Pediatric Anesthesia and Critical Care Journal 2015; 3(2):85-88
doi:10.14587/paccj.2015.16

PACCJ

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

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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 tertiary care public hospital in New Delhi, India.

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, 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. This adds to the challenges faced by the anaesthesia team working in a limited facility.

Case report

A three month-old, term female child, weighing four kilograms, was referred to our hospital from a peripheral centre following repeated hospitalizations for fever, cough, and respiratory distress. She was admitted five times in three months for recurrent pneumonia. Cardiac anomalies were ruled out. Computed tomography (CT) scan of chest was advised, and the patient was referred to our hospital for further management. On arrival, the child was found awake, underweight, febrile, and in respiratory distress. There were marked intercostal and subcostal retractions. Air entry was decreased in the right hemithorax. There were crepitations in the bilateral lower zones and expiratory wheeze. Saturation was 85% on room air and 92% under oxygen hood. There was no cyanosis. Routine haemotological studies and biochemical investigations were normal. Under the oxygen hood, arterial blood gas (ABG) analysis revealed pH 7.46, paO_2 66 mmHg, and $paCO_2$ 27 mmHg. Chest

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.

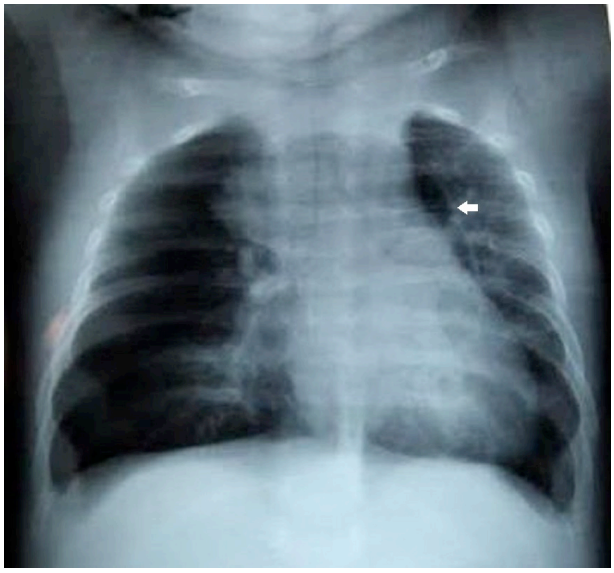

Fig. 1. Preoperative chest x-ray AP view showing hyperlucency of right upper and middle zone with leftward mediastinal shift, and segmental collapse in right upper zone; arrow indicates visceral pleura of herniated part of hyperinflated right lung.

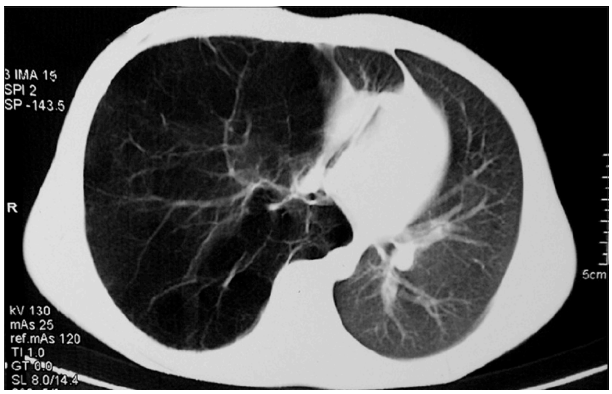

Fig. 2. NCCT chest showing hyperinflated right middle lobe with parenchymal herniation to left side through the anterior mediastinum.

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_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. Maintainance 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).

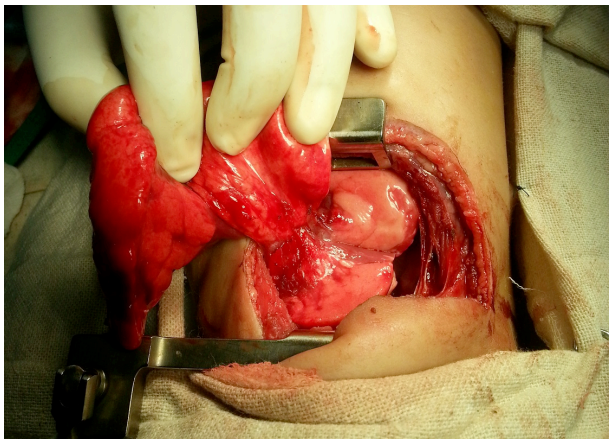

Fig. 3. Emphysematous lobe delivered through thoracotomy.

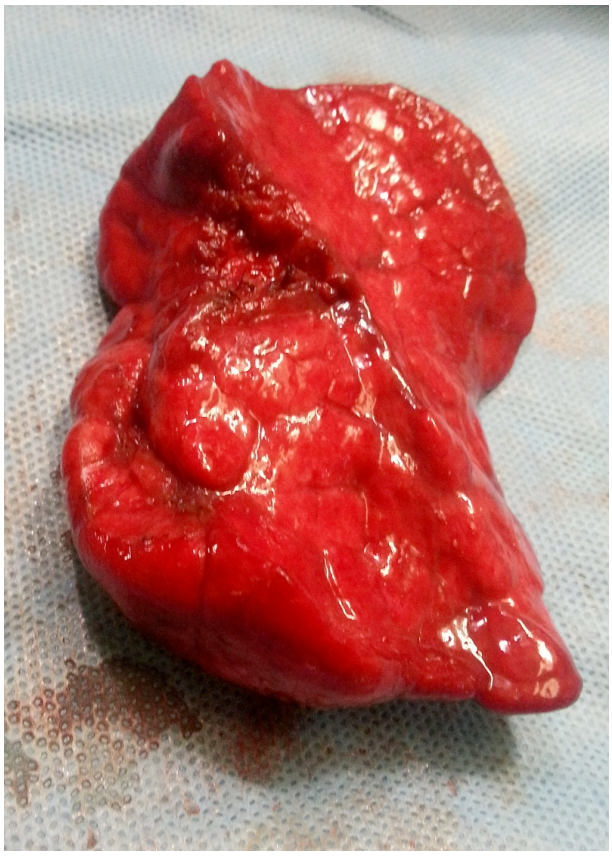

Fig. 4. Specimen of the resected emphysematous right middle lobe

Right-sided intercostal drain was placed with an underwater seal. Intraoperative spO_2 ranged from 85-94%, but improved with recruitment of the remaining lung. Blood and fluid loss were judiciously replaced. At the end of the surgery, intercostal nerve block was performed by surgeon with bupivicaine 0.25% (2 mL). The patient was ventilated in the postoperative period for four hours. Postoperative CXR revealed expanded left lung and right upper and lower lobe, and normal position of mediastinum. The child was discharged on the sixth post-operative day, after complete resolution of pneumonia.

Discussion

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.[1,2] It 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.[1-3] CLE can be hypoalveolar or polyalveolar, based on number of alveoli within each acinus.[4] Usually one lobe is affected, but bilateral involvement is seen in 20%.[1-3] Congenital heart disease is associated in 12-14% of these patients.[3] 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[5] and pneumothorax, even resulting in wrongful placement of a chest drain.[2] CT and MRI help in diagnosis of CLE, but the single photon emission tomography ventilation-perfusion lung scan is confirmatory. [2] 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 pnuemonia or respiratory failure is controversial. Clearing the infection prior to surgery can improve pulmonary mechanics,[6] 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.[7] 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_2O;[8] Tempe et al recommended an airway pressure less than 20 cmH_2O.[2] 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_20[9] 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 vitals, till affected lobe was extrathoracic. Pressure regulated volume controlled and high frequency ventilation are other useful techniques.[2] Gupta et al[10] employed endobronchial intubation until resection of the affected lobe and controlled ventilation throughout the surgery. One lung ventilation may be required in thoracoscopic surgeries. Nitrous oxide was avoided in this surgery as it is known to diffuse rapidly into closed cavities, causing further hyperinflation. Options for analgesia include intravenous fentanyl or ketamine, caudal epidural analgesia, and intercostal nerve blocks. Fluid balance is an important consideration. There is usually dramatic improvement in the status of the patient once the lobectomy is accomplished and the compressed lung is reexpanded. Infants can usually be extubated at end of procedure. It was decided to electively ventilate our patient in view of poor preoperative respiratory condition and since the case was taken up as an emergent case in early hours of the morning.

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