Anaesthetic Management of Congenital Lobar Emphysema : A Report of Two Cases

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Introduction

Congenital lobar emphysema (CLE) is a rare cause of sudden respiratory distress in the newborn and infants [1]. Hyperinflation and air trapping cause expansion of the affected lobe along with the compression of other lung tissue, mediastinal shifting and impaired venous return. Anaesthesia for CLE, is challenging, as often children are sick and the laid down principles of anaesthesia such as keeping the airway pressures low, cannot be adhered to. Attempts to maintain spontaneous respiration may not be successful and institution of positive pressure ventilation has the potential of worsening the condition.

The aim of this paper is to present two case reports with unusual clinical features, each highlighting two different aspects of the disease. The first case had a short history, rapid downhill course with features of life threatening mediastinal shift whereas the second case had an eventful induction of anaesthesia. Anaesthetic management of cases with CLE is outlined and the relevant literature is discussed briefly.

Case Reports

Case No. 1

6 weeks old male child weighing 4 kg presented with history of fever and rapid breathing rate for last four days. Examination revealed the child in respiratory distress with intercostals retraction and tachypnoea of 60-64 per minute. The oxygen saturation (SpO₂) on room air was 90%. The face was congested and the neck veins were engorged, suggesting presence of superior vena cava (SVC) obstruction. Auscultation revealed reduced air entry on the left side and bronchial breathing over right hemi thorax. An urgent arterial blood gas (ABG) analysis showed respiratory acidosis (pH-7.2, PO₂ – 101.3 mmHg, CO₂ – 68.2 mmHg, BE – 1.3). Hb was 8.2 gm% and chest radiograph revealed right upper lobe collapse with hyperinflation of left upper lobe and gross mediastinal shift to the right side. A diagnosis of CLE was made and in view of his condition the child was put on ventilator and an emergency thoracotomy was planned.

The child was transported from the neonatal intensive care unit (NICU) in an incubator on T piece. Inside the theatre, monitoring in form of electrocardiography (ECG), non-invasive blood pressure (NIBP) and pulse oximeter was established and the child was induced with oxygen and halothane via the existing endotracheal tube. Ventilation was assisted gently and thereafter, thorough tracheal suction was done. As the secretions were thick it was decided to remove the tube and perform a rigid bronchoscopy to facilitate bronchoscopic lavage and suctioning of tenacious, viscid and purulent secretions. As suctioning was completed, the chest lift became better and SpO₂ also improved to 95%. Thereafter, the child was given atracurium and a double lumen tube (DLT) was passed in order to isolate the left side of lung. After checking the proper placement of the DLT, the child was positioned for left sided thoracotomy. For analgesia, a mixture of 4 ml of 0.25% bupivacaine and 0.2 mg of morphine was deposited in the caudal epidural space.

Anaesthesia was maintained with oxygen, halothane, fentanyl and increments of atracurium. Trachea was found to be full of purulent secretions and repeated suctioning was necessary. Nitrous oxide was added after the lobe was isolated and clamped and branches of pulmonary artery and vein were ligated and divided. The emphysematous left upper lobe was excised and then the collapsed right and left lower lobes were allowed to expand. At the conclusion of surgery, chest physiotherapy was done and secretions were sucked out further. The DLT was replaced with a single lumen tube and the anaesthetic agents were terminated followed by reversal of residual neuromuscular blockade with neostigmine and atropine. The child was electively ventilated for 48 hours in NICU and was weaned off successfully thereafter. Postoperatively chest radiograph was repeated in the evening, which revealed complete correction of mediastinal shift and remarkably enough, resolution of the right upper lobe collapse.

Case No. 2

A male term infant aged 6 weeks weighing 3.1 Kgs presented with complaints of rapid breathing rate and fever of three weeks duration. He was treated in a peripheral hospital initially for pneumonia and then for pneumothorax. However

there was no improvement even after medical therapy and ICD insertion. He was then referred to the present hospital. The child looked restless, had a tachypnoea of 55 per minute and required oxygen with nasal prongs to keep saturation (SpO_2) above 95%. On examination of respiratory system, there was no air entry on the left side of the chest whereas on the right side, conducted sounds were heard. Heart sounds were normal but better heard on the right side. The chest radiograph was repeated which raised the suspicion of CLE and it was later confirmed by CT scan. There was minimal mediastinal shift. Arterial blood gas analysis (ABG) on room air was consistent with hypoxia and there was no acidosis. Other biochemical parameters were within normal limits.

With a diagnosis of CLE, excision of left upper lobe under general anaesthesia was planned. Under SpO_2 and ECG monitoring, induction was started with 100% oxygen and halothane however the child became apnoeic almost immediately and respiration had to be assisted. Care was taken to keep the inflation pressures low. Mask ventilation was easy and there was no change in saturation. Immediately prior to intubation, two drops of 4% lignocaine jelly was smeared with gloved finger onto the posterior pharyngeal wall down to the epiglottis. During intubation, there was severe bradycardia with bronchospasm, which was transient and responded to injection atropine 20 $\mu\text{g}/\text{Kg}$. Anaesthesia was maintained as in the first case. However as the child remained apnoeic no muscle relaxants were required. SpO_2 , ECG, NIBP, capnography and temperature were monitored. During surgery, there were no adverse events except fall in temperature 36.4°C to 33.4°C despite active warming measures. During the course of surgery, tracheal suctioning was required repeatedly. Nitrous oxide was added after the bronchus and pulmonary vessels were ligated. After the excision of the emphysematous upper lobe and ruling out any leaks an ICD seal was placed. Blood loss was insignificant. Analgesia was provided with fentanyl and the intercostal nerves were infiltrated with bupivacaine from the pleural side just before closure. At the conclusion of surgery, the child was electively ventilated primarily because of hypothermia for six hours in neonatal intensive care unit (NICU) and was weaned off the ventilator successfully.

Discussion

CLE is an uncommon condition, which affects boys twice as much as girls. It is characterized by over inflation, irreversible distension and rupture of alveoli of one or more lobes, secondary to bronchial obstruction. The cause of obstruction is unknown in most of the cases although many show evidence of deficient and disordered cartilage. Some patients have widespread lung disease with poor lung recoil throughout [2]. Most commonly affected lobe is the left upper lobe as seen in both the cases, followed by right middle and right upper lobes.

CLE usually appears between the neonatal period and the first six months of age. In about 50% of cases symptoms are present in the neonatal period with respiratory distress and radiographic features of CLE

[3]. In the remainder, symptoms are evident before 6 months. Initially the child has rapid respiratory rate, tachycardia and retractions, which worsens as progressive accumulation of gas occurs in the affected lobe. There is asymmetric expansion of the thorax, wheezing, displacement of apical impulse, hyper-resonance to percussion and diminished breath sounds and heart sounds. Chest radiograph may initially show opacification followed by over-distension of the affected lobe along with atelectasis in the other lobes and shift of mediastinum. The X-ray can also help in differentiating lobar emphysema and pneumothorax or congenital cysts by the presence of faint bronchovascular markings and herniation of the affected lobe across the midline. A stable child may be subjected to further investigations like radioisotope perfusion scan studies, angiography and CT imaging. Differential diagnosis of cyst like lesions on the plain chest radiograph include infected sequestered lobe, bronchopulmonary dysplasia, congenital cysto-adenomatoid malformations (CCAM), single or multiple developmental cysts, congenital diaphragmatic hernia, pneumatocele, lung abscess and encysted empyema [3]. Management options can be both, conservative as well as surgical, usually the latter. Conservative management may be successful with spontaneous resolution. The outlook following the surgery is generally good provided there are no associated abnormalities. However, if there is rapid deterioration, chest tube placement, aspiration of trapped air and vigorous mechanical ventilation have been tried as palliative measures but these are associated with much higher mortality than thoracotomy and lobectomy.

Preoperative evaluation depends on the degree of the distress and should be aimed at establishing cardio-respiratory stability of the child. The progression of distress and the need of supplemental oxygen are key components of the examination and many a times, infants who deteriorate rapidly, constitute a surgical emergency to relieve the expanding lobe with its ventilatory and cardiac impairment. In such situations, emergent palliative measures such as those mentioned above, leave very little time for further preoperative preparations. Cardiac evaluation is important since approximately 14% of patients are likely to have congenital heart diseases [4].

Induction of anaesthesia is the most critical phase in the management of children with CLE. The crying and struggling infant can increase the amount of trapped gas. Similarly, positive pressure ventilation or the positive airway pressure to assist the ventilation by the anaesthesiologist can also increase the emphysema. The aim should be to achieve a smooth inhalational induction with oxygen and halothane with minimum positive pressure ventilation until the chest is open. Controlled

or assisted ventilation is added as and when unacceptable hypoventilation develops while intubation is being performed. An alternative induction approach especially for the unstable infants is, sedation with intravenous ketamine and local anaesthetic infiltration of the incision site [5]. After the intra-thoracic pressure has been released, general anaesthetic can then proceed with any other appropriate technique. In addition to the problems of thoracotomy and neonatal anaesthesia, there may be sudden expansion of the emphysematous lobe during induction following assisted ventilation. This is a real possibility, as these children, who often have poor cardio-respiratory reserve, lose their spontaneous respiratory efforts even after minimal assistance. This was noticed in both the cases, though fortunately there was no worsening. In such situations, gentle manual ventilation should be done with a close watch on the haemodynamic parameters and oxygenation. If the lobe expands suddenly, the surgeon should be ready to open the chest immediately and relieve the pressure. The use of muscle relaxants depends upon the patient's tolerance of positive pressure ventilation. High frequency ventilation has been used successfully in the patients with CLE, as low airway pressures are especially suitable [6]. It is recommended if the circumstances permit and the user is familiar with the technique. After induction, a thorough application of local anaesthetic jelly (lignocaine 2-4%) to the posterior pharyngeal wall, will further smoothen the course of anaesthesia. This can also be done if awake intubation is contemplated and is commonly practiced by the author.

Commonly, single lumen tubes are used for the purpose of tracheal intubation. Even though lung isolation is desirable, double lumen tubes are not freely or commercially available. In small infants, lung isolation is technically difficult and not commonly practiced [7]. As an alternative, selective main stem bronchus intubation can be done. A detailed discussion on the problems of one lung ventilation and lung isolation is beyond the scope of this paper. It will be sufficient to mention that lateral decubitus position can cause significant ventilatory changes in infants which is further complicated by opening of pleura, retraction of lungs and surgical manipulation and standard strategies should be adopted to overcome the detrimental effects of lateral decubitus position and ventilation-perfusion mismatches and to ensure optimal oxygenation.

During maintenance of anaesthesia nitrous oxide should be avoided initially as it can expand the emphysematous area. However, there is a belief that nitrous oxide is unlikely to enter the affected area as it is poorly perfused (personal communication, Dr Ian McKenzii, Consultant Paediatric Anaesthetist, Royal Children Hospital, Melbourne, Australia). Blockage of

the tube with secretions and sometimes blood is common and requires repeated and meticulous suctioning. Pre-operative bronchoscopy was done in the first case to suck out thick secretions and it has a definite role in aiding the diagnosis, ruling out any foreign body and removing any cause of bronchial obstruction in addition to bronchoscopic lavage and suction. Pulse oximetry remains the cornerstone of monitoring to detect rapid changes in the oxygenation in the preoperative period as well as during induction [8]. Facilities for airway pressure monitoring are desirable whereas EtCO₂ is useful in detecting sudden changes in ventilation. Effective analgesia can be provided with epidural narcotics administered caudally. Another attractive option is to block the intercostals nerves under direct vision before the thorax is closed. In most of the cases the trachea can be extubated at the end of the surgery. Elective postoperative ventilation is usually not required especially if excision is limited to one lobe and the other lung is in good functional state. However in our cases, elective ventilation was necessitated for reasons other than those because of the disease or surgery. These children do well postoperatively, however they have reduced forced vital capacity not only in the immediate postoperative period but also later in the childhood [9].

In summary, patients with CLE present a myriad of problems to the anaesthesiologist, which are highlighted in the reports above. It is further emphasized that induction remains the most critical phase during anaesthesia more so in patients with mediastinal shift.

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