

Case report

Esophageal lung: a rare case of communicating bronchopulmonary foregut malformation

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ABSTRACT:

Esophageal lung is a type of communicating bronchopulmonary foregut malformation (CBPFM). CBPFM are rare and esophageal lung where there is anomalous origin of the main stem bronchus from the esophagus is even rarer. We report a case of a 5 month old male child, a known case of tubercular meningitis with subdural empyemas, who was referred to us with cough and increased respiratory activity since 15 days and history of recurrent pneumonias since birth. Chest x-ray revealed a hypoplastic lung and further investigations with CT thorax and barium esophagogram led to the diagnosis of esophageal lung. This case emphasizes the need to have CBPFM in differential diagnosis of patients with recurrent pneumonias and further to include barium esophagogram in the investigation protocol of these patients.

Keywords: Esophageal lung, barium esophagogram, recurrent pneumonias

INTRODUCTION:

Esophageal lung is a rare communicating bronchopulmonary foregut malformation with only about 20 cases reported in literature and is characterized by abnormal connection of the main stem bronchus, usually the right with the esophagus instead of the trachea¹. It is associated with congenital anomalies of the upper gastrointestinal tract, ribs and vertebrae and the pulmonary and systemic vascular system and usually the patients present with repeated chest infections due to aspiration². Diagnosis is made radiologically (by CT and upper GI contrast studies) and accurate preoperative imaging diagnosis is essential before planning surgical intervention. So we have decided to describe the presentation, radiological features and

the imaging protocol to be used for this rare entity which every radiologist must be aware of.

CASE REPORT:

A 5 month old male child, a known case of tubercular meningitis presented with cough and increased respiratory activity since 15 days. The patient had a history of repeated hospitalization for fever with productive cough and respiratory distress. The baby was a full term normal delivery (FTND) with normal immunization and development for age. Chest X-ray showed increased opacification of the right lung with reduced lung volume, air bronchograms, and mild compensatory hyperinflation of the left lung [Fig I]. USG revealed lower lobar consolidation with no effusion. The child was advised a CT thorax for further management. CT thorax revealed hypoplasia of the right lung with consolidation and multiple air

bronchograms. Tracheal bifurcation was absent with direct continuation of left main stem bronchus. Lower sections revealed anomalous origin of the right main bronchus from the lower third of the esophagus which was coursing laterally to supply right lung [Fig II]. This was later confirmed by giving barium oral contrast given via a nasogastric tube. Contrast opacified the bronchial tree on the right thus confirming the bronchoesophageal communication [Fig III]. Volume rendered CT showed anomalous origin of the right bronchus from the lower esophagus and only left bronchus at the bifurcation [Fig IV]. The hypoplastic right lung was supplied by right pulmonary artery and veins with no anomalous arterial supply or venous drainage. The ipsilateral right pulmonary artery was also narrowed. At surgery, right posterolateral thoracotomy was done which revealed hypoplastic consolidated lung. A cartilaginous short right main stem bronchus supplying the right lung was seen communicating with the esophagus. Right pneumonectomy with resection of the esophageal bronchus and repair of the esophagus at the site of bronchial communication was done.

DISCUSSION:

Congenital bronchopulmonary foregut malformation (CBPFM) include anomalies in which there is an abnormal patent communication between isolated portion of respiratory tissue (lung, lobe, segment) and gastrointestinal tract. They result from anomalous budding of the embryonic foregut and tracheobronchial tree³. Most common associations being cardiac anomalies, tracheoesophageal fistula and esophageal atresia. Its association with esophageal atresia is incompatible with life⁴. They are subdivided into communicating and noncommunicating types. Noncommunicating types

include foregut duplication cysts, diverticulae, intralobar or extralobar pulmonary sequestrations while in communicating variety there is communication between the respiratory and gastrointestinal systems⁵. CBPFM classification based on anatomic features and supported by a proposed embryogenesis theory is as follows⁶.

- Group I (16%): anomaly is associated with esophageal atresia and tracheoesophageal fistula.
- Group II (33%): one lung originates from the lower esophagus and is termed as esophageal lung as in our case.
- Group III (46%): an isolated anatomic lung lobe or segment communicates with the esophagus or stomach and is termed as esophageal bronchus.
- Group IV (5%): A portion of the normal bronchial system communicates with the esophagus. The portion of the lung served by the communicating bronchus receives systemic blood supply.

Another classification by Braimbridge and Keith⁷ is based on the morphology of the fistulous tract (neck, diverticulum, cyst) and the blood supply.

- Type I is associated with a wide-necked congenital diverticulum of the oesophagus with an inflammatory fistula at the tip.
- Type II is a simple fistula running directly from the oesophagus to a lobar or segmental bronchus.
- Type III consists of a fistulous track connecting the oesophagus to a cyst in the lobe, which in turn communicates with the bronchus.

- In type IV the fistula runs into a sequestered segment; the sequestration connects by one or more tracks with the bronchus.

Clinically these patients present with chronic cough, recurrent pneumonias, hemoptysis and respiratory distress due to bronchoesophageal communication. In 65% of cases, patients have cough on swallowing liquids (Ono's sign) or food in the sputum which was not seen in our case ⁷. The condition is more common in females as compared to males, with a ratio 1.5 to 1. The right lung is more commonly involved as in our case because of proximity of the right main stem bronchus with the esophagus ⁸. As against our case 75% of patients present in adulthood. The late onset of symptoms may be due the presence of a membrane or valve within the fistulous tract that ruptures or becomes incompetent, the oblique upward course of the fistula or the closure of the fistula due to contraction of its muscular coat during swallowing ².

Differential diagnosis include pulmonary sequestration, congenital cystic adenomatoid malformation, and iatrogenic, inflammatory, or



neoplastic fistulas. In sequestration anomalous systemic arterial supply with only lobar involvement is seen. In our case entire right lung was hypoplastic with right main stem bronchus arising from esophagus and no systemic arterial blood supply. The two main methods of treatment are division and suturing of the ends of the fistula and complete resection ⁹.

CONCLUSION:

Esophageal lung is a rare entity which is compatible with life if not associated with other anomalies and usually presents with repeated chest infections and coughing especially after drinking. Hypoplastic consolidated lung supplied by a mainstem bronchus arising from the esophagus on CT scan proven by barium contrast opacifying the ipsilateral bronchial tree on esophagogram are the characteristic radiological features. Thus esophagography with barium is the modality of choice for demonstration of the fistula. Even though it is a benign disease, serious complications can occur and so early recognition and proper imaging diagnosis can guide early surgical intervention and better clinical outcome.

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Figure I: Chest X -ray shows increased opacification of the right lung with reduced lung volume, air bronchograms, and mild compensatory hyperinflation of the left lung.

Figure II: Coronal CT scan (soft tissue window) reveals anomalous origin of the right main bronchus from the lower third of the esophagus and coursing laterally to supply right lung.

Figure III: CT esophagogram reveals opacification of the bronchial tree on the right thus confirming the broncho-esophageal communication.

Figure IV: Volume rendered CT shows anomalous origin of the right bronchus from the lower esophagus and only left bronchus at the bifurcation.

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