

Bilateral congenital lobar emphysema: A rare cause for respiratory distress in infancy

Ahmad M. Abushahin, Amjad S. Tuffaha, Najeh K. Khalil¹, Adel M. Ismeal²

Divisions of Pediatrics
Pulmonology, ¹Pediatric
Critical Care, ²Pediatric
Surgery, Hamad
Medical Corporation,
Doha-Qatar

**Address for
correspondence:**

Dr. Ahmad M. Abushahin,
Department of Pediatrics,
Division of Pediatric
Pulmonology,
Hamad Medical
Corporation,
Doha-Qatar.
E-mail: amabushahin@
yahoo.com

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Abstract:

We report a rare case of bilateral congenital lobar emphysema in a 2-month-old male infant who presented with severe respiratory distress leading to respiratory failure. Plain chest X-ray and later high-resolution CT scan of the chest revealed that both the right middle and the left upper lobes were emphysematous. Surgical excision of the affected lobes has been done successfully in two sequential operations of right middle lobectomy followed by left upper lobectomy which resulted in significant improvement of respiratory status. In this report, the presentation, diagnosis, and surgical management of this rare condition are discussed.

Key words:

Congenital lobar emphysema, Infant, lobectomy, respiratory distress

Congenital lobar emphysema (CLE) is a rare anomaly of lung development which is usually unilateral.^[1] Bilateral involvement is exceptionally rare and has been reported only in few cases in the literature.^[2-4] CLE usually results in progressive respiratory distress in early infancy,^[1] and it often presents a diagnostic and therapeutic dilemma. We report a case of bilateral CLE in a 2-month-old male infant presented with acute respiratory distress. In this report, the presentation, diagnosis, and surgical management of this rare condition are discussed.

Case Report

A 2.7-kg male infant who was born normally at term with no antenatal care presented at the age of 15 days with cough and respiratory distress that required hospitalization. He was treated as acute bronchiolitis (positive direct fluorescent antibody (DFA) testing for Respiratory Syncytial Virus) for one week; and then was discharged home fairly well. He was readmitted two weeks later with continuing breathing difficulties. On admission, he was found to be tachypneic at a rate of 65-70/min, pulse rate of 167/min, and blood pressure of 84/50 mmHg. His temperature was 36.4°C and his O₂ saturation was 92 to 94% on room air. Chest examination revealed subcostal and intercostal retractions, decreased air entry in both sides with expiratory wheeze bilaterally. Rest of systemic examination including cardiovascular examination was within normal. Infant was supplemented with oxygen, intravenous fluids, and a trial of inhaled bronchodilator. Respiratory status worsened

significantly and infant was transferred to our pediatric ICU where he was intubated and mechanically ventilated. Plain chest X-ray of the first admission and the second admission [Figure 1] revealed marked hyperinflation of the right middle lobe and to a lesser extent the left upper lobe, with atelectatic changes of the adjacent lung lobes. These persistent findings raised the possibility of CLE. High-resolution CT scan of the chest was done and revealed emphysematous changes in the right middle lobe and left upper lobe with mild displacement of the mediastinum to the left side and atelectatic changes of the ipsilateral remaining lung lobes [Figure 2]. Flexible bronchoscopy showed bronchomalacia affecting the left main bronchus and bronchus intermedius. Echocardiography ruled out cardiac anomalies. Viral respiratory studies were negative. Infant continued to do poorly on mechanical ventilation, and after consulting with pediatric surgery, surgical intervention in two sequential operations to remove the affected lobes was agreed on. Because the right middle lobe was occupying most of the thoracic cavity and causing mediastinal shift to the left, right thoracotomy was performed first. The right middle lobe looked emphysematous at time of resection. Postoperative chest X-ray showed expansion of the right upper and lower lobes, and as expected, the left upper lobe emphysema became more pronounced [Figure 3]. At this stage, the infant continued to require ventilatory support, and five days later, left upper lobectomy was performed. The left upper lobe appeared emphysematous as well at time of resection. Histopathological

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Figure 1: Preoperative chest X-ray. The left upper lobe and right middle lobe are hyperlucent with compressive atelectasis of right upper and left lower lobes



Figure 2: CT scan of the chest confirms the diagnosis of right middle lobe and left upper lobe emphysema with mild displacement of the mediastinum to the left side and compressive atelectasis in the right upper lobe and left basal segment

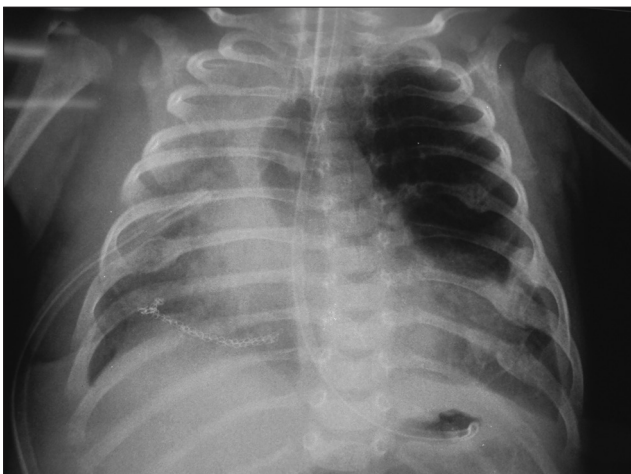


Figure 3: Post right thoracotomy, more pronounced left upper lobe emphysema

examination of the excised right middle and left upper lobes revealed changes consistent with CLE in both lobes [Figure 4].

On day three post resection of the left upper lobe, the infant was successfully extubated to CPAP for few days and then to room air. Tachypnea and retractions significantly improved with good and equal air exchange in both sides of the chest. At time of discharge, 14 days post first surgical intervention, chest X-ray revealed good bilateral lung inflation with no residual atelectasis, emphysema, or mediastinal shift [Figure 5]. Our infant was seen as an outpatient at 2 and 6 months of age. He had normal O₂ saturations in room air and his respiratory rate was 40 to 45/min. He was feeding well and gaining weight.

Discussion

CLE is a rare anomaly of lung development characterized by progressive over-inflation of one or more lung lobes.^[1] The prevalence of CLE is 1 in 20 000 to 1 in 30 000 and the incidence is estimated to be 1 in 70 000 to 1 in 90 000.^[5,6] It is usually unilateral and the most commonly affected lobe is the left upper (42%), followed by the right middle (35%) but any lung lobe can be affected.^[2] Bilateral involvement is extremely rare and has been reported only in few cases in the literature.^[2-4]

The massive over distension of the affected lobe and subsequent compression of the surrounding structures usually result in progressive respiratory distress in the newborn period and early infancy.^[1]

CLE is most often detected in neonates. Antenatal detection of CLE and other lung anomalies can be done at midgestation using ultrasonography and fetal MRI which was missed in our case. Early detection of the lesion identifies fetuses that should be delivered in a center offering high-level neonatal intensive care and where early postnatal surgery can be performed if needed.^[7]

In our case, the patient presented in the neonatal period with signs of respiratory distress that was progressive and required PICU care and mechanical ventilation. Bilateral CLE, although rare, should be considered in the differential diagnosis of respiratory distress in newborn period and early infancy, particularly if the chest images supporting this possibility as in our case. Management of CLE is usually resection of the affected lobe,^[8] but in asymptomatic and mildly symptomatic patients, conservative approach is warranted.^[9] Surgical approach of bilateral CLE is controversial. Some reports are advocating the two-step sequential resection of the emphysematous lobes,^[2,10] while others support the one-step operation in which the affected lobes are excised simultaneously.^[3,4] Considering that our infant was in acute respiratory failure and on ventilatory support, and taking into consideration that simultaneous bilateral thoracotomy is considered to be a high-risk procedure with significant postoperative pain,^[10] we felt that the two-stage surgical approach was safer. The right middle lobe was removed in the first operation as it was the most offending lobe. This was followed 5 days later by left upper lobectomy. We felt that our infant, who presented with severe respiratory embarrassment, tolerated well sequential thoracotomies with excellent outcome.

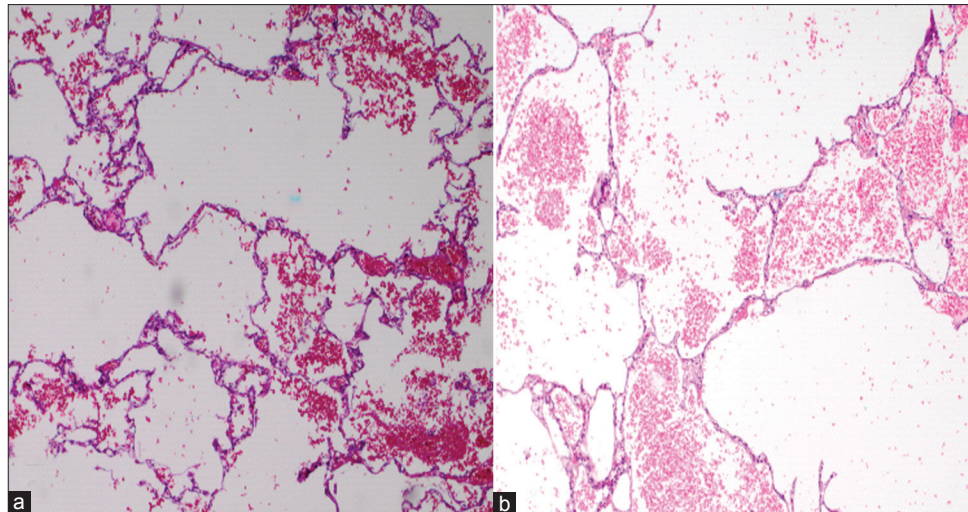


Figure 4: Histopathological specimen stained with hematoxylin and eosin x 100 of excised right middle lobe (a) and left upper lobe (b). Note the marked degree of alveolar destruction consistent with CLE



Figure 5: Postoperative follow-up chest X-ray showed good lung inflation in both sides with no residual atelectasis, emphysema, or effusion

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