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Anaesthetic management of a child with congenital lung cyst posted for thoracotomy

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ABSTRACT

Congenital pulmonary airway malformations are rare development disorders of the lung mimicking conditions such as congenital lobar emphysema, bronchogenic cyst, bronchopulmonary sequestration etc. They can be detected prenatally during ultrasonogram, or may present later in infancy or childhood as respiratory distress, infection or rupture of the cyst causing pneumothorax. Surgery is the mainstay of treatment. Here we discuss the perioperative management of a ten-year-old child with a lung cyst who initially presented with a pneumothorax like picture. The child was posted for elective thoracotomy and lung cyst resection under general anaesthesia. The main anaesthetic concern was rupture of the cyst with positive pressure ventilation and resultant haemodynamic compromise. We avoided positive pressure ventilation and maintained the child on spontaneous ventilation till thorax was opened and also avoided nitrous-oxide intra-operatively. The intra-operative period was uneventful. The child was extubated on table and post-operative period was uneventful.

Keywords: CPAM, CCAM, anaesthesia, ventilator strategy

INTRODUCTION

Congenital pulmonary airway malformations (CPAM), previously called Congenital cystic adenomatoid malformations (CCAM) are a spectrum of diseases which have rare incidence and varied presentation. They are most commonly detected at or before birth, but if asymptomatic, has been shown to present as late as 13 years of age with symptoms of pneumonia, infected CPAM, respiratory distress or pneumothorax. Surgical resection of such lesions has been determined to be the definitive treatment. Here we discuss the anaesthetic management of a child with CPAM.

CASE REPORT

A 10-year-old female child presented with acute onset of fever, breathlessness and lethargy of one-day duration to paediatric ED. On examination, the child was febrile, dyspnoeic, tachypnoeic (RR of 50/min) with signs of respiratory distress (suprasternal and intercostals retraction) and no cyanosis. She had a low pulse volume with tachycardia and cool peripheries. On auscultation, air entry was diminished on the left side.

Initial investigations revealed increased Total Count of 14,600 cells/cu mm with other laboratory parameters being normal. Chest X-ray mimicked left-pneumothorax like picture (Fig 1). After initial supportive treatment of the child with fluids, antibiotics, vasopressors and antipyretics, a left-sided intercostal drainage of the pneumothorax was planned and (L) ICD inserted. Surprisingly, the repeat radiograph after ICD insertion showed persistent air collection even with the ICD in situ (Fig 2). This led to the suspicion of a lung cyst, for which a CECT thorax was done, which revealed the presence of a large emphysematous bulla in the left lower lobe of the lung (Fig 3).

Figure 1: Chest X-Ray before ICD insertion

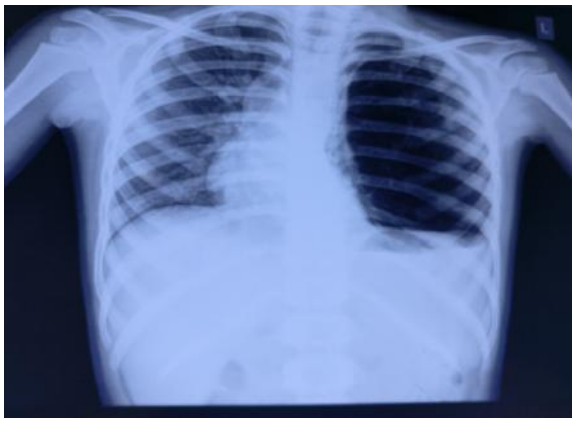


Figure 2: Chest X-Ray after ICD insertion

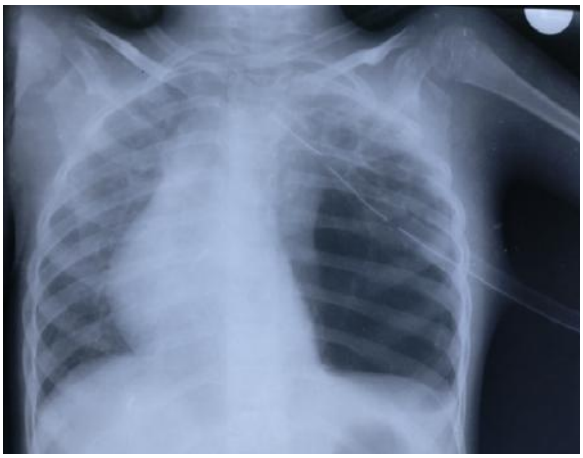
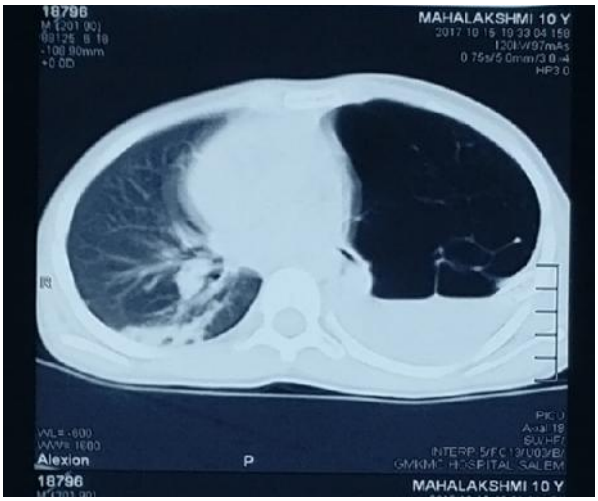


Figure 3: CT thorax showing large left emphysematous bulla



The child was planned for a thoracotomy and resection of the bullous lesion under suitable anaesthesia after initial supportive care.

In the pre-anaesthetic assessment clinic, the child was awake, alert and afebrile. Her BP was 100/60mmHg, HR 132/min, respiratory rate 25/min with a room air saturation of 93%. Cardiovascular examination was normal except for the tachycardia. Respiratory system examination showed shift of trachea to the right, decreased air entry on the left and a left sided ICD which was non-functional. Airway examination was normal with adequate mouth opening and a Mallampati grade II. The child weighed 25kg. Laboratory investigations showed Hb 9.9gm/dl, PCV 30%, platelet count- 5L/cu mm. Bleeding time was 1 minute and clotting time 5 minutes 30 seconds. Renal parameters and ECG were normal. Chest X-ray showed a large air column on the left side with ICD in-situ and mediastinal shift to the right. The child was assessed under ASA PS III.

The procedure and expected complications were discussed with the parents and an informed high-risk consent was obtained. Adequate blood products were reserved. Postoperative ventilator availability was ensured. Patient was kept nil per oral, 8 hours for solids and 3 hours for clear fluids.

In the OR, Anaesthesia machine was checked and all emergency drugs and equipments were kept ready. Baseline monitors such as NIBP, SpO2, ECG were applied and vitals recorded. A 22G IV cannula was secured in the right forearm and lactated Ringers solution @ 4ml/kg/hr started.

Since the most feared complication of a lung cyst is rupture during positive pressure ventilation, we planned induction of general anaesthesia with preservation of spontaneous ventilation. The child was preoxygenated with 100%O_2 (4L/min) for 5 minutes, premedicated with Inj.Glycopyrrolate 0.2mg IV and induced with Inj. Ketamine 60mg IV and Sevoflurane 3-4% using Jackson-Rees circuit. Nitrous oxide was omitted to avoid expansion of the cyst due to diffusion of N_2O into the cyst. After achieving adequate depth of anaesthesia, laryngoscopy was done and vocal cords were sprayed with two actuations of 10% lignocaine spray. The child was then intubated with 5mm armoured cuffed ETT- bilateral air entry confirmed (air entry diminished on left side due to the lung cyst). Anaesthesia maintained using O_2: Sevoflurane 3-5%. Intercostal nerve block was performed on the operative side from 2nd to 5th intercostal spaces using 12 ml of 0.25% Bupivacaine before skin incision.

The child was positioned in the right lateral decubitus position and a postero-lateral thoracotomy incision was made. Spontaneous ventilation maintained until the thorax was entered after which the patient paralysed, positive pressure ventilation was initiated and the patient connected to paediatric closed circuit - Drager Anaesthesia workstation with Tv- 200ml, RR- 14/min and PEEP 0 cm H_2O.

Intraoperatively, a 20 ×15cm cystic lesion was found arising from the lower part of the upper lobe with the upper lobe collapsed (Figure 4). The cyst was ligated and removed (Figure 5).

Figure 4: Intraoperative finding- lung cyst

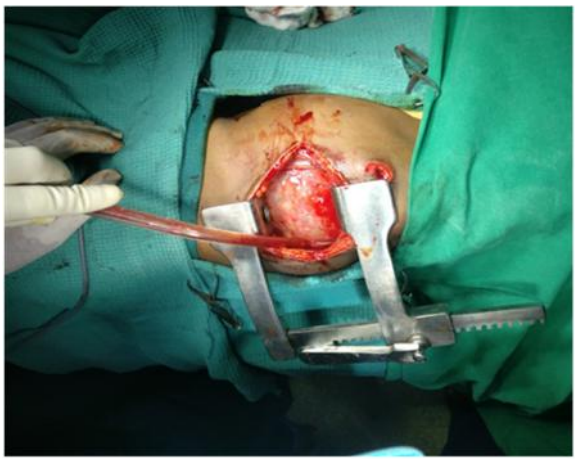
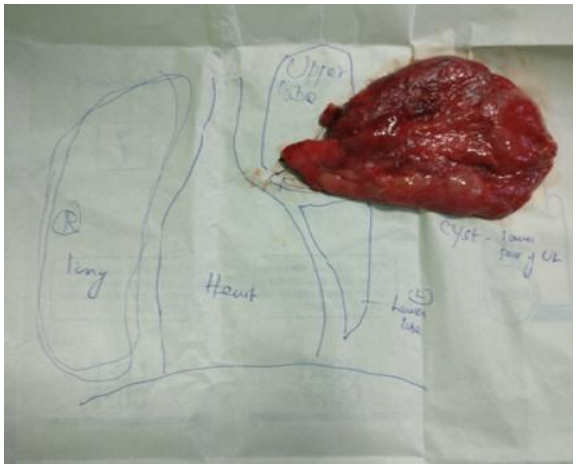


Figure 5: Resected specimen



The intraoperative hemodynamic parameters were stable (Figure 6) and the procedure was uneventful. Estimated blood loss was around 100ml. 300 ml of warmed RL was infused. Urine output was 100 ml. Inj. Paracetamol 250 mg iv was given for analgesia towards the end of the procedure.

Figure 6: Intraoperative vital parameters

Figure 7: Child in recovery position post-operatively

At the end of the surgery, the child was reversed after complete return of consciousness and after return of adequate muscle power. Child was extubated after careful suctioning and nursed in the recovery position (Figure 7)

Figure 8: Child on pulmonary rehabilitation

Postoperative period: Child was shifted to the PACU for postoperative observation. She was advised O_2 by mask at 4L/min, strict monitoring of vitals and Inj. Paracetamol 250mg IV eighth hourly. She was also advised early mobilisation, chest physiotherapy and spirometry (Figure 8). The histopathological examination of the excised cyst showed tissue to be consistent with congenital pulmonary adenomatoid malformation. Child recovered well and was discharged on the 10th postoperative day.

DISCUSSION

Congenital cystic adenomatoid malformation of the lung is a rare hamartomatous disease of the lung causing abnormal fetal lung development where there is an imbalance between lung proliferation and apoptosis leading to an adenamotoid proliferation and cyst formation. It is non-hereditary in origin, which can be diagnosed as early as during prenatal ultrasonogram to later in childhood[1]

Children may present asymptomatically, with respiratory distress or with recurrent pulmonary infections. Physical signs include signs of respiratory distress such as tachypnoea, grunting and use of accessory muscles of respiration; signs of pneumothorax/air trapping like tracheal deviation, shifted heart sounds, and decreased air entry on the affected side etc. The most common complications associated with CCAM are pulmonary infections which do not respond to treatment, or less frequently, hydropneumothorax, hemoptysis and chronic cough. The cystic lung lesion may expand rapidly causing pulmonary hypoplasia, mediastinal shift and a possibility of spontaneous pneumothorax. The most dreaded complication is the malignant potential that some types of CCAM possess.

Stocker *et al*., suggested a new name, congenital pulmonary airway malformation and classified CCAM into five types based on the site of the defect in the tracheobronchial tree. CCAM type 0 originates from tracheal or bronchial tissue, type 1 originates from distal bronchi or proximal bronchioles, types 2 and 3 have an acinar origin; type 4 have an acinar-alveolar origin[2]

The differential diagnosis of CCAM includes other pulmonary malformations such as Congenital Lobar Emphysema, Bronchogenic cyst and Bronchopulmonary sequestration.

The mainstay of treatment of CCAM is surgical. Surgical resection of the affected lobe (lobectomy) is the standard treatment of choice, to avoid complications such as residual lesion, recurrent infections and malignant transformation. Though, in instances where the lesion is bilateral or involving multiple lobes, parenchyma-saving procedures have been tried and met with similar outcomes[3]

Anaesthetic challenges associated with management of such cystic lung lesions may be due to hemodynamic compromise at induction and the risks of positive pressure ventilation. Administration of anaesthetic agents may cause fall in tone of the thoracic muscles, abolishing the recoil and making the intrathoracic pressures positive. This may cause compression of the remaining lung parenchyma or other vital structures such as the heart. Similar effects may be produced by over distension of cystic structures by positive pressure ventilation before the chest is opened. Several strategies have been suggested to avoid over distension of the cystic lung lesions. Strategies such as gentle manual ventilation to maintain the airway pressure at 20-25 cm H2O before thoracotomy while keeping an eye on the vital signs, selective endobronchial intubation of the healthy lung till the lobectomy/ pneumonectomy is performed after which the endotracheal tube is withdrawn in the mainstem bronchus, high frequency jet ventilation, thoracic epidural catheter via the caudal insertion site, adequate analgesia and retaining spontaneous ventilation until thoracotomy, avoiding intubation until thoracotomy, a flexible ultra-thin bronchoscope into the bronchus of affected side have been suggested[4].

Nitrous oxide should be avoided as it diffuses more rapidly into cystic air spaces and cause its more rapid expansion. An arterial line should be used for beat-to-beat monitoring. A double lumen tube may be used in older children for differential lung ventilation. Awake intubation can be tried in selected cases. A multimodal approach for analgesia should be undertaken to provide complete pain relief.

Intraoperative hydration should be maintained with an isotonic fluid such as Ringer's Lactate, 10-20 ml/kg initial bolus (depending on initial hydration status) plus 4ml/kg for each ml of blood lost consequently. All measures are to be taken to keep the child warm intraoperatively.

After removal of the cyst, the lungs are to be mechanically inflated to rule out any air leaks in the tracheobronchial tree and to expand previously collapsed lung tissue. The decision regarding extubation is taken depending upon the preoperative condition, presence of infection and the intraoperative events.

The outcome of surgery depends both on the nature of the lesion as well as clinical presentation. An elective surgery is generally well tolerated with good recovery.

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