

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

\*Behera PK<sup>1</sup>, Tripathy KP<sup>2</sup>

<sup>1</sup>Associate professor, Dept of Medicine, KIMS, Bhubaneswar, Odisha, India

<sup>2</sup> Professor, Dept of Medicine, KIMS, Bhubaneswar, Odisha, India

---

**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 hemoptysis. Here we report a case of intralobar pulmonary sequestration in a male of 36 years who continued to have hemoptysis 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.<sup>1</sup> 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.<sup>2</sup> Cases mostly present with repeated chest infections, pleural effusion or minor hemoptysis but cases with severe and fatal hemoptysis 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 hemoptysis 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-tubercular drugs and had completed full course of Anti tubercular therapy twice, once 5 years 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 built 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.

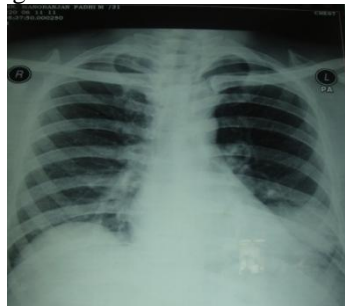
On investigation, Hb. was 9.5gm%. TLC - 12,000/cmm, ESR 40mm, blood urea 30mg/dl, serum creatinine 1mg/dl. FPG 90mg/dl. X Ray of chest showed a homogenous opacity in the left lower lobe adjacent to cardiac shadow. CT angiography of chest showed the opacity to have a separate blood supply from abdominal 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 sequestered 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.<sup>3</sup> Rokitansky and Rektorzic described the first case in 1861. They proposed that the sequestration was due to a separated but normally developed lung fraction.<sup>4</sup> 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 intercostals arteries.<sup>4</sup> 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.<sup>5</sup> Intralobar pulmonary sequestration is three to six times more common than the extralobar type.<sup>6</sup> 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.<sup>1</sup> The symptoms in Intralobar sequestration typically occurs during early childhood but around 50% cases the diagnosis is made after the age twenty.<sup>7</sup> Commonest symptoms are cough, expectoration, recurrent attacks of pneumonia. Pleural effusion occur in 4% of cases.<sup>1</sup> Minor hemoptysis is common. More severe hemoptysis, bleeding into pleural space, oesophagus or into the sequestration itself has been reported.<sup>8</sup> In 15% cases the anomaly is asymptomatic and normal longevity has been reported. In majority of cases the arterial supply is from thoracic aorta (73.9% cases) followed by abdominal aorta.<sup>1</sup> The caliber of the vessels varies from 1-15mm. The venous return from the intralobar sequestration is usually via the pulmonary veins. Extralobar sequestrations most commonly (77.4%cases) 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.

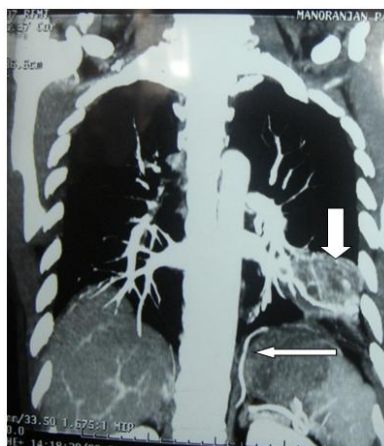
Intralobar sequestrations are not associated with other cardio-pulmonary anomalies but extralobar sequestrations may be found in association with cardiac or more frequently diaphragmatic anomalies in 50% of cases.<sup>9</sup> There have been few reports of malignant neoplasms being involved in or near sequestered 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.<sup>10</sup> 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.<sup>11</sup> 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.



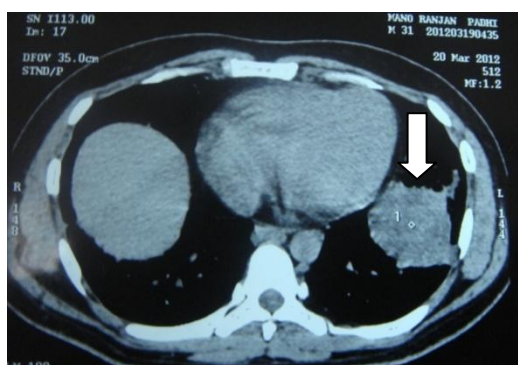
**Fig 1.**Showing homogenous opacity in Lt Lower zone



**Fig 2.**CT Angiography showing an aberrant feeding artery from abdominal aorta.



**Fig 3** CT Angiogram showing sequestered segment(thick and short arrow) and the feeding artery from abdominal aorta (thin and long arrow).



**Fig 4** Contrast CT Scan showing sequestered lung segment ( arrow)

### References

- [1]. Savic B, Birtel FJ, Tholen W et al. Lung sequestration: report of seven cases and of 540 published cases. *Thorax*.1979; 34:96-101.
- [2]. Sade RM, Clous M. The spectrum of pulmonary sequestration. *Ann Thoracic Surgery*.1974; 18: 644-58.
- [3]. Kravitz R M. Congenital malformations of the lung. *Ped Clin N Am*. 1994;41/3:453-473.
- [4]. Halkie N, Cuenoud P F, Cortesy M E, Ksontini R, Boumghar M. Pulmonary sequestration: a review of 26 cases. *Eur J Cardiothorac. Surg*. 1998; 14:127-133.
- [5]. Yong W, Fan L. Pulmonary sequestration : a retrospective analysis of 2625 cases in China. *Eur. J. Cardiothorac. Surg*.2011;40:39-42
- [6]. Sugio K, Kaneko S, Yokoyama H, Ishida T, Sugimachi K, Hasuo K. Pulmonary sequestration in older child and in adults. *Int Surgery*. 1992;77:102-107
- [7]. Lin CH, Lin CT, Chen Y, Phen HC, Chen HC, Wong PY. Pulmonary sequestration. *Chin Med J.(Taipei)*1994;53:168-172
- [8]. Frazier AA, Chirstension MR, Stoker JJ,Tempelton PA. Intralobar sequestration: Radiopathologic correlation. *Radiographics*.1997; 17:725-745
- [9]. Avishai V, Dolev E, Weissberg D, Zaidel L, Priel IE. Extralobar sequestration presenting as massive hemothorax. *Chest* 1996;109 /3:843-845.
- [10]. Yagy H, Adachi H, Furukawa K, et al. Intralobar pulmonary sequestration presenting increased serum CA 19-9 and CA 125. *Intern Med*.2002;41:875-8
- [11]. Naidich DP, Rummanink WM, LeflleurRS, Estioko MR, Brown SM. Intralobar pulmonary sequestration: MR Evaluation. *J Comp Assisted Tomo*.1987;11: 531-533.