

Hybrid Lesion: Extralobar Sequestration with Cystic Adenomatoid Malformation – Misdiagnosed as Pulmonary Tuberculosis

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ABSTRACT

We report a rare case of bronchopulmonary extralobar sequestration in a 9-year girl mimicking pulmonary tuberculosis. Patient had recurrent chest infections and two episodes of massive hemoptysis. Preoperative x-ray chest showed an opacity in left lower lobe and computed tomography-aortogram showed that the lesion had systemic arterial supply from celiac trunk. Diagnosis of pulmonary sequestration was made and left lateral muscle sparing thoracotomy was planned. Intraoperatively, extralobar sequestration was found with dual systemic blood supply. Histopathology confirmed it to be a hybrid lesion.

Key Words: *Bronchopulmonary sequestration. Pulmonary sequestration. Extralobar. Intralobar. Hybrid.*

INTRODUCTION

Pulmonary sequestrations are rare abnormalities which constitute up to 6.4% of all congenital pulmonary malformations.¹ Pulmonary sequestration is defined as "the presence of a mass of abnormal pulmonary tissue that does not communicate with the tracheobronchial tree through a normally located bronchus and is supplied by an anomalous systemic artery".^{1,2} Extralobar sequestration is differentiated from intralobar, as it has its own separate visceral pleura. It is speculated that sequestration results from a supernumerary lung bud that is separate from normal tracheobronchial tree.³

We describe a rare case of an extralobar pulmonary sequestration coexistent with cystic pulmonary adenomatoid malformation (CPAM) and having dual systemic blood supply in a 9-year girl, who was initially misdiagnosed as having pulmonary tuberculosis.

CASE REPORT

A 9-year girl was referred to Pediatric Surgery Department, Mayo Hospital, after her computed tomography (CT) scan thorax revealed a multi cystic lesion. She had complaints of cough, fever and tachypnea, off and on, for last 2 years. Despite having negative mantoux test and no acid fast bacilli (AFB) in her sputum, she was started on anti-tuberculosis therapy (ATT) by a pediatrician, as she was resident of

Bajaur Agency and there was history of contact with her mother who had suffered from pulmonary tuberculosis and her chest X-ray (CXR) showed an opacification in left lower hemithorax. When her symptoms did not improve even after a 9-month course of ATT and she had two episodes of massive hemoptysis, she was referred to Lahore, where initially she was considered as having multidrug resistance (MDR) tuberculosis. Further investigations revealed congenital cystic lesion of lung on CT scan thorax and she was referred to pediatric surgery department. On admission in June 2017, her general and systemic examinations were unremarkable except reduced breath sounds in left lower zone of chest. Her mother had no history of antenatal checkups and she was delivered at home. She had normal developmental milestones other than recurrent chest infections.

Her full blood count and biochemical tests were normal. CT scan thorax revealed a well-defined area of multi-cystic lesion in the posterior segment of left lower lobe, measuring 4x4 cm and showing no post-contrast enhancement or air fluid level (Figure 1A). Due to history of massive hemoptysis, CT aortogram was done and it revealed systemic arterial supply of the lesion from celiac trunk and venous drainage to the left inferior pulmonary vein (Figure 1B). Diagnosis of bronchopulmonary sequestration (BPS) was made. Pre-operatively, bronchoscopy revealed normal endobronchial appearance; and lavage did not reveal any AFB. Left lateral muscle sparing thoracotomy was planned after informed consent and blood arrangement. Intraoperatively, the lesion was separate from normal lung and had a dual blood supply from celiac trunk and thoracic aorta (Figure 1C). It was excised (Figure 2A). Postoperative recovery was uneventful and chest drain was removed on second postoperative day. Patient was

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Figure 1A: CT scan chest showing a well-defined, multi-cystic lesion in the posterior segment of left lower lobe.



Figure 1B: CT aortogram showing the blood supply of the lesion.



Figure 1C: Peroperatively, the lesion was separate from normal lung and had a dual supply from coeliac trunk and thoracic aorta.

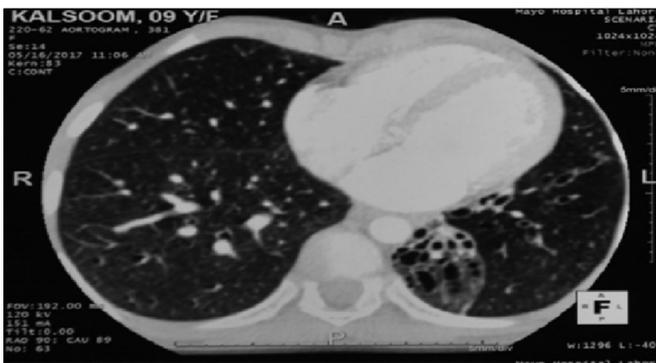


Figure 2A: Gross appearance of the lesion.

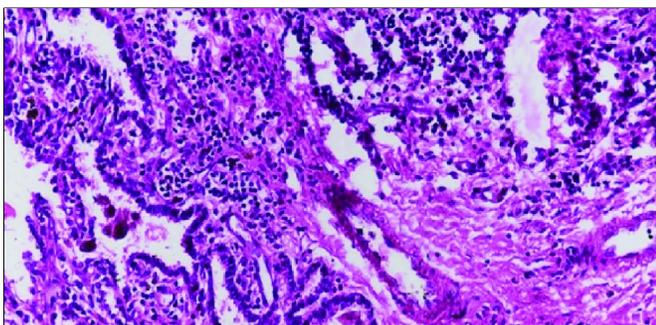


Figure 2B: Microscopy of the lesion.

discharged on day 5; and did not complain of any restriction of activities in follow-up. Histopathology revealed unevenly dilated air spaces, lined by cuboidal epithelium suggesting CPAM and no epithelioid cells (Figure 2B). Ethical approval was sought from the Institute and informed consent was taken from her father to publish this case report.

DISCUSSION

Rektorzik described BPS in 1861, but Pryce was the first to use the term of sequestration and described its variants: extralobar and intralobar sequestration.⁴

BPS has three main features: no communication with normal tracheobronchial tree, systemic blood supply, and bronchial anomaly with parenchymal dysgenesis.⁵⁻⁸

Extralobar sequestration commonly occurs in male infants on left side.^{1,3} We report a case of extralobar sequestration in female child on left side between left lower lobe and diaphragm. Other reported rare locations are suprarenal, intradiaphragmatic, intrapericardial and mediastinal.^{1,5,6}

It is associated with diaphragmatic hernia, broncho-pulmonary malformations, foregut duplication, vertebral anomalies and rarely, like in this case, with CPAM. When extralobar sequestration is associated with CPAM, it is called a hybrid lesion and it may point to a common embryological origin. Nowadays, more cases are being reported and this has led to changes in classification of cystic lung lesions.^{8,9} Conran was first to report this association to be around 50%.²

Stocker classified CPAM into five types from type 0 to IV. Type 0 (dysplasia) leads to *in utero* death. Type I (macro cystic) and Type II (micro cystic) present in newborns with respiratory distress and rarely, late with infection. Type II may have associated anomalies like cardiovascular anomalies, congenital diaphragmatic hernia, extralobar sequestration and renal anomalies. Type III is mainly adenomatoid mass and may present with severe respiratory distress or still birth. Type IV has peripheral cysts, presents in later age and does not have associated anomalies, but may later on undergo plumopulmonary blastema formation. Our case was most likely type IV.¹⁰

Extralobar sequestration may be complicated by tuberculosis, but it is rare that it may be misdiagnosed as a case of pulmonary tuberculosis.¹¹ Extralobar sequestration is rarely associated with massive hemoptysis.¹⁰ Main cause of hemoptysis in our patient was associated bronchiectasis.

In this case, histopathology report confirmed that this patient was misdiagnosed as pulmonary tuberculosis.

The gold standard to evaluate the blood supply in BPS is the arteriography. It is commonly from thoracic aorta but can be from abdominal aorta, pulmonary artery, intercostal or subclavian arteries. Dual arterial supply is noted in 5% of cases,^{5,6} and in our case it was from

celiac trunk and thoracic aorta. It is important to note that CT aortogram may not demarcate all the branches of arterial supply to these lesions and high index of suspicion is needed at the time of surgery.

Although embolization has been reported to treat these lesions, yet higher recurrence rates after embolization have been noted. In symptomatic patients; surgery is the only treatment option. Thoracoscopic removal has gained popularity widely; but in a few centres, thoracotomy is still being used as an alternate.¹²

It is important to have a high suspicion of these lesions in children presenting with recurrent chest infections; and it is necessary to rule out these lesions before starting ATT.

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