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Case report

Pulmonary sequestration presenting as a massive haemoptysis in adult: A case report

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ABSTRACT

Introduction: Pulmonary sequestration is a rare congenital lung malformation characterized by non-functioning mass of pulmonary tissue that lacks normal communication with bronchial tree and receives one or more aberrant systemic arterial supply. It usually presents in children with recurrent chest infections. It is uncommon in adult and remains asymptomatic or present with recurrent chest infections, rarely with more severe symptoms like a massive haemoptysis as in seen in our case.

Case presentation: A 22-year-old male presented with a chief complain of multiple episodes of massive haemoptysis. After evaluation with chest x-ray, CT chest and angiogram, he was diagnosed to have intralobar pulmonary sequestration. He underwent successful thoracotomy and left lower lobectomy. He is asymptomatic after one year of follow up.

Discussion: Pulmonary sequestration is rare in adult and can present with various symptoms like chest pain, cough, sputum production, recurrent infection and rarely haemoptysis. Intralobar sequestration of left lower lobe is the most common as seen in our case. The diagnosis can be made by Computed Tomogram chest with angiogram. Surgical intervention is the definitive curative treatment. Post-operative outcome is excellent with early patient satisfaction and promising outcome in long term follow-up.

Conclusion: Bronchopulmonary sequestration can rarely present in adults presenting with massive haemoptysis. CT chest is the best modality for diagnosis. Early surgical intervention is definitive treatment with good long term outcome.

1. Introduction

Pulmonary sequestration (PS), a rare congenital lung malformation, is symbolized by non-functioning lung tissue with no apparent communication with tracheobronchial tree and one or more aberrant systemic arterial supply. It represents 0.51–6.4% of all congenital lung malformations. [1,2]. No single embryonic hypothesis is clearly adapted for it, and no chromosomal abnormality has yet been identified. Majority regard the disease part of an anomaly spectrum, with normal vessels supplying anomalous pulmonary tissue at one end, and deviant vessels supplying normal lung tissue at the other [3,4].

Here, we report a case of intralobar pulmonary sequestration in an adult presented to us with a chief complain of massive haemoptysis. He was successfully managed with surgical intervention. This case has been reported in accordance to SCARE criteria [5].

2. Case presentation

A-22-year-old male presented to emergency department with history of multiple episodes of massive haemoptysis within 24 h. The patient had no recent history of cough, fever, shortness of breath, chest pain. He had history of recurrent lower respiratory tract infections until five years of age. He also received anti-tubercular treatment at the one year of age. There were no other associated significant co-morbidities or trauma. He denied smoking, alcohol and illicit drug use. Family history was unsupportive of his diagnosis. His vital signs were within normal limits. On physical examination there was occasional added sounds on left lower zone, rest of the systemic examinations were within normal limits. His initial laboratory investigations were within normal limits except for haemoglobin (9 g/dl). Chest X-ray revealed ill-defined density in left lower zone (Fig. 1). Contrast enhanced computed tomogram (CECT) chest with angiogram showed consolidation and haemorrhage in left

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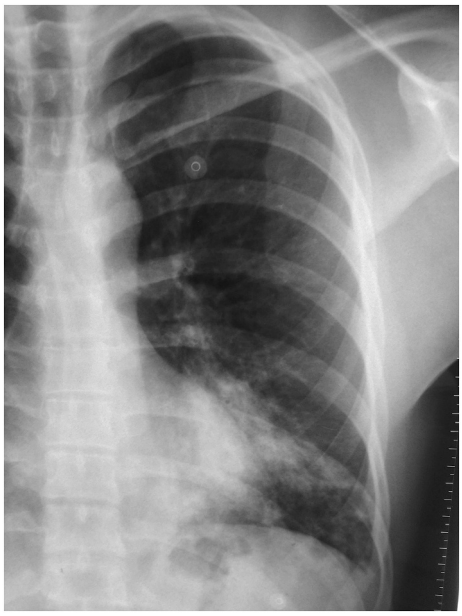

Fig. 1. Chest x-ray showing haziness in left lower zone.

lower lobe posterior segment (Fig. 2) with large aberrant systemic arterial supply from distal descending thoracic aorta to the lower lobe suggestive of PS (Fig. 3). The venous drainage was to the pulmonary veins. A diagnosis of intralobar PS was established and planned for left lower lobectomy. Initially he refused for any surgical intervention and later again presented after one month with similar history of massive haemoptysis. This time he consented for surgical intervention. The thoracotomy was done with standard posterolateral incision. After mobilization of inferior pulmonary ligament, a large 8 mm arterial supply to the left lower lobe from distal descending thoracic aorta was identified (Fig. 4). The arterial supply was doubly ligated, transfixed and cut. The left lower lobectomy was completed in the usual fashion. There was no postoperative complication and was discharged on fourth post-operative day in satisfactory condition. Histopathological examination showed extensive areas of haemorrhage and congestion along with few areas having fibrosis and lymphoid follicles and areas showing thrombus and clot in bronchioles lumina. He is asymptomatic after one year of follow up.

3. Discussion

PS in adult is rare and almost half of them remain asymptomatic. In symptomatic cases recurrent infection, cough and chest pain are common presentations. [2] It can present with haemoptysis, however massive haemoptysis as in our case is not frequently reported.

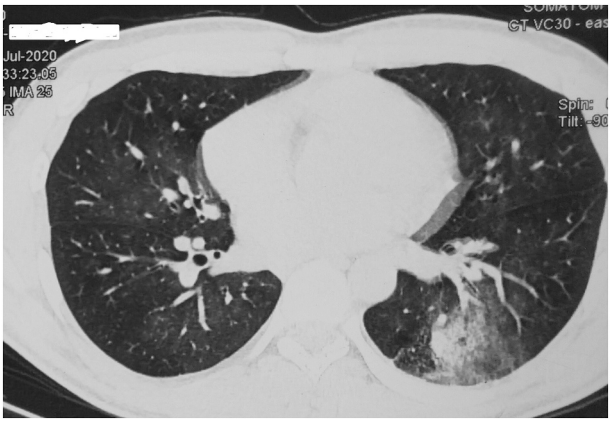

Fig. 2. CECT chest showing haemorrhage and consolidation in left lower lobe posterior segment.

PS is divided into two forms: intralobar sequestration (ILS) and extralobar sequestration (ELS).ELS is enclosed within its own pleural membrane maintaining complete anatomical separation from adjacent lung tissue, whereas in ILS the lesion lies within the pleural membrane of the normal lung. The arterial blood supply to the sequestered lung is by aberrant systemic arteries mostly from the thoracic or abdominal aorta. These arteries may arise above or below the diaphragm, and are frequently small and variable. The venous drainage of ELS is predominantly to systemic veins whereas in ILS it is predominantly to pulmonary veins. [6] ILS is more common and mostly involves the left hemithorax. ELS has male predominance and is commonly associated with other congenital anomalies like diaphragmatic hernias, other lung anomalies like dysplasia, pericardial defects, cardiac malformations and communications with the foregut. ILS has no sexual predominance and usually cause problem after two years of age [7–9]. There was no history of any congenital anomalies in our patient consistent with ILS.

The clinical presentation of PS is often with nonspecific symptoms and diagnosisis difficult without radiological investigation for vasculature and pulmonary parenchyma. Chest x-ray in ILS shows variable parenchymal changes like well-defined triangular shaped mass pointing medially, air fluid level due to bronchial communication, area of hyperlucency, air bronchogram and consolidation suggesting pneumonia [10].CECT chest with angiogram best demonstrates parenchymal abnormalities and aberrant vascular supply. Magnetic resonance imaging, too can detect the parenchymal changes and aberrant vascular supply, thus avoiding contrast [11].When non-invasive imaging fails conventional angiogram still has a place in diagnosis of PS, especially if coil embolisation is planned.

Surgical resection is the current standard and curative treatment for PS even in asymptomatic cases. The documentation and control of the aberrant artery branch, above or below the diaphragm, are essential for counteracting haemorrhage. Postoperative results are characteristically excellent [8].

A comparison review of previously published series from different countries [10,12–15], validates left lower lobe predominance of ILS among the adult population, and more than 80% of patients presenting with respiratory symptoms. Without significant postoperative complications, surgical resection of the sequestered lesion was achieved in approximately 90% of cases. As in line with aforementioned findings, our patient was a male of 22 years presenting with massive haemoptysis. The sequestration was intralobar and was successfully managed with surgical resection providing a promising outcome on long term.

In a case series of 29 patients with pulmonary sequestration by Tashtoush B et al., mean age of presentation in adults was 42 with 52% being male. Seventeen percent of them presented with haemoptysis and 86% was diagnosed with CT angiogram. Majority of cases involved left lower lobe [16]. Our patient was male who presented with massive haemoptysis and ILS was present in his left lower lobe which was diagnosed with CT chest and CT angiogram.

Endovascular coil embolization has recently been reported as alternative treatment modality for pulmonary sequestration in both paediatrics and adult. [17]. However, retained nonaerated pulmonary parenchyma can become nidus for infection and abscess formation as a result of necrosis after coil embolization. Another feared complication is inadvertent coil embolization of spinal artery. A large scale comparison of surgical and endovascular intervention for PS in adults has not been done. Our patient was successfully treated with surgical intervention which is regarded the gold standard for the pulmonary sequestration.

The other alternative approach to the management in this case can be minimal invasive video assisted lobectomy. However in view of very large feeding artery and limited resources in our set up, we opted for open surgery.

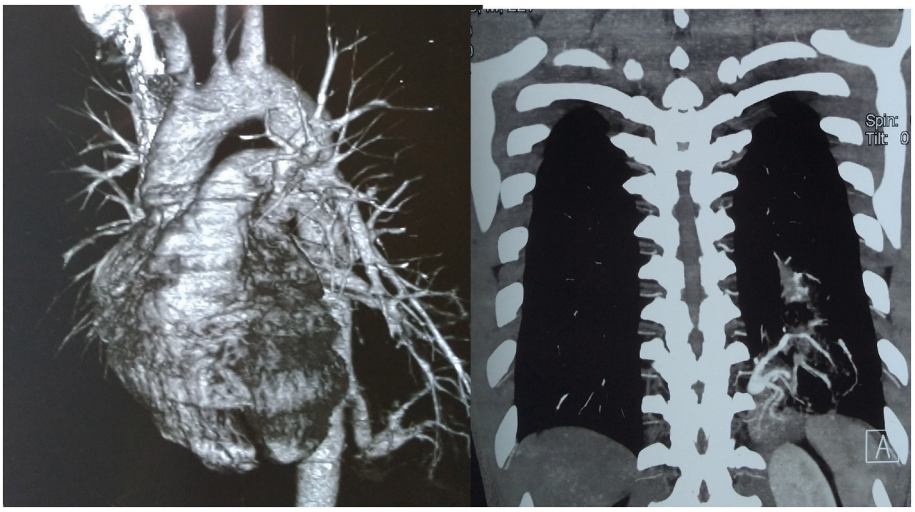

Fig. 3. CT angiogram showing aberrant arterial supply (arrow) to lower lobe from distal descending thoracic aorta.

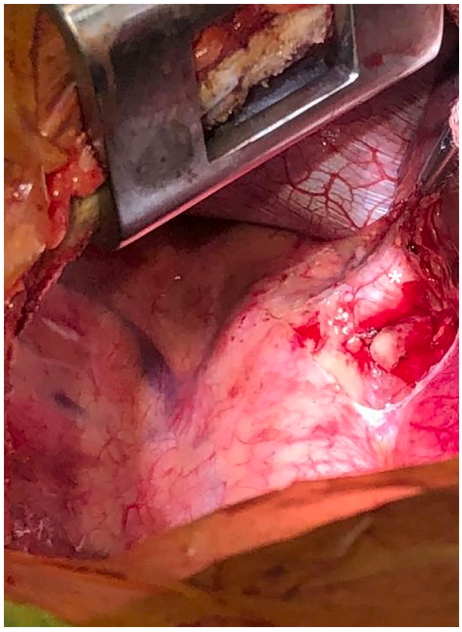

Fig. 4. Intraoperative image of aberrant arterial supply (asterisk) from distal descending thoracic aorta.

4. Conclusion

Pulmonary sequestration can present with massive haemoptysis in adult and should be kept in differential diagnosis. CT chest is the best modality for the diagnosis and surgical resection is curative with good long term outcomes.

Ethical approval

The ethical approval has been exempted from our institution for the case report.

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Registration of research studies

Not applicable.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

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CRediT authorship contribution statement

Lokesh Shekher Jaiswal (LSJ), Durga Neupane(DN) = Study concept, Data collection, and surgical therapy for the patient.

Lokesh Shekher Jaiswal(LSJ), Durga Neupane(DN) = Writing- original draft preparation.

Lokesh Shekher Jaiswal (LSJ), Durga Neupane(DN) = Editing and writing.

Lokesh Shekher Jaiswal (LSJ), Durga Neupane(DN) = senior author and manuscript reviewer.

Both the authors read and approved the final manuscript.

Declaration of competing interest

None.

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None.

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