Case Report

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Congenital Lobar Emphysema: Anaesthetic Challenges and Review of Literature

Anaesthesia Section

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ABSTRACT

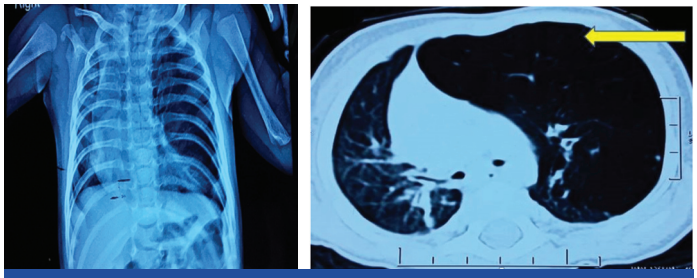
Congenital Lobar Emphysema (CLE) is a developmental anomaly, characterized by hyperinflation of one or more pulmonary lobes. It presents in infancy with variable degree of respiratory distress due to compression atelectasis. It is most often associated with mediastinal shift with subsequent hypoxia. CLE poses a diagnostic and therapeutic dilemma. We report a case of five-month-old infant of CLE requiring left lobectomy, who was previously being treated for pneumonia which was unresponsive to medical therapy. Anaesthetic challenges experienced during the case and a brief review of literature is presented.

Keywords: Lobectomy, Pneumonia, Positive pressure ventilation

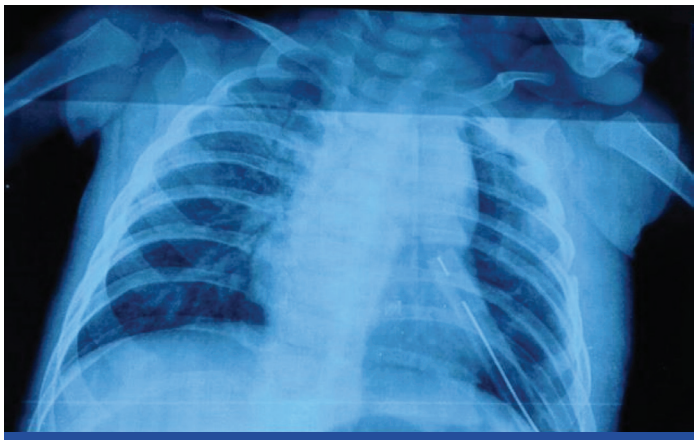
CASE REPORT

A five-month-old, male child, weighing 5 kg was referred to our hospital for breathlessness. The child had repeated hospitalizations since four months, due to recurrent episodes of fever and gradually worsening respiratory distress. Initial diagnosis of pneumonia was made but the child was unresponsive to medical therapy. On examination, the child was irritable, febrile with marked intercostal and subcostal retractions. His respiratory rate was 52 breaths/minute and heart rate was 130 beats/minute. On auscultation, decreased air entry was noted in left hemithorax and heart sounds were shifted to right side. Oxygen saturation (SpO_2) in room air was 90%. Chest X-ray showed hyperlucency of left upper lobe with mediastinal shift towards right [Table/Fig-1]. Computed Tomography (CT) scan of chest revealed hyperlucent left upper lobe crossing the midline and subsequent collapse of right upper lobe [Table/Fig-2]. Echocardiography was normal. A diagnosis of CLE of left upper lobe was made and lobectomy was planned. Routine haematological and biochemical investigations were normal. Preoperative Arterial Blood Gas analysis (ABG) revealed: pH 7.45, PaO_2 67 mmHg and $PaCO_2$ 28 mmHg. Oxygen supplementation was continued in preoperative period. The child was kept fasting, according to standard guidelines. No premedication was advised.

In the operating room, standard monitoring {Noninvasive Blood Pressure (NIBP), pulse oximetry, Electrocardiogram (ECG), capnography, temperature} was initiated. Baseline parameters were recorded as: Heart Rate (HR)-150 beats/minute, Blood Pressure (BP)-95/69 mmHg, Respiratory Rate (RR)-64/minute and SpO_2 92%. Child was preoxygenated. Peripheral intravenous access was in situ. Glycopyrrolate 0.04 mg and fentanyl 10 µg was administered. Anaesthesia was induced with graded concentration of sevoflurane (1%-8%) in 100% oxygen (O_2). Adequate anaesthetic depth was achieved while maintaining gentle assisted ventilation using Jackson-Rees circuit and tracheal intubation was accomplished using 3.5 mm ID portex uncuffed tracheal tube. Surgical team was kept on standby for emergency thoracotomy, if needed. Left femoral artery was cannulated for invasive BP monitoring and intraoperative Arterial Blood Gas (ABG) analysis. The child was placed in right lateral position. Rectal paracetamol 80 mg was given. Anaesthesia was maintained with sevoflurane 2%-3% in 100% O_2. Local anaesthetic, 4 ml bupivacaine 0.25%, was infiltrated along the line of incision in fourth intercostal space. A left thoracotomy was performed. Gentle assisted manual ventilation was continued until the emphysematous lobe popped out through left thoracotomy incision. Atracurium 2.5 mg was then administered. Positive Pressure Ventilation (PPV) was initiated, using pressure controlled mode keeping peak inspiratory pressure around 20 cmH_2O. Intraoperative analgesia was maintained with ketamine 0.5 mg/kg given intermittently. Intraoperative ABG was normal. After left upper lobectomy was accomplished, Nitrous oxide (N_2O) was added in a ratio of O_2:N_2O (50%:50%). After resection of the affected lobe, left lower lung was expanded using manual recruitment maneuver before closure and presence of leaks was checked with saline. Surgery lasted for two hours with stable haemodynamics. Intraoperatively, SpO_2 remained between 92%-96%. Left Intercostal Drain (ICD) was inserted. Intercostal blocks were given by surgeon under direct vision with 3 ml of 0.25% bupivacaine in third, fourth and fifth Intercostal Space (ICS) for postoperative analgesia. Fluid (Ringer lactate) and blood were replaced judiciously as per losses. Residual neuromuscular blockade was reversed with glycopyrrolate



[Table/Fig-1]: Preoperative chest X-ray showing hyperinflated left lung and shifting of mediastinum. **[Table/Fig-2]:** Hyperinflated left lobe of lung crossing midline.



[Table/Fig-3]: Postoperative chest X-ray showing normal lungs and mediastinum.

0.05 mg and neostigmine 0.3 mg. Trachea was extubated after recovery of airway reflexes and adequate spontaneous respiratory efforts. Infant was observed for 15 minutes in the Operating Room (OR) and then shifted on oxygen to paediatric Intensive Care Unit (ICU) for observation under continuous SpO_2 and ECG monitoring without any evidence of respiratory distress. Postoperative chest radiograph done next day revealed an expanded left lung and normal position of mediastinum [Table/Fig-3]. ICD was removed at 72 hours. Postoperative period remained uneventful and the infant was discharged on day eight.

DISCUSSION

CLE also known as infantile lobar emphysema, is a rare, idiopathic congenital malformation characterized by abnormal multilobar or unilobar overdistension of lung with an otherwise normal pulmonary architecture [1]. It usually presents as life threatening respiratory distress during infancy secondary to hyperinflated lung due to ball valve effect [2]. This result in compression atelectasis of ipsilateral or contralateral side with mediastinal shift, hypoxia and associated hypotension [3]. Reported incidence is 1:70,000 to 1:90,000 live births [4]. Despite medical advancements, CLE poses a diagnostic and therapeutic dilemma. Early recognition and surgical management can be life saving.

CLE is a rare entity with male predominance. It is usually unilateral affecting more often the left upper lobe (43%) followed by right middle lobe (32%). Bilateral involvement has been reported to be 20% [5]. Progressive hyperinflation of the lobe occurs either due to anomalies of bronchial cartilage or due to external bronchial compression with resultant air trapping on expiration [4,6].

Prenatal diagnosis of CLE is generally missed. It is detected during early infancy, when progressive hyperinflation of the affected lobe causes cardiopulmonary compromise due to compression of remaining ipsilateral as well as contralateral lung tissue. Associated congenital heart disease is present in approximately 15% of patients with CLE [7].

As in this infant, respiratory distress is the most common mode of presentation. Clinical signs include tachypnoea, tachycardia and chest retraction which worsens with progressive accumulation of gas and subsequently leads to respiratory failure. Physical examination may reveal asymmetric expansion of thorax, hyperresonant percussion note, rhonchi and diminished breath sounds over the affected hemithorax. Mediastinal shift, displaced apical impulse, flattening of diaphragm and at times “tension emphysema” due to hyperaeration of affected lobe is evident on chest radiograph. This leads to ventilation-perfusion mismatch (V/Q) with resultant hypoxia and ultimately decreased respiratory reserve that makes anaesthetic management challenging in these patients. Diminished cardiac output may be seen secondary to decreased venous return due to raised intrathoracic pressure following airtrapping in the affected lobe.

Infants with CLE present a diagnostic challenge. The disease is frequently confused with pneumonia [8] and pnemothorax [6]. At times, patients get an ICD inserted due to misdiagnosis which further worsens the situation [9]. Our child was also being treated for pneumonia. Chest radiograph done in peripheral hospital was helpful but not conclusive. CT or Magnetic Resonance Imaging (MRI) scan is mandatory for diagnosing CLE when in doubt as was done later in our patient since child was not responsive to medical therapy. Confirmatory test is single photon emission tomography V/Q lung scan which reveals hypoperfusion of affected lobe and hyperperfusion of normal lobe [6]. Fluoroscopy, bronchoscopy, angiography, radio-isotopic evaluation and lung scintigraphy are other diagnostic modalities available. All patients should have cardiac evaluation by echocardiography [10].

Controversy still exists regarding the management of CLE. Although conservative management is suitable for milder cases, lobectomy is the mainstay of treatment. Mortality associated with conservative therapies is 50%-75% and these patients invariably need surgical intervention later [2]. Anaesthetic management in these children with reduced respiratory reserve is challenging. Lateral decubitus position adds to the already existing V/Q mismatch. Due to close proximity of Functional Residual Capacity (FRC) to residual volume in this position, there is more propensity of airway closure in the dependant lung making this position unfavourable [11].

Induction of anaesthesia is crucial in children with CLE as excessive crying and struggling can increase the amount of trapped gas due to ball valve effect. Also, PPV leads to overinflation of the emphysematous lobe resulting in mediastinal shift and haemodynamic compromise, even cardiac arrest [12]. Thus, it is imperative to avoid PPV and provide gentle assisted manual ventilation when required, while maintaining airway pressure at 20-25 cmH_2O until thoracotomy. Monitoring of vital parameters during this phase is must.

Inhalational induction is the technique of choice in patients with CLE as described in the literature. N_2O is avoided during this surgery as it is known to diffuse rapidly in a closed cavity leading to further compression and mediastinal shift. Anaesthesia was induced in our child with sevoflurane in 100% O_2 without muscle relaxant. The optimum ventilatory mode is also debatable in patients with CLE. Though avoidance of PPV is advocated by several authors until thoracotomy [8]. Prabhu M et al., achieved anaesthetic induction using gentle Intermittent Positive Pressure Ventilation (IPPV) following muscle relaxation in their patient [2]. Pressure Regulated Volume Control (PRVC) is an useful option, if available [2,13]. Goto H et al., advocated the use of high frequency ventilation successfully [14]. We used assisted ventilation until thoracotomy followed by pressure controlled ventilation. Our patient experienced mild hypoxia and raised End Tidal Carbondioxide ($EtCO_2$) on few occasions intraoperatively which was corrected by switching off N_2O and removal of retractors by the surgeon.

Suitable options for maintenance of anaesthesia are inhalational agents along with intermittent Neuromuscular Blocker (NMB) agent. Pre-emptive analgesia in the form of local infiltration of local anaesthetic along the line of incision, blunts the incision response thereby decreasing the requirement of inhalational agents. Intermittent doses of intravenous ketamine (0.5 mg/kg-1 mg/kg) has been described [7,13]. Longer acting opioids are avoided due to the risk of postoperative respiratory depression. Caudal thoracic epidural catheter provides stable haemodynamics along with excellent analgesia without any risk of postoperative respiratory depression. However, kinking of catheter is a probable issue [15]. We used intercostal nerve blocks in our patient.

Extubation is preferred at the end of surgery. Elective postoperative ventilation is generally not required except in cases where more than one lobe has been excised. Adequate analgesia is warranted for the infant to breathe spontaneously. Lung protective strategies should be continued, if elective ventilation is needed [2].

CONCLUSION

A low threshold is required for suspicion to diagnose CLE promptly for any infant presenting with progressive respiratory distress. Early recognition and timely management can be life saving. Surgical management poses several specific anaesthetic challenges. Maintaining spontaneous ventilation using inhalational agents is desirable during induction of anaesthesia. However, gentle IPPV with a pressure limit <25 cmH_2O may be performed during hypoventilation under deep anaesthetic plane, until thoracotomy.

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