A case of congenital lobar emphysema with pneumonia. An anaesthetist’s challenge

R. Nandi, S. Singh, K. N. Saxena

Department of Anaesthesiology, Critical Care and Pain, Lok Nayak Jai Prakash Hospital, Maulana Azad Medical College, New Delhi, India

Corresponding author: R. Nandi, Department of Anaesthesiology, Critical Care and Pain, Lok Nayak Jai Prakash Hospital, Maulana Azad Medical College, New Delhi, India. Email: delphinium80@gmail.com

Abstract

Congenital lobar emphysema (CLE) is a rare pulmonary anomaly that causes respiratory distress in infancy. CLE presents significant challenges in diagnosis and management, particularly in low-resource settings. We report the case of a three-month old infant with CLE and pneumonia. After a delay in diagnosis, emergency lobectomy was performed under general anesthesia in a tertiary care public hospital in New Delhi, India. Initially, resolution of pneumonia was sought, but continued compression atelectasis warranted emergency surgery. Ventilatory management of our patient of CLE with pneumonia was especially challenging and is discussed.

Keywords: Congenital lobar emphysema, recurrent pneumonia, positive pressure ventilation

Introduction

Congenital lobar emphysema (CLE) is a rare idiopathic overdistension of a pulmonary lobe. CLE poses diagnostic and therapeutic dilemmas. A delay in diagnosis and surgery may lead to deterioration in the patient.

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Key points

Congenital lobar emphysema (CLE) presents significant challenges in diagnosis and management, particularly in low-resource settings. We report the successful management of a three-month old infant with CLE and pneumonia. After a delay in diagnosis, emergency lobectomy was performed under general anesthesia in a tertiary care public hospital in New Delhi, India.
X-Ray (CXR, Figure 1) showed hyperlucency of right upper and middle zone with leftward mediastinal shift, and segmental collapse in right upper zone. Non-contrast CT of chest (Figure 2) revealed hyperinflated right middle lobe with herniation to left side through the anterior mediastinum.

Other slices showed subsegmental collapse of right upper lobe and consolidation collapse in apical segments of bilateral lower lobes. CLE of the right middle lobe with pneumonia was diagnosed and lobectomy was planned after resolution of chest infection. In spite of medical management of pneumonia over four days, the child worsened. ABG under oxygen hood showed poor oxygenation (paO\textsubscript{2} 57mm Hg).

Urgent right middle lobectomy was pursued. In the operating theater, intravenous access was checked, and infusion of 5% dextrose in 0.9% saline was commenced. Monitoring included noninvasive blood pressure, pulse oximetry, electrocardiogram, capnography, and temperature. Patient was preoxygenated and intravenous fentanyl 8 mcg was given.

Anesthesia was induced with 6% sevoflurane in 100% oxygen via Jackson-Rees circuit. Tracheal intubation was done after achieving adequate anesthetic depth with maintenance of spontaneous breathing, and with surgical team on standby for emergency thoracotomy. The infant was placed in left lateral position. Bupivicaine 0.25% (2 mL) was infiltrated along the line of incision.

Manually assisted spontaneous ventilation continued until the emphysematous lobe was delivered through the thoracotomy (Figure 3). Intravenous atracurium 2 mg was then administered and positive pressure ventilation (PPV) instituted. Maintenance of anesthesia was with sevoflurane in 100% oxygen; analgesia, with fentanyl; and muscle relaxation, with atracurium.

Right middle lobectomy (Figure 4) was accomplished in two hours with stable haemodynamics (though tachycardia persisted, 150 bpm).
Congenital lobar emphysema, recently termed as congenital lobar overinflation, is a rare anomaly characterized by overdistension of an otherwise anatomically normal pulmonary lobe with atelectasis of the neighboring lung and mediastinal shift. The incidence of CLE is 1 in 70,000 to 1 in 90,000, with male predominance. Congenital heart disease is associated in 12-14% of these patients. Clinical signs of CLE include tachycardia, tachypnea, and chest retractions which progress to respiratory distress and respiratory failure. Asymmetric expansion of the hemithorax, rhonchi, displacement of apical impulse, hyper resonant percussion on affected side, and diminished breath sounds and heart sounds may be noted. Lobar hyperinflation, atelectasis of contralateral lung, mediastinal shift, and flattening of ipsilateral diaphragm are typically seen on CXR. Thus, the disease is often confused with pneumonia and pneumothorax, even resulting in wrongful placement of a chest drain. CT and MRI help in diagnosis of CLE, but the single photon emission tomography ventilation-perfusion lung scan is confirmatory. This scan may reveal hypoperfusion of affected lobe due to compression of vasculature and hyperperfusion of normal lobes by shunted blood. While conservative management for CLE has been described, lobectomy is the mainstay of treatment. The management of a child with CLE with pneumonia or respiratory failure is controversial. Clearing the infection prior to surgery can improve pulmonary mechanics, but unrelieved compression atelectasis can be detrimental as in our patient. Anesthetic management of these infants, often with concurrent pneumonia and respiratory distress (as in our patient) is challenging. PPV may lead to overinflation of emphysematous lobes and the domino effect of mediastinal shift and cardiac arrest. Lateral decubitus position poses another challenge. In addition to nerve and compression injuries, ventilation and perfusion impairment in an already compromised infant must be considered. Inhalational induction of anesthesia with maintenance of usually presents in the neonatal period. Exact etiology of the disease is not known, but in 25% cases, it may be associated with intrinsic bronchial narrowing or external bronchial compression resulting in air trapping by a ball valve mechanism. CLE can be hypoalveolar or poly-alveolar, based on number of alveoli within each acinus. Usually one lobe is affected, but bilateral involvement is seen in 20%. Congenital heart disease is associated in 12-14% of these patients. Clinical signs of CLE include tachycardia, tachypnea, and chest retractions which progress to respiratory distress and respiratory failure. Asymmetric expansion of the hemithorax, rhonchi, displacement of apical impulse, hyper resonant percussion on affected side, and diminished breath sounds and heart sounds may be noted. Lobar hyperinflation, atelectasis of contralateral lung, mediastinal shift, and flattening of ipsilateral diaphragm are typically seen on CXR. Thus, the disease is often confused with pneumonia and pneumothorax, even resulting in wrongful placement of a chest drain. CT and MRI help in diagnosis of CLE, but the single photon emission tomography ventilation-perfusion lung scan is confirmatory. This scan may reveal hypoperfusion of affected lobe due to compression of vasculature and hyperperfusion of normal lobes by shunted blood. While conservative management for CLE has been described, lobectomy is the mainstay of treatment. The management of a child with CLE with pneumonia or respiratory failure is controversial. Clearing the infection prior to surgery can improve pulmonary mechanics, but unrelieved compression atelectasis can be detrimental as in our patient. Anesthetic management of these infants, often with concurrent pneumonia and respiratory distress (as in our patient) is challenging. PPV may lead to overinflation of emphysematous lobes and the domino effect of mediastinal shift and cardiac arrest. Lateral decubitus position poses another challenge. In addition to nerve and compression injuries, ventilation and perfusion impairment in an already compromised infant must be considered. Inhalational induction of anesthesia with maintenance of
spontaneous respiration has been recommended in CLE. However, complete avoidance of PPV may not be possible as hypoventilation and apnea often develop under deep levels of anesthesia. In our case, poor respiratory reserve of the patient (due to atelectasis and infection) necessitated some positive pressure ventilation to maintain saturation. There is no consensus on the airway pressure to be maintained during manual ventilation. Cote et al suggested gentle manual ventilation before thoracotomy with positive airway pressure kept at 20-25 cmH\textsubscript{2}O; Tempe et al recommended an airway pressure less than 20 cmH\textsubscript{2}O. We attained adequate depth using sevoflurane and intubated the trachea without muscle relaxant, with surgical team on constant stand-by. Muscle paralysis at induction and pressure controlled ventilation with airway pressure below 20-25 cmH\textsubscript{2}O\textsuperscript{0} could have been beneficial for our patient as she had respiratory distress (high work of breathing). This modality, however, was not available in our set up for an infant. Thus, spontaneous ventilation was maintained with gentle manual assistance, and a vigil on vital signs, till affected lobe was extrathoracic. Pressure regulated volume controlled and high frequency ventilation are other useful techniques.


References

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