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BRONCHOPULMONARY FOREGUT MALFORMATION TYPE III CASE SERIES OF AN OFTEN MISDIAGNOSED ENTITY

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Abstract : Pulmonary sequestration occurs when a disturbance in embryonic development produces a cystic mass of nonfunctioning lung tissue. Most often the mass is supplied by an anomalous systemicartery and has its own bronchial system, which usually does not communicate with the normal bronchial tree. Though traditionally classified as intralobar (ILS) and extralobar (ELS) sequestrations, it is postulated that they belong to a broad spectrum of bronchopulmonary-foregut malformations. These Bronchopulmonary foregut malformations were classified by Shrikant et al and our discussion is pertaining to type III of this set of malformations. The treatment of ILS in adults is often misdiagnosed and hence delayed. We present a case series of six cases over the last five years. All of these cases were on medical management before they presented to our hospital and on presentation to us they were diagnosed to have communicating bronchopulmonary foregut malformation type III. Of these 5 cases were surgically treated while one patient was lost in follow up as he was not willing for surgery.

Keyword : Sequestration, Intralobar, Cystic Adenomatoid malformation, Bronchopulmonary foregut malformation

INTRODUCTION

Bronchopulmonary sequestrations are a rare group of congenital malformations with a subset of those have a communication with esophagus, making such cases rarer. Because of the rarity, the diagnosis of these conditions is often not looked into and patients undergo prolonged refractory course of medications before someone looks for such etiology. We present our experience, emphasizing the importance of early diagnosis and intervention to avoid complications and morbidity of the patient. Between 2010 and 2015, six patients presented to us with recurrent episodes of coughandrespiratory infection refractory to medical treatment. These were ultimately diagnosed to have communicating bronchopulmonary foregut malformation type III. Of these five cases were operated and one patient did not want to get operated and hence was lost in follow up. Inadequate preoperative study and incorrect diagnosis could increase the risk of perioperative morbidity and mortality.

As surgical treatment is available and curative it is all the more important to diagnose this condition.

PATIENTS AND METHOD Between 2010 and 2015, 6 patients were diagnosed to have communicating bronchopulmonary foregut malformation type III. Five were males and one patient was female with ages ranging from 16 years to 41 years. All patients underwent a clinical examination, chest radiograph, CT scan of thorax, bronchoscopy [Table 1] as part of clinical workup.

TABLE 1

Case	Age	Sex	PRESENTATION	History of ATT	Chest Xray	CT THORAX		
						LUNG LOBE INVOLVED	ABBERENT BLOOD SUPPLY	OESOPHAGEAL COMMUNICATION
1	23	M	Cough since childhood, haemoptysis - 5 yr	no	Right lower zone cystic changes.	RML, RLL	Not defined	Between lower esophagus and RLL bronchus
2	21	M	Cough since-childhood, left pleural effusion 8yr back	Yes	Left lower zone bronchiectasis.	LLL	Not defined	Not defined
3	21	F	Cough since- 3 yrs	Yes	Consolidation and Fibro cavitatory lesion in the Right Lower Zone.	RLL & RML	Not defined	Not defined
4	41	M	Cough since-15yrs cough on ingestion of liquids - 2 months. Intercostal chest drainage 5 yrs back for right sided empyema.	Yes	Right lower zone ?mass lesion with air fluid levels*	RLL	Dilates systemic arterial collaterals and Pulmonary arteries entering the sequestration	Fistulous communication between anterolateral wall of esophagus and sequestered segment
5	16	M	Cough since-childhood, haemoptysis - 5 yr	nil	Right middle and lower lobe bronchiectasis	Segment 3 & RLL	Not defined	Not defined

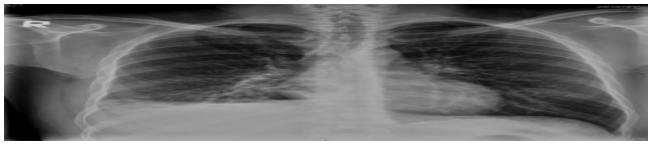
6	22	M	Cough since-childhood and cough on ingestion of food/ water	nil	Left multi lobar bronchiectasis	LLL & Lingula	Two communic Systemic artery branch from splenic artery ations between the lower esophagu s into the LLL & Lingula

*Barium swallow of the patient showed Fistula between the lower one third of thoracic esophagus to an ill defined cavity in the right lower para-mediastinal region. RLL –Right lower lobe, RML – Right middle lobe, LLL – Left lower lobe

Presentation :

All patients had complaint of chronic recurrent episodes of cough with expectoration. Two patients had complaint of hemoptysis. Two patients had cough in association with ingestion of foo or water. Three patients were treated in past empirically as Tuberculosis and completed a course of ATT but these patients were sputum negative for AFB on presentation here..

Segment affected:



All cases had lower lobe involvement with Right : Left – 4:2 ratio. Two cases also had associated middle lobe involvement.

Figure 1A

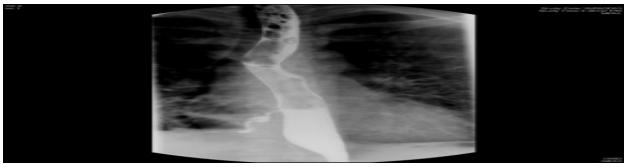


Figure 1B

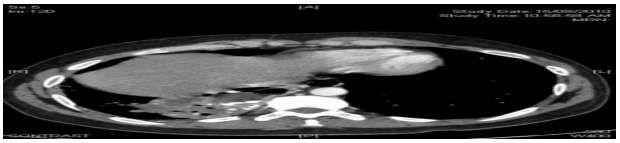


Figure 1C

Chest x-ray : [Figure 1A]

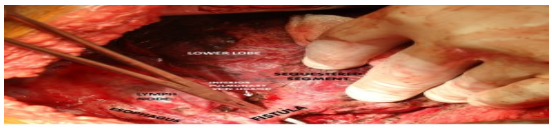
Features of bronchiectasis of surrounding lobe or consolidation of surrounding lobe are seen in all cases. Figure 1A shows x-ray of a patient with Right lower lobe intralobar sequestration with foregut malformation which shows bronchiectatic changes in Right lower lobe in x-ray.

Barium Swallow:

Two patients present with cough in relation to food or liquid ingestion underwent barium swallow study. Both patients demonstrated a fistulous tract from lower esophagus that communicated with lobe of lung with bronchiectatic features. [Figure 1B].

CT Thorax: [Figure 1C]

--> Involved lobes show bronchiectasis and consolidation or collapse. --> Esophageal communication was apparent in 50% of the cases. --> Anomalous systemic arterial supply was defined in two of the six cases.



TREATMENT: [Table 2]

Of the 5 patients who proceeded with definitive treatment were all operated and Lobectomy and esophageal repair. A well defined fistulous tract was visualized intraoperatively in all five cases communicating the affected lobe to esophagus. [Figure 2]

Post op Period:

Three patients had postoperative leakage from the esophagus, detected on barium study on 7th post op day which healed conservative managementonly .

Histopathology

Four of cases had proven Intralobar Sequestrationwith bronchiectasis. All cases had esophageal communication. One case, had associated Cystic Adenomatoid malformation.

TABLE 2

Case	Operation done	OPERATIVE FINDINGS	BIOPSY REPORT	POST OPERATIVE PERIOD
1	RIGHT MIDDLE AND LOWER LOBECTOMY WITH GI REPAIR	A 6 cm diameter thin walled diverticulum in the esophagus 5 cm above the diaphragmatic hiatus stuck to the posterior aspect of the lower lobe and having a fistula between it and the substance of the lower lobe.	Intra lobar sequestration	Uneventful
2	LEFT LOWER LOBECTOMY WITH GI REPAIR	Once cm diameter fistula between the esophagus and the bronchus just below the level of the inferior pulmonary vein.	Intra lobar sequestration	Minimal leak from the esophagus in 7th POD Barium swallow. Managed conservatively
3	RIGHT MIDDLE AND LOWER LOBECTOMY WITH GI REPAIR	There was a fistulous tract from the esophagus into the lower lobe. The middle and lower lobes were consolidated and infected.	Bronchiectatic abscess with esophageal fistula	Contained leak visualized in 7th POD Barium study
4	RIGHT LOWER LOBECTOMY WITH GI REPAIR	Large communication between the medial and anterior segments of the lower lobe and the lower esophagus.	Intra lobar sequestration with cystic adenomatoid malformation	7th POD Barium study showed a esophageal leak into a intra thoracic collection. Managed by percutaneous drainage and feeding jeujunostomy.
5	RIGHT LOWER LOBECTOMY WITH GI REPAIR	Fistula from the esophagus just above level of the inferior pulmonary vein directly entering the basal bronchus posteriorly.	Intra lobar sequestration	Uneventful

DISCUSSION Bronchopulmonary sequestration (BPS), is a rare congenital malformation of the lower respiratory tract. It consists of a nonfunctioning mass of lung tissue that lacks normal communication with the tracheobronchial tree, and that receives its arterial blood supply from the systemic circulation(1).

Communicating bronchopulmonary foregut malformations (CBPFMs) are characterized by a fistula between an isolated portion of respiratory tissue (ie, a lung, a lung lobe, or a segment) and esophagus or stomach. Shrikant et al had classified these into Group I (16%): anomaly is associated with esophageal atresia and tracheoesophageal fistula. Group II (33%): one lung originates from the lower esophagus. Group III (46%): an isolated anatomic lung lobe or segment [sequestration] communicates with the esophagus or stomach. Group IV (5%): A portion of the normal bronchial system communicates with the esophagus(2). We present a cases of Type III CBPFM with communication between lower lobe and esophagus. These communications are explained by presence of abnormal collection of pluripotent cells with respiratory potential, arising from the dorsal side of developing esophagus, distal to normal lung bud, which develop into the lung tissue, retaining the communication with the esophagus(foregut). These anomalies have their own arterial supply, venous drainage and pleural investment (3).

Treatment of these anomalies involves essentially the lobectomy with gastrointestinal repair. Failure to diagnose the anomaly may lead to improper treatment leading to prolonged morbidity and surgical complications.

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