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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.

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 on 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 airspace 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

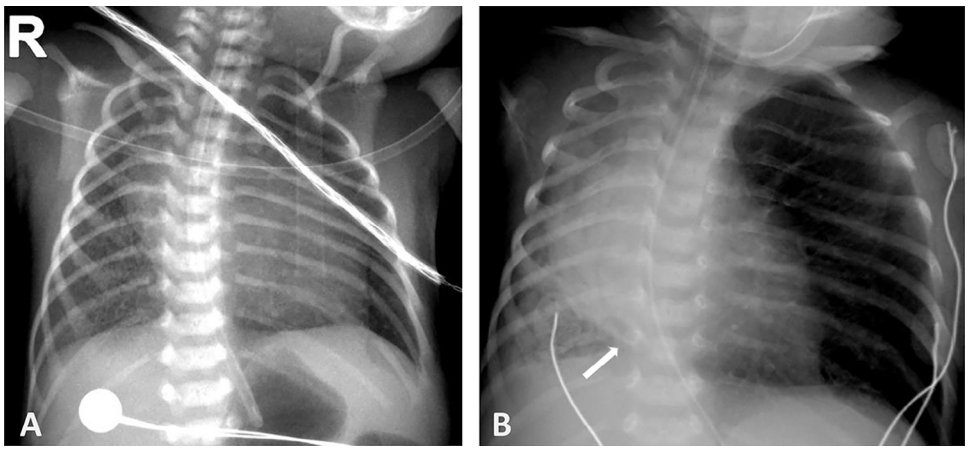

Fig. 1 – Preoperative (A) and postoperative (B) chest radiographs. The preoperative image shows mild interstitial thickening with normal lung aeration and the postoperative image shows opacification of the right lung with a dilated airway directed abnormally to the lower esophagus (arrow).