Congenital lobar emphysema: anaesthetic considerations

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Congenital lobar emphysema (CLE) is a rare clinical entity that usually presents as acute respiratory distress. It is potentially reversible, though possibly life-threatening, cause of respiratory distress in the neonate. It poses dilemma in diagnosis and management. We are presenting a 40 days old baby who presented with a sudden onset of respiratory distress related to CLE affecting the left upper lobe. Lobectomy was performed under general anaesthesia with spontaneous and controlled lung ventilation. Strategies to prevent hyperinflation and anaesthetic consideration of various techniques adopted for lung separation in infants have been reviewed.

Keywords:
congenital lobar emphysema, gentle manual ventilation, inhalational induction

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Introduction

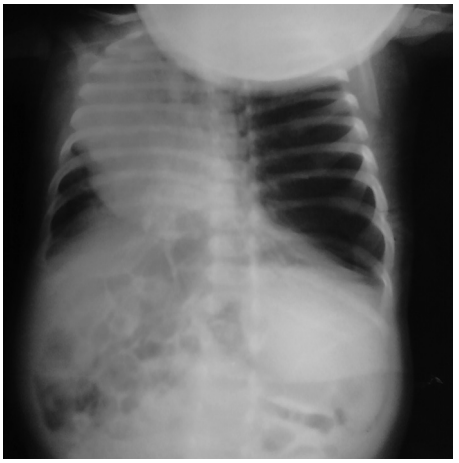
Congenital lobar emphysema (CLE), which is also known as congenital lobar overinflation or infantile lobar emphysema, is defined as an idiopathic postnatal abnormal overdistension of an otherwise anatomically normal lobe of the lung, characterized by expiratory air trapping within the lobe producing compression and atelectasis of adjacent lobes as a result of check valve mechanism [1–3]. It is uncommon, but when present it occurs more commonly in male children. Usually, only one lobe is involved, with the left upper lobe being affected in about 50% of cases, the right middle lobe in 30% and the right upper lobe in 20% [4]. Incidence of CLE is reported to be between one in 70 000 and one in 90 000 live births [5]. As yet, very few cases have been reported. In our institute, a case of CLE was reported after a gap of 8 years. Its early recognition and surgical intervention can be life saving.

CLE is a rare entity and usually presents as acute respiratory distress during infancy. Presenting features in infants can be dyspnoea, tachypnoea, retraction, wheezing, coughing, cyanosis and asymmetric breath sound. In these infants, there is an increased intrathoracic pressure because of hyperinflation of one or more pulmonary lobes, leading to mediastinal shift and atelectasis of the ipsilateral or contralateral lobes of the lung, impaired venous return, secondary hypoxia and hypotension. Chest radiographs help to diagnose but are not definitive [6]. A computed tomography (CT) scan confirms the diagnosis and may rule out associated anomalous vascular slings [7]. All patients should have adequate preoperative cardiac evaluation by echocardiography and CT scan to rule out any associated congenital heart disease or vascular anomalies.

Case history

A 40-day-old, full-term, male baby weighing 3.5 kg delivered vaginally at a peripheral maternity hospital to a gravida two mother was presented at our hospital with respiratory distress. Since birth, the baby had respiratory distress, which gradually worsened. Fifteen days after birth, the baby had severe respiratory distress with fever for which parent sought the advice of a paediatrician. On clinical examination, a diagnosis of pneumonia was made. However, medical management of pneumonia failed to resolve the problem and respiratory distress worsened. The paediatrician advised a chest radiography. The chest radiograph (Fig. 1) revealed hyperinflated left lung with shifting of the mediastinum towards the right side. There was no pneumothorax. A CLE was suspected; hence, contrast-enhanced CT scan of the chest was advised. A CT scan (Fig. 2) revealed grossly hyperinflated left lung upper lobe with attenuated pulmonary vascularities

Figure 1



Chest radiography.

and nonvisualization of the bronchi suggesting CLE. Hence, for further evaluation and management, the patient was referred to our hospital. After evaluating all investigations and clinical examinations conducted by the paediatric surgeon, left lobectomy was planned.

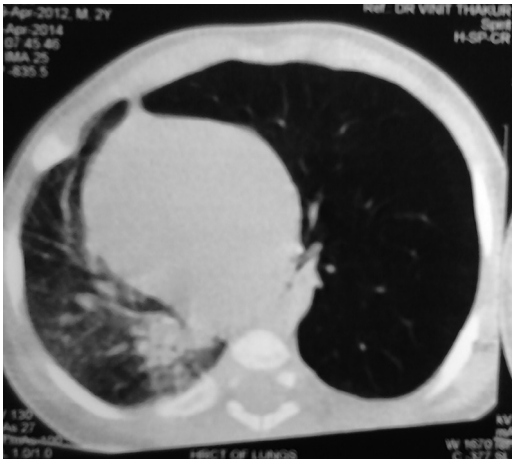
Preoperative examination revealed a heart rate of 150 beats/min, a respiratory rate of 56 breaths/min, presence of intercostal and sternal recession and hyper-resonant percussion on the left side. On auscultation, there was decreased breath sound on the left side of the chest. The cardiovascular system was normal. Oxygen saturation (SpO_2) was 88% in air. Routine haematological and biochemical investigations were within normal limits. Preoperative echocardiography was advised to rule out any associated cardiac anomalies. Echocardiography was normal. Arterial blood gas (ABG) analysis revealed a pH of 7.45, pO_2 of 70 mmHg and pCO_2 of 28 mmHg in air. The child was scheduled for left lobectomy the next day.

In the operation theatre, ECG, saturation and temperature monitoring were initiated and a peripheral line was secured. Standard precautions to avoid hypothermia were taken and premedication with intravenous atropine 0.04 mg and fentanyl 7 μg was given. A surgeon was made to scrub in and be ready to perform an emergency thoracotomy if the need arose. Induction was performed by a mask with 100% O_2 and sevoflurane with gradually incremental concentration up to 7%. After achieving an adequate depth of anaesthesia, the trachea was intubated with size 3.5 mm endotracheal tube (ETT) fixed at 10 cm. Gentle assisted ventilation was performed with hand using a Jackson Rees circuit; neonate was placed in the right lateral position and anaesthesia was maintained with sevoflurane 2–3% in 100% O_2. Local anaesthetic 0.125% (4 ml) bupivacaine was infiltrated in the line of incision before initiating surgery. A left upper lobectomy was performed through the 4th intercostal space with stable haemodynamic throughout the 2-h surgical procedure. After lobectomy, the left lower lobe was expanded using gentle manual recruitment and a left intercostal drainage tube was inserted. Once resection of the affected lobe was completed, controlled lung ventilation with atracurium as the neuromuscular blocking agent was initiated. Thereafter, nitrous oxide was added. Blood and fluid loss was monitored and replaced with 100 ml of warmed Isolyte-p and 50 ml of fresh whole blood. Blood gases intraoperatively and postoperatively were within normal limits. At the end of surgery, intercostal block was given with 4 ml of 0.125% bupivacaine, and residual neuromuscular block was reversed with neostigmine 0.2 mg along with atropine 0.04 mg intravenously. The infant was extubated when spontaneous respiration was sufficient to maintain SpO_2 above 90% in air. Later, the baby was kept in an oxygen-enriched environment in the Paediatric ICU under continuous SpO_2 and ECG monitoring. Postoperative analgesia was provided with rectal paracetamol. At 72 h the chest drain was removed after full expansion of the residual lung (Fig. 3). Rest of the postoperative period was uneventful, and the child was discharged after 7 days.

Discussion

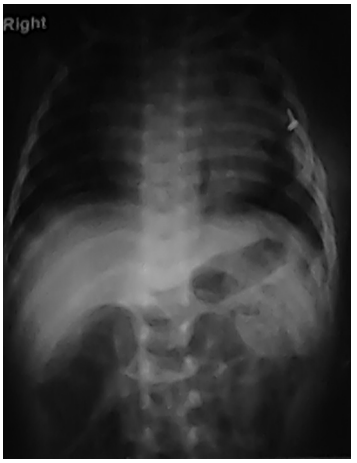
CLE may be diagnosed during prenatal ultrasonography, but most commonly it is missed and is detected in the neonatal period when progressive hyperinflation causes symptoms due to compression of the remaining ipsilateral as well as contralateral lung leading to cardiopulmonary compromise. Less severe CLE may not present until later in infancy or childhood.

Figure 2



Computed tomography scan.

Figure 3



Chest radiography on fifth postoperative day.

The child usually presents with rapid respiratory rate, tachycardia and chest retraction, which worsens with progressive accumulation of gas in the affected lobe and progresses into respiratory distress and respiratory failure. There is increased intrathoracic pressure because of hyperinflation of one or more pulmonary lobes resulting in mediastinal shift and atelectasis of the ipsilateral or contralateral lobes of the lung. This causes displacement of heart sounds, decreased venous return and varying degree of ventilation–perfusion mismatch further leading to hypoxia. Chest radiographs help in diagnosis but may not be definitive [6]. It is well known that the condition is often confused with pneumothorax, and patients have an inter costal drainage (ICD) tube inserted, which does not help, and instead may further increase the respiratory distress [8]. A CT scan usually confirms the diagnosis and also helps in ruling out associated anomalous vascular slings. In our present case, the treating physician had high degree of suspicion of CLE and had thus advised for CT scan well on time.

To have successful anaesthetic management of such cases, it is necessary to avoid overdistension of the emphysematous lobe before opening the chest, as it may result in severe respiratory and haemodynamic disturbances. Intermittent positive pressure ventilation (IPPV) should be best avoided as it can result in overdistension of the lung, as the critical airway pressure is not known [5].

The physiological considerations for anaesthetizing these infants undergoing thoracic surgery in the lateral decubitus position include ventilation and perfusion impairment of the lung. In contrast to adults, infants with unilateral lung disease do not improve their oxygenation when the healthy lung is dependent and the diseased lung is nondependent [9], as the chest wall of infant is more compliant, and thus cannot completely support the dependent lung. Hence, the functional residual capacity (FRC) is closer to the residual volume and airway closure becomes more likely in the dependent lung [10].

It is usually advisable to use inhalation induction, as spontaneous ventilation should be maintained until either the chest is opened or one lung ventilation (OLV) of the contralateral lung is achieved [11]. Positive-pressure ventilation is best avoided, but gentle assisted manual ventilation keeping airway pressure at 20–25 cm of H_2O may be needed if hypoventilation occurs during induction due to poor respiratory reserve [3]. As N_2O diffuses in a closed cavity, thus causing further compression and mediastinal shift, it should be avoided. Overdistension of the lung can be life threatening; hence, a standby surgical team is desirable for immediate thoracotomy during induction if need arises. Use of OLV is not common for open thoracotomies in neonates and infants as the surgeon is usually able to manually retract the lung and also because ETT size is too small for bronchial blocker and double-lumen ETT is too big for infants. Maintenance of anaesthesia is usually performed with inhalational agents, and opiates such as morphine are best avoided because of the fear of postoperative respiratory depression. Most of the patients are successfully extubated at the end of the surgical procedure. Need for elective postoperative ventilatory support arises mostly in those cases where more than one lobe has been excised or when analgesia is inadequate.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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