

Case Report

Congenital lobar emphysema of right middle lobe-transition from an opaque lung to hyperlucent lung in a neonate

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ABSTRACT

Congenital Lobar Emphysema (CLE) is a rare congenital malformation of the lung characterized by over distension of a lobe of a lung due to partial obstruction of the bronchus. CLE often presents a diagnostic and therapeutic dilemma. We report a case of a 3 week old neonate who presented with sudden onset of respiratory distress related to CLE affecting the right middle lobe. Chest X-ray showed overexpansion of the right middle lobe, collapse of the ipsilateral lower lobe and shift of mediastinum. Lobectomy was performed successfully under general anaesthesia. Chest X-ray done at 6 hours of age showed an opaque right middle lobe which progressively became hyperlucent at 3 weeks of age requiring surgical intervention.

Keywords: Congenital lobar emphysema, Lobectomy, Neonate, Respiratory distress, Opaque lung

INTRODUCTION

Congenital Lobar Emphysema (CLE) is a developmental anomaly of the lower respiratory tract characterized by hyperinflation of one or more of the pulmonary lobes and is usually unilateral.^{1,2} CLE is a rare congenital malformation, with an incidence of 1 in 70000 to 1 in 90000 live births.³ We report a case of a 3 week old neonate who presented with sudden onset of respiratory distress related to CLE affecting the right middle lobe which was operated successfully.

Partial bronchial obstruction causes inspiratory air entry but collapse of bronchial lumen during expiration results in lobar air trapping and emphysema. Left upper lobe is most commonly affected followed by right middle lobe.¹

Congenital lobar emphysema is an uncommon but potentially life threatening anomaly affecting infants. It

usually presents in the neonatal period with respiratory distress. Chest X-ray and Computed Tomography (CT) scan of chest are diagnostic and shows hyperlucent affected lobe with herniation, shift of the mediastinum to the opposite side and collapse of the remaining part of the ipsilateral lung.³ Congenital Heart Disease (CHD) is associated with 12 to 20% of cases of CLE.

CASE REPORT

A 3 week old male neonate weighing 3.46 kg was admitted to our hospital with sudden onset of respiratory distress, noticed few hours prior to admission. There was no history of cough, vomiting or any abnormal movements. He was earlier admitted to our NICU at 6 hours of age and was treated for pneumonia.

This baby was born to a 28 year old G2P1L1 mother by cesarean section (Ind: previous cesarean birth) at 39

weeks of gestation and weighed 3240 grams. There was no history of maternal fever or prolonged rupture of membranes. Liquor was clear. Apgar scores were 8 & 9 at 1 and 5 minutes of age. He developed respiratory distress soon after birth and was admitted to our NICU. Chest X-ray showed consolidation of right middle lobe (Figure 1). Septic screen was negative and blood culture was sterile. He was treated for pneumonia with antibiotics (cefotaxime, amikacin) for 1 week and was discharged on day 8 of life.



Figure 1: Chest X-ray at 6 hours of age shows an opaque right middle lobe.

On examination, the baby had severe respiratory distress with RR of 82 per minute with sub costal retractions. On auscultation, air entry was decreased on the right side. Blood pressure was 86/48 mm of Hg, HR 152 per min and SpO₂ 84% in room air. Chest X-ray (Figure 2) showed hyperinflation of right middle lobe, collapse of right lower lobe and mediastinal shift to the left. A provisional diagnosis of CLE was made and CT scan of chest was done.



Figure 2: Chest X-ray shows marked overdistension of the right middle lobe, collapse of right lower lobe, herniation of lung and mediastinal shift to the left.

CT scan of chest showed a hyperlucent, emphysematous right middle lobe (attenuated but intact pattern of organized vascularity) with midline herniation, compression of the remaining lung and shift of mediastinum suggestive of CLE of right middle lobe (Figure 3).



Figure 3: CT scan shows a hyperlucent, emphysematous right middle lobe with herniation and compression of the remaining lung.

He underwent right middle lobectomy under general anaesthesia the following day (Figure 4). Thoracotomy was done through 5th intercostal space on the right side. Right middle lobe was isolated and clamped, pulmonary vessels were ligated and divided with significant improvement in oxygenation. The baby was electively ventilated for 12 hours and weaned off respiratory support. Oral feeds were started on the 3rd day and the baby was discharged uneventfully on the 8th postoperative day.



Figure 4: Resected middle lobe of right lung.

DISCUSSION

Congenital Lobar Emphysema (CLE) is a developmental anomaly of the lower respiratory tract characterized by

hyperinflation of one or more of the pulmonary lobes. It is more common in male children (male:female ratio of 3:1), is usually unilateral and affects left upper lobe (43%) more often followed by right middle lobe (32%).¹ Several factors have been associated with its development. In 50% of cases, there is decreased bronchial cartilage tissue which produces a ball valve effect with consequent air trapping and overinflation.⁴ Vascular rings that produce compression, bronchial stenosis, bronchial torsion, bronchogenic cysts, polyalveolar lobe and congenital CMV infection have also been reported.⁵⁻⁸ CLE is described in twins and occasionally from same family⁹ but in 40% of cases, the cause is unknown. The age of onset of symptoms ranges from a few days after birth to 6 months.

CLE often presents a diagnostic and therapeutic dilemma.

^[6] The basic investigation in CLE is the chest radiograph from which a diagnosis can be made often. Other diagnostic modalities used for diagnosis of CLE are CT chest, bronchoscopy, pulmonary angiography and V/Q scintigraphy.¹⁰ In this patient, a CT scan of chest was done to confirm the diagnosis and to delineate bronchial anatomy before surgery. This baby developed respiratory distress soon after birth and chest X-ray showed consolidation of right middle lobe which was misinterpreted as pneumonia. In fact he had retained lung fluid due to bronchial obstruction.

Review of literature showed similar case reports of opaque lungs in the initial radiographs which subsequently showed hyperlucent lungs suggestive of CLE.¹¹ Emphysematous lung with herniation and shift of mediastinum mimics pneumothorax and there are case reports of chest drains being inserted.^[10] We suspected CLE due to vascular markings but missed the opaque lung which was seen on the first X-ray done at 6 hours of age during first admission.

Resection of affected lobe is the commonest mode of treatment. In patients with mild symptoms, conservative measures are sometimes taken, but these may fail in the presence of inter-current infections. Resection of vascular rings is required where this is the cause. In over 85% of cases, the long-term outcome after surgery is good. In CLE, it is necessary to avoid further inflation and gas trapping in the diseased lung as this may compromise the normal lung reducing the cardiac output.¹²

In conclusion, a 3-week-old baby presented with sudden onset of respiratory distress related to CLE affecting the right middle lobe. Lobectomy was performed and the child was discharged on the 8th postoperative day. It should be noted that CLE at onset can appear as opaque lungs in neonates due to retained lung fluid and close

follow up of such neonates can help identify evolving emphysema which often requires surgical intervention.

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