

A rare case of esophageal lung in a neonate

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Abstract

We report a rare case of esophageal lung in a neonate who presented with repeated chest infections and respiratory distress. Chest radiograph revealed increased opacification of the right lung with reduced lung volume and air bronchograms. Further evaluation with computed tomography (CT) showed the presence of only left mainstem bronchus at the tracheal bifurcation. Right mainstem bronchus originated from distal esophagus and aerated the right lung. Nasogastric tube was inserted into the stomach with injection of small amount of dilute barium through it, which established the communication of right mainstem bronchus with esophagus. Advanced CT scan imaging by virtual bronchoscopy and volume rendering further delineated the anatomical abnormality precisely prior to surgery. Surgical findings confirmed the diagnosis.

Key words: Bronchopulmonary foregut malformation; esophageal lung; virtual bronchoscopy; volume rendering

Introduction

Oesophageal lung is a rare congenital malformation. We came across a neonate who presented with repeated chest infections and respiratory distress. On evaluation by CT we could demonstrate broncho-oesophageal communication. The same was confirmed by surgery.

Case Report

A full term female baby was born after an uneventful pregnancy. At 1 month of age, the child presented with severe respiratory distress and chest infection. On clinical examination, the child looked sick and had tachypnea. She had similar episodes in the past where the child was treated with antibiotics. A plain radiograph of the chest showed increased opacification of the right lung with reduced

lung volume, air bronchograms, and mild compensatory hyperinflation of the left lung [Figure 1]. Following this, a computed tomography (CT) scan was performed, which showed hypoplasia of the right lung with multiple air bronchograms. In the mediastinum, two air-filled tubes were visualized, that is, trachea and esophagus [Figure 2A]. Only left mainstem bronchus was visualized at the tracheal bifurcation. The right mainstem bronchus originated from distal esophagus and coursed superiorly to aerate the right lung [Figure 2B]. Nasogastric tube was inserted that delineated the retracted air filled esophagus on the right side of the trachea. Dilute barium was slowly injected through the nasogastric tube, which established the diagnosis of bronchoesophageal communication by opacifying the bronchial tree on the right side [Figure 3]. Imaging advances such as volume rendered CT and virtual bronchoscopy showed anomalous origin of the right mainstem bronchus from the esophagus [Figure 4A] and

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Figure 1: Chest radiograph shows increased opacification of right lung with air bronchograms, mediastinal shift, and herniation of the contralateral lung towards right



Figure 3: Dilute barium has entered the right bronchial tree, thus, establishing the bronchoesophageal communication

only one opening at the tracheal bifurcation [Figure 4B]. The right lung was supplied by pulmonary artery and veins. There was no anomalous systemic arterial supply or venous drainage.

At surgery, a hypoplastic consolidated lung was discovered on the right side. There was a firm cartilaginous tube (right mainstem bronchus) coming from the distal esophagus and entering into the right lung [Figure 5]. Right pneumonectomy with *enbloc* resection of esophageal bronchus was performed, and abnormal opening in the esophagus was ligated and repaired.

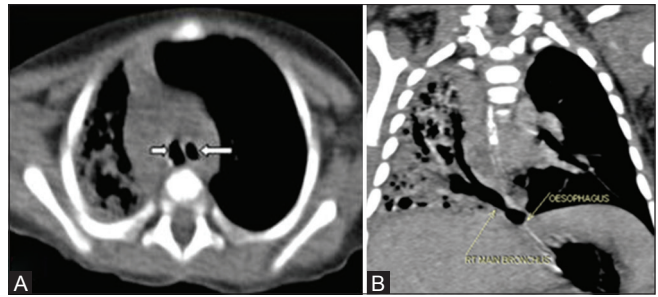


Figure 2 (A and B): (A) Axial image shows two closely placed air-filled tubes esophagus (small arrow) and trachea (large arrow) in the mediastinum. (B) Coronal reformatted image depicts the origin of right the mainstem bronchus (small arrow) from distal esophagus (large arrow) with associated collapse-consolidation of the right lung

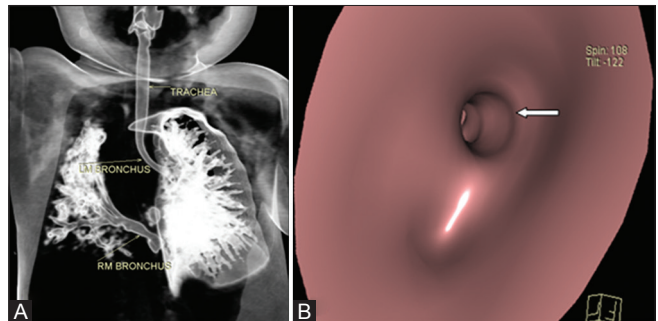


Figure 4 (A and B): (A) Volume rendered computed tomography scan shows the single left mainstem bronchus at carinal level. The right mainstem bronchus originates from the distal esophagus and courses superiorly to aerate the right lung. (B) Virtual bronchoscopy shows the single opening of the left mainstem bronchus (arrow) at the tracheal bifurcation. Right mainstem bronchus is not visualized

Discussion

Bronchopulmonary foregut malformations are rare. Esophageal lung is an entity belonging to this group. Anomalous origin of the main bronchus from the esophagus is termed as esophageal lung. Sometimes only a segmental bronchus may arise from the lower esophagus with rest of the lung being normal. Diagnosis is usually established within the first 8 months of life, even though late presentations in adulthood are also described. Earlier the presentation, worse the prognosis. Females are more commonly affected.^[1] Because of the proximity of the right mainstem bronchus with esophagus, right side is frequently affected.^[1,2] Furthermore, clinical manifestations may vary from recurrent chest infections to severe respiratory failure, depending on the type of malformation.^[1]

Patients presenting with recurrent chest infections or cough on intake of fluids or food should be investigated with chest radiograph followed by contrast study of esophagus and chest CT. Vascular studies can be further conducted for preoperative mapping as well as to rule out sequestration.^[1] Barium esophagogram is the investigation of choice in such cases and should be the ideal choice in patients with recurrent refractory chest infections.^[1]



Figure 5: Operative findings: Small consolidated right lung with a cartilaginous right mainstem bronchus(long arrow), communicating with the esophagus(short arrow)

Embryologically, the respiratory system develops as a ventral diverticulum from the foregut. As laryngotracheal tube elongates, the tracheoesophageal ridge develops and separates the laryngotracheal tube from rest of the foregut. Later, tracheoesophageal ridge fuses to form the septum and divides the foregut into ventral and dorsal portions. The ventral part is the trachea and dorsal portion forms the esophagus. Any abnormal development of tracheoesophageal groove along with the differential elongation of trachea and esophagus results in these anomalies.^[2,3]

Known associations of esophageal lung include cardiac anomalies, esophageal atresia, and tracheoesophageal fistula. When associated with esophageal atresia, they are incompatible with life.^[3,4] The two subdivisions include noncommunicating and communicating types. Foregut duplication cysts, diverticulae, intralobar or extralobar pulmonary sequestrations come under the noncommunicating variety. Communicating type is one where there is communication between the respiratory and gastrointestinal systems.^[5]

In 1966, Braimbridge and Keith^[6] suggested a classification for congenital fistulas depending upon the communication between esophagus and bronchus/sequestered lobe. Further, Srikanth *et al.* classified the communicating bronchopulmonary foregut malformations as follows:^[7,8]

- 1A: Total sequestered lung communicating with the foregut, associated with esophageal atresia and tracheoesophageal fistula to the distal pouch
- 1B: Sequestered anatomic lobe or segment communicating with the foregut, associated with esophageal atresia and tracheoesophageal fistula to the distal pouch
- II: Total sequestered lung communicating with the lower esophagus; absent ipsilateral mainstem bronchus
- III: Isolated anatomic lobe or segment communicating with the foregut

IV: Portion of the normal bronchial system communicating with the esophagus

Our case falls into category IV where the entire lung was aerated by the right mainstem bronchus, which originated from the lower esophagus.^[9] Internal bronchial anatomy appeared normal. Diagnostic challenge is to differentiate it from bronchopulmonary sequestration. In sequestration, there is only lobar involvement with systemic blood supply. In our case, the entire lung was hypoplastic and consolidated with the left mainstem bronchus arising from the distal esophagus. No systemic arterial supply was observed. The differential diagnosis includes pulmonary sequestration, congenital cystic adenomatoid malformation, and iatrogenic, inflammatory, or neoplastic fistulas.^[10]

The two main methods of treatment are division and suturing of the ends of the fistula and complete resection.^[11] Till today, less than 20 cases of esophageal lung have been reported,^[12] and most of the cases were associated with esophageal atresia. In our case, there was no esophageal atresia as the nasogastric tube easily passed into the stomach.

Conclusion

Bronchopulmonary foregut malformations are rare entities where there is abnormal communication between the respiratory and upper alimentary system. Early recognition of this rare entity could guide an appropriate treatment with better clinical outcome.

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Conflicts of interest

There are no conflicts of interest.

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