



Case report

An atypical case of congenital lobar emphysema in an adult, non-smoker patient presenting with pneumothorax

Zekarias T. Asnake^{*}, Joshua K. Salabei, Jordan Pierce, Angela Fernandez, Ramin Ahmad, Zeeshan H. Ismail, Calestino Mathew

University of Central Florida College of Medicine, Graduate Medical Education, North Florida Regional Medical Center, Internal Medicine Residency Program, 6500 W Newberry Rd, Gainesville, FL, 32605, USA

ARTICLE INFO

Keywords:

Congenital lobar emphysema

CLE

Video assisted thoracoscopy surgery

VATS

Pneumothorax

ABSTRACT

Congenital lobar emphysema (CLE) is a developmental anomaly of the lower respiratory tract characterized by hyperinflation of one or more pulmonary lobes in the absence of extrinsic bronchial obstruction. We present a case of a 24-year-old male, nonsmoker who presented with shortness of breath and severe left sided chest pain. A chest x-ray was significant for a very large left-sided pneumothorax and chest CT showed lobar emphysematous changes. Video assisted thoracoscopic surgery (VATS) and lobectomy was subsequently performed after persistence of pneumothorax despite chest tube insertion and conservative management. Surgical pathology of resected specimen showed chronic emphysematous changes with patchy chronic organizing pneumonitis. Histology showed advanced emphysematous changes of pulmonary parenchyma consistent with congenital lobar emphysema. This finding combined with features seen on computed tomography of the chest led to the diagnosis of congenital lobar emphysema. This case demonstrated that CLE can be a cause of tension pneumothorax in adults in rare cases.

1. Introduction

Congenital lobar emphysema (CLE) is a rare developmental lung anomaly, characterized by hyperinflation of one or more pulmonary lobes, most commonly the left upper lobe. It is typically detected in the neonatal period, although rare cases have been reported in early adulthood [1]. CLE affects males more than females (ratio of 3:1) and is commonly associated with bronchial abnormalities/obstruction and vascular dysfunction, resulting in ventilation-perfusion mismatch, hyperinflation, and dyspnea [2]. We report a rare case of CLE diagnosed in an adult non-smoker who presents with respiratory distress caused by a tension pneumothorax. This is a peculiar presentation of CLE in adulthood which has only been rarely reported to date [3].

2. Case presentation

A 24-year-old male with no significant past medical history presented to the emergency department with shortness of breath and left side chest pain without any precipitating event. He denied any fever, cough, or trauma such as barometric injury, including recent skydiving

and airplane travel. He also denied cigarette smoking, vaping, and history of lung infection. Family history was not pertinent. Physical exam on presentation was significant for a heart rate of 108 beats per minute, blood pressure of 123/72, temperature of 98.2 Fahrenheit, respiratory rate of 46 breaths per minute, and oxygen saturation of 87% on room air. Pulmonary examination revealed absent breath sounds on the left upper chest with good air entry and normal breath sounds on the right. Laboratory values on presentation are shown in Table 1. Of note, alpha-1 antitrypsin level was normal. Chest x-ray (CXR) at presentation showed a very large left side pneumothorax (Fig. 1A). An emergency chest tube was placed leading to improvement of symptoms. However, a follow up CXR done after chest tube insertion showed persistence of pneumothorax. (Fig. 3) Computed tomography (CT) of the chest showed emphysematous changes in the left upper lobe (Fig. 1B). The patient then underwent successful video assisted thoracoscopic surgery (VATS) with left upper lobectomy. Intraoperative inspection of the lung was notable for 80–90% of the left upper lobe demonstrating emphysematous changes. The diseased lobe was resected (Fig. 2A) and submitted to pathology. Macroscopic and microscopic tissue analyses were consistent with chronic emphysematous changes with patchy chronic organizing

^{*} Corresponding author. UCF College of Medicine HCA GME Consortium, 6500 W Newberry Rd, Gainesville, FL, 32605, USA.

E-mail address: Zekarias.Asnake@hcahealthcare.com (Z.T. Asnake).

Table 1
Pertinent laboratory data at the time of presentation.

Lab values	Level on admission	Reference range
WBC	6.8	(4.5–11.0 × 10 ³ /mm ³)
RBC	5.32	(4.40–5.90 × million/uL)
Platelet count	199	(150–450 × 10 ³ /mm ³)
Sodium	141	(136–145 mmol/L)
Potassium	3.3	(3.5–5.1 mmol/L)
Blood urea nitrogen	9	(7–18 mg/dL)
Creatinine	0.92	(0.55–1.30 mg/dL)
AST	15	(15–37 units/L)
ALT	24	(12–78 units/L)
Troponin	0.017	(0.000–0.045 ng/mL)
Alpha-1 antitrypsin	135.9	(90–200 mg/Dl)
INR	1.1	(0.9–1.1)
D-dimer	141	(215–500 ng/mL)

WBC-White blood cell count; RBC-Red blood cell count; AST-Aspartate transaminase; ALT-Alanine transaminase; INR-International normalized ratio.

pneumonitis and foci of compression atelectasis with no evidence of malignancy (Fig. 2B and C). The patient improved clinically post-surgery and he was subsequently discharged in stable conditions. Follow up evaluation at our outpatient pulmonology clinic three months after discharge revealed patient remained stable.

3. Discussion

The pathophysiology of CLE is not fully understood. The most identified factor in 25% of the patients is congenital bronchial cartilage defects [4]. Meanwhile, the remaining 75% of cases are caused by multiple causes associated with intrinsic or extrinsic obstruction, such as mucus plugging, vascular anomalies, mucosal folds, and/or intrathoracic masses [4]. The polyalveolar lobe theory, where there is an increase in the number of alveoli per lobe, has been proposed to underlie the pathophysiology of CLE [5].

CLE commonly affects the left upper lobe, and one study showed that 60% of cases had left upper lobe involvement and 29% had right middle lobe emphysema [6]. CLE has a wide range of presentations, and it poses a diagnostic and therapeutic dilemma [7,8]. Nearly half of patients are symptomatic at birth, while the other half mostly develop symptoms in the first 6 months of life. In infancy, wheezing, chronic cough, and recurrent respiratory tract infections can be seen [9]. Respiratory distress occurs because of over distension of the lobe with compression

of the healthy portions, and in some cases, compression of the contralateral lung. Therefore, early detection and management are vital to prevent mortality in those patients. Rarely, CLE can be diagnosed in young adults and these patients can be mistakenly diagnosed with pneumothorax and pneumonia instead of CLE [10]. The initial diagnostic choice for CLE is with plain chest radiography which shows hyperinflation of the affected lobe. CT scans are useful for confirmatory purposes. Other imaging modalities include magnetic resonance imaging and ventilation perfusion scan, but they are only adjunct to the diagnosis of CLE.

Pulmonary function tests in CLE can be normal in asymptomatic or mild disease, but obstructive pattern is a feature of moderate to severe disease.

The accepted treatment modality of CLE is resection of the affected lobe which is usually curative. Early surgery is recommended to prevent the complications of lobar over distension and video assisted thoracoscopic surgery (VATS) seems to be an emerging and advantageous approach. However, there are reports of successful conservative approach that can be used in stable and asymptomatic patients, which can lead to improvement thus avoiding surgery [11]. However, further studies are required to determine the long-term outcome of conservative management, as the disease is rare in adults.

CLE is usually diagnosed in infancy and, rarely, in adults. When diagnosed in adults, the presenting symptom is usually respiratory distress caused by ventilation-perfusion mismatch as a result of compression, caused by hyperinflation of one or more emphysematous lobes, atelectasis on the ipsilateral or contralateral lung and causing mediastinal shift [2]. In rare cases, the formation of overt pneumothorax can occur. In such cases, a clear demarcation by the pleural membrane will be seen on CXR (Fig. 1 A). Also, the onset of symptoms caused by a pneumothorax is sudden onset, as opposed to the gradual progression of symptoms usually seen in respiratory distress caused by compression atelectasis in CLE. Thus, our case is unique in that CLE was diagnosed in adulthood and the presenting symptom of respiratory distress was caused by a pneumothorax. This highlights that CLE should be considered in adult nonsmokers presenting with a pneumothorax and with negative findings of other possible connective tissue disease involvement such as the lack of alpha-1 antitrypsin deficiency.

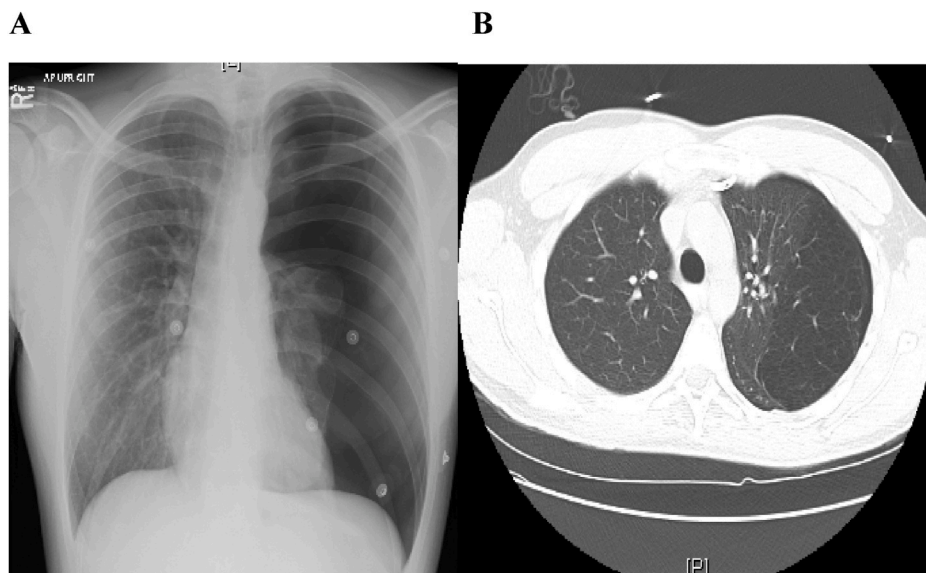


Fig. 1. (A) Initial anterior-posterior chest radiograph at presentation showing very large pneumothorax on the left. (B) Computerized tomographic axial image revealing emphysematous changes in the left upper lobe. In contrast, the pulmonary parenchyma of the right lung is normal in appearance.

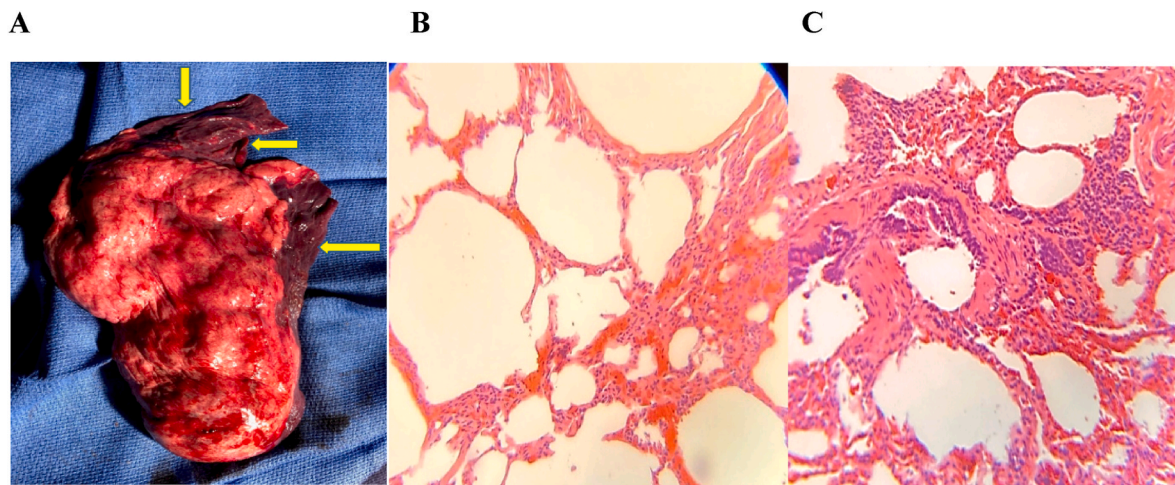


Fig. 2. (A) Gross specimen of the left upper lobe status post lobectomy. The yellow arrows delineate the only remaining healthy lung tissue. The remainder of the specimen is abnormal and representative of gross anatomy consistent with chronic emphysematous disease. (B) Hematoxylin & eosin stain 10 × magnification demonstrating moderate emphysematous changes with inflammatory neutrophilia (pneumonitis) noted in lung tissue. (C) Hematoxylin & eosin stain at 45 × magnification showing advanced emphysematous changes of pulmonary parenchyma in congenital lobar emphysema.

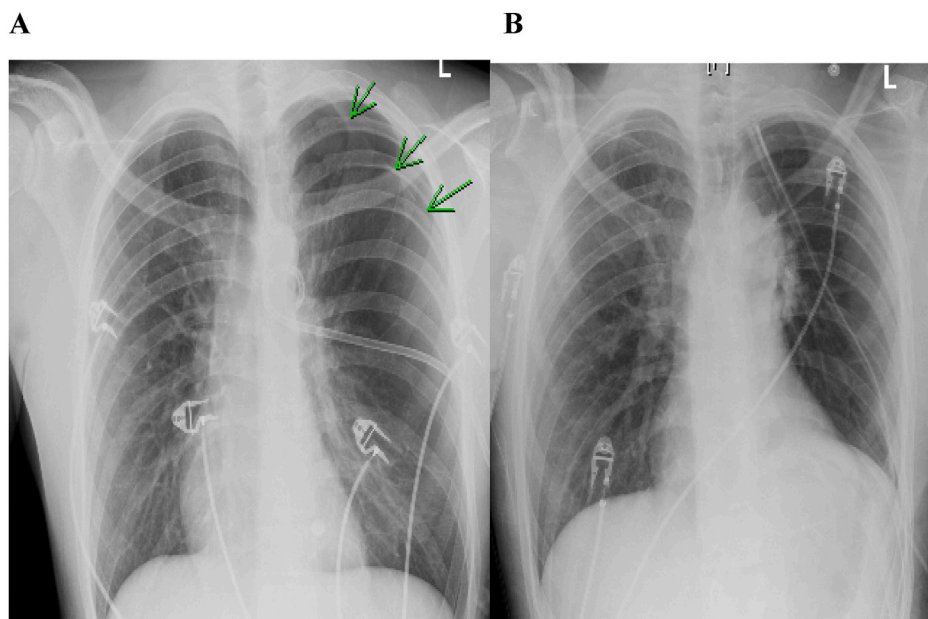


Fig. 3. (A) Post chest tube CXR showing persistent left apical pneumothorax. Green arrows indicating pleural demarcation. (B) Post VATS and lobectomy CXR showing reduced left lung volume with left side chest tube in place and resolution of pneumothorax.

Disclaimer

This research was supported (in whole or in part) by HCA Healthcare and/or an HCA Healthcare affiliated entity. The views expressed in this publication represent those of the author(s) and do not necessarily represent the official views of HCA Healthcare or any of its affiliated entities.

Declaration of competing interest

None.

References

- [1] M. Khalid, B. Khan, Congenital lobar emphysema in adult: a rare case report, *Respiratory Medicine CME* 3 (2010) 150–152.
- [2] C.D. Kennedy, P. Habibi, D.J. Matthew, I. Gordon, Lobar emphysema: long term imaging follow up, *Radiology* 180 (1991) 189–193.
- [3] T. Muramatsu, M. Furuichi, T. Nishii, S. Ishimoto, M. Shiono, Lobar emphysema with pneumothorax in an adult: report of a case, *Surg. Today* 43 (2013) 539–541.
- [4] M. Sadaqat, J.A. Malik, R. Karim, Congenital lobar emphysema in an adult, *Lung India* 28 (2011) 67–69.
- [5] A. Hislop, E. Hey, L. Reid, The lungs in congenital bilateral renal agenesis and dysplasia, *Arch. Dis. Child.* 54 (1979) 32–38.
- [6] O.A. Bawazir, Congenital Lobar emphysema: thoracotomy versus minimally invasive surgery, *Ann. Thorac. Med.* 15 (2020) 21–25.
- [7] O.F. Demir, M. Hangul, M. Kose, Congenital lobar emphysema: diagnosis and treatment options, *Int. J. Chronic Obstr. Pulm. Dis.* 14 (2019) 921–928.
- [8] J. Palla, M.M. Sockrider, Congenital lung malformations, *Pediatr. Ann.* 48 (2019) e169–e174.
- [9] A. Bush, J. Harcourt, R.J. Hewitt, A.G. Nicholson, Congenital lung disease, in: R. W. Wilmott, R. Deterding, A. Li, et al. (Eds.), *Kendig's Disorders of the Respiratory Tract in Children*, ninth ed., Elsevier, Inc, Philadelphia (PA), 2019, pp. 321–322.
- [10] R. Subramanyam, A. Costandi, M. Mahmoud, Congenital lobar emphysema and tension emphysema, *J. Clin. Anesth.* 29 (2016) 17–18.
- [11] J.M. Laberge, I. Bratu, H. Flageole, The management of asymptomatic congenital lung malformations, *Paediatr. Respir. Rev.* 5 (Suppl A) (2004) S305–S312.