Case Report

Esophageal Lung: A rare congenital malformation cured successfully

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ABSTRACT

Communicating broncho-pulmonary foregut malformations are rare developmental anomalies, ''esophageal lung/bronchus'' being one of the entities in this group. Anomalous origin of the main bronchus from the esophagus is even rare and is known as ''esophageal lung'' or ''total pulmonary sequestration''. Similar anomalies can involve a segmental bronchus only, with the remainder of the lung being normal. We report a 1 year old female presenting with recurrent cough and chocking episode, diagnosed as having right bronchus communicating to the lower esophagus. The rarity of this condition and advanced age of presentation are the highlights of this case. Child was managed successfully with a nearly total pneumonectomy and esophageal repair.

Keywords: Bronchopulmonary foregut malformation, esophageal lung, bronchoscopy.

Case Report

1 year old girl from Afghanistan referred at Madhukar Rainbow Childrens hospital to us with complains of repeated episode of cough, choking while eating solid food since age of 6 month. She had multiple hospital admission in view of same and use to get recovered for brief period. She was also unable to gain weight (current weight 7.3 kg) despite good appetite.

At time of birth, she was full term with birth weight of 3 kg. There was no history of NICU admission or other even in perinatal period. She had a chest X-ray showing increased opacification of the right lung (Figure1) with reduced lung volume, compensatory hyperinflation of the left lung and CT chest (Figure 2) was suggestive of Hypoplastic right lung with shift of mediastinum to the right. Clinically air entry was decreased on right side with suprasternal retraction.

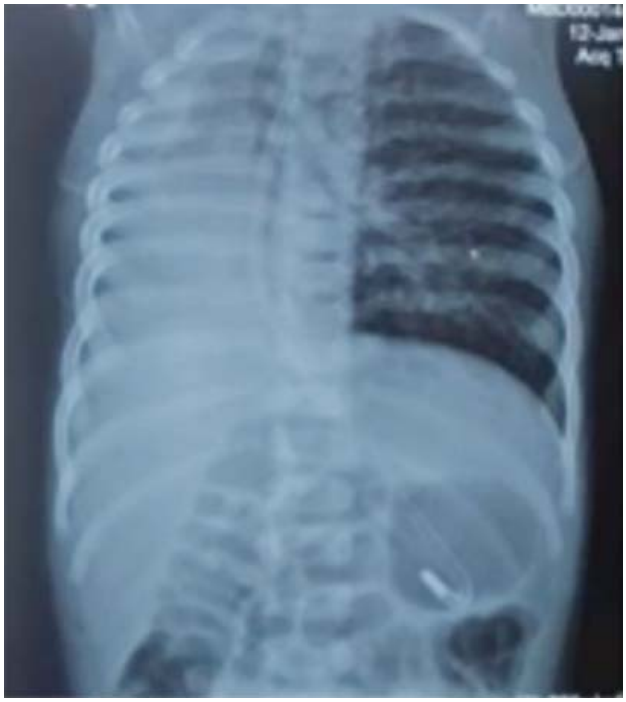

Fig 1: Chest X-ray showing right opacification

Patient was admitted in PICU and Flexible bronchoscopy with upper GI endoscopy was done beside. Left bronchus was patent, without any

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anatomical abnormality. There was no evidence of right bronchial opening. Endoscopy revealed an esophageal opening (Figure 3) going to right lung.

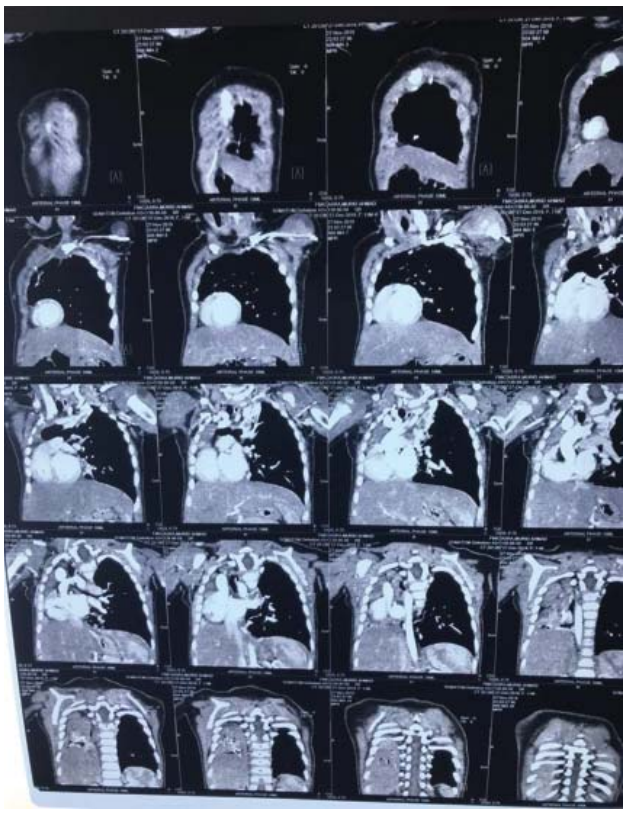

Fig 2: CT chest showing hypoplastic right lung and dextroverted heart

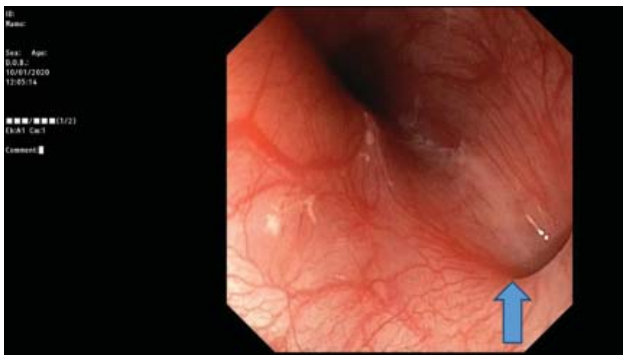

Fig 3: Esophageal opening leading to right bronchus on endoscopy (shown by blue arrow)

2 D ECHO showed Dextroverted Heart with normal function. After a team meeting and counseling with parents, child was taken for surgery. All initial finding were confirmed and since right lung was small it was surgically removed with esophageal repair. This was checked for any leak with upper GI Dye study. Post-surgical stay was uneventful. Child was discharged in stable condition with consistent weight gain. Family was very pleased with the services at our hospital specially Pediatric Intensive care bronchoscopy and Pediatric pulmonary, pediatric gastroenterology and pediatric surgical services. They were very thankful that by multispecialty team approach at Madhukar Rainbow Childrens hospital their child was able to survive and get cured of this rare congenital abnormality (Esophageal lung) causing failure to thrive and repeated infections and hospitalizations.

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 main stem 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].

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 no communicating and communicating types. Foregut duplication cysts, divericulae, intralobar or extralobar pulmonary sequestrations come under the no communicating 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/sequestrated lobe. Further, Srikanth *et al*. classified the communicating bronchopulmonary foregut malformations as follows[7,8]:

1. Total sequestered lung communicating with the foregut, associated with esophageal atresia and tracheoesophageal fistula to the distal pouch
2. Sequestered anatomic lobe or segment communicating with the foregut, associated with esophageal atresia and tracheoesophageal fistula to the distal pouch
3. Total sequestered lung communicating with the lower esophagus; absent ipsilateral mainstem bronchus
4. Isolated anatomic lobe or segment communicating with the foregut
5. 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].

Surgical repair is the treatment of choice. 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.

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