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Anesthetic considerations and challenges in infants with congenital pulmonary airway malformation

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Keypoints

Congenital Pulmonary Airway Malformation (CPAM) formerly called Congenital Cystic Adenomatoid Malformation (CCAM) is a rare congenital lung lesion. Authors report two cases of CPAM posted for thoracotomy with excision.

Abstract

Congenital pulmonary airway malformation (CPAM) is a rare entity. Anesthetic management for lobectomy is challenging due to sudden hemodynamic collapse which may occur during induction and ventilatory problems due to hyperinflation of cystic lesions with positive pressure ventilation. We discuss successful anesthetic management of two infants with CPAM posted for pneumonectomy with a brief mention of the various options available for one lung ventilation in this age group.

Keywords: infant, congenital pulmonary airway malformation, anesthesia, one lung ventilation, double lumen tube

Introduction

Congenital Pulmonary Airway Malformation (CPAM) formerly called Congenital Cystic Adenomatoid Malformation (CCAM)[1] is a rare congenital lung lesion with an estimated incidence of 1:25,000 to 1:35,000 live births[2,3]. It is characterized by overgrowth of terminal bronchioles with reduced alveoli which do not participate in gas exchange. The mortality is high with 12.5 %death rate in newborns[3]. We report two cases of CPAM posted for thoracotomy with excision.

Case reports

The first baby was a 2 month old female child born at term by vaginal delivery. An antenatal ultrasound (USG) done at seven months of gestation did not reveal any abnormality. Apgar score was 8/10 at 5 minutes of life. She weighed 3.7 kg and presented with gradually increasing chest indrawing and decreased feeding since a month of life. On examination baby was tachypneic but maintained saturation on room air. Computerized tomography (CT) chest revealed CPAM involving right lung with herniation towards left side. Trachea, heart and mediastinum were shifted to left side (Figure 1)

The second baby was a 45 day old female child weighing 3.4 kg, born at term by vaginal delivery. Apgar score was 9/10 at 5 minutes of life. She presented with gradually increasing respiratory distress and decreased feeding since a month. On examination child had respiratory distress. Room air saturation was maintained at 96%. Chest radiograph showed consolidation of left lower lobe. CT chest revealed multiple cysts ranging from 2-4 cm in left upper lobe, lingula and apical segment of left lower lobe, suggestive of CPAM. Left lower lobe basal atelectasis was present, and right lung was clear.

Antenatal USG performed at 3, 5 and 9 months of gestation had missed the diagnosis.

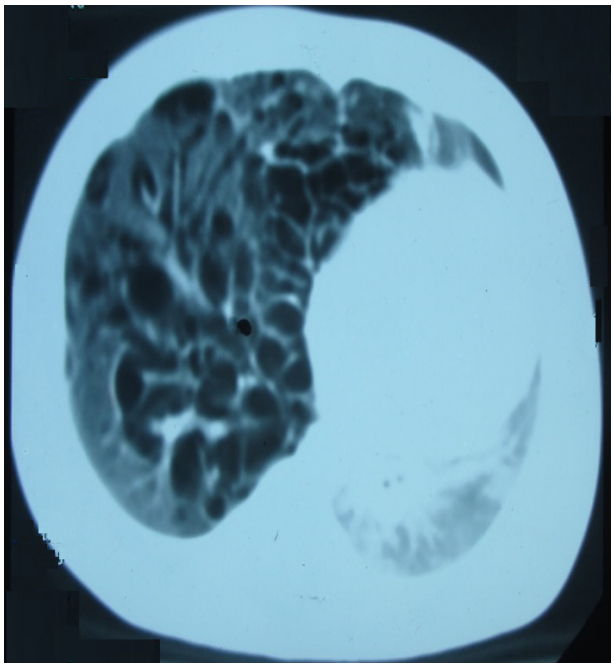

Figure 1. CT chest showing emphysematous hyperinflation of right lung with mediastinal shift

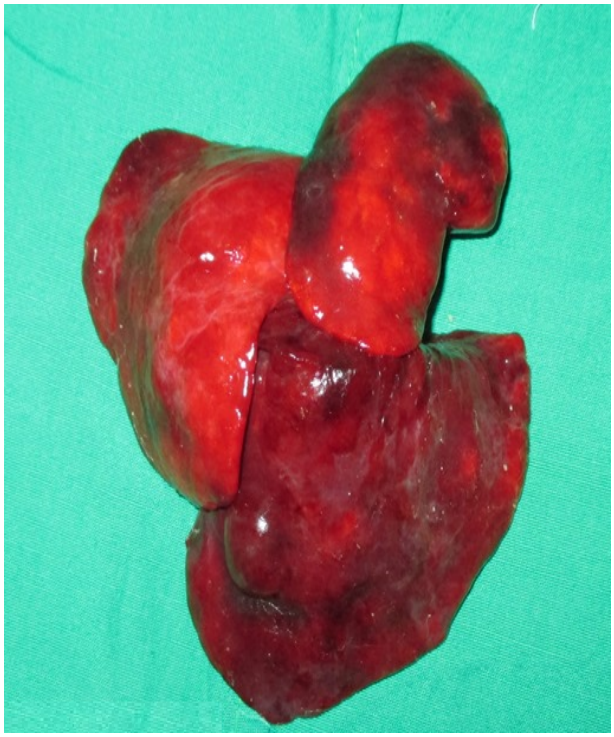

Figure 2. Resected specimen of upper lobe, lingual and apical segment of left lung

Both babies were posted for thoracotomy with excision of cysts. General anesthesia with epidural analgesia was planned in both cases. On shifting to operation theatre, the babies were placed on a warming mattress. Both babies had 24 G IV cannula in situ and second access was taken after induction. All standard monitors were attached including cardioscope, pulse oximeter, non invasive blood pressure and temperature probe. Following preoxygenation anesthesia was induced with sevoflurane. IV glycopyrrolate 4 mcg/kg, IV fentanyl 2 mcg/kg was administered. IV atracurium 0.5 mg/kg was given for muscle relaxation and trachea was intubated with4 mm ID uncuffed endotracheal tube by oral route in both patients. 10 Fr infant feeding tube was inserted to decompress the abdomen. Anesthesia was maintained on (O_2 + air) 50:50 and sevoflurane at 1 MAC with intermittent doses of IV atracurium.

Under all aseptic precautions, caudal epidural space was located using a 20G Tuohy's needle. After confirmation of the space, 24G epidural catheter was inserted and threaded up to the thoracic level. The catheter was fixed after tunneling. Test dose of 0.1ml/kg of 1% lignocaine with adrenaline was administered following which bolus dose of bupivcaine 0.125% 2 cc was injected through the caudal catheter.

Intra-operative traction of lung and bronchus resulted in intermittent fall in saturation. During this time, anesthesia was maintained with 100% O_2 and sevoflurane. In the first child right pneumonectomy was performed and in the second case, left lobectomy was performed (Figure 2).

Intra-operatively hemodynamic parameters were well maintained. Maintenance fluid (ringer lactate with 2% dextrose) was given at the rate of 6 ml/kg/hr. Blood loss was replaced with packed red blood cells. Duration of surgery was approximately 90 min. At completion of surgery neuromuscular blockade was reversed and trachea was extubated.

Post-operative analgesia was maintained with 6 hourly bolus doses of 0.125 % bupivacaine. Post-operative chest radiograph showed good lung expansion and both babies were discharged home after a week.

Discussion

CPAM is a rare developmental lung anomaly. 80% cases present in newborn period with same frequency in premature and term infants. It is usually unilateral and restricted to a single lobe[3]. It develops during the pseudoglandular and saccular period of lung development[4].15-50% of CPAM decrease in size before birth significantly[5]. Complete post-natal resolution is rare and half of these children subsequently require surgery[6]. USG, CT, MRI and chest radiograph are used to identify and locate lung abnormality and to identify other thoracic abnormalities. Associated renal, intestinal, bony and cardiac anomalies are present in up to 25% of patients with CPAM.[3]

Patients commonly present with respiratory distress, retraction, grunting, recurrent cough and cold[6,7]. Cystic expansion and compression of structures lead to pulmonary hypoplasia, mediastinal shift with possibility of spontaneous pneumothorax. Hence symptomatic lesions require urgent radiological evaluation with surgical excision of lesions[3].

For asymptomatic patients some centers follow conservative management[8,9]. Surgery is deferred if patient is asymptomatic at birth. However CT scan should be performed within one month post-natally to demonstrate nature of abnormality and connection with tracheobronchial tree.Long-term out-come is good for surgically managed patients despite slight decrease in lung volume[5].

Monitoring of vital parameters during neonatal and infant surgery is essential, especially during thoracotomy as the child is at great risk[10].

During anesthesia due to positive pressure ventilation there is risk of rapid inflation of cysts with sudden mediastinal shift and cardiac arrest[11]. Therefore lungs should be ventilated with minimal airway pressure to avoid such complications before the chest is opened. Surgeon presence during induction is mandatory for emergency wide bore needle decompression of hyperinflated cysts causing hemodynamic instability. One lung ventilation (OLV) can be employed as a ventilatory strategy. OLV improves surgical access and may reduce blood loss. It also minimizes trauma to the limited residual normal lung tissue and protects normal lung from contralateral contamination. This is challenging in an infant but can be achieved with a double lumen tube (DLT) (Marraro DLT for neonates and infants), double endotracheal tube intubation, selective mainstem bronchial intubation, bronchial blockers [Fogarty catheters (3, 4,5 Fr G), Arndt endobronchial blocker (smallest 5 Fr G)] or surgical retraction. Physiological considerations of OLV in neonates and infants should be kept in mind and include higher oxygen consumption, immature alveoli and increased chest wall compliance which impedes the oxygenation of the healthy dependent lung unlike adults. Additional ventilatory problems due to positioning, retraction, compression make OLV more difficult to institute in this age group. Where OLV may not be possible, a micro cuff endotracheal tube can be used. However the smallest available size is 3.5 mm ID and manufacturer's recommend downsizing by 0.5 mm for appropriate size selection. We were unable to use this tube in both our babies as they were too small for the smallest size micro cuff tube. During maintainence of anaesthesia, hyperinflation of cysts can be prevented by avoiding the use of nitrous-oxide before delivery of affected lobe. It has faster diffusion capacity leading to hyper-inflation, compression of normal lung and mediastinal shift[12]. Surgery can be associated with additional haemodynamic effects like decreased venous return, aortic compression, mediastinal retraction, mechanical arrhythmias and bleeding.

Conclusion

Thorough knowledge, anticipation of complications with efficient monitoring intra-operatively and post-operatively along with good analgesia lead to successful management in such cases.

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