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Case Report

The utility of CT virtual bronchoscopy in the esophageal lung diagnosis: A case report

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ABSTRACT

The esophageal lung is a variant of the communicating bronchopulmonary foregut malformation (CBPFM). It needs a high index of suspicion for diagnosis because it is a rare condition and does not have specific symptoms. A CT scan or an esophageal contrast study, showing direct communication between the airways and the esophagus or stomach, confirms the diagnosis. Patients with esophageal lung need flexible bronchoscopy for evaluating tracheobronchial anomalies.

We present a three-month-old boy with a right esophageal lung in which the CT virtual bronchoscopy showed an absence of the right main bronchus at the carina level. This case report highlights the importance of CT virtual bronchoscopy as an alternative to flexible bronchoscopy for the diagnosis of tracheobronchial anomalies associated with CBPFM.

1. Introduction

The esophageal lung is a rare congenital malformation in which the main bronchus, most commonly the right, originates from the esophagus with the absence of the ipsilateral main bronchus at the level of the carinal bifurcation. This anomaly is a variant of communicating bronchopulmonary foregut malformations (CBPFM) [1].

The affected lung is typically hypoplastic and will require a pneumonectomy. This procedure can eventually lead to post-pneumonectomy syndrome, especially if it is on the right side [2,3].

CBPFMs can be associated with gastrointestinal, diaphragmatic, vascular, vertebral, and cardiac anomalies.

The diagnosis of CBPFM is challenging because it is a rare malformation that has no specific symptoms. It presents with chronic cough, feeding problems, recurrent chest infections, and shortness of breath in orally fed infants. However magnetic resonance imaging and prenatal ultrasound can detect this anomaly antenatally [4].

Although esophageal contrast and bronchoscopy are used to confirm the diagnosis of esophageal lung, the use of computed tomography (CT) scan with virtual bronchoscopy (VB) as a diagnostic tool has not been emphasized enough in previous reports.

VB is a postprocessing technique to generate high-resolution images of the tracheobronchial tree. It is a breakthrough in CT technology that allows for reconstructing the airways in three dimensions (3D). This technique allows noninvasive visualization of the inside of the tracheobronchial tree in children, reminiscent of a flexible bronchoscopy [5–7].

Here, we report a male infant with a right esophageal lung. The diagnosis of group II CBPFM was confirmed with esophageal

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contrast study and CT virtual bronchoscopy.

2. Case report

The patient was 3 months old, born as a twin preterm at 32 weeks' gestation by cesarian section because of maternal pregnancy-induced hypertension. His Apgar score was 9 and 9 at 1 and 5 minutes, respectively. His birth weight was 1.6 kg, and his head circumference and height were normal for his age. He was admitted to the neonatal intensive care unit because of low birth weight and tachypnea. Initially, started on nasal cannula oxygen, which was weaned into room air the next day. Vital signs were normal. Systemic examination was normal except for left inguinal and umbilical hernias. Orogastric tube feeding began with gradual increment and received a course of IV antibiotics for 7 days. On day 18, he started oral feeding, then experienced apnea with desaturation. The patient started on antibiotics for suspected nosocomial sepsis. His laboratory investigations were normal. Blood culture was negative, but his urine culture was positive for Enterobacter. CXR (Fig. 1) shows total collapse of the right lung with mediastinal shift. The ultrasound of the brain and abdomen was normal, and echocardiography showed dextrocardia and a large PDA with normal cardiac function. He was discharged home in good condition on day 21.

He was admitted with apnea at the age of 40 days. There was no history of fever, cough, vomiting, or choking during feeding. No history of poor feeding or decreased activity. He was active, had no respiratory distress, and his vital signs were normal. A chest examination revealed a decrease in air entry on the right side. Oxygen saturation was 96% in room air. Echocardiography showed dextrocardia. The results of a lumbar puncture were normal. He had no more apnea during hospitalization and was discharged home in good condition.

One month later, he was admitted as aspiration pneumonia when he presented with a history of cough and fever of three days' duration associated with a brief attack of rolling up of eyes. The mother reported having a history of crying during feeding, but without vomiting, choking, or decreased activity. He was active, and not in respiratory distress. His vital signs and oxygen saturation were normal. On auscultation, breath sounds were decreased on the right side compared with the left side. The COVID-19 test, and blood culture were negative. A chest X-ray was performed, which showed total collapse of the right lung with mediastinal shift to the right, and an air bronchogram highlighted the airway orientation, which appeared abnormally pointing downward and medially. A CT scan (Fig. 2) was performed showing total collapse of the right lung in addition to the absence of the right main bronchus. There is marked hypoplasia of the right pulmonary artery compared to the left side. VB (Fig. 3) shows the abrupt blunt end of the right bronchus with normal airway of the left lung. CT did not show clear communication between the esophagus and the lung. However, the upper GI study (Fig. 4) shows the contrast opacified airway communication between the esophagus and the right main bronchus at the lower part of the esophagus with focal airway stenosis at the site of communication.

An echocardiography showed evidence of mild pulmonary hypertension with an estimated PAP of 30 mm Hg in addition to

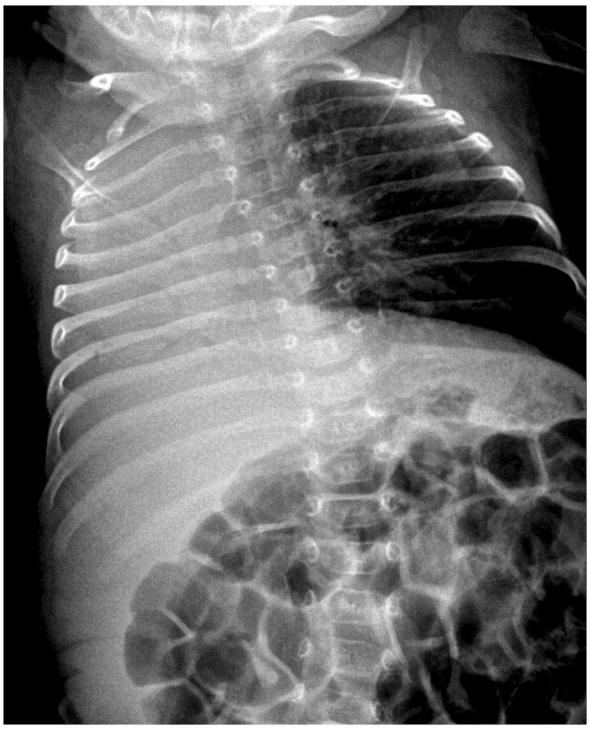

Fig. 1. Chest X-ray shows a total collapse of the right lung with mediastinal shift. Note the convergence and downward orientation of the right lung airway toward the lower esophagus.

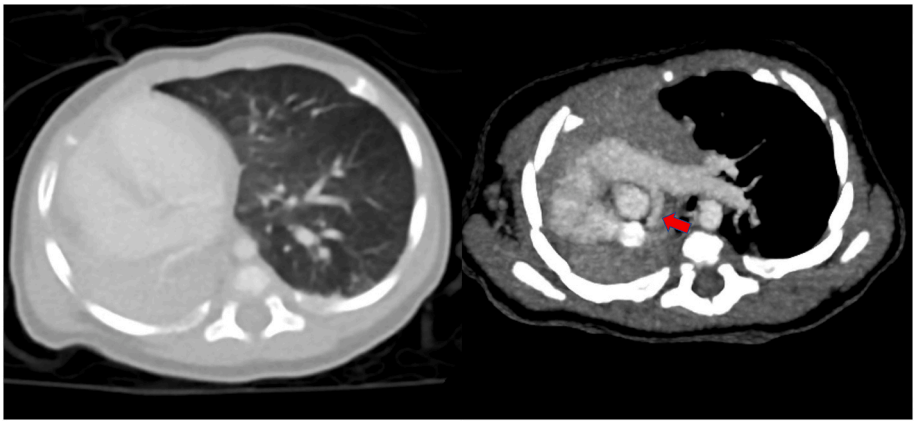

Fig. 2. The chest CT scan shows total collapse of the right lung with marked hypoplasia of the right pulmonary artery (red arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

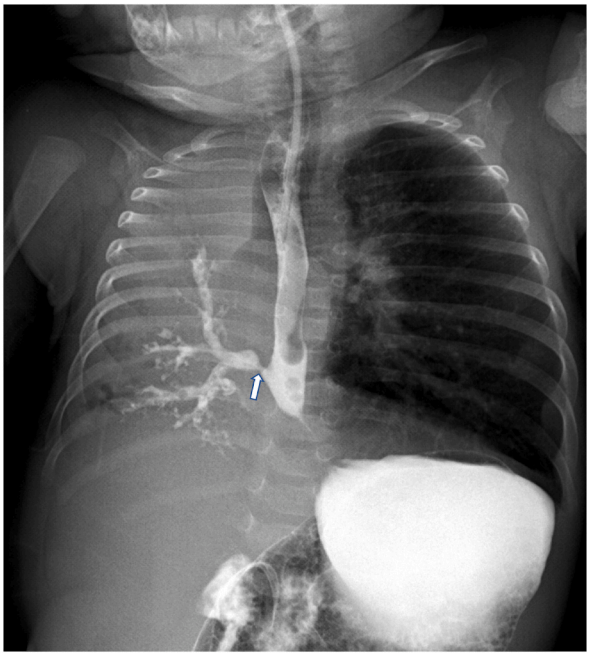

Fig. 3. An upper GI study shows abnormal direct communication between the right main bronchus and esophagus at its lower part with focal stenosis at the site of communication (arrow).

dextrocardia. He was treated with intravenous antibiotics and diagnosed with an isolated right esophageal lung. Both parents were informed and educated about the diagnosis and surgical intervention that this patient required, but unfortunately the parent refused surgery despite multiple counseling. He was discharged from the hospital in stable condition, tolerating oral feeding in his left lateral position to reduce the risk of aspiration to the right esophageal lung. He is currently 17 months old, on a regular follow-up in the clinic, and has two lower respiratory tract infections. His growth parameters were normal, and there was no major respiratory compromise.

The other twin was diagnosed with total anomalous pulmonary venous drainage. He underwent corrective surgery at the age of 3 months, but unfortunately died of multiorgan failure on postoperative day 5. The mother is 42 years old. She had regular antenatal care and was diagnosed with pregnancy-induced hypertension and gestational diabetes. There was no parental consanguinity. The other siblings are healthy, except for one who has Down syndrome and associated congenital heart disease.

3. Discussion

The esophageal lung is a rare congenital malformation in which there is an abnormal origin of the right or left lung from the lower esophagus. The esophageal lung is more common on the right side [8]. When the esophageal lung is associated with airway stenosis, it results in long-term morbidity. Although a patient with esophageal lung has nonspecific symptoms and can mimic other lung diseases,

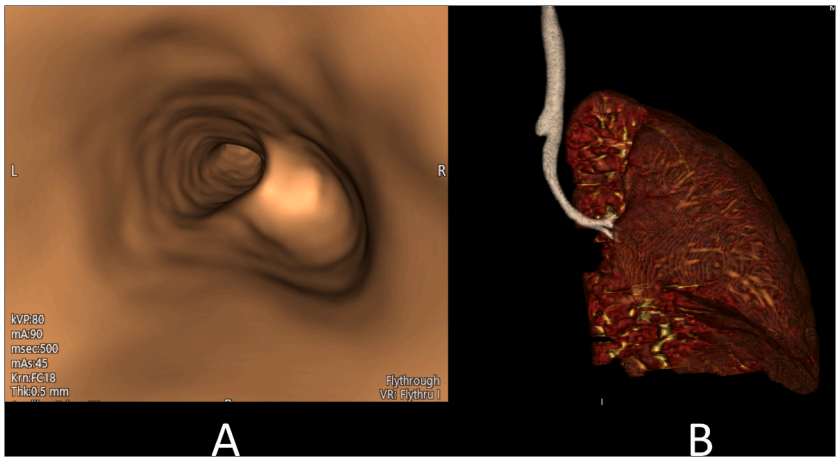

Fig. 4. CT Virtual bronchoscopy (A) with 3D reconstruction of the airway (B). Note the blind end of the right main bronchus with normal left main airway structures.

it should be suspected in a patient with persistent unilateral lung collapse, especially when presented with respiratory distress, cough, and recurrent pneumonia after feeding. One clue to the diagnosis on chest radiographs is to observe the direction of airway convergence, whether it is toward the hilum in normal cases or directed downward and medially toward the esophagus (Fig. 1).

Previous studies showed that CBPFM can be associated with other congenital anomalies like VACTERL (vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities), pulmonary artery sling, esophageal atresia, duodenal atresia, and laryngeal cleft [9–11]. However, our case did not have any of these associated anomalies.

Early diagnosis of the esophageal lung is important to prevent complications related to recurrent aspiration and infection. Diagnosis is suspected when parenchymal opacification is seen on chest radiographs and is confirmed by esophageal contrast study and bronchoscopy. An esophageal contrast study is the modality of choice to demonstrate direct communication between the bronchial tree and the esophagus, but its diagnostic utility is limited in group I CBPFM due to proximal esophageal atresia. Therefore, the diagnosis requires preoperative flexible bronchoscopy or intraoperative esophageal contrast study. A chest CT scan is important for mapping the vascular supply and drainage of the lung, detecting airway malformations, and evaluating the degree of parenchymal disease [12]. Preoperative echocardiography is an important tool for detecting associated cardiovascular malformations [13].

Our case falls under group II according to Srikanth's classification [8], in which the whole right lung communicates with the lower esophagus with the ipsilateral absence of the right main bronchus from the trachea. This patient presented with recurrent infection of the lower respiratory tract with consolidation of the right lung. The diagnosis was confirmed by the presence of abnormal communication between the lower esophagus and the right lung on the esophageal contrast study, and the absence of the right main bronchus at the level of the carina was confirmed by chest CT with 3D reconstruction and virtual bronchoscopy. Hypoplasia of the right pulmonary artery is present, but otherwise no other associated vascular abnormalities are seen.

Here, we emphasize the importance of preoperative flexible bronchoscopy or VB to be included in the diagnostic approach for CBPFM to assess associated airway abnormalities, as was shown by Atzori et al. [14] in their study, preoperative bronchoscopy detected abnormalities in the respiratory system, changing surgical plan in one quarter of patients. Moreover, our case also demonstrated that the images obtained using VB (Fig. 4) clearly showed the absence of the right main bronchus at the expected location, and thus it obviates the need for flexible bronchoscopy.

VB is an alternative modality to flexible bronchoscopy for the diagnosis of CBPFM in an unstable neonate who cannot tolerate flexible bronchoscopy and cannot tolerate the esophageal contrast study, which bears the risk of leaking into the airways [15]. VB can also evaluate the airways beyond obstruction, which cannot be done by flexible bronchoscopy [5,6,16,17]. Another advantage of VB it does not require general anesthesia and the expertise needed for flexible bronchoscopy. Other potential diagnostic roles of VB are in broncho-esophageal fistulas, post-lung transplantation anastomosis, suspected foreign body aspiration, and inhalation injuries. Virtual bronchoscopy has the disadvantage of not being able to detect small mucosal lesions, and patients will be exposed to radiation. Other drawbacks of VB include its inability to determine mucosal architecture, vascularity, color, and false positive findings caused by secretions and artifacts [7,18,19].

Treatment of the esophageal lung involves resection of the hypoplastic lung. This procedure is well tolerated in newborns and infants, but has long-term complications such as chest wall asymmetry, scoliosis, and post-pneumonectomy syndrome, especially if performed on the right side. It can be done by open thoracotomy or by thoracoscopy. Another surgical option is bronchial reimplantation [20–23].

Post-pneumonectomy syndrome occurs because of rotation of the heart and mediastinal shift to the side of the pneumonectomy, and this leads to obstruction of the large airways and blood vessels, which may lead to respiratory failure, dyspnea, and stridor. Physicians who care for these patients must have a high index of suspicion of this complication, as it can appear months or years after pneumonectomy. It is more common following right pneumonectomy, especially in infants, because of the rapid growth of their thoracic cage, the high elastic recoil and compliance of the lung and mediastinum [24]. Treatment of this complication usually

involves correcting the mediastinal shift. Correcting the mediastinal shift achieved by installing the expandable prosthesis prophylactically at the time of pneumonectomy or as therapeutic when the patient has symptoms of post-pneumonectomy syndrome [2,3,25–27]. Here, we recommend a right pneumonectomy with the installation of an expandable prosthesis to prevent post-pneumonectomy syndrome, but the parents, unfortunately, refused the surgery.

4. Conclusions

This case report highlights:

- Esophageal lung should be suspected in a patient with persistent unilateral lung collapse, especially when associated with respiratory distress, cough, and recurrent pneumonia after feeding.
- The importance of CT virtual bronchoscopy in the diagnostic approach for respiratory tract anomalies associated with CBPFM.

Informed consent

We have obtained written informed consent for the publication of this manuscript and the accompanying images.

Declaration of competing interest

We declare that we have no conflict of interest.

Acknowledgments

We are grateful to the parents for consent to the publication of the case.

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