Esophageal lung with rare associated vascular and anorectal malformations

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ABSTRACT
Esophageal lung is a rare communicating bronchopulmonary foregut malformation in which the main bronchus arising from the trachea is absent. The affected lung is usually hypoplastic and aerated via an anomalous airway originating from the esophagus. Other anomalies such as esophageal atresia with tracheoesophageal fistula or VACTERL (vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities) association can co-exist. The initial radiographic findings may be normal, but subsequent imaging usually shows progressive and recurrent lung collapse, probably because of recurrent aspiration through the anomalous airway and poor compliance of the affected lung during breathing. In this report, we describe a neonate with esophageal lung and rare associated anomalies, including anorectal malformation, pulmonary artery sling, and inferior vena cava interruption with azygous continuation. To our knowledge, this is the first report of esophageal lung with such associations.

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Introduction
Esophageal lung is a rare congenital malformation in which the main airway of one lung (usually the right) originates from the esophagus and the ipsilateral main bronchus from the trachea is absent [1]. Clinical presentation usually includes opacification of the lung airspace because of recurrent aspiration through the anomalous airway. If there are associated congenital anomalies, such as esophageal atresia, the affected lung is persistently collapsed and nonresponsive to ventilation. Computed tomography (CT) can show the absent main bronchus, demonstrate the anomalous airway connection to the esophagus, and identify other associated airway and vascular anomalies. An upper gastrointestinal (GI) contrast study is the modality of choice for identification of the origin of the

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anomalous esophageal airway and can assess the condition of the esophagus and whether or not there is associated esophageal atresia.

**Case report**

The patient was a 1-month-old boy born via a normal spontaneous vaginal delivery at 36 weeks’ gestation to a 28-year-old mother with gestational diabetes and polyhydramnios. There was no family history of congenital anomalies or metabolic disease. At birth, his vital signs and growth parameters were normal for age and sex. His oxygen saturation was 95% on room air and there was no respiratory distress. He had no dysmorphic features, and the rest of the clinical examination was unremarkable except for an imperforate anus. After delivery, he was admitted to the neonatal intensive care unit for close observation and monitoring before surgery to repair the anorectal malformation. Initial routine blood and urine investigations were unremarkable. Ultrasonography of the brain and abdomen was normal. Echocardiography showed a small ventricular septal defect and a patent foramen ovale, normal atrioventricular and ventriculoarterial concordance with good biventricular function, and a left pulmonary artery arising distally from the right pulmonary artery with no stenosis. His initial chest radiograph was normal (Fig. 1A).

On the second day of life, the patient underwent colostomy surgery for a high anorectal malformation, and was then transferred to the neonatal intensive care unit for ventilatory support. He was extubated on postoperative day 3. He subsequently developed tachypnea with decreased air entry on the right side. Orogastric tube feeding was started on postoperative day 5 and increased over the next 5 days to 40 mL every 8 hours. Intravenous antibiotic cover was provided during the postoperative period.

Subsequent chest radiographs showed a recurrent right lung collapse with a compensatory overinflated left lung (Fig. 1B). The right lung collapse persisted despite chest physiotherapy, 3% nebulized saline, and noninvasive respiratory support.

Chest CT with contrast showed agenesis of the right main bronchus with an anomalous origin of the right lung bronchus from the distal part of the esophagus near the gastroesophageal junction (Fig. 2A). The right lung was mildly hypoplastic and supplied by a single normal-appearing right pulmonary artery. No anomalous systemic arterial supply or venous drainage was noted. However, the left pulmonary artery was found to be arising distally from the right pulmonary artery and curving posterior to the left main bronchus and anterior to the esophagus (ie, pulmonary artery sling, Fig. 2B). The trachea and left main bronchus were unremarkable (Fig. 3). Abdominal CT showed absence of the infrahepatic inferior vena cava with azygos continuation. No splenic, renal, or skeletal abnormalities were noted.

A further upper GI study using water-soluble contrast medium clearly showed opacification of the entire right lung air space by contrast material because of reflux of contrast through the anomalous airway communication with the lower part of the esophagus (Fig. 4). The remaining part of the esophagus was unremarkable, with no focal narrowing or abnormal communication with the upper tracheobronchial system.

The patient was referred to a tertiary center for treatment of a right esophageal lung associated with pulmonary artery sling, an interrupted inferior vena cava with azygos continuation, and anorectal malformation.

**Discussion**

Bronchopulmonary foregut malformations include a wide spectrum of airway abnormalities that occur because of abnormal separation between the primitive foregut and esophagus. The spectrum includes 4 main types, that is, foregut anomalies, vascular abnormalities, lung parenchymal abnormalities, and airway anomalies. Each type has its own spectrum of pathologies and presentations. Examples of these anomalies include congenital pulmonary airway malformation, bronchogenic cyst, pulmonary sequestration, congenital lobar emphysema, bronchogenic cyst, esophageal atresia with or without

![Fig. 1 – Preoperative (A) and postoperative (B) chest radiographs. The preoperative image shows mild interstitial thickening with normal lung aeration and the postoperative image shows opacification of the right lung with a dilated airway directed abnormally to the lower esophagus (arrow).](image-url)
tracheoesophageal fistula, and bronchial atresia or stenosis. Overlap between these anomalies (hybrid lesions) can exist, with more than 1 pathology coexisting within the same lesion [2].

Communicating bronchopulmonary foregut malformation (CBPFM) occurs when there is a congenital communication between the lung and foregut because of a focal mesodermal defect [3,4]. Srikanth et al. [1] devised a system whereby CBPFM can be classified into 4 main groups. Group I is associated with esophageal atresia and tracheoesophageal fistula and contains 2 subdivisions, that is, group IA, in which the mainstem bronchus is absent and the entire lung arises from the esophagus or stomach, and group IB, in which only a portion of 1 lung is communicating with the esophagus. Group II is characterized by the absence of a mainstem bronchus arising from the trachea and the presence of 1 hypoplastic lung (usually the right) arising from the esophagus. Group III occurs when an isolated part of the lung is communicating with the esophagus and group IV occurs when there is communication between a normal bronchial system and the esophagus.

When a primitive mainstem bronchus is joined to the esophagus, the anomaly is known as “esophageal lung” [4], and when only a lobar bronchus arises from the esophagus, the anomaly is called “esophageal bronchus” [5]. Esophageal lung was first described by Keely et al. in 1960 [6] and is considered a rare type of CBPFM, with only about 25 cases described in the English literature. Other congenital anomalies can co-exist in association with esophageal lung. For example, esophageal lung with esophageal atresia and tracheoesophageal fistula has been described in about half of the reported cases [4,7,8]. Other associated conditions, such as duodenal atresia, duodenal stenosis, and congenital heart disease, have been reported [9,10].

The clinical presentation depends on the type of abnormality, site of communication, and associated anomalies. Most cases present early in life, although there has been 1 report of late presentation at the age of 20 years [10]. The initial presentation varies from asymptomatic to life-threatening with recurrent aspiration and collapse of the affected lung. The mortality rate has been reported to be 28% if CBPFM is associated with long-segment congenital tracheal stenosis [11].
The diagnosis is usually made at the time of radiological investigation for persistent or recurrent lung collapse despite ventilatory support. CT can easily identify the absence of a mainstem bronchus arising from the trachea and can be used to evaluate the site of origin of the anomalous airway. Associated thoracic and extrathoracic vascular anomalies may be identified when intravenous contrast medium is used. An upper GI contrast study using oral water-soluble contrast medium is the golden standard method for showing the anomalous origin of the airway from the esophagus and identifying the condition of the remaining esophagus. In a retrospective study, magnetic resonance imaging of surgically proven esophageal bronchus was able to identify a bronchial abnormality in the form of a T2-hyperintense tubular structure directed from the bronchus to the gastroesophageal junction in the prenatal period.

Our patient was considered to have group II CBPFM on the basis of the absent main bronchus and anomalous origin of the lung airway from the esophagus. The initial preoperative chest radiograph showed minimal interstitial thickening, but not to the degree that requires medical attention. However, the postoperative chest radiographs did show recurrent total lung collapse with no improvement despite ventilatory support. CT with contrast clearly showed the airway abnormality with an absent right main bronchus and abnormally dilated airway structures that converged to a single airway directed toward the lower esophagus. The left pulmonary artery was arising anomalously from the right pulmonary artery and coursing to the left lung, while passing posterior to the left main bronchus and anterior to the esophagus. The right pulmonary artery was slightly small in size and supplied a hypoplastic lung. There was no abnormal systemic vascular supply to the right lung or any abnormal draining pulmonary veins. The visualized portion of the abdomen showed azygos continuation of the inferior vena cava and absence of the hepatic and infrahepatic segments. No associated cardiac, renal, or musculoskeletal abnormalities were identified. An upper GI contrast study clearly outlined the anomalous right lung airway arising from the lower part of the esophagus. No abnormal narrowing in the rest of the esophagus and no fistula communicating with the trachea or left bronchus was found. There have been case reports of CBPFM with VACTERL (vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities) association [13–15], as well as a report of CBPFM with pulmonary artery sling [16]. However, to our knowledge, this is the first reported case of CBPFM presenting as imperforate anus, a right esophageal lung with an absent right bronchus (group II), pulmonary artery sling, and an interrupted inferior vena cava with azygos continuation.

Successful treatment of esophageal lung depends on how much of the lung is affected and the nature of the associated congenital anomalies. Resection and reimplantation of the anomalous airway into the trachea can be considered in some cases. However, pneumonectomy performed via a thoracotomy without cardiopulmonary bypass may be safer than bronchial reconstruction in patients with severe lung hypoplasia [17].

**Conclusion**

This report highlights the importance of considering esophageal bronchus in the differential diagnosis of nonresolving lung collapse, especially in the neonatal period. Early identification of this congenital anomaly can help to prevent further destruction of the lung and preserve its function. CT with contrast and upper GI contrast studies are the modalities of choice for identifying the abnormality and the associated anomalies that can contribute to the outcome of treatment.

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