ORIGINAL RESEARCH PAPER

Internal Medicine

SEQUESTRATION OF LUNG

KEY WORDS:

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ABSTRACT

Pulmonary sequestration is a condition in which a segment of dysplastic lung tissue or lobe is present without any communication with the rest of the tracheobronchial tree, and which receives an anomalous systemic blood supply separate from the rest of the lung. It is a piece of nonfunctional lung tissue and a foregut embryonic remnant. It may present as haemoptysis, and this condition may mimic other respiratory diseases like Tuberculosis, Bronchiectasis and Malignancy. We present a case of a 16-year-old female, who came with complaints of massive haemoptysis. A CT Thorax done showed sequestration of the right lower lobe of lung, which was operated upon in a higher centre with the aid of Video Assisted Thoracic Surgery. (VATS)

INTRODUCTION

Pulmonary sequestration (PS) is a rare congenital broncho-pulmonary malformation and belongs to one of the congenital cystic lung lesions, accounting for 0.15%~6.4% of all congenital pulmonary malformations.[1][2] As this may present with haemoptysis, primary care physicians attending such patients should be aware of this condition, so that this can be kept as a differential diagnosis and ruled out. Once the diagnosis is confirmed, prompt and early treatment of complications with definitive surgical management using VATS may reduce the morbidity and mortality significantly.

Case Summary

A 16-year-old female, an athlete by profession, with no previous comorbid conditions or addictions got admitted with complaints of cough and scanty haemoptysis for the last 5 days along with low grade fever for 10 days.

There was no previous history of hospitalization. Haemoptysis was not associated with chest pain, dyspnoea, or bleeding from any other site. There was no history of tuberculosis in the family or history of close contact with patient of active tuberculosis. On admission patient was conscious, oriented, BP was 110/70 mmHg, pulse rate 120/minute, respiratory rate 24/minute, saturation on room air was 95%. There was no pallor, icterus, clubbing, cyanosis, or any palpable lymphadenopathy. Chest X ray on admission showed right lower zone opacity. (figure 1). Patient was provisionally diagnosed as right mid zone pneumonia and treatment was started with Injection Ceftriaxone 1gm intravenous twice daily, Clarithromycin 500 mg twice daily along with other supportive care.

The patient was reviewed by the chest physician and a CT scan of the thorax was advised. Sputum for Acid fast bacilli was negative (table 1). CT thorax showed large heterogenous enhancing lesions in the right lower lobe, bronchial wall thickening and peri bronchial centrilobular nodules suggestive of an intra lobar sequestration. (Figure 3-4). Opinion of the chest physician was taken. Sputum for acid fast bacilli was negative. Patient had one episode of massive haemoptysis during her stay in hospital. The BP crashed, the pulse rate rose, and she went into hypovolaemic shock – something called Pulmonary Apoplexy. She was treated with whole blood, iv fluids, tranexamic acid, and other styptic agents. The haemoptysis gradually abated, the BP and Pulse rate normalised, and the storm eventually got settled. On the fourth day, after stabilisation she was discharged with a diagnosis of lung sequestration right lower lobe and probable pulmonary tuberculosis and advised to take opinion from another tertiary care hospital. She went to CMC Vellore, where a diagnosis of combined congenital pulmonary airway malformation (CPAM) and intra lobar sequestration was made. She underwent right uniportal VATS (video assisted thoracic surgery)- wedge resection. Operative findings revealed systemic artery supplying the segment of lung which was coursing on the inferior pulmonary ligament from across the diaphragm. The posterior basal segment was completely diseased. After the surgery she has been following up with us and has been by and large asymptomatic with no fresh bouts of haemoptysis.

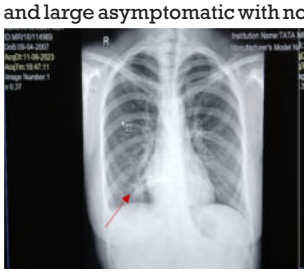

Figure 1- Right lower zone opacity

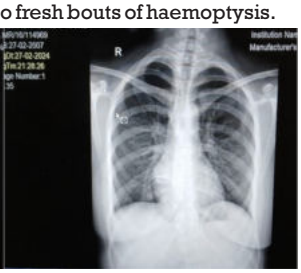

Figure 2 – Resolved Opacity

Table 1: Lab Investigations

DATE	PARAMETERS	VALUE	NORMAL RANGE
23.01.2023	URIC ACID	5.58	2.5 3.5
	SERUM PROTEIN	7.36	6.6 8.3
	SERUM GLOBULIN	3.72	2.5 3.5
	LIPASE	13.3	0 39
	TOTAL BILIRUBIN	0.83	0.2 1
	DIRECT BILIRUBIN	0.62	0.1 0.5
	INDIRECT BILIRUBIN	0.21	
	AST	23.5	0 45
	ALT	13.3	0 40
	ALP	94.3	35 125
	SERUM SODIUM	131	136 146
	SERUM POTASSIUM	4.5	3.5 5.5
	C REACTIVE PROTEIN	0.71	0.08 0.79
	SERUM CREATININE	0.62	0.5 1.5
	PLATELET COUNT	3.13 LACS	1.5 LACS 4.1 LACS
	TOTAL LEUCOCYTE COUR	14600	4000 11000
	HEMOGLOBIN	11.4	11.5 16.5
	NEUTROPHILS	87	60 70
	LYMPHOCYTE	9	30 40
	APTT	30.5	23 33.4
	INR	0.98	0.8 1.2
	URINE RBS	NIL	
	SPUTUM FOR MYCOBACTERIUM TUBERCULOSIS	NOT DETECTED	
	HIV 1 AND II ELISA	NEGATIVE	
26.01.2023	TOTAL LEUCOCYTE COUR	9000	4000 11000
	PLATELET COUNT	2.93 LACS	1.5 LACS 4.1 LACS
	HEMOGLOBIN	9.8	11.5 16.5

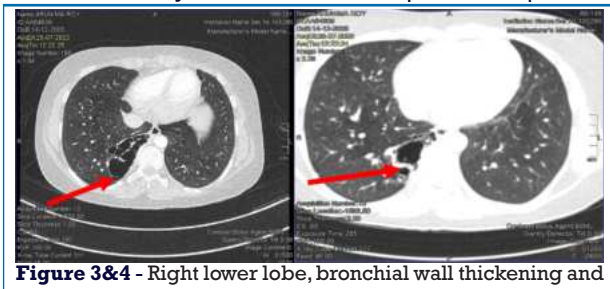

Figure 3&4 - Right lower lobe, bronchial wall thickening and peri bronchial centrilobular nodules suggestive of an intra lobar sequestration.

DISCUSSION

Pulmonary sequestration, as described by Pryce in 1946, is a condition in which a segment or a lobe of dysplastic lung tissue exists with no communication with the rest of the tracheobronchial tree and receives an anomalous systemic vascular supply, separate from the rest of the lung. It is, therefore, a non-functional tissue. It accounts for 0.15% to 6.40% of all congenital lung malformations.[1][2][18]

Anatomically, classified as intra lobar and extra lobar sequestration. Intra lobar sequestration presents within the visceral pleura of the functioning lung. It is more common than the extra pleural sequestration. Venous drainage is into the pulmonary vein. Extra lobar sequestration has a separate visceral pleura and venous drainage. Venous drainage is into the systemic veins, frequently the lower lobe vein, azygos vein, hemiazygos vein. Arterial supply is from systemic vessels–most commonly descending thoracic aorta (73%), the cranial portion of the abdominal aorta, celiac trunk, splenic artery (21%), as well as intercostal arteries.[3]

Intra lobar sequestration is commonly found in left lower lobe. Intra lobar sequestrations is the predominant variant in the adults, commonly diagnosed before 20 years of age, median age at diagnosis is 42 years. [7][8] Extra lobar sequestration is commonly located between the diaphragm and the lower lobes, rarely infra-diaphragmatically.[4]

Pulmonary sequestration is a congenital anomaly of the primitive foregut. It develops from an accessory supernumerary lung bud below the normal lung bud which continues to migrate caudally with the oesophagus in extra lobar sequestration, then derives its blood supply from the primitive splanchnic vessels surrounding the foregut. If the lung bud arises before the development of pleura, it results in intra lobar sequestration.[5][6]

Histologically, lymphocytic inflammation and fibrosis are seen in lung parenchyma in the sequestered segment. Also, the presence of cystic air spaces lined by cuboidal or columnar epithelium, alveoli with emphysema-like hyperinflation are not uncommon. A granulomatous reaction can sometimes be seen in response to superimposed infection in intra lobar sequestrations.

Extra lobar sequestration presents, clinically, with respiratory distress, high output congestive heart failure (due to right-to-left shunt), and occasional spontaneous pulmonary or pleural haemorrhage. They rarely get infected as they are separated from the tracheobronchial tree by their pleural investment.

Intra lobar sequestration frequently remains asymptomatic and presents with recurrent pneumonia in a localized segment of the lung with persistent cough, exertional shortness of breath and haemoptysis. Most common causative agent is Pseudomonas aeruginosa, Other agents include tuberculosis, *Nocardia,* or *Aspergillus*.[16][17] Haemoptysis is the most common complication in adulthood.[10]

Antenatal diagnosis is made by Doppler ultrasound at 18 to 19 weeks of gestation.[11] Definitive diagnosis requires the establishment of a systemic arterial supply and venous drainage of the sequestered lung tissue. Multi-planar CT with 3-dimensional reconstruction is the study of choice to demonstrate the lesions with its anomalous vascular supply. On Contrast enhanced CT, the sequestered lung tissue may present as a cyst, a mass, a lamellar lesion, a capsulated lesion with an air-fluid level (and thus, falsely interpreted as hydatid cyst or abscess), focal atelectasis, or bronchiectasis. Biopsy of the pulmonary sequestration is confirmatory tissue diagnosis, it is associated with a high risk of bleeding from aberrant vessels and not used routinely.[12][13] The pulmonary function test reveals the obstructive disease in 8.8% of patients.[14] Bronchoscopy can be done to exclude other potential causes of haemoptysis and confirm absence of any communication of affected lung tissue with bronchial arteries. CT/MR angiography may be required to clinch the diagnosis.

Intra or extra lobar sequestration requires prompt management in a tertiary care hospital equipped to provide vigorous resuscitation and critical care. Thoracoamniotic shunting is recommended in a foetus less than 30 weeks gestation showing signs of hydrops. Post-delivery of the baby in large sequestration, supportive management with ventilator support, high-frequency oscillatory ventilation or extracorporeal membrane oxygenation (ECMO) may be necessary.[15] Patient needs to be managed conservatively with IV antibiotics for superadded infections, intravenous fluids and other resuscitative measures for fluid deficit and hypotension if need be. Pulmonary lobectomy is the treatment of choice for established pulmonary sequestration.[16] It is useful even in asymptomatic patients to avoid recurrent infections and prevent fatal complications e.g., haemoptysis, and progressive inflammation of the lung parenchyma. Resection can be achieved by open thoracotomy or video-assisted thoracoscopic procedures (VATS). The postoperative complication rate is 25 to 28%. Complications include prolonged air leak, bronchopleural fistula, haemoptysis, empyema, and arrhythmia.[17]

Endovascular embolization and coiling is a recently developed therapeutic alternative.[9] It reduces blood flow to the sequestered tissue, leading to necrosis, fibrosis and progressive involution of the sequestrated lobe. Embolization is associated with recurrence rate of 25% to 47%.[18]

CONCLUSION

This case highlights the importance of thorough diagnostic work up after making a differential diagnosis of sequestrated lung especially in paediatric and adolescent patients. Once the diagnosis is confirmed, pulmonary lobectomy with VATS and wedge resection is the best treatment option currently. Although rare, it should be considered in young patients presenting with recurrent respiratory infection and haemoptysis. Intra lobar sequestration is the predominant variant in adults and has better prognosis with early diagnosis and prompt management. This young girl with Intrapulmonary lung sequestration got successfully and timely operated by the VATS technique in an advanced centre for Pulmonary medicine and lives on to tell her tale.

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