Intralobar Pulmonary Sequestration in an Adult: An Unusual Case Report with Brief Review of Literature

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Abstract: Intralobar pulmonary sequestration is a rare congenital abnormality of the lower respiratory tract which becomes symptomatic early in life. Clinical presentation may vary from asymptomatic state with incidental diagnosis while imaging to severe catastrophic hemoptyisis. Here we report a case of intralobar pulmonary sequestration in a male of 36 years who continued to have hemoptyisis for fourteen years was misdiagnosed as a case of pulmonary tuberculosis and received complete anti tubercular therapy in a peripheral hospital.

Keywords: Intralobar, extralobar, pulmonary sequestration.

I. Introduction

Pulmonary sequestration is a relatively rare entity comprising of 0.15 -6.4% of all congenital pulmonary malformations.¹ It was first identified by Pryce in 1946 as a clinical and pathological entity and related it embryologically to accessory lung. Since then several case reports and reviews have appeared in literature with presentations in childhood as well as late in third decade. ² Cases mostly present with repeated chest infections, pleural effusion or minor hemoptyisis but cases with severe and fatal hemoptyisis have also been reported. Contrast CT Scan with angiography helps to make the diagnosis and delineate anatomical features for operation planning. Definitive treatment involves resection of the affected lung segment.

II. Case Report

31 years Hindu male, Businessman by profession, presented to us with complaints of coughing out of blood on and off at intervals of few months for nearly fourteen years. It was associated with intermittent cough and scanty expectoration, at times with blood tinged sputum, but hemoptyisis was never massive. There was also history of intermittent low grade fever, but no history of weight loss, loss of appetite or night sweats. He was neither a known case of rheumatic heart disease nor there was any history of dyspnea on exertion, orthopnea or paroxysmal nocturnal dyspnea. He was non-diabetic, non-hypertensive. There was history of contact with pulmonary TB as his father was a known case of PTB . There was no significant occupational exposure. He was married with two children. He was treated with antibiotics from time to time along with anti-tubecular drugs and had completed full course of Anti tubecular therapy twice ,once 5years back and again one year back though there was no report of sputum being positive for Mycobacterium Tuberculosis.

On clinical examination he was of average body build with pulse rate 87/min, BP 110/70 mm Hg and mild pallor. There was no icterus, cyanosis or clubbing. Jugular venous pressure was not raised and no dependant edema was present. No significant lymphadenopathy was found.

On examination of respiratory system coarse crepitations heard over left infra-axillary and infrascapular area. No other significant findings noted. Examination of CVS, abdomen and CNS revealed no significant findings. With this clinical presentation and examination findings provisional diagnosis of non resolving left lower lobe pneumonia , left lower lobe bronchiectasis , left lower lobe pulmonary TB or malignancy was thought of including rare presence of intralobar pulmonary sequestration.

On investigation, Hb. was 9.5gm%. TLC - 12,000/cmm, ESR 40mm, blood urea 30mg/dl, serum creatinine 1mg/dl, FPG 90mg/dl. X Ray of chest showed a homogenous opacity in the left lower lobe adjacent to cardiac shadow. CT angiography of chest showed the opacity to have a separate blood supply from abdominal aorta confirming the diagnosis of intralobar pulmonary sequestration. The case was treated conservatively with antibiotics and referred to cardiothoracic surgeons where he was planned for surgical resection of the sequestrated segment.

III. Discussion

Pulmonary sequestration is a rare congenital malformation characterized by a mass of non-functioning lung tissue separated from the normal bronchopulmonary tree and vascularised by an aberrant systemic artery.³ Rokitansky and Rektorzic described the first case in 1861. They proposed that the sequestration was due to a separated but normally developed lung fraction.⁴ Since then, several theories were put forward to explain the
genesis of this anomaly. Pryce et al were the first to use the term sequestration and the traction theory put forward by Pryce is considered to be the most accurate explanation for this pathology. They identified the association between congenital isolation and vascularisation by an aberrant artery of the systemic circulation mostly from thoracic aorta, celiac trunk or from intercostal arteries. Pulmonary sequestrations are classified into two types- intralobar sequestration and extralobar sequestration. Intralobar sequestration is an abnormal region within the normal pulmonary parenchyma without its own pleural covering. Extralobar sequestration corresponds to a true accessory lung with its own pleural covering. Intralobar pulmonary sequestration is three to six times more common than the extralobar type. Overall the sequestrations are located more in left lung (65%) than right lung. Intralobar lung sequestrations are mostly present in lower lobes (97.75% of cases) and there is no specific sex distribution with respect either to the occurrence of the anomaly or to its location. The symptoms in Intralobar sequestration typically occur during early childhood but around 50% cases the diagnosis is made after the age twenty. Commonest symptoms are cough, expectoration, recurrent attacks of pneumonia. Pleural effusion occurs in 4% of cases. Minor hemoptysis is common. More severe hemoptysis, bleeding into pleural space, oesophagus or into the sequestration itself has been reported. In 15% cases the anomaly is asymptomatic and normal longevity has been reported. The arterial supply to intralobar sequestration is usually via the pulmonary veins. Extralobar sequestrations most commonly located between the diaphragm and the lower lobe and the distribution and location are identical in both sexes. The arterial supply to extralobar sequestrations is most commonly from descending thoracic aorta followed by abdominal aorta. In contrast to intralobar pulmonary sequestration extralobar pulmonary sequestration is usually diagnosed in infancy secondary to respiratory distress or failure to thrive.

Intralobar sequestrations are not associated with other cardio-pulmonary anomalies but extralobar sequestrations may be found in association with cardiac or more frequently diaphragmatic anomalies in 50% of cases. There have been few reports of malignant neoplasms being involved in or near sequestrated segments. Recent data has shown that CA 19-9 and CA 125 can also be elevated in cases of benign lung disease including pulmonary sequestration. The typical radiographic appearance of a bronchopulmonary sequestration is that of a soft tissue or cystic mass in lower lobes in association with a systemic arterial feeder. Definitive diagnosis of pulmonary sequestration requires demonstration of abnormal arterial supply from systemic circulation mostly aorta by angiography or noninvasive tests like CT angiography or MR angiography. Definitive treatment involves resection of the affected lung segment.

**Fig 1.** Showing homogenous opacity in Lt Lower zone

**Fig 2.** CT Angiography showing an aberrant feeding artery from abdominal aorta.
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Fig 3 CT Angiogram showing sequestrated segment (thick and short arrow) and the feeding artery from abdominal aorta (thin and long arrow).

Fig 4 Contrast CT Scan showing sequestrated lung segment (arrow)

References