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Bronchiectasis without lower respiratory symptoms in the presence of multisystem anomalies – a clinical clue to diagnose esophageal lung anomaly

Amit Gupta[1], Aman Snehil[2], Sujeet Kumar[1], Roshan Chanchlani[1], Reyaz Ahmad[1], Keerti Swarnkar[3], Garima Goel[4]

[1]Department of Pediatric Surgery; [2]Department of Radiodiagnosis; [3]Department of Pediatrics; [4]Department of Pathology, All India Institute of Medical Sciences, Bhopal, Madhya Pradesh, India

Correspondence: Amit Gupta, Department of Pediatric Surgery, All India Institute of Medical Sciences, Room no.1004, Academic Block, Hospital Building, Bhopal 462020, Madhya Pradesh, India.
E-mail: amitpedsurgeon@gmail.com

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Abstract

Esophageal lung is a type of Group-II communicating bronchopulmonary foregut malformations (CBPFM) usually diagnosed beyond neonatal period during investigation for recurrent respiratory symptoms and persistent radiographic features suggesting pneumonia or bronchiectasis. In our case, we noticed bronchiectasis and disproportionately severe volume loss in an infant with associated multisystem anomalies in the absence of "significant" lower respiratory tract symptoms. A detailed evaluation with repeat imaging confirmed a Group-II CBPFM, a congenital pathology instead of an infective cause. Pneumonectomy is a more prudent option instead of undertaking major airway reconstruction for the dysplastic "dysfunctional" tissue. Amongst the various associated anomalies reported till now, the associated rib and renal anomalies noted by us have not been described earlier to the best of our knowledge.

Introduction

Communicating bronchopulmonary foregut malformations (CBPFM) are a spectrum of rare congenital anomalies characterized by communication between esophagus or stomach and an "isolated" portion of respiratory tract [1]. Srikanth *et al*. classified CBPFM into four groups [2]. We describe an infant with Group-II CBPFM, also referred to as *esophageal lung*, where the main bronchus of the affected lung arises from the distal esophagus instead of trachea. The patient was referred with a contrast enhanced computed tomography (CECT) chest showing bronchiectasis on the affected side. The associated multiple skeletal and renal anomalies are being reported for the first time to the best of our knowledge.

Case Report

A 15-month-old girl weighing 7.9 kg (<25th centile), presented to Pediatrics Department with recurrent episodes of high-grade fever and failure to thrive since 1-month of age. However, during any of these episodes, she never had significant lower respiratory symptoms in the form of cough, cold, hemoptysis or labored breathing with chest retractions which indicate lower respiratory tract infections or inflammatory pathology. Right hemithorax was small with crowding of ribs and scoliosis, dull on percussion and breath sounds were absent; heart sounds were shifted to right side. Serial chest X-rays during these episodes showed volume loss in right hemithorax, a persistent multi-loculated cystic lesion nearly constant in extent, marked ipsilateral mediastinal shift, crowding of ribs, scoliosis and a hemivertebra at L-1. An ultrasonography (USG)

abdomen had shown non-visualized left kidney and a CECT chest suggested hypoplastic right lung with cystic bronchiectatic changes. The child was referred to Pediatric Surgery for further management. Bronchiectatic changes on CECT with significant volume loss were incongruent with the absence of significant lower respiratory symptoms leading to a diagnostic dilemma. Therefore, a repeat CECT Chest was done which revealed following interesting findings to reach a diagnosis: i) The right main bronchus arising from the lower esophagus (D-8 vertebral level) confirmed on administering oral contrast and the trachea continued into left main bronchus (D-5 vertebral level) without any division to the right; ii) near total collapse/consolidation of right lung with bronchiectatic changes; iii) arterial supply from a diffusely attenuated right pulmonary artery with absent superior branch and venous drainage by an inferior pulmonary vein; the superior pulmonary vein was not seen; iv) ipsilateral mediastinal shift; and v) compensatory hypertrophy of the left lung (Figure 1 a,b). An upper gastrointestinal (UGI) contrast study demonstrated spillage into the right main bronchus & its divisions through a communication from lower esophagus (Figure 1c, Video clip). A diagnosis of CBPFM Group-II was made. Additionally, the CT also showed 13 pair of ribs and multiple vertebral segmentation anomalies involving C-3/C-4 and D1/D-2 with scoliotic deformity (Figure 2 a,b). Since the left kidney was not visualized on USG Abdomen, further evaluation with CECT abdomen showed inferior crossed fused ectopia of left kidney (Figure 2b). 2-D Echocardiography showed normal cardiac anatomy and function. A right pneumonectomy was done with intercostal drainage (ICD). Per-operatively right lung was small and densely adherent to chest wall with a single lobe and liver like consistency without any aeration; vascular supply was from single hypoplastic artery and vein; the bronchial communication (dysplastic right bronchus) with esophagus was disconnected and esophageal defect was repaired (Figure 3 a-c). The histopathology revealed fetal lung histology with alveolar spaces lined by cuboidal epithelium and dysplastic bronchus with cartilage (Figure 3 d,e). The patient was started on nasogastric feeds from second postoperative day (POD). The ICD tube was removed and full oral feed was started on POD-8 after confirming no leak from esophageal repair on UGI contrast study. At 6-months follow-up, the patient is thriving well with catch up growth evident by weight gain (10.8 kgs; 50th centile).

Discussion

Bronchopulmonary foregut malformations (BPFM) are a spectrum of congenital cystic lung malformations including congenital pulmonary adenomatoid malformations (CPAM), pulmonary

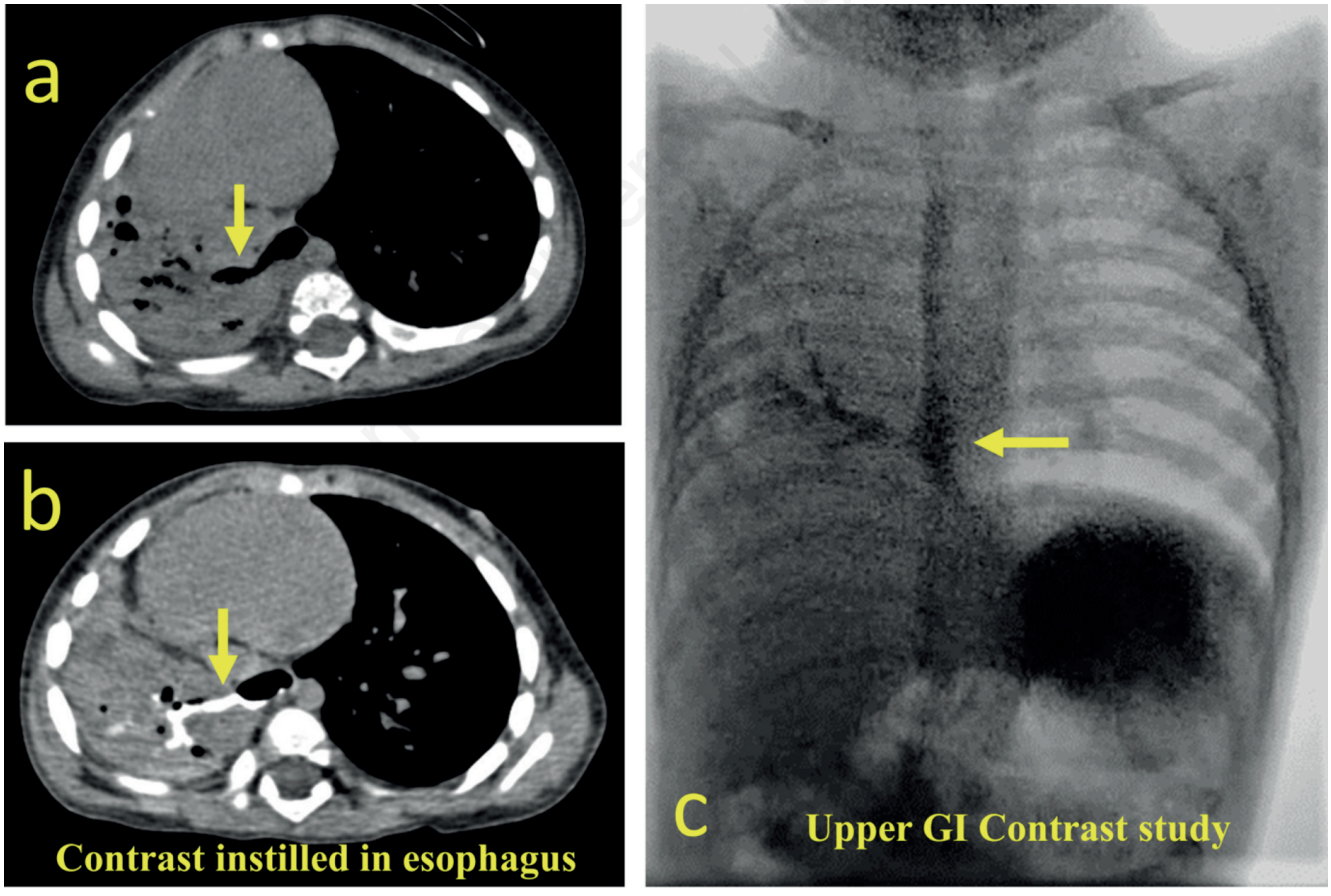

Figure 1. a) Right main bronchus noted at a lower level arising directly from lower esophagus at D-8 vertebral body approximately 2.8 cm proximal to gastroesophageal junction; the bronchial diameter at its origin measures approximately 8 mm. b) On oral contrast administration, there is opacification of esophagus with spilling of contrast into the right main bronchus and its divisions; this likely represents an esophageal bronchus, Group-II type of communicating bronchopulmonary foregut malformation. c) Upper gastrointestinal contrast study demonstrating the spillage and arborization of contrast from the lower esophagus on the right-side suggesting communication with the right airway.

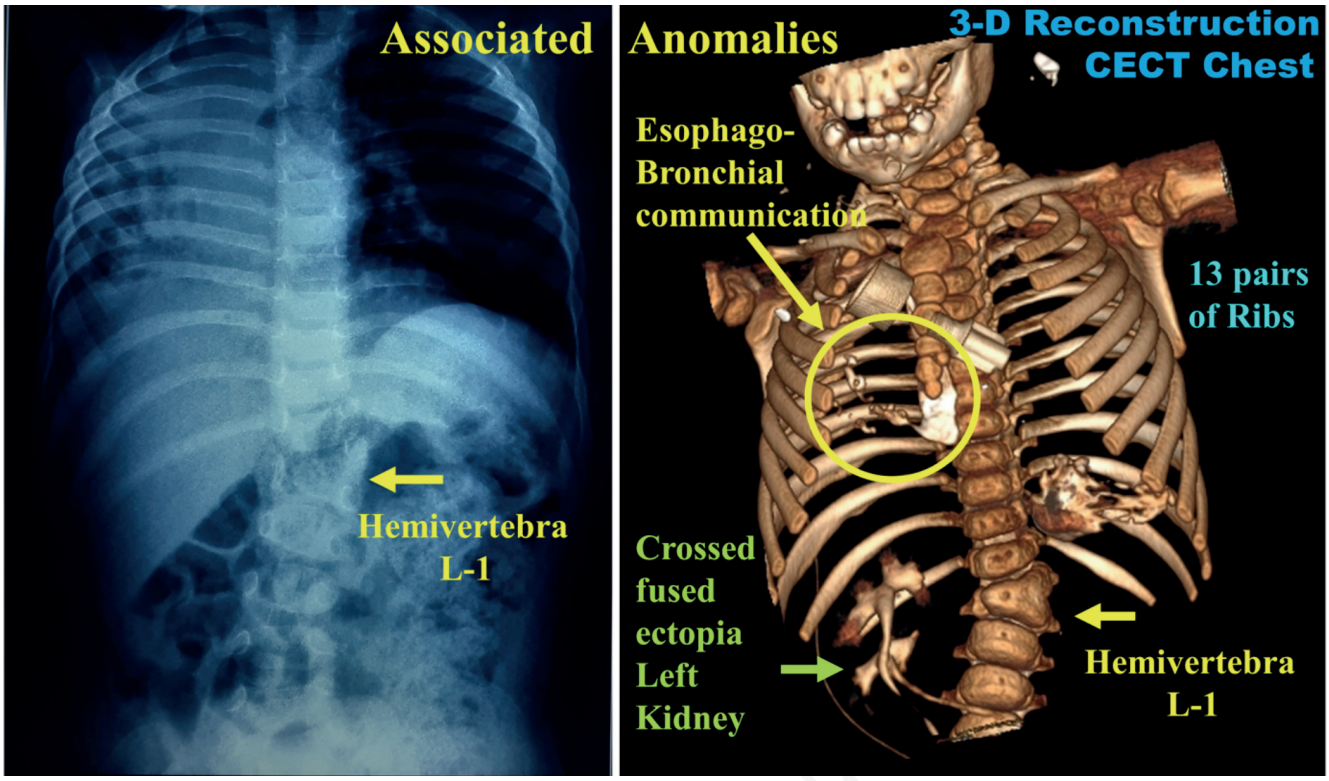

Figure 2. Associated anomalies. Lumbar vertebral anomaly on plain X-ray, 13 pairs of ribs and crossed fused ectopia of left kidney on 3-D reconstruction of contrast enhanced computed tomography chest and abdomen.

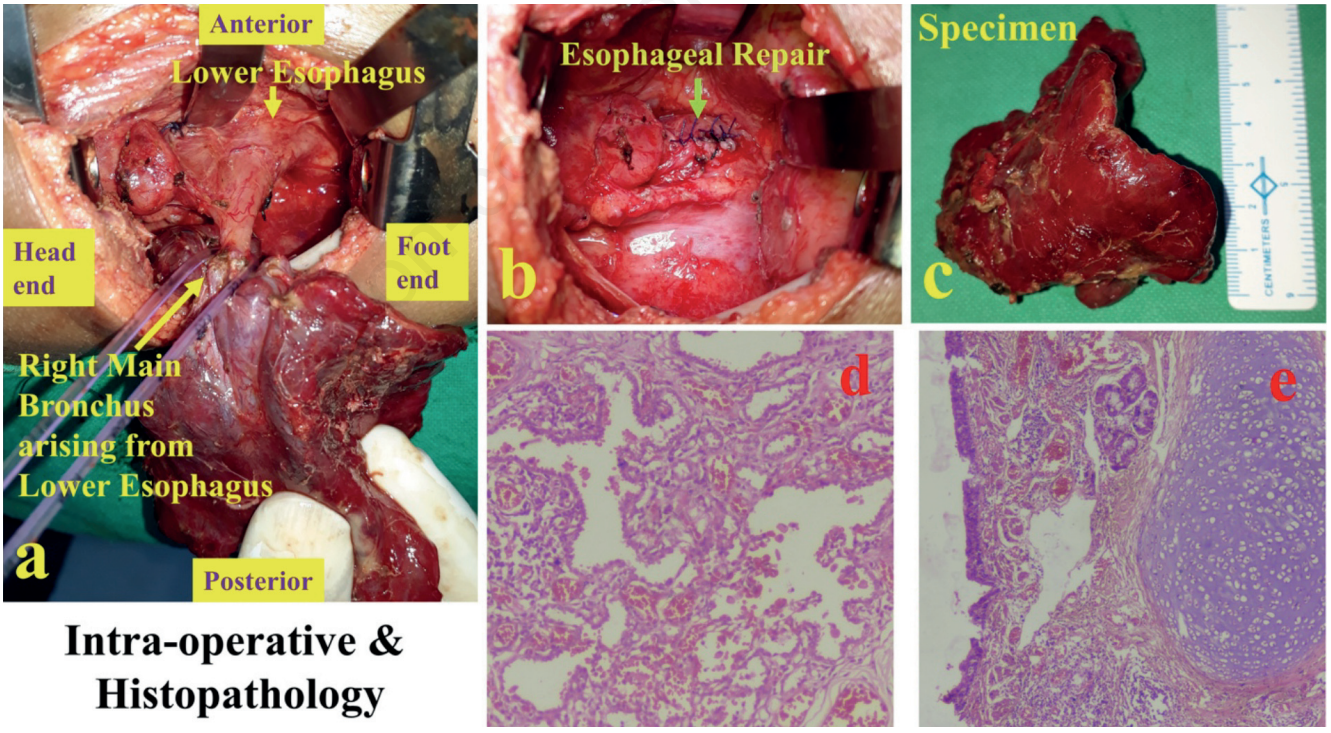

Figure 3. a) Right posterolateral thoracotomy demonstrating the dysplastic right lung with the main bronchus, looped in the feeding tube, arising from the lower esophagus. b) Repaired defect in the lower esophagus following disconnection of the right main bronchus during pneumonectomy of the dysplastic right lung. c) Resected specimen of right pneumonectomy; the blood supply was from single hypoplastic pulmonary artery and vein. d) Histopathology of dysplastic right lung showing alveolar spaces resembling fetal lung tissue. e) Histopathology of dysplastic bronchiole lined by respiratory epithelium and cartilage.

sequestration, duplication cysts and tracheoesophageal fistula or broncho-esophageal fistula [5]. CBPFM results from anomalous communication between esophagus or stomach and bronchial tree [2,3]. The embryogenesis proposed by Srikanth et al describes the communication to result from a focal mesodermal defect in the tracheoesophageal groove and the involved portion of lung may be pulled away by the differential elongation of the esophagus, isolating it from the main respiratory tract [2]. There is a strong predilection of this anomaly to occur on the right side due to the proximity of the right main bronchus to the esophagus [2,4,6,7]. Esophageal lung refers to a variant where the entire lung is dysplastic and communicates with the esophagus through its main bronchus.

Srikanth *et al*. classified CBPFM into 4 groups based on its association with esophageal atresia and tracheoesophageal fistula (EA/TEF), the portion of bronchus involved in the communication, isolation of the portion of lung served by the bronchus and blood supply (they also noted the incidence) as follows: Group-I (16%): These patients always have EA/TEF associated with group-II anatomy (IA) or group-III anatomy (IB) as described. Group-II (33%; the second most common): The "isolated" entire lung communicates through the main bronchus (esophageal lung). Group-III (46%; the most common): a lobar or segmental bronchus connects the respective isolated anatomic portion of lung supplied by pulmonary artery, aorta or both. Group IV: (5%; the less common). Communication with a bronchus of structurally normal bronchial tree, but the portion of the lung served by the involved bronchus receives systemic blood supply [2].

Diagnosis of group-I is made in association with EA/TEF. Other varieties are usually diagnosed later as they present with a background of recurrent respiratory symptoms and non-resolution of opacity on plain chest radiograph. However, antenatal diagnosis with fetal MRI has also been reported [8]. The index case presented here is a Group-II malformation.

On a detailed analysis of clinical presentation, the following important clinical features emerged as strong indicators of diagnosis: i) Bronchiectasis without any respiratory symptoms and normal appearing contralateral lung with compensatory hypertrophy; ii) associated anomalies evident on serial X-ray chest, USG and CECT. The dysplasia in right lung appeared as bronchiectasis, right main bronchus joined the distal esophagus and the trachea continued into left mainstem bronchus without any division at carina. The right lung was poorly aerated, small and solid with no lobulations supplied by hypoplastic pulmonary vessels. Associated compensatory hypertrophy of the left lung without emphysema was an additional corroboratory finding for a congenital lesion [9]. The association with 13 pairs of ribs has been noted for the first time.

To the best of our knowledge, only 63 cases of CBPFM have been reported in English literature till date, of which only 18 cases are group-II [8]. The highlighting features in our case were the absence of lower respiratory symptoms in clinical presentation and the associated anomalies not described earlier.

"Isolation" of the malformed right lung from the normal tracheobronchial tree prevented irritation of the respiratory tract, likely accounting for the absence of respiratory symptoms in our case. Bronchiectasis on imaging may be explained by dysplasia and chronic inflammation due to spillage of the esophageal contents producing constitutional symptoms (recurrent episodes of fever, loss of appetite and failure to thrive) [10]. Considering the significant volume loss disproportionate to the short duration of postnatal illness, the onset appears to be prenatal resulting in small dysplastic lung.

Multiple congenital anomalies have been reported in association with CBPFM. Group-I is always associated with EA/TEF [6,9,11-13]. In the index case, we noticed following associated anomalies - scoliosis with vertebral segmentation anomalies C-3/4 and D-1/2, 13 pairs of ribs and inferior crossed fused ectopia of left kidney. No such combination of anomalies has been reported till now in association with CBPFM.

Yang et al. conducted a systematic review of 61 cases. Although female predominance has been quoted in literature, they found no sex predilection. Right side was predominant (72%) with majority of patients being neonates (62%) and <1-year age (75%). Cardiovascular anomalies were the most common (18%) associated malformation followed by VACTERL (V=vertebral, A=anorectal, C=cardiac, T=tracheal, E=esophageal, R=renal, L=limb anomalies) association (10%), skeletal malformation (10%), diaphragmatic hernia (3.3%) and anorectal malformation (3.3%) [7]. Plain chest radiograph showed opacification of ipsilateral lung and mediastinal shift uniformly in all (n=18;100%) group-II CBPFM, the diagnosis of CBPFM was confirmed by UGI contrast study in 62%, CT in 11.5%, bronchoscopy in 1.6% and intraoperatively in 13%; five cases (8.2%) were diagnosed prenatally on fetal MRI [8].

Pneumonectomy is the mainstay of treatment for esophageal lung and is well-tolerated [8,11]. There are also reports of successful thoracoscopic pneumonectomy [14]. The term "pneumonectomy" per se may raise concern regarding post-pneumonectomy syndrome, but the reports of its occurrence in relation to CBPFM are very few [15]. In the systematic review by Yang *et al*., only one out of thirty-one patients with group1A/II CBPFM developed post-pneumonectomy syndrome [8]. It appears that the associated hypertrophied contralateral lung occupies most of the mediastinum already establishing a state of equilibrium from prenatal period and the removal of hypoplastic affected lung occupying very little space does not result in significant mediastinal shift to result in post-pneumonectomy syndrome. Our patient also remained absolutely stable postoperatively. There are reports of successful tracheobronchial reconstructions [16]. Developmentally anomalous bronchus and dysplastic "dysfunctional" lung may not be suitable for the tracheobronchial reconstruction [17]. In the recent systematic review by Yang *et al*., the results of reported reconstruction were unsatisfactory [8]. Therefore, pneumonectomy seems to be a more suitable surgical option compared to a major reconstruction of tracheobronchial tree with dysplastic tissues except in very selective patients [8,11,17].

Conclusion

Through this case report we wish to highlight that bronchiectasis on imaging in early childhood with absence of significant lower respiratory symptoms in the presence of other congenital anomalies strongly indicates a diagnosis of esophageal lung. It should alert the radiologist to look for characteristic imaging features of esophageal lung. The associated anomalies noted by us have not been reported earlier. Although UGI contrast study is confirmatory, CECT or MRI reveal anatomic details and also detect associated malformations. Pneumonectomy is a more prudent option than a major tracheobronchial reconstruction for a congenitally malformed lung and bronchus with uncertain functional capacity.

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