

CASE REPORT

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Fatal congenital lobar emphysema in a puerpera: a case report and literature review

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Abstract

Background: Congenital lobar emphysema (CLE) is a developmental lung abnormality usually diagnosed in the neonatal period and is rarely observed in adults. Adults with CLE are usually asymptomatic and only a small fraction may present with coughing, recurrent pneumonia and respiratory distress. In imaging studies, the most frequently affected lobe of CLE is the left upper lobe, followed by the right middle lobe. However, multilobar involvement with severe mediastinal shift is extremely rare.

Case presentation: We report a case of fatal CLE in a 28-year-old puerpera with postpartum respiratory failure. Chest computed tomography (CT) revealed emphysema of the right upper, middle and lower lobes resulting in adjacent atelectasis. Hyperinflation of the right upper lobe crossed the midline, leading to a deviation of the mediastinal structure to the left hemithorax and severe compression of the left lung.

Conclusions: Early and timely diagnosis of CLE with routine follow-up is necessary for patients. CLE, especially with multilobar involvement or mediastinal shift, could be life-threatening and should be promptly and aggressively treated to prevent severe complications.

Keywords: Congenital lobar emphysema, Adults, Computed tomography, Case report

Background

Congenital lobar emphysema (CLE) is a rare developmental lung abnormality usually diagnosed in the neonatal period, with an incidence of 1/20,000–30,000 [1]. Half of the CLE cases are idiopathic, and absence, hypoplasia or dysplasia of bronchial cartilage may occur in one-quarter of the patients [1]. Nearly all infants with CLE will develop symptoms and are diagnosed within the first 6 months of life [2, 3]; CLE in adults is quite rare. Adults with CLE are usually asymptomatic, and only a small fraction may present with coughing, recurrent

pneumonia and respiratory distress [4, 5]. Imaging studies are regarded as the gold standard for the diagnosis of CLE, which is characterized by overinflation of the involved lobes [1]. The most frequently affected lobe is the left upper lobe, followed by the right middle lobe [6, 7]. However, multilobar involvement is extremely rare. To the best of our knowledge, CLE in adults with multilobar involvement has only been reported in a few cases [7, 8].

Here, we reported a typical and fatal CLE with all three lobes of the right lung involved and severe mediastinal shift in a puerpera. This case suggests us that CLE in adults can be life-threatening; thus, early and timely diagnosis of CLE with routine follow-up is crucial.

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Case presentation

A 28-year-old puerpera suffering from severe dyspnoea was admitted to the emergency department. She had given birth 3 days prior. A closed thoracic drainage procedure was performed for the diagnosis of right pneumothorax in a local hospital failed to relieve her symptom. She was transferred to our center due to declining respiratory status. The patient had a history of fatigue and exertional dyspnoea from her childhood, though she had never sought medical care previously. The dyspnoea suddenly worsened in the third trimester. She was a non-smoker and free from other medical history.

This patient was lethargic on admission, and auscultation of the chest showed an absence of breath sounds in the right lung and decreased breath sounds in the left lung. Arterial blood gas revealed a PO_2 of 106.3 mmHg and a PCO_2 of 113.1 mmHg upon 53% oxygen concentration inhalation. Emergency tracheal intubation was performed considering the diagnosis of respiratory failure and her state-of-consciousness. She was then transferred to the intensive care unit.

Chest radiograph revealed increased radiolucency of almost the whole right lung, and the mediastinum shifted to the left (Fig. 1). Chest computed tomography (CT) displayed emphysema of the right upper, middle and lower lobes resulting in adjacent atelectasis. Hyperinflation of the right upper lobe crossed the midline, leading to a deviation of the mediastinal structure to the left hemithorax and severe compression of the left lung (Fig. 2).

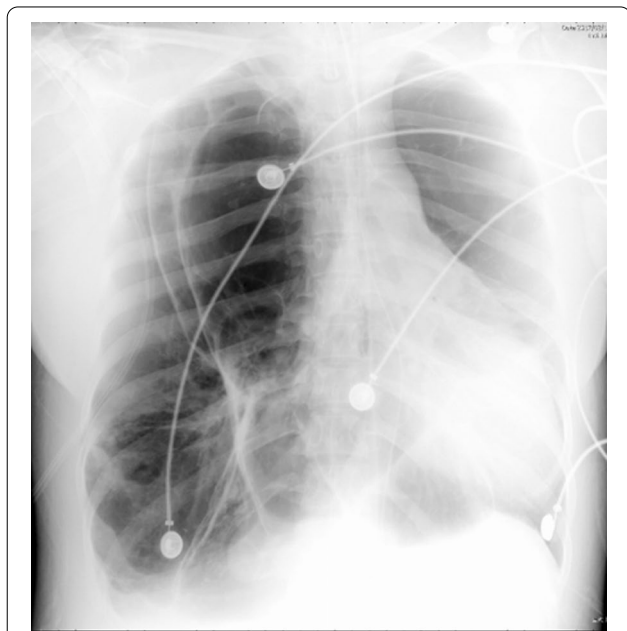


Fig. 1 Chest X-ray revealing increased radiolucency of almost the right lung, and the mediastinum shifted to the left

Flexible bronchoscopy revealed no obstructive intrabronchial lesions. The clinical diagnosis of congenital lobar emphysema (CLE) was established in combination with the clinical history and imaging examinations.

Since the patient could not tolerate one-lung ventilation, we planned to perform pulmonary resection assisted by extracorporeal membrane oxygenation. However, her legal representative refused the recommendation, and unfortunately, she eventually succumbed to respiratory failure and pulmonary infection after failed conservative management.

Discussion and conclusions

CLE is a developmental abnormality usually diagnosed in the neonatal period and is rarely diagnosed in adulthood. Few cases of adults with CLE have been reported before, and we have summarized the characteristics of them in Table 1 [5, 7–11]. The gold standard for the diagnosis of CLE relies on imaging examinations [1]. CLE is characterized by overinflation of the involved lobes, with or without attenuated and displaced pulmonary vessels on imaging studies [4, 6, 8]. Multilobar involvement in CLE is extremely rare. Shen et al. reported CLE in an elderly patient with involvement of the right upper and middle lobes [8]. Our case presented a typical and fatal CLE with all three lobes of the right lung involved and severe mediastinal shift in a puerpera.

The treatment choice for adults is dependent on the clinical severity, mainly including either conservative management for patients with mild symptoms or lobectomy for patients with disease progression [1].

Pregnancy-specific conditions like amniotic fluid embolism, preeclampsia can lead to respiratory failure in the peripartum period. Besides, pulmonary conditions including asthma, infection, thromboembolism and CLE are also causes of peripartum respiratory failure [12]. This patient developed respiratory failure after giving birth, indicating that severe CLE can be life-threatening in the peripartum period. Therefore, early and timely diagnosis of CLE is extremely important, and adult patients diagnosed with CLE should be followed routinely even when they are in stable condition. Pulmonary resection is an alternative treatment modality [1, 7]. For this patient, we considered surgical intervention to correct the mediastinal shift and assist in re-expansion of the compressed lung, since a previous report indicated positive re-expansion of a long-term compressed lobe after surgery in a patient with CLE [7].

Finally, the lessons we can learn from this case include that early and timely diagnosis of CLE with routine follow-up is necessary. CLE, especially with multilobar involvement or mediastinal shift, can be life-threatening

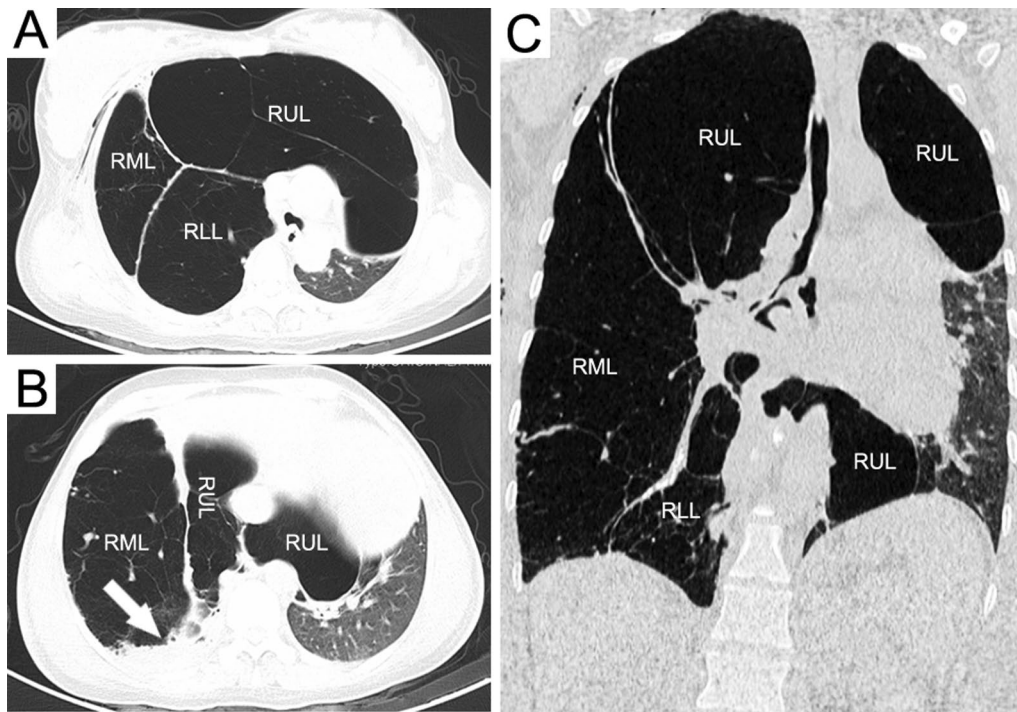


Fig. 2 Chest computed tomography (CT) displaying emphysema of the right upper, middle and lower lobes resulting in adjacent atelectasis (arrow); hyperinflation of the right upper lobe crossed the midline, leading to a deviation of the mediastinal structure to the left hemithorax and severe compression of the left lung. A and B, Axial chest CT scans; C, Coronal chest CT scan. RUL, right upper lobe; RML, right middle lobe; RLL, right lower lobe

Table 1 Characteristics of reported congenital lobar emphysema in adults

Literature	Patient	The way of diagnosis	Involved lobes	Mediastinal shift
Sadaqat et al. [9]	A 19-year-old man	Chest radiograph and Chest CT	Right upper lobe	Yes
Ogul et al. [10]	A 22-year-old man	Chest CT	Right middle lobe	Yes
Muramatsu et al. [11]	A 31-year-old woman	Chest radiograph and Chest CT	Right middle lobe	No
Sasieta et al. [7]	A 37-year-old man	Chest radiograph and Chest CT	Right middle lobe	Yes
King et al. [5]	A 28-year-old woman	Chest radiograph and Chest CT	Left lower lobe	No
Shen et al. [8]	A 78-year-old man	Chest radiograph and Chest CT	Right upper and middle lobes	No
Our case	A 28-year-old puerpera	Chest radiograph and Chest CT	Right upper, middle and lower lobes	Yes

and should be promptly and aggressively treated to prevent severe complications.

Abbreviations

CLE: Congenital lobar emphysema; RUL: Right upper lobe; RML: Right middle lobe; RLL: Right lower lobe.

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Not applicable.

Authors' contributions

LL designed the study. FG, LX and YY collected data. FG, LX and QP analyzed data and wrote the case report. FG, LX and YY contributed to the discussion of

results and the review of the manuscript. All authors read and approved the final manuscript.

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Availability of data and materials

All data and materials are provided in the manuscript.

Declarations

Ethics approval and consent to participate

The present case study was approved by Institutional Review Board of West China Hospital.

Consent for publication

Written informed consent was obtained from the patient's husband for publication of this case report and any accompanying images.

Competing interests

The authors declare that they have no competing interests.

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