Intralobar Pulmonary Sequestration Masquerading as Lung Abscess: A Rare Paediatric Presentation

Dr. Shilpi Sahu, Dr. Reeta Dhar, Dr. Reenal Patel

1Associate Professor, Department of Pathology, MGM Medical College, Navi Mumbai, Maharashtra, India
2Professor & Head, Department of Pathology, MGM Medical College, Navi Mumbai, Maharashtra, India
3MBBS, Department of Pathology, MGM Medical College, Navi Mumbai, Maharashtra, India

Abstract: Pulmonary sequestration refers to lack of normal airway connection and aberrant vascular supply, and it may be either intralobar or extralobar. Bronchial atresia involving the lower lobes, especially the left lower lobe, is often associated with aberrant systemic arterial connection, forming an “intralobar sequestration” in which a region of parenchyma is functionally isolated from the remainder of the lung. Pulmonary sequestration is an uncommon condition that accounts for 0.15-6.4% of all pulmonary malformations and is typically diagnosed in childhood. We present a rare case of intralobar pulmonary sequestration in a 25 months old female involving the right lower lobe with secondary infection.

Keywords: Intralobar pulmonary sequestration (IPS), right lower lobe, lobectomy, histopathology.

1. Introduction

Pulmonary sequestration is a disease; accounting for only approximately 0.15-6.4% of all congenital pulmonary malformations. Pulmonary sequestration is an uncommon congenital usually cystic mass of non-functioning primitive lung tissue that does not communicate with the tracheobronchial tree or the pulmonary arteries. It is supplied by an anomalous artery arising from aorta & venous drainage is via the azygos system, pulmonary veins or the inferior vena cava. The two forms of pulmonary sequestration include, intrapulmonary which is surrounded by normal lung tissue & extra pulmonary which has its own pleural investment. Sixty percent of these lesions are diagnosed within the first decade of life. Symptoms may vary and typically are related to chronic respiratory infection although sequestrations may be discovered incidentally on radiographic studies. We describe a case where early surgical intervention and histopathological diagnosis aided in preventing the complications of pulmonary sequestration.

2. Case Report

A twenty five months-old-female patient was admitted with complaints of chronic cough, fever and shortness of breath on and off since one year. No significant past history was noted. On general examination, she was afebrile, pulse was 90 beats/min, respiratory rate was 32 cycles/min and blood pressure was 90/70mm Hg. She was pale and responsive with diminished activity in mild respiratory distress. Positive physical findings were limited to the right side of the chest, which revealed slightly decreased expansion, increased vocal and tactile fremitus, dull note on percussion and rales on auscultation. A clinical suspicion of Lung abscess was made. Routine laboratory studies were within normal range except for mild thrombocytosis.

HRCT was performed to the patient to visualize the parenchyma of the lung, and it revealed multiple cystic areas showing small fluid levels with adjacent fibrotic scarring at apical segment of right lower lobe. Differential diagnoses of Post-Infectious, Pneumatocoele and Cystic Bronchiectasis with secondary infection were made. The HRCT findings and the history of recurrent pulmonary infections suggested a possibility of pulmonary anomaly.

Thus, right lower lobectomy via a posterolateral thoracotomy was performed. Histopathological examination revealed following features.

a) Gross
We received a specimen of right lower lobe measuring 7x4x3cm. Pleural surface was unremarkable. Cut section showed multiple cystic spaces, largest measuring 4cm in diameter with multiple small cystic spaces and lower part showed solid areas.

b) Microscopy
Hematoxylin and Eosin stained sections studied following histologic features- Mucocele, Obstructive parenchymal changes, Airspace enlargement, abundant mucus with chronic inflammatory infiltrates, pulmonary hyperplasia and cystic maldevelopment.
B. Mucocele consisting of a dilated large airway filled with mucus and mucin-filled macrophages.

C. Presence of microcystic maldevelopment and chronic inflammatory infiltrate.

D. Giant cell inflammatory infiltrate within the distended airway.

3. Discussion

The term sequestration, which is derived from the Latin verb *sequestare*, meaning "to set apart", was first used by Pryce in 1946, when he described a case of ILS and aroused clinical interest in the entity. Pulmonary sequestration, as first described by Rektorzik in 1861, a malformation comprised of dysplastic lung tissue with no normal communication with the tracheobronchial tree and with an anomalous systemic arterial supply. The etiology of this defect is thought to be congenital. There are two types of pulmonary sequestration: intralobar and extralobar. Intralobar Pulmonary sequestration is three to six times more common than the extralobar type. In intralobar pulmonary sequestration, the pulmonary tissue is isolated from the normal lung tissue; however, the pleural covering remains contiguous with that of lung.

The term “intralobar bronchopulmonary sequestration of the lung” has been used to describe the association of a congenital cyst of the lower lobe of a lung with an anomalous artery, which arises from the aorta usually at the level of the diaphragm. The original report by Huber in 1777 went unnoticed until Harris described the condition in 1940. Haight warned that failure to recognize the possibility of the entity may result in the surgeon's cutting the large vessel inadvertently. However, in our case; the patient is doing well post-operatively.

Achieving a diagnosis of pulmonary sequestration can range in difficulty depending upon the type of anomaly and presenting symptoms. Computed tomography will typically suffice in most cases with some debate still held over the need for angiography. In our experience, however the chest CT scan could delineate the anatomic features notable for operative planning but did not give a definite diagnosis.

Most frequently supported theory of how a sequestration arises is that an accessory lung bud develops from the ventral aspect of the primitive gut. The pleuripotent tissue from this additional lung bud migrates in a caudal direction with the normally developing lung. It receives its blood supply from vessels that connect to the aorta & that cover the primitive gut. The attachments to the aorta remain to term resulting in the systemic arterial supply of the sequestration. Early embryonic development of the accessory lung bud results in formation of sequestration within normal lung tissue & encasement within the same pleural covering which results in an intrapulmonary variant.

4. Conclusion

Angiography is the gold standard for the diagnosis of pulmonary sequestration but since it was not done in this case, the best standard possible for confirming pulmonary sequestration was histopathological examination which ruled out the remaining differential diagnoses. In this case report, the patient had involvement of right lower lobe of lung and the lesion was diagnosed at an early age by histopathological studies which makes this case rare and unique. So, early suspicion, surgical resection and confirmation of the pulmonary sequestrations should be considered as the standard of care for such patients.

We conclude in our rare case report that given the potential for recurrent infections and life-threatening hemoptysis in these patients, operative resection of affected lung and early confirmation of the suspected sequestration cases by histopathological examination is the preferred approach.

References

[1] Philip T. Cagle, Color atlas and text of pulmonary pathology, Section-24, Paediatric pulmonary pathology, Chapter 133-Bronchial atresia and Intralobar Sequestration (Lippincott Williams & Wilkins) 597-599.


