Congenital lobar emphysema: A modified approach to anesthetic management

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Abstract

Congenital lobar emphysema (CLE) is a potentially reversible, though life-threatening cause of respiratory distress in neonates. It is a rare developmental anomaly of the lower respiratory tract characterized by hyperinflation of one or more pulmonary lobes. It is a rare condition presenting in 1 in 20,000-1 in 30,000 cases. Conventionally, intermittent positive pressure ventilation (IPPV) is carried out during anesthesia, but avoided in cases of CLE as it further inflates the diseased lobes compromising the functioning of the normal lung, which calls for modification of the existing anesthetic management strategies.

Keywords: Congenital lobar emphysema, one lung ventilation, thoracotomy

Introduction

Congenital lobar emphysema (CLE) is a potentially reversible, though life-threatening cause of respiratory distress in neonates. CLE is a developmental anomaly of the lower respiratory tract characterized by hyperinflation of one or more pulmonary lobes. It is a rare condition presenting in 1 in 20,000-1 in 30,000 cases. Conventionally, intermittent positive pressure ventilation (IPPV) is carried out during anesthesia, but avoided in cases of CLE as it further inflates the diseased lobes compromising the functioning of the normal lung, which calls for modification of the existing anesthetic management strategies.

Case Report

A 10-month-old male child presented to the pediatric outpatient department with fever and cough, difficulty in breathing of 1 month duration. History informed by the mother revealed that the baby was normal till 9 months after which he developed above said symptoms. Developmental history was normal. No history of any hospital admissions in the past.

On general physical examination, the child weighed 8 kg with no evidence of cyanosis, clubbing, lymphadenopathy or edema with a pulse rate of 140/min. During respiratory system examination, tachypnea was noted with intercostal retraction and auscultation revealed decreased breath sounds over left upper lobe with room air saturation of 95%. Cardiovascular examination revealed shifting of apex beat to right with no murmurs. Chest radiograph demonstrated hyperinflated left upper zones with mediastinal shift to the right. Clinical diagnosis of CLE was made and the child started on intravenous (IV) fluids, antibiotics and nebulization.
A thoracic computed tomography scan confirmed the diagnosis [Figure 1].

After taking informed consent from the parents, thoracotomy with left upper lobectomy under endotracheal general anesthesia was planned.

Baseline values of vitals were recorded as pulse rate 140/min, saturation 95% on room air, blood pressure 110/70 mmHg and EtCO₂ 45 mmHg. The child was premedicated with glycopyrrrolate 0.01 mg/kg, pentazocine 0.5 mg/kg and midazolam 0.05 mg/kg. Preoxygenation using face mask with 100% O₂ for 3 min was carried out followed by induction with thiopentone 5 mg/kg. Succinyl choline 1.5 mg/kg was administered to facilitate intubation. Deliberate endobronchial intubation was performed to the right side using 4.5 mm internal diameter uncuffed oral endotracheal tube. Tube position confirmed by auscultation and connected to anesthesia ventilator. Depth of anesthesia was maintained using sevoflurane 0.2–1% in 50% oxygen. IPPV carried out using low tidal volumes (4–6 ml/kg) on pressure regulated volume control mode. Atracurium 0.5 mg/kg IV administered as muscle relaxant. IV hydration maintained with Ringer lactate 70 ml in the 1st hour and 30 ml in the next hour. Intra operative period was uneventful with saturation maintaining at 95–96%.

Surgery lasted 90 mins during which left thoracotomy with left upper lobectomy was performed under one lung ventilation and an intercostal drainage tube placed at the left 5th intercostal space in the mid axillary line. Intra-operative findings revealed an emphysematous left upper lobe [Figure 2]. At the end of the procedure, an intercostal block administered using 0.25% bupivacaine 2.5 ml in two intercostals spaces adjacent to the surgical incision. Reversal achieved with neostigmine and Glycopyrrolate. Extubation was done once adequate respiratory efforts were confirmed with the child maintaining a room air saturation of 100%.

Child was observed in the pediatric intensive care unit. On the first postoperative day, the child developed fever and treated appropriately. Chest X-ray done on the second postoperative day revealed an adequate left lung expansion. Child was discharged on the seventh postoperative day.

**Discussion**

Congenital lobar emphysema is a rare congenital anomaly characterized by over inflation of the pulmonary lobe. Most patients develop symptoms in the neonatal period and the male: female ratio is 3:1.[2] The cause of CLE is difficult to determine and none is identified in 50% of the cases.[3] In other 50% of the cases there is a decrease in bronchial cartilage tissue. This defect produces a ball valve effect with consequent over inflation.[4] The age of onset of symptoms ranges from a few days after birth to 6 months.[8]

Respiratory distress is the commonest presentation. Other symptoms and signs include dyspnea, wheezing, grunting respiration, tachypnea and sometimes progressive cyanosis. Upper lobe of left lung is most commonly affected (41%), followed by right middle lobe (34%) and right upper lobe (21%). A diagnosis of CLE can be made by a chest radiograph which is readily available but there are reports of it being confused with pneumothorax or pneumonia.[6] Operative surgery with lobectomy is the commonest mode of treatment.[7] Extracorporeal membrane oxygenation has been used to maintain oxygenation in children with persisting pulmonary acquired interstitial emphysema.[9]

Anesthetic concerns are during induction and intubation as these are very critical phases. Induction of anesthesia should aim to avoid crying and struggling because these can increase the amount of trapped gas in the emphysematous lobes.[9] Positive pressure ventilation to assist the ventilation also increases the emphysema. Even

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**Figure 1:** Computed tomography scan showing the emphysematous left upper lobe with mediastinal shift

**Figure 2:** Emphysematous lobe seen prior resection
though lung isolation is desirable, pediatric size double lumen tubes are not freely available and technically difficult hence not commonly practiced. As an alternative, selective main stem bronchus intubation can be done. During ventilation it is necessary to avoid further inflation and gas trapping in the diseased lung since this may compromise the normal lung. It is important to avoid IPPV until the diseased lobe is isolated because further inflation of the diseased side leads to a decrease amount of functional lung tissue and increase in intra thoracic pressure due to emphysema which further decreases the respiratory reserve. However, gentle manual ventilation maintaining the airway pressure of 20–25 cm of H₂O before thoracotomy keeping an eye on the vitals has been the method of induction used by Coté and Payne et al. 

In this case an endobronchial intubation along with IPPV was done keeping in mind the above said concerns which lead to an uneventful anesthetic management of the case.

Conclusion

Anesthetic management in a case of CLE is indeed a difficult issue and various modes of management have been described in the literature. Anesthetic induction with inhalational agents is desirable; however, if IPPV is necessary, gentle manual ventilation or pressure controlled ventilation with a pressure limit of 20–25 cm of H₂O can be carried out until thoracotomy. Successful anesthetic management of CLE can be achieved by proper understanding of pathophysiology, good perioperative monitoring, and adopting novel management strategies.

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