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CASE REPORT

Intralobar Pulmonary Sequestration: GMC Jammu Experience

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Abstract

Pulmonary sequestration is a rare entity comprising a small portion of all congenital pulmonary malformations which is characterized by aberrant formation of nonfunctional lung tissue that has no communication with the bronchial tree and receives systemic arterial blood supply. We reviewed three cases, who presented to CTVS Department at GMC Jammu each of which presented with cough, expectoration and hemoptysis and were subsequently diagnosed as intralobar pulmonary sequestration (ILS). The aim of this case series is to increase awareness about the condition and to review criteria of its definitive diagnosis and subsequent treatment

Key Words

Bronchopulmonary sequestration, Intralobar sequestration (ILS), Congenital pulmonary malformations

Introduction

Bronchopulmonary sequestration is a rare lung abnormality which is, benign in nature anatomically it is classified into intralobar and extralobar sequestration; intralobar sequestration (ILS) is more common and accounts for 75% of sequestrations and 0.15% to 1.7% of all congenital lung abnormalities (1). ILS is characterized by the presence of nonfunctional parenchymal lung tissue, receiving systemic arterial blood supply. It lacks normal communication with tracheobronchial tree. Failure to diagnose and treat this condition can lead to recurrent pneumonia and fatal hemoptysis (2).

Case Reports

We present three patients (two males and one female) who were diagnosed as having intralobar pulmonary sequestration. All of them were between age group of 15-20 and presented with history of recurrent chest infections. Cough expectoration and intermittent fever was present in all the patients. One out of three was having hemoptysis as presenting symptom. Computed tomography (CT) of chest revealed a multiloculated mass in the right lower lobe of the lung in all three patients (*Figure 1&2*). None of the patients underwent a selective angiogram

Operative Findings

All three patients underwent standard posterolateral thoracotomy and right lower lobectomy. Two out of three had classical features suggestive of intralobar pulmonary sequestration (*Figure 3*) whereas in the third case who had presented with hemoptysis had a cystic swelling with compression atelectasis of the surrounding

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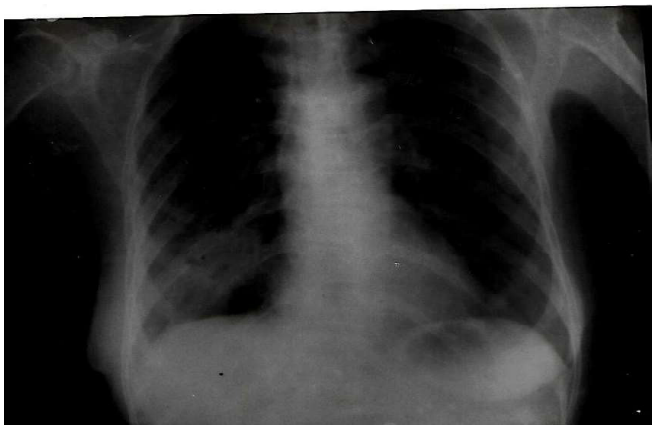

Figure 1: Chest X-ray of Patient Showing Lesion in the Right Lower Zone

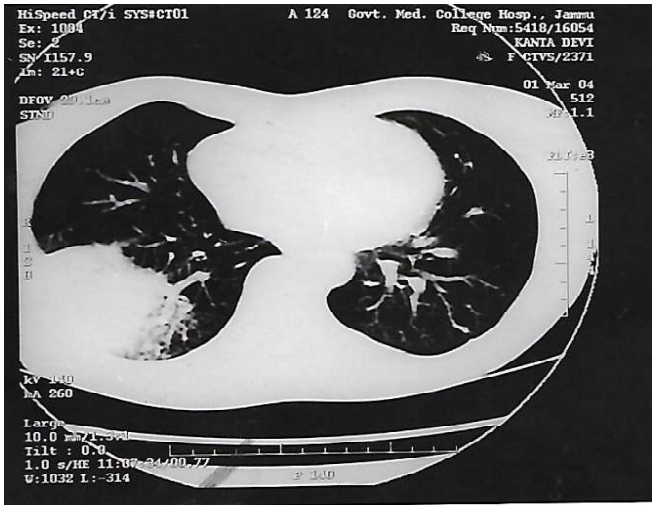

Figure 2: CT Scan Chest Showing the Sequestration in the Right Lower Lung

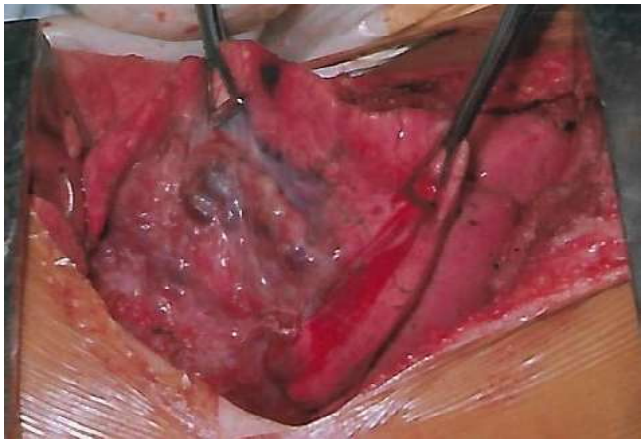

Figure 3: Operative Picture Showing the Sequestrated Segment

lung tissue along with bleeding vessels around the sequestrated segment. Two out of three patients had aberrant blood supply from descending aorta and was lodged around the inferior pulmonary ligament and third case the blood supply was in form of vascular plexus arising from thoracic aorta.

Post operatively, two patients had uneventful recovery whereas one patient had prolonged air leak for four weeks and required prolonged chest tube drainage. Two patient's resected specimens showed tubercular granulomas and they were put on antitubercular treatment and responded well as per follow-up.

Discussion

Intralobar pulmonary sequestration described by Rektorzik in 1861 is a relatively rare congenital anomaly which can present with an incidental pulmonary lesion on imaging and can be otherwise asymptomatic (3). There are two types of pulmonary sequestrations: Intralobar sequestration (ILS), which is surrounded by normal lung tissue; and extralobar sequestration (ELS), which has its own pleural investment. Intrapulmonary sequestration is four times more common than the extralobar type. It presents late in childhood or adolescence with recurrent pulmonary infection while extralobar sequestration more commonly presents in newborn with respiratory distress, cyanosis, and infection (4,5).

Many believe that ILS is not a congenital but acquired entity. Bronchial obstruction, pneumonia, pulmonary artery occlusion, pleuritis, and parasitisation of pulmonary ligament have been proposed to explain the acquired origin theory of the development of ILS (6).

Most of the ILSs are located in the medial and posterior basal segments of the left lung. Overall, 98% occur in the lower lobes (7). Bilateral involvement is uncommon. However, in our case all the lesions were found in the right lung and lower lobes. The ILS is contained within the visceral pleura with venous drainage into the pulmonary veins. The ELS is separated from the normal lung and is outside the visceral pleura with venous drainage into a systemic vein. The arterial supply in both ILS and ELS is from thoracic aorta in 75% of cases whereas in rest it is from abdominal aorta and its branches (8).

For achieving a diagnosis of pulmonary sequestration, chest X ray and computed tomography will typically suffice in most adult cases (9). In our experience, the chest CT scan was clearly sufficient to make the diagnosis plus delineate the anatomic features notable for operative planning. However, angiography remains the gold

standard for identifying pulmonary sequestration as it confirms the anatomy, delineates the arterial supply, and the venous drainage (10).

Definitive treatment involves resection of the affected lung segment as has been done in the all three cases. Open thoracotomy remains a safe approach which gives wide access for accurate preoperative identification of the arterial blood supply and great care should be given to securing the systemic arterial branches at the time of operation, which can be quite large in diameter. Complete thoracoscopic resection has been reported with low morbidity and mortality (11,12).

Two out of three cases in our series were found to have evidence of Mycobacterium tuberculosis in the specimen and were treated with antitubercular treatment. Yatera et al. (13) established the presence of homogenates of Mycobacterium tuberculosis in the resected specimen of sequestration and subsequently treated the patient with antitubercular drugs and 6 years later her clinical status remained excellent.

Conclusion

Correct and prompt suspicion of non-resolving radiological lesions is needed to provide correct and early diagnosis of bronchopulmonary sequestrations. Early surgical resection should continue to be the standard of care in both adolescent and adult patients with this disease process. Excised specimen can reveal a surprise as seen in our cases.

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Conflicts of Interest

There are no conflicts of interest.

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