Congenital Lobar Emphysema: An Anaesthetic Challenge

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Abstract

Congenital Lobar Emphysema (CLE) is a rare, life-threatening yet potentially curable cause of respiratory distress in neonates and infants presenting with varying degrees of severity. There is hyperinflation due to air trapping in the affected lobe(s) resulting in a mediastinal shift, hemodynamic instability, and hypoxia. We report a case of CLE in a 3-month-old male baby who was posted for thoracotomy and left upper lobe lobectomy. Here, we discuss the anaesthetic challenges and management including endobronchial intubation, controlled ventilation, and the successful outcome.

Keywords: Congenital Lobar Emphysema, Endobronchial intubation, Lobectomy

Introduction

Congenital Lobar Emphysema is a rare cause of sudden respiratory distress in a newborn or infant. It is a developmental anomaly of the lower respiratory tract system involving hyperinflation due to air trapping of the histologically normal, affected lobe(s) and resultant compression of the remaining lung tissue[1,2,3]. Congenital lobar hyperinflation is the currently recommended term as it involves the pathologically healthy lung tissue[4]. It is the 2nd leading cause of early mortality in infants after anomalies of the cardiovascular system. The reported incidence is 1:20,000 to 1:30,000 live births with males being affected three times more than females[5,6]. The involvement of left upper lobe (LUL) is 43% followed by right middle lobe – 32% although bilateral involvement is unknown[7,8]. Etiology is unknown in 50% of cases but several intrinsic and extrinsic causes have been described[9]. Surgery is considered the first choice of treatment in severe cases[10]. We report the anaesthetic management in an infant diagnosed with CLE, posted for LUL lobectomy with a review of the literature.

Case Report

A 3-month-old male baby weighing 5.4 kgs presented with symptoms of respiratory distress and fever and signs of lower respiratory tract infection (LRTI). Oxygen saturation was 93% on room air. The chest x-ray revealed hyperinflated LUL and mediastinal shift to the right (figure 1), which was further confirmed with high-resolution computed tomography (HRCT) (figure 2). A diagnosis of CLE was made, and treatment initiated with supplemental oxygen by mask, antibiotics, bronchodilators, nebulization and intravenous fluids.

Surgery was scheduled after 3 days when the infective phase subsided. During the preanaesthetic check-up, the baby was active, afebrile, and had an O2 saturation of 93% on room air. Oxygen by mask, antibiotics, bronchodilators, nebulization and intravenous fluids were confirmed intra-operatively by endoscope. Ventilation of (RUL) bronchus was achieved with a 3.5mm micro-cuff endotracheal tube (Haylard - Kimberly Clark) that was advanced into the right main bronchus by turning the baby’s head to the left side[11]. The right main bronchus intubation and correct placement proximal to RUL branch was confirmed with a flexible endoscope.

Blood pressure, end-tidal Carbon Dioxide, temperature, Oxygen saturation, and urine output) was established. Surgical readiness for emergency thoracotomy at induction was ensured. Induction with spontaneous inhalation of Sevoflurane and Oxygen using Jackson-Rees circuit, aided with Atracurium (0.5 mg/kg) and gentle intermittent positive pressure ventilation was done. The baby was intubated with a 3.5mm micro-cuff endotracheal tube (Haylard - Kimberly Clark) that was advanced into the right main bronchus by turning the baby’s head to the left side[11].

The baby was premedicated using Atropine 0.1mg IV, Midazolam, 0.01mg/kg IV and Ketamine, 1mg/kg IV. In the operation theatre, standard monitoring (heart rate, blood pressure, end-tidal Carbon Dioxide, temperature, Oxygen saturation, and urine output) was established. Surgical readiness for emergency thoracotomy at induction was ensured. Induction with spontaneous inhalation of Sevoflurane and Oxygen using Jackson-Rees circuit, aided with Atracurium (0.5 mg/kg) and gentle intermittent positive pressure ventilation was done. The baby was intubated with a 3.5mm micro-cuff endotracheal tube (Haylard - Kimberly Clark) that was advanced into the right main bronchus by turning the baby’s head to the left side[11].

The right main bronchus intubation and correct placement proximal to RUL branch was confirmed with a flexible endoscope. Ventilation of (RUL) bronchus were confirmed intra-operatively by auscultation. Standard precautions were taken for temperature control (ambient OT temperature maintained at 27⁰ Celsius, baby’s head and limbs were wrapped in Ganggee rolls; warming mattress, convective forced-air warmer, warm fluids, heat and moisture exchanging filters were also used). The preoperative maintenance fluids were continued @ 5 -7 ml/kg/hour with extra boluses of 10-20 ml whenever required, with the concomitant watch on...
Discussion

**CLE** is a rare congenital anomaly characterized by hyperinflation of the affected, histologically normal pulmonary tissue with compression atelectasis of the contralateral lung associated with mediastinal shift [12].

A definitive causative agent cannot be identified in approximately 50% of cases. Airway obstruction can be intrinsic or extrinsic with the former being more common. Common intrinsic causes are bronchomalacia, bronchial stenosis or cysts [13]. Bronchomalacia is reported in at least 25% cases resulting in a ball-valve mechanism wherein the air can go in during inspiration but cannot be expelled during expiration, causing air trapping in the affected lobe [14,15]. Extrinsic cause includes compression due to abnormal vessel [13]. The resultant compression atelectasis due to the overinflated lobe causes ventilation-perfusion mismatch and hypoxia. There is increased intrathoracic pressure causing impaired venous return and hemodynamic instability [16].

The baby usually presents with respiratory distress, signs of LRTI and displaced cardiac sounds. It is most commonly confused with pneumothorax with some reports having mentioned wrongful placement of ICD [17]. Other differential diagnoses of CLE are lung cysts, congenital diaphragmatic hernia, and congenital cystic adenomatoid malformation.

The common diagnostic modalities include chest x-ray; [18] HRCT and (MRI) help to confirm the diagnosis. Bronchoscopy is increasingly being employed as a diagnostic as well as atherapeutic tool [19]. Single photon emission tomography V/Q lung scan reveals hypo-perfusion of affected lobe due to compression of vasculature and hyperperfusion of normal lobe by shunted blood. Other diagnostic modalities include fluoroscopy, angiography, radio-isotope evaluation, and lung scintigraphy [11,18,20,21]. Its prenatal diagnosis is difficult due to its low prevalence; and also, the requirement of ventilation for it to manifest. Fetal MRI is helpful to establish a diagnosis.

Surgical excision of the affected lobe is the commonest mode of treatment in severe cases [10].

The Preoperative evaluation aims at optimizing the cardio-respiratory status of the baby.

The anaesthetic challenges in a case of CLE comprise of the ventilatory management of a baby undergoing thoracic surgery in the lateral decubitus position. Induction is crucial as the patients are in respiratory distress; the crying and struggling can worsen the emphysema. Nitrous oxide must be avoided as it can rapidly diffuse into closed cavities leading to further compression and mediastinal shift. A High degree of suspicion of pneumothorax with surgical standby for emergency thoracotomy at induction is prudent to deal with any hemodynamic catastrophe.

We intubated with the aid of muscle relaxant using gentle intermittent positive pressure ventilation, keeping airway pressures below 20-25 mm of Hg [17]. This helped to deliver less tidal volume at a controlled rate thereby avoiding over-aeration of the lungs. Pressure regulated volume control (PRVC) mode is desirable if available [17].

One lung ventilation is desirable, although lung isolation is technically difficult in small infants. Moreover, double lumen tubes are not freely available. We achieved OLV by doing selective right sided endobronchial intubation [11]. The RUL blockage was avoided by endoscopic guided placement and fixation of the tube. Kamra et al have reported rigid bronchoscopic placement of Fogarty catheter as abronchial blocker in the diseased lung [22]. However, it may pose the risk of proximal displacement in trachea and inability to apply suction or continuous positive airway pressure (CPAP) as it is a closed tip. In the past, anaesthetic management of CLE has been described with tracheal intubation and spontaneous ventilation till lobectomy. However, it demands deeper planes of anaesthesia which may aggravate hypoxia and hypotension. Whereas, our approach of gentle manual ventilation allows control over lung expansion and ventilatory rate while

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maintaining hemodynamic stability. In our case, intraoperative and postoperative analgesia was provided through thoracic epidural catheter via caudal insertion site. Raghavendra et al. have also reported similar placement while retaining spontaneous ventilation until thoracotomy[23].

Other methods of providing intra-op and post-op analgesia include parenteral narcotics/ketamine, non-steroidal anti-inflammatory drugs, intercostal nerve blocks. Inflammatory drugs, intercostal nerve blocks under direct vision and rectal suppositories. Most cases get extubated postoperatively in OT. Elective ventilation is indicated when more than one lobe is excised or if the infant is very sick preoperatively and has poor spontaneous efforts.

Conservative management has a role especially when the patients are mildly symptomatic and have tolerated the respiratory exacerbations adequately. Use of flexible bronchoscope has been described to directly and dynamically evaluate the airway anomalies and identify the reversible cause in patients with suspected CLE[19]. Use of flexible ultra-thin bronchoscope passed into affected bronchus for emergency relief of initial respiratory distress has been reported[24].

Conclusion

In conclusion, an infant presenting with respiratory distress should be viewed with ahighe degree of suspicion of CLE. Early recognition and prompt intervention can treat this potentially life-threatening yet reversible illness. Anaesthetic management pertaining to varying modes of ventilation, lung isolation techniques and adequate postoperative analgesia is crucial for the successful outcome of the case.

References


Conflict of Interest: Nil

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