Case Report

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Anaesthetic Management of Congenital Lobar Emphysema- A Case Report

Paediatric Surgery Section

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ABSTRACT

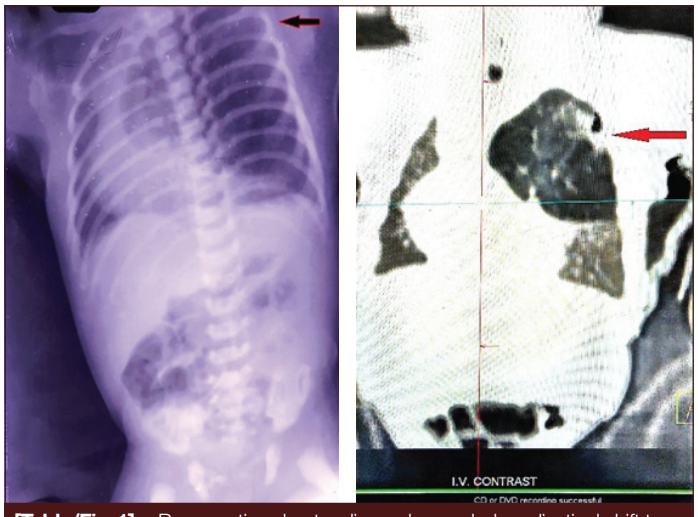
Congenital Lobar Emphysema (CLE) is a rare congenital disorder causing overinflation of a lobe or lobes of the lung. The left upper lobe is most commonly involved. Surgery is the preferred treatment in cases of CLE, with the patients recovering well after surgery, even with excision of more than one lobe. Here authors report a case of 2-week-old female infant with worsening respiratory distress that did not resolve despite supplemental oxygen and non invasive ventilatory support and was subsequently diagnosed with CLE. The pathology in CLE is the overinflation of the affected lobes and consequent collapse of the normal lobes, leading to hypoxia and reduced gas exchange. The emphysematous lobe causes the mediastinum to shift to the other side and severe cases may have a haemodynamic compromise. Anaesthesia in such infants is a precarious proposition. The infants may also have other congenital abnormalities. Furthermore, the over distended lobe will cause several problems affecting the infant's cardiorespiratory physiology. Hence, the preanaesthetic evaluation, anaesthesia plan and execution should be meticulously planned.

Keywords: Difficult airway, Lobectomy, Paediatric anaesthesia, Pneumonia, Thoracotomy

CASE REPORT

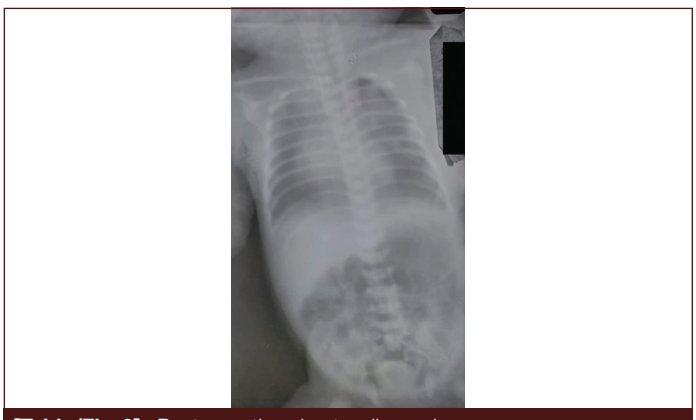
A 1-day-old female neonate delivered by lower segment caesarian section at term, weighing 2.75 kg, was shifted to the Neonatal Intensive Care Unit (NICU) as she was having persistent respiratory distress with tachypnoea and subcostal retraction. The baby was suspected of having congenital pneumonia and was being treated for the same with antibiotics and O_2 therapy. A chest X-ray was taken, which revealed of mediastinal shift to the right [Table/Fig-1]. Hence, the intercostal drain was inserted. The baby was managed conservatively as it was showing signs of improvement.

From the second week onwards, there was continuous deterioration in the baby's condition with worsening tachypnoea, intercostal and subcostal retraction. Chest X-ray revealed worsening mediastinal shift. The Computed Tomography (CT) scan revealed a hyperinflated left upper lobe with a collapsed left lower lobe and a rightward shift of the mediastinum [Table/Fig-2]. The baby also started to desaturate, necessitating bubble Continuous Positive Airway Pressure (CPAP). Hence, it was decided to be taken up for surgery. Blood investigations were unremarkable. Echocardiography revealed an ostium secundum atrial septal defect with left to right shunt.



[Table/Fig-1]: Preoperative chest radiograph revealed mediastinal shift to the right. **[Table/Fig-2]:** Contrast computed tomography- coronal section- The emphysematous lobe is indicated by the arrow. (Images from left to right)

The baby was medicated with 0.1 mg atropine, 12.5 mcg fentanyl, 0.2 mg midazolam and induced with sevoflurane. A 3.5 mm uncuffed tube was secured without the use of relaxants. The baby was kept in spontaneous ventilation with O_2, air and sevoflurane. Muscle relaxant and positive pressure ventilation were avoided to prevent further distension of the affected lobe and prevent reduce venous return. It was decided to push the endotracheal tube deeper if the surgeon needed one lung ventilation, as bronchial blockers or Fogarty's catheter were not available at the hospital. The surgeon was asked to give intercostal nerve blocks before giving the incision. Once the thoracotomy was performed, atracurium 2 mg was given for muscle relaxation. The vitals were stable intraoperatively, except when there was traction on the lung, when the End-tidal Carbon Dioxide (EtCO2) dropped drastically, but it would return to normal once the traction was reduced. After lobectomy, airleaks were checked for with valsalva and the thoracotomy was closed. The baby was shifted to the Neonatal Intensive Care Unit (NICU) with the endotracheal tube insitu for elective postoperative ventilation. Extubation was done the next day, and the baby was put on nasal prongs. The baby was maintaining SpO_2 of 98% with nasal prongs. The lung expansion was satisfactory and the baby had an uneventful postoperative stay [Table/Fig-3].



[Table/Fig-3]: Postoperative chest radiograph.

DISCUSSION

The CLE is a rare congenital disorder causing overinflation of a lobe or lobes of the lung. Nelson RL was first to describe congenital cystic disease in 1932 of the lung [1]. The CLE is a rare disease and has an incidence of 1 in 20,000 to 1 in 30,000 [2,3], although the incidence may be underestimated [2]. Boys are twice more likely to be affected than girls [4]. It occurs due to the overinflation of a lobe or lobes of the lungs, leading to ipsilateral collapse of the other lobes and contralateral shift of the mediastinum and contralateral lung collapse [5]. The aetiology of CLE is not well established, but there are multiple hypotheses like bronchial abnormalities, defects of alveoli and cartilaginous disorder leading to the collapse of the bronchi [2]. In 1970 Hislop A and Reid L first described CLE is a polyalveolar disease after examining the excised lung specimen [6]. This was later confirmed in some cases in the next decade. The left upper lobe is most commonly involved, followed by the right middle and right upper lobes. Lower lobes aren't usually involved [2]. Most patients with respiratory distress in the first six months of life are 80%, and around 50% present in the first week of life [3].

Although, the diagnosis of CLE can be made by chest X-ray, it requires a high index of suspicion as the disease has a very low incidence. The normal lobes appear opaque as they are collapsed due to the pressure from the overinflated lung and can mimic pneumonia like picture. Along with the radiolucent overinflated lobe, can be confused with spontaneous pneumothorax and insertion of an intercostal drain may be done, which can cause lung injury, thereby worsening the respiratory distress [2,3]. As described in a report by Sreevastava DK and Kiran S, which was also same as the present study [4]. Around 5-40% of the CLE cases have accompanying anomalies, with more than half being cardiac anomalies. Surgery is the preferred treatment in cases of CLE, with the patients recovering well after surgery, even with excision of more than one lobes [2,3]. The massive mediastinal shift caused by the emphysematous lobe can impair the venous return and cardiac output; keeping the patient in spontaneous ventilation before the thoracotomy may help in maintaining the venous return to the heart [7]. The mediastinal shift also can lead to difficult intubation. Hence, relaxants to aid intubation are better avoided. As the patient needs to be positioned laterally with the pathological lung being on top, the ventilation perfusion matching will be further affected and may require higher FiO_2 to maintain saturation [8]. Using nitrous oxide can cause further inflation of the affected lobe, hampering the oxygenation and haemodynamics further, hence avoided [2]. But nitrous oxide can be used once the pathological lobe has been isolated by clamping the bronchus [4,7]. The anaesthesia provider needs to be vigilant during lung manipulation, as it can cause changes in the haemodynamics and ventilation. Elective ventilation may be done postoperatively to provide adequate time for the collapsed lobe to expand.

CONCLUSION(S)

The CLE is a rare disease and needs a high index of suspicion for diagnosis. An early diagnosis and prompt surgical intervention in these cases have very favourable outcomes. Anaesthesia in such infants is complicated by heart disease, unstable haemodynamics, difficult intubation and needs a detailed anaesthesia plan. A thorough preanaesthetic evaluation includes an echocardiogram to exclude any significant congenital heart disease and evaluate venous return and ventricular activity. Care should be taken during the induction of anaesthesia to maintain adequate blood pressure; keeping the patient in spontaneous respiration will help. As there is bound to be a contralateral mediastinal shift, difficulty in siting the endotracheal tube must be anticipated. Avoiding the use of muscle relaxants helps in steering away from a potential off "cannot intubate cannot oxygenate scenario" as well as in augmenting the venous return. These infants need postoperative ventilation for the lungs to expand well and can be extubated once there is improvement seen in the X-ray and in blood gases. The postoperative recovery in these infants is usually good, and has normal development.

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