cystic lesions containing air or fluid, as in this patient, or a well-defined cystic mass. Though intralobar sequestration has many characteristic clinical and radiologic features, many mimics have been described in the literature; thus making it a diagnostic challenge for the physicians. Therefore, careful evaluation must be done to rule out alternative pathologies and thus ensure a correct diagnosis.
Bronchopulmonary sequestration is treated by surgical excision, which is curative and is associated with minimal morbidity. Complete excision of intralobal sequestration usually requires lobectomy or segmental resection and is best suited for symptomatic patients. Even asymptomatic patients with intralobal sequestration should have elective resection in order to prevent recurrent infections.\textsuperscript{3,8,9}

It is concluded that recurrent chest infections in a healthy individual may be an indicator of underlying bronchopulmonary sequestration. A correct, timely diagnosis and treatment can effectively cure the disease with minimal morbidity. Clinicians should be familiar of all the aspects of this rare entity in order to pick-up this otherwise challenging diagnosis correctly. It is further concluded that non-resolving consolidation on a chest radiograph should never be ignored lest the diagnosis of a curable disease like sequestration is missed and left untreated.

REFERENCES


