Anesthetic management of a baby with a large CCAM

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Introduction

CCAM (congenital cystic adenomatoid malformation) is a congenital anomaly of the lung resulting from abnormal fetal lung development. There is increased cell proliferation but decreased apoptosis, resulting in adenomatoid proliferation and cyst formation. The lesion is connected to the airway, but a normal intrapulmonary bronchial system is missing. Blood supply is from the pulmonary circulation. Majority of lesions are left-sided. CCAMs represent up to 25% of congenital lung anomalies and may be associated with other anomalies. CCAM classifications are based on the cyst size. CCAMs may be present before birth and may regress or increase in size. CCAMs may also be associated with other congenital defects. Associated (renal, intestinal, bony, cardiac) anomalies are present in up to 25% of patients with CCAM. CCAM malformations are based on the cyst size. Small CCAMs may remain asymptomatic and present later in life or are found incidentally. CCAM is relatively common among congenital pulmonary malformations. It can lead to respiratory distress early in life and require semi-emergent surgery. Surgical findings: left upper lobe consisted of multiple cysts with elements of emphysema. Left lower lobe was small and atelectatic. Fissure had to be created between left upper and lower lobe through the tissue of left lower lobe as there was no fissure at all. This contributed to reoccurrence of left lung air leak and resulted in prolonged hospital stay of the patient.

Case Presentation

4 week old previously healthy baby was admitted to our hospital for respiratory distress due to RSV infection. CXR demonstrated mediastinal shift due to mass occupying lesion in left chest (Fig 1). Chest CTA was performed to clarify the anomaly (Fig 2). Transthoracic ECHO showed dextrocardia by dextroposition, normal cardiac anatomy, small pericardial effusion and elevated flow velocities in SVC and IVC due to external compression. The child did not have other associated congenital defects. After RSV resolution the patient was unable to leave the hospital due to respiratory distress and feeding difficulties. At 7 weeks of age baby presented to OR for left thoracotomy with left upper lobectomy. Baby arrived to OR tachypneic, on high flow nasal cannula, with markedly decreased breath sounds bilaterally. Body Wt 4.8 kg. Anesthetic management goals were to prevent worsening of left lung hypoplasia/pulmonary hypertension and mediastinal shift and to isolate left lung. We did induction as a combination of a low dose of IV Midazolam to facilitate mask acceptance and inhalational Sevoflurane with preserved spontaneous respirations. Additional IV access was obtained, peripheral A-line placed with patient breathing spontaneously. Then we relaxed the baby with a non-depolarizing neuromuscular blocker, gently took over ventilation, performed direct laryngoscopy and placed 5 Fr Arndt endobronchial blocker (BB) first, then regular ETT next to the blocker. We positioned BB in the left mainstem bronchus under guidance of fiberoptic bronchoscopy. After the patient was positioned in right lateral decubitus, we reconfirmed the position of the BB and inflated the cuff. We did not suction the lumen of BB in order to deflate left lung but instead relied on absorption atelectasis as the patient was ventilated with 100% O2 up to this point. Surgical findings: left upper lobe consisted of multiple cysts with elements of emphysema. Left lower lobe was small and atelectatic. Fissure had to be created between left upper and lower lobe through the tissue of left lower lobe as there was no fissure at all. This contributed to reoccurrence of left lung air leak and resulted in prolonged hospital stay of the patient.

Discussion

CCAM is relatively common among congenital pulmonary malformations. It can lead to respiratory distress early in life and require semi-emergent surgery. For thoracic cases like we presented above single lung ventilation (SLV) helps with surgical approach, protects healthy ventilated lung and maximizes V/Q match. Routine adult approach to SLV is not suited for small pediatric patients. Way of lung isolation with positioning of regular ETT in one of the mainstem bronchi is not ideal but still used commonly, particularly in emergency situations. Traditional placement of BB through the lumen of ETT is often not an option due to small internal diameter of the endotracheal tube. Technique of BB placement extraluminally from ETT works very well for neonates, babies and small toddlers.

References