Congenital lobar emphysema developing massive hemoptysis in adulthood treated by thoracoscopic lobectomy.

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Background

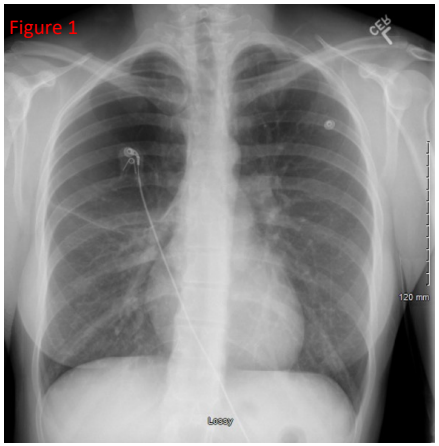
- Congenital lobar emphysema (CLE) arises mainly from intrinsic anomalies involving abnormal airway branching, usually with a defect in cartilage formation
  - This results in progressive hyperinflation of the emphysematous lobe, compression of remaining lung parenchyma, and displacement of mediastinal structures
- CLE is usually diagnosed within days of birth
  - Almost all cases are identified by six months of age.
  - Very rarely an adolescent or adult CLE patient develops persistent cough, dyspnea or recurrent pulmonary infections requiring resection via lobectomy.
- Herein we present a case of a healthy adult with known CLE who presented with serious hemoptysis and was managed successfully by thoracoscopic lobectomy.

PATIENT PRESENTATION

A 26-year-old Caucasian female presented to the emergency department after reporting eight episodes of hemoptysis the prior 10 hours. She was a never smoker with a known history of CLE discovered incidentally more than 10 years earlier. Her lifestyle had been normal and active and had not had pulmonology follow-up since that time.

On presentation vital signs were within normal limits. Physical examination revealed distant breath sounds in the right upper lung field. Chest radiograph revealed decreased bronchial and vascular markings suggestive of a right upper lobe emphysematous bulla (Figure 1). Chest computerized axial tomographic angiography (CTA) confirmed the presence of the large bulla occupying approximately 75% of the right lung upper lobe parenchyma (Figure 2).

During her initial 24 hours in the hospital she experienced several additional episodes of hemoptysis. Bronchoscopy identified fresh clot in the upper lobe bronchial os. Review of imaging confirmed that there was neither a vascular malformation, nor obvious target for embolization of systemic (bronchial) vasculature. Therefore, the surgical team recommended urgent right upper lobectomy due to the ongoing hemoptysis



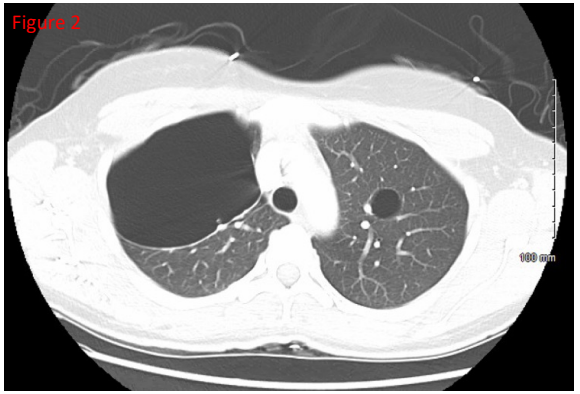


Figure 1: Anterior/Posterior plain chest x-ray with right upper lobe bleb

Figure 2: Axial chest CT with right upper lobe congenital bleb and smaller left upper lobe bleb

OPERATIVE COURSE

Under general anesthetic, the thoracic surgical team performed a VATS right lung upper lobectomy using standard techniques. Final pathologic specimen revealed focal large bullous formation with extensive hemorrhage. The patient was discharged on room air on postoperative day four, and four months postoperatively she remains symptom free with normal radiographs.

DISCUSSION

CLE is a rare developmental anomaly affecting approximately 1:20-30,000 births. This case is unusual because the patient was diagnosed incidentally at 16 years of age, whereas 90% or more of patients are diagnosed before six months of age. Furthermore, surgical resection is very rarely required in adult CLE patients, and when so the indication is typically for infection, dyspnea or spontaneous pneumothorax. We have found no other cases of massive hemoptysis leading to surgical intervention in adult patients. The final pathologic specimen did not identify a tumor or vascular anomaly, so we suggest that erosion of thin, poorly formed airway cartilage into the surrounding pulmonary vasculature was the etiology of her hemoptysis.

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