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Esophageal lung in a 4-month-old infant

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ABSTRACT

Esophageal lung is an extremely rare congenital malformation characterized by an abnormal connection between the esophagus and the bronchial tree, without communication with the airway. We present the case of a 4-month-old female patient who was admitted to the hospital with a diagnosis of pneumonia. Complementary studies confirmed the presumptive diagnosis of esophageal lung. Surgical treatment was then decided upon, and a pneumonectomy with esophageal repair was performed. The patient progressed favorably.

Keywords: *lung diseases; congenital anomalies; esophageal diseases.*

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INTRODUCTION

Esophageal lung (EL) is an extremely rare congenital malformation in which there is an abnormal connection between the esophagus and the bronchial tree. The main bronchus originates from the esophagus.[1]

Due to its low incidence, with fewer than 20 cases reported in the medical literature, EL represents a diagnostic and therapeutic challenge for pediatricians and pulmonologists.[2]

The objective of this publication is to report a clinical case of a girl diagnosed with EL who was successfully treated, highlighting the most relevant aspects of her clinical presentation, diagnostic approach, and surgical management.

CLINICAL CASE

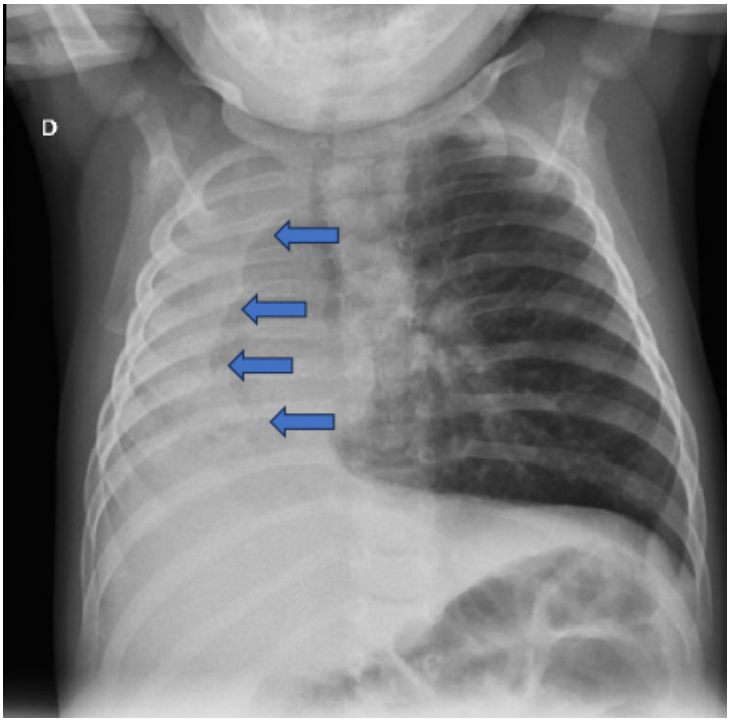
We present the case of a 4-month-old girl who was referred from another institution with a diagnosis of pneumonia and atelectasis vs. right lung hypoplasia. Relevant background information included that she was a full-term newborn, 37 weeks, 2880 g. She was delivered by cesarean section due to fetal arrhythmia; her mother was 15 years old, used marijuana and tobacco during pregnancy, and had no documented prenatal ultrasounds.

She received treatment for syphilis with 3 doses. Serology tests for Chagas disease, HIV, hepatitis B, and toxoplasmosis were negative. The patient was hospitalized in the neonatal unit for 20 days due to right renal agenesis associated with left renal hypoplasia, maternal syphilis, and a single umbilical artery. She was fed orally and had no other history of respiratory pathology. The mother denied respiratory symptoms during feeding.

Upon admission to our institution, the patient was cardiorespiratory sufficient, tachypneic, with oxygen saturation of 96-98% on room air, in good general condition, alert, and responsive. On auscultation, she had subcrepitant rales in the right lung field. On feeding, she presented with tachycardia, tachypnea, and intercostal retractions, with an oximetry reading of 96%.

A chest X-ray showed an opacity in the right lung field with mediastinal shift, retraction of the right costal arches, and a left pneumonocele (*Figure 1*). A chest CT scan showed dextroposition, absence of the right main bronchus, lung parenchyma with alveolar consolidation and air bronchogram,

Figure 1. Chest X-ray



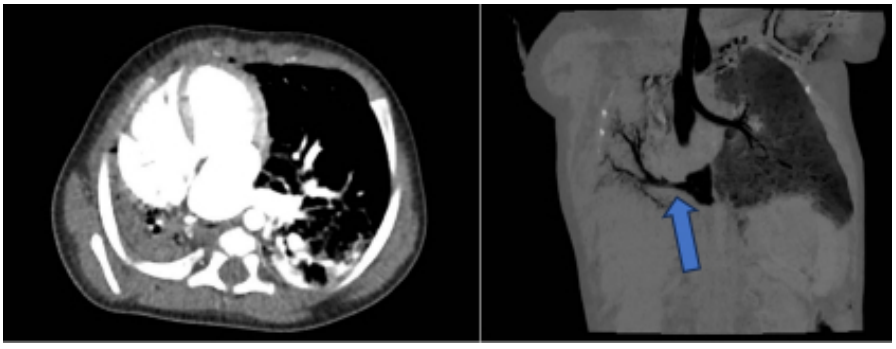
Note the ipsilateral mediastinal shift with retraction of the right ribs and left pneumonocele (arrows).

and a bronchus communicating with the lower esophagus, a small-caliber right pulmonary artery, and a single small-caliber right inferior pulmonary vein (*Figure 2*).

Respiratory endoscopy and esophagogram were performed, confirming the absence of the right main bronchus and esophageal communication at the lower third level, with emergence of an anomalous bronchus toward the right lung parenchyma (*Figure 3*).

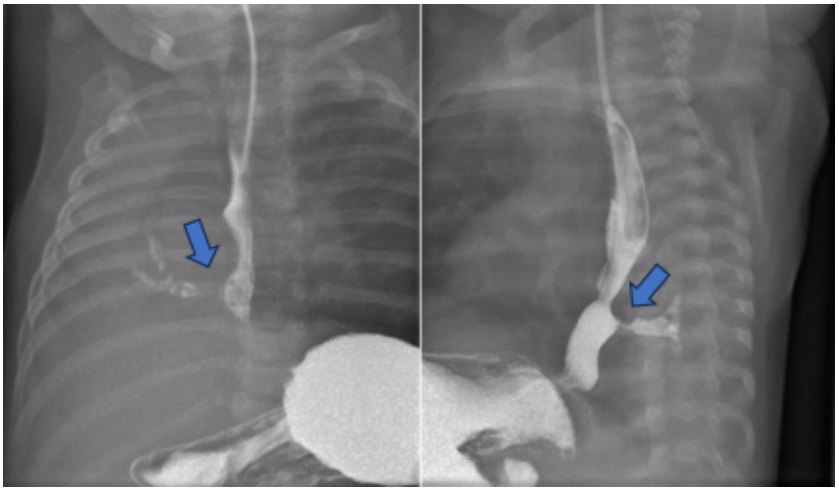
With a presumptive diagnosis of EL, we decided on a surgical intervention. A thoracotomy and right pneumonectomy with primary esophageal repair were performed. The patient developed a small esophageal leak that resolved spontaneously. On the 21st postoperative day, the pleural drain was removed after an esophagogram confirmed the absence of a fistula, and oral feeding was resumed. The pathological anatomy report stated "Sequelae in the lung with extensive mixed inflammatory process, fibrosis, and type 2 pneumocyte hyperplasia. Bronchial lumen of the hilum with partial squamous metaplasia."

Figure 2. Chest tomography



2a. Right lung hypoplasia, cardiac dextroposition. Cystic images in the consolidated right lung are suggestive of bronchiectasis. 2b Tomographic reconstruction showing the presence of the right bronchial tree originating from the lower esophagus (arrow).

Figure 3. Esophagogram, anteroposterior and lateral views



A fistula (arrow) is evident between the lower esophagus and the right bronchial tree.

DISCUSSION

EL is an extremely rare malformation of the primitive anterior intestine in which a bronchus originates abnormally from the esophagus rather than the trachea (*Figure 4*). It is more common on the right side, with only two left-sided cases reported in the literature.[2]

Its etiology is unknown, but it is thought to result from a defect in the separation of the primitive anterior intestine into the esophagus and the trachea during early embryonic development.[3]

Several associated congenital anomalies have been reported, particularly esophageal atresia with fistula. Other associated anomalies described include systemic and/or pulmonary vascular anomalies, diaphragmatic anomalies, upper gastrointestinal tract anomalies, and vertebral anomalies.[2-4]

It is included within the communicating bronchopulmonary malformations of the foregut, in which there is a congenital communication between the respiratory tract and the esophagus or stomach. Our case corresponds to Group II, in which one lung arises from the lower esophagus. The ipsilateral main bronchus is absent, and the trachea extends to form the contralateral main bronchus.[5,6]

The clinical manifestations of EL are heterogeneous and vary with patient age, the size of the abnormal lung, and the presence of associated complications.

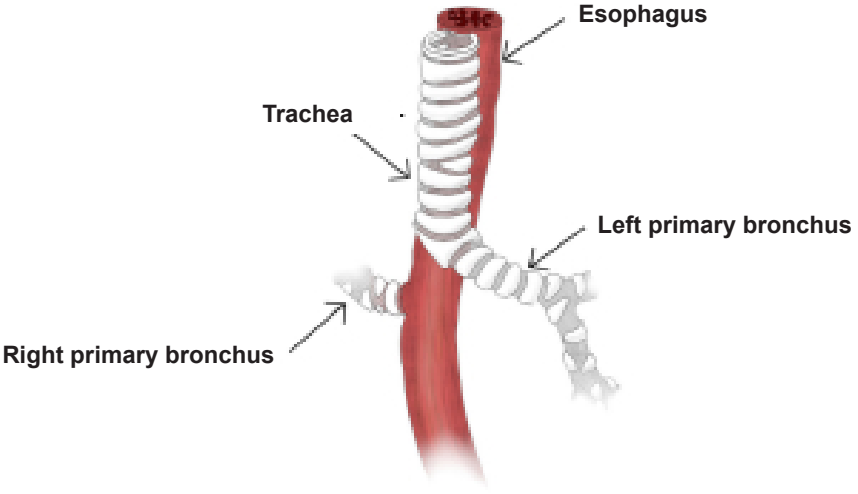
They may occur in the neonatal period[7] or later. They usually include cough, recurrent pneumonia, and hemoptysis.

The diagnosis is usually confirmed by endoscopy of the digestive or respiratory tract in a patient with clinical or radiological suspicion. Tomography is helpful because, in addition to establishing the presence of esophageal communication, it provides information on the vascularization of the hypoplastic lung and the presence or absence of bronchiectasis.[8] The diagnosis in our patient was made during the first episode of pneumonia, and the findings of the chest tomography, combined with a high index of suspicion, were fundamental.

The primary differential diagnosis is the esophageal bronchus (EB), a bronchopulmonary malformation of the foregut. The main difference is that in EB, communication occurs between an affected lung lobe (usually the upper lobe) and the airway. In contrast, in EL, the entire lung is connected to the esophagus, with no communication with the airway. Other differential diagnoses include pulmonary hypoplasia and agenesis, pulmonary sequestration, cystic adenomatoid malformation, and complex bronchopulmonary malformations of the scimitar syndrome spectrum.

Treatment consists of resection of the affected lung (pneumonectomy), although successful reimplantation of the lung into the trachea

Figure 4. Esophageal lung



Bronchus originating abnormally from the esophagus rather than the trachea (original drawing).

has been reported.[9] In our case, the affected lung already showed images consistent with cystic bronchiectasis, suggesting irreversible parenchymal damage, so resection was decided upon.

The importance of early diagnosis and treatment lies in preventing recurrent pneumonia that can affect the contralateral lung. Given the low incidence of this condition, clinical suspicion is essential for timely diagnosis. ■

REFERENCES

1. Thakur N, Agarwal D, Narayan S, Shukla D. Recurrent Peumonia in an Infant with an Esophageal Lung. *Indian Pediatr*. 2020;57(3):266-7.
2. Sugandhi N, Sharma P, Agarwala S, Kabra SK, Gupta AK, Gupta DK. Esophageal lung: presentation, management, and review of literature. *J Pediatr Surg*. 2011;46(8):1634-7. doi: 10.1016/j.jpedsurg.2011.04.057.
3. Bokka S, Jaiswal AA, Behera BK, Mohanty MK, Khare MK, Garg AK. Esophageal lung: A rare type of communicating bronchopulmonary foregut malformation, case report with review of literature. *J Indian Assoc Pediatr Surg*. 2015;20(2):92-4. doi: 10.4103/0971-9261.151558.
4. Singh VK, Dhua AK, Jana M, Bhatnagar V. Esophageal Lung-Clinical Images. *J Indian Assoc Pediatr Surg*. 2018;23(2):109-10. doi: 10.4103/jiaps.JIAPS_167_17.
5. Srikanth MS, Ford EG, Stanley P, Mahour GH. Communicating bronchopulmonary foregut malformations: classification and embryogenesis. *J Pediatr Surg*. 1992;27(6):732-6. doi: 10.1016/s0022-3468(05)80103-4.
6. Kumar A, Babbar S. Esophageal lung complicated by recurrent pneumonia. *Pediatr Pulmonol*. 2023;58(9):2663-5. doi: 10.1002/ppul.26537.
7. Trisno SL, Higano NS, Kechele D, Nasr T, Chung W, Zorn AM, et al. Case Report: Esophageal bronchus in a neonate, with image, histological, and molecular analysis. *Front Pediatr*. 2021;9:707822. doi: 10.3389/fped.2021.707822.
8. Alem N, Alsufiani H, Alsaadi A, Aljohani S, Arkoubi M. The utility of CT virtual bronchoscopy in the esophageal lung diagnosis: A case report. *Respir Med Case Rep*. 2022;37:101658. doi: 10.1016/j.rmcr.2022.101658.
9. Lallemand D, Quignodon JF, Courtel JV. The anomalous origin of bronchus from the esophagus: report of three cases. *Pediatr Radiol*. 1996;26(3):179-82. doi: 10.1007/BF01405293.