

CASE REPORT

Congenital Lobar Emphysema (CLE)

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INTRODUCTION

Congenital lobar emphysema (CLE) is a life-threatening yet potentially reversible cause of respiratory distress in the neonate. CLE presents with overexpansion of a pulmonary lobe and resultant compression of the rest of the ipsilateral lung. The symptoms vary according to the severity of the disease and the degree of the ventilatory compromise. We present a case of CLE which was successfully managed by lobectomy through posterolateral thoracotomy immediately after definitive diagnosis was made.

CASE REPORT

A male baby was born on 8 February 2008 through caesarian section at NESCOM Hospital; his birth weight was 2.7kg, and he was discharged on fourth day with an apparently good condition. According to the mother, the baby was doing well on



Fig 1: Hyperinflated left lung with right sided mediastinal shift. Collapse/consolidation right upper lobe



Fig 2A: Hyperinflated left lung with herniation to right side. Collapse/consolidation right middle lobe

breastfeeding and was also gaining weight, when on 19 March 2008, he developed dyspnea and became unconscious. He was rushed to NESCOM Hospital, where he was admitted and managed by the child specialist. The chest x-ray revealed pneumatocele in left upper zone with no mediastinal shift.

The next day, the baby developed severe respiratory distress with x-ray findings of left tension pneumothorax on the left side with marked mediastinal shift towards right. Chest intubation was done and the patient was put on Inj Meronem™ 100mg I/V q8h, Inj Vancomycin™ 120mg I/V q8h. The condition did not improve and the patient was shifted to KRL Hospital with the provisional diagnosis of septicemia and pneumothorax.

The patient was evaluated by the pediatric surgeon and a diagnosis of congenital lobar



Fig2B: Hyperinflated left lung with herniation to right side. Mediastinum is pushed to right.

emphysema (CLE) was made and posterolateral thoracotomy planned with left upper lobectomy.

Investigations were carried out; the results were as under;

Chest x-rays in two planes were ordered. (Fig 1, 2A&B)

WBC 111900/cm, RBC 3085mil/cm, HB 11.7g/dl, Hct 35%, MCV 90, MCH 30pg/dl, Platelets 91000/cm³.

Differential Count: Neutrophils 35%, Lymphocytes 62%, Monocytes 03%, Eosinophils 0%, Basophils 01%, ESR 12mm/hr.

His electrolytes revealed Na 136mmol/l, K 3.6, Cl 99, Urea 13 mg/dl, Creatinine 0.6mg/dl and ALT 15u/l.

The baby was premedicated with atropine 100 µg. Monitoring included pulse oximetry, NIBP, ECG and ET_{CO}₂. The body temperature was maintained by wrapping the baby in soft cotton and by placing electric heating pad under him. Inhalation induction was carried with sevoflurane in 100% O₂, intubated with ETT#3.5, relaxed with Inj. Acuron™ in repeated small doses, analgesia with Inj Nalbin™ 200µg with Gravinate™ 2mg and IPPV carried out with sevoflurane in N₂O/O₂. Pediatric saline 20ml/hr and packed RBCs 40ml were transfused.

Left upper lobectomy was done. Left upper lobe was found to be fluid-filled and grossly overdistended.

At the end of the surgery, the baby was kept intubated for elective ventilation in the pediatric ICU. After about 2 hours, the patient had good respiratory efforts, ventilation was disconnected and oxygen given by T-tube. The ETT was taken out after another 2 hours with the patient maintaining good ventilation with O₂ through nasal prongs. Oral feeding was started on the 2nd postop day. The patient had uneventful postoperative recovery.

DISCUSSION

Congenital lobar emphysema (CLE) is a life-threatening yet potentially reversible cause of respiratory distress in the neonate. CLE presents with overexpansion of a pulmonary lobe and resultant compression of the rest of the ipsilateral lung. A mediastinal shift away from the increased-volume lung can also compress the contralateral lung. The abnormality is related to intrinsic bronchial narrowing. In these cases there are weakened or absent bronchial cartilages so that there is inspiratory air entry but collapse of the narrow bronchial lumen during expiration. This bronchial defect results in lobar air trapping¹.

CLE affects mostly the left upper lobe (41%) followed by right middle lobe (34%) and right upper lobe (21%). It presents in the newborn with a fluid-filled, over-distended lobe. The diagnosis can be made in utero or shortly after birth, but less severely affected patients may present in infancy or childhood. CLE has 2 forms:

- Hypoalveolar (fewer than expected number of Alveoli)
- Polyalveolar (greater than expected number of alveoli)

Approximately 10% of patients have associated anomalies, primarily congenital heart disease². Mostly, a single lobe is involved however, patients can show multiple lobar involvement. Microscopically, cartilage plates in the bronchi are absent at the level where the cartilage is expected².

Congenital Lobar Emphysema (CLE)

Clinically subtle or obvious respiratory distress is observed in an otherwise normal infant, with asymmetry of chest and abdominal retractions on inspiration. The thorax on the involved side is hyper-resonant with decreased or absent breath sounds with trans-illumination. Hypoxemia (in severely affected patients) may occur. The diagnosis is often suspected upon in utero sonography if an overexpanded lobe filled with fluid is identified. Progressive respiratory distress from birth reflects the degree of emphysema; symptoms are at their worst in the first month. Occasionally, patients present in later childhood or even during adulthood.³⁻⁵

Radiography of the chest in anteroposterior and lateral projections identifies the involved lobe, the degree of involvement, and the effect on surrounding structures. If a decubitus position radiograph is obtained, the involved lung does not collapse. CT scan can provide details about the involved lobe and its vascularity as well as information about the remaining lung. MRI can be used as an adjunct to identify vascular supply and distribution to the involved lung.

Differential diagnosis / other problems to be considered include pneumothorax, bronchial mucous plug, extrinsic bronchial compression, agenesis/hypogenesis of the contralateral lung, bronchial hypoplasia with air trapping peripherally and congenital cystic adenomatoid malformation.^{2,6}

Emergency surgical lobectomy is the only treatment for CLE with severe respiratory distress, but nonsurgical management may be appropriate in infants with only mild to moderate respiratory distress. Maintaining ventilator pressures and volume as low as possible avoids producing ventilator-related

hyperexpansion of an affected lobe. Management by more conservative, gentle ventilation technique is often successful. By following this course fewer surgeries result, because after diagnosis and initial treatment the affected lobe only rarely continues to expand, and infants with CLE who are not clinically in respiratory distress and who are able to feed and grow do not necessarily need surgery^{1,6,7}.

REFERENCES

1. Congenital Lobar Emphysema. Virtual Children's Hospital, University of Iowa. <http://indy.radiology.uiowa.edu/Providers>. May 13, 1998.
2. www.learningradiology.com/lectures/chestlectures/conglobaremphysemapppt.htm
3. Doull IJ, Connett GJ, Warner JO. Bronchoscopic appearances of congenital lobar emphysema. *Pediatr Pulmonol*. Mar 1996;21(3):195-7.
4. Koontz CS, Oliva V, Gow KW, Wulkan ML. Video-assisted thoracoscopic surgical excision of cystic lung disease in children. *J Pediatr Surg*. May 2005;40(5):835-7.
5. Lacy DE, Shaw NJ, Pilling DW, Walkinshaw S. Outcome of congenital lung abnormalities detected antenatally. *Acta Paediatr*. Apr 1999;88(4):454-8.
6. Schwartz MZ, Ramachandran P. Congenital malformations of the lung and mediastinum--a quarter century of experience from a single institution. *J Pediatr Surg*. Jan 1997;32(1):44-7.
7. www.bchealthguide.org/kbase/nord/nord922.htm

