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Management of Esophageal Lung in a Patient With VACTERL Anomalies

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Abstract

Esophageal lung is a rare type of bronchopulmonary foregut malformation where an anomalous main bronchus arises from the esophagus rather than the trachea. This differentiates from an esophageal bronchus where a lobar bronchus arises from the esophagus. Fewer than 30 of these anomalies have been reported in the literature. A female infant was born at 35 weeks gestational age and found to have multiple congenital abnormalities including cleft palate, long-gap esophageal atresia, tracheoesophageal fistula (TEF), imperforate anus, and renal anomalies. She initially underwent thoracoscopic ligation of TEF with colostomy and mucus fistula creation. Bronchoscopy found no right mainstem bronchus, and subsequent computed tomography scan was consistent with possible esophageal bronchus. Esophagoscopy through the gastrostomy discovered a fistulous connection between the distal esophagus and anomalous main right bronchus.

Right thoracotomy with pneumonectomy of the hypoplastic lung was performed. She subsequently underwent vaginostomy for hydrometrocolpos. Her proximal esophagus was diverted with a cervical esophagostomy for control of secretions. She also required tracheotomy and is currently at home on minimal ventilator settings awaiting staged reconstruction.

Esophageal lung is a rare congenital abnormality with few reported cases. Surgical treatment with pneumonectomy is often required, and pediatric surgeons should be familiar with these congenital bronchopulmonary malformations.

Keywords

pediatric surgery, esophageal lung, congenital anomalies

Esophageal lung is a rare bronchopulmonary foregut malformation where an anomalous main bronchus arises from the esophagus rather than the trachea.[1] This is differentiated from an esophageal bronchus, defined by a lobar bronchus that arises from the esophagus. Fewer than 30 cases of esophageal lung have been reported in the literature.[2] This report describes a case of esophageal lung associated with VACTERL abnormalities.

A female infant, born at 35 weeks gestational age, was found to have multiple congenital abnormalities including cleft palate, long-gap esophageal atresia, tracheoesophageal fistula (TEF), imperforate anus, and renal anomalies. On initial chest x-ray, there was some aeration of the right lung, and a thoracoscopic ligation of TEF was performed. Colostomy, mucus fistula creation, and gastric tube placement were also performed during this index operation due to her imperforate anus.

After the initial operation, persistent right lung atelectasis prompted further investigation. Bronchoscopy identified the fistula tract to the proximal esophageal pouch that had been previously clipped thoracoscopically, but the right mainstem bronchus was unable to be identified (Figure 1). A subsequent CT scan demonstrated a possible anomalous bronchus from the right lung to the distal atretic esophagus (Figure 1). Esophagoscopy was then performed through the gastrostomy site and confirmed the diagnosis of the esophageal lung.

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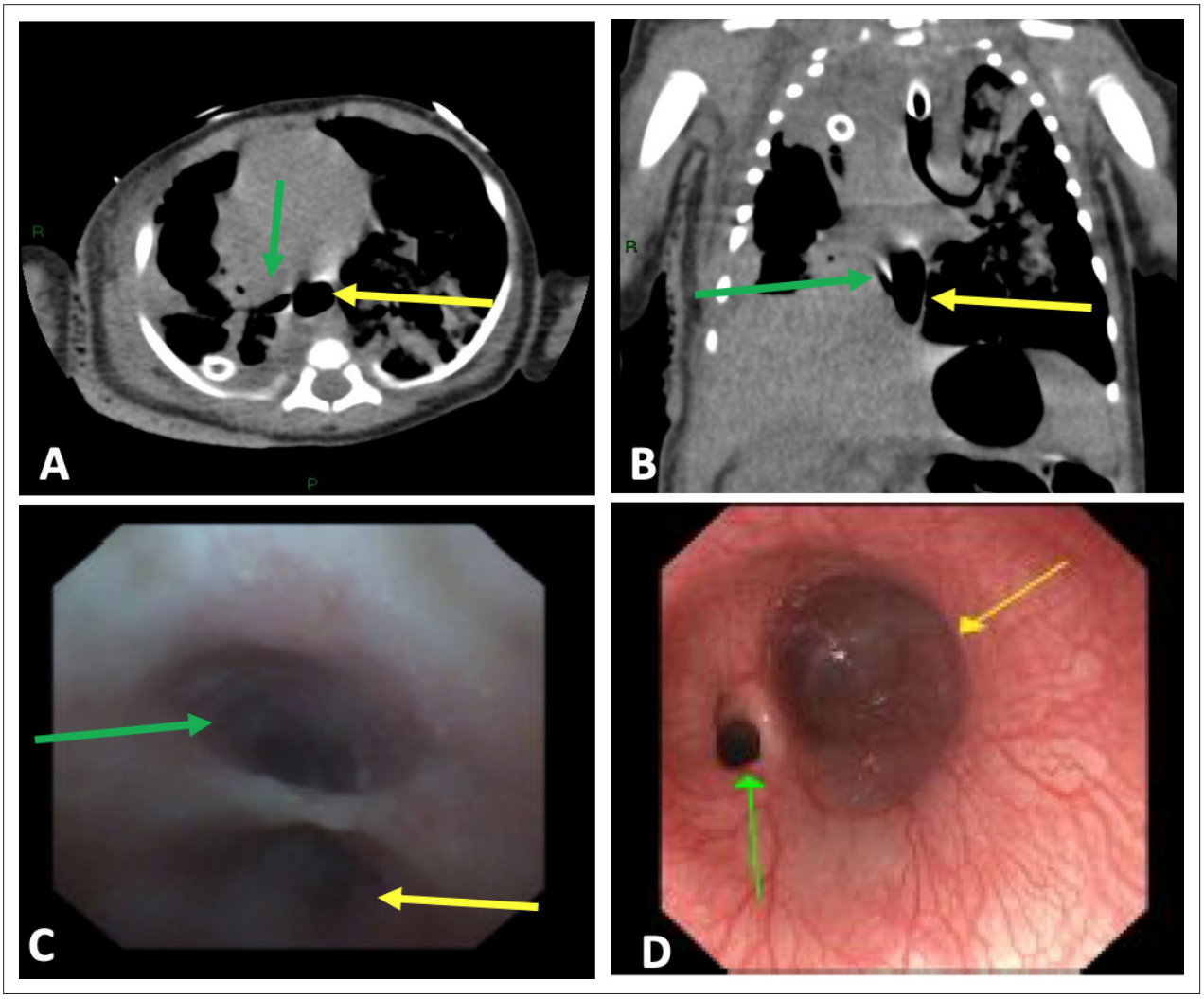

Figure 1. Diagnostic images. (A and B) Computed tomography scan showing anomalous main bronchus (green arrow) in connection with esophagus (yellow arrow). (C) Bronchoscopic view of the left main stem bronchus (green arrow) with ligated tracheoesophageal fistula (yellow arrow). (D) Esophagoscopic image showing esophagus (yellow arrow) and anomalous right main bronchus fistulous connection (green arrow).

A right thoracotomy was performed, and a hypoplastic lung was identified with the anomalous main bronchus connecting to the distal atretic esophagus (Figure 2). Due to its limited functional nature and complete lack of connection to the airway, pneumonectomy was performed. She also subsequently underwent vaginostomy for hydrometrocolpos with Urology.

During her hospitalization, excessive secretions caused recurrent aspiration pneumonia despite medical management with Robinul and Replogle tube in the proximal esophageal pouch. To mitigate this, the proximal esophagus was diverted with a cervical esophagostomy to prevent aspiration with plans for future repair.

Eventually, the patient was weaned off the ventilator and extubated to the nasal cannula. She was initially discharged from the hospital on the day of life 110 but has had 2 readmissions for respiratory-related complications. She eventually required tracheostomy for the management of ongoing respiratory issues. Currently, the patient is at home on minimal ventilator settings awaiting staged repair of her esophagus followed by repair of her anorectal malformation.

VACTERL association is a term used to describe nonrandomly associated congenital defects including vertebral, anal, cardiac, TEF, renal, and limb anomalies.[3] Though diagnostic criteria of this condition are vague, patients should have at least 3 of these defects for VACTERL diagnosis.[3] The patient in this report was found to have anal atresia, TEF, and renal anomalies, thus meeting criteria for diagnosis.

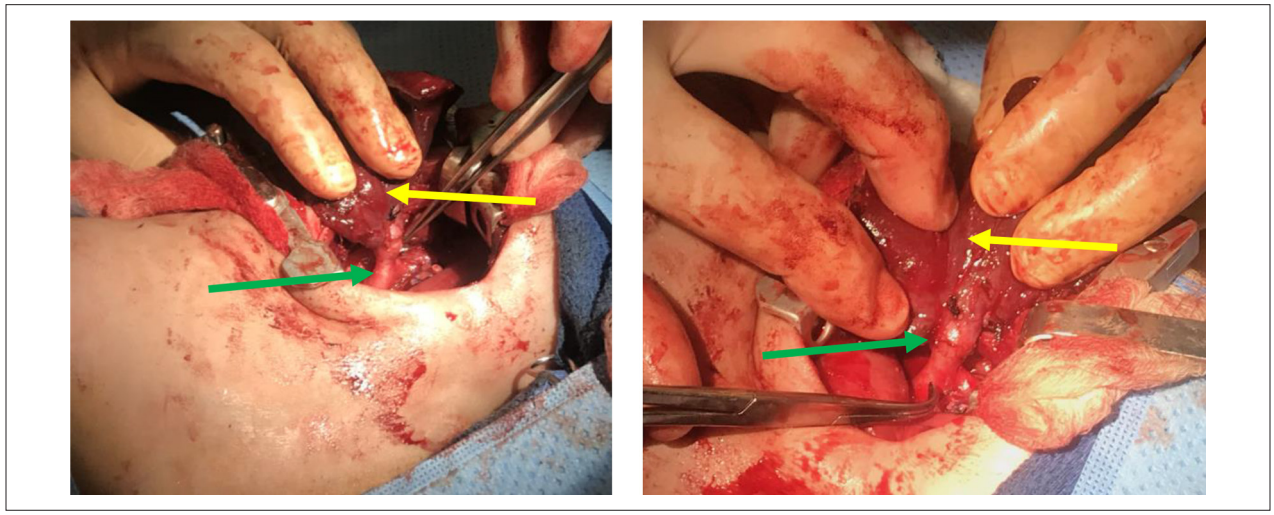

Figure 2. Intraoperative photos: right thoracotomy incision showing hypoplastic right lung (yellow arrow) with anomalous main bronchus (green arrow) connected to the esophagus.

Esophageal lung is a rare congenital bronchopulmonary abnormality with few reported cases, not typically associated with VACTERL. This anomaly is thought to occur due to abnormal separation of the esophagus and primitive foregut.[2] Other diagnoses such as congenital lobar emphysema, pulmonary sequestration, and congenital pulmonary airway malformation develop along similar pathways.[2] An analysis of the limited previously reported cases of the esophageal lung has shown it to occur most often on the right, such as in this case.[1]

Our patient's esophageal lung was identified after multiple diagnostic and imaging modalities during the first month of life. Other case reports have identified this condition later in life due to recurrent pulmonary infections.[1,2,4] Diagnosis of this condition can be difficult due to the anatomical variations. An upper gastrointestinal contrast study can help identify connections with the alimentary tract, though this was not available in this case due to the long-gap esophageal atresia.[2] Bronchoscopy and CT scan can also help delineate the bronchial anatomy. As mentioned above, our patient eventually required esophagogastroscopy through her gastrostomy tube site to achieve a definitive diagnosis due to her multiple anatomical abnormalities.

Though a few cases have reported successful bronchial reconstruction, surgical treatment with pneumonectomy is the mainstay of treatment with esophageal lung.[2] Significant hypoplasia along with a lack of normal blood supply, such as in the patient presented here, would indicate the need for pneumonectomy rather than reconstruction.

Though rare, pediatric surgeons should be familiar with esophageal lung and esophageal bronchus as part of the spectrum of congenital bronchopulmonary malformations. Surgical intervention, most often thoracotomy and pneumonectomy, should be considered at the time of diagnosis. More cases and additional investigations are needed to evaluate the long-term effects of these rare malformations.

Declaration of Conflicting Interests

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