ANESTHESIA EXPERIENCE IN A CHILD AFFECTED BY CONGENITAL CYSTIC ADENOMATOID MALFORMATION AND RETINOPATHY OF PREMATURITY FOR EYE LASER PROCEDURE

- A Case Report -

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Abstract

Congenital Cystic Adenomatoid Malformation or bronchopulmonary displasia (congenital lung cysts disease) is a rare condition. Intrauterine death affect severe cases, those infants surviving till birth lead a chronic course of respiratory illness amenable to corrective thoracic surgery or other corrective measures for non pulmonary conditions. This report describes an experience with one girl that needed anesthesia for congenital retinopathy's laser treatment.

Key Words: Congenital Cystic Adenomatoid Malformation bronchopulmonary displasia, Congenital retinopathy, Anesthesia: Sevoflurane, Ketamine.

Introduction

Congenital Cystic Adenomatoid Malformation (CCAM) was reviewed recently[1]. It was first described in 1949[2]. It is a rare congenital malformation of the lung representing 25% of congenital lung malformations and 95% of congenital lung lesions[2,3]. This lesion occurs more often in males (1.8:1), and is primarily unilateral, but may occur bilaterally[4].

Associated anomalies are rare. Since the technological advancement of ultrasound examination, CCAM has been increasingly diagnosed on routine prenatal examinations. Some CCAM lesions present only at birth with respiratory distress symptoms but are confirmed by an abnormal chest radiograph or a more definitive computed tomography scan. However, there are CCAM lesions that are not identified on routine ultrasound examination, and present without symptoms at birth. These lesions may not be identified until later in life.

When diagnosed perinatally with or without symptoms, most CCAM lesions are manageable if proper assessment, diagnosis, and interventions took place. So, infants with the more severe cases of CCAM are expected to survive[1-4].

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Case Report

A 27 weeks gravid premature girl, weighing 1000 g at birth, delivered to G7 P6 +1 mother who had eclampsia at 72 weeks of gestation. At the age of 4 weeks the girl was intubated and ventilated for 26 days due to respiratory impairment. Later she was diagnosed as a case of: (1) retinopathy of prematurity. (2) Congenital bronchopulmonary displasia. No other medical diseases were documented. She was referred from peripheral hospital to ophthalmic unit at King Fahad Medical City for laser surgery to control congenital retinopathy. The girl was admitted to the NICU, and investigated and found to have right lung cyst. The treating physician asked an anesthesia consultation for laser surgery.

Upon examination in her incubator, the patient was breathing spontaneously, active and on oxygen supplements 4 L per minute via mask. Auscultation of the chest did not elicit abnormality. Vital signs; HR:170 per minute. Chest was quite with breathing sounds. Blood pressure: 85/40 mmHg. RR-35 m. She was on oral feeding.

Laboratory investigations show: urea:1.4 mmo L^{-1}. creatinine 19.0 µmol.L^{-1}. K: 5.5 mEq.L^{-1} Na: 141 mEq.L^{-1} Hb: 11 g.dL^{-1} platelets:497 x10.e9.L^{-1} Coagulation profile PT:12.4, APTT:40.1.

The pulmonologist who cared for the patient advised that the lung cysts are small in size. A pediatric surgical consultation for management of the lung cyst obtained, and advised that there is no need for urgent surgery, but advised that if pneumothorax develops during laser surgery then immediate intervention should take place.

Anesthetic management (18-8-2007):

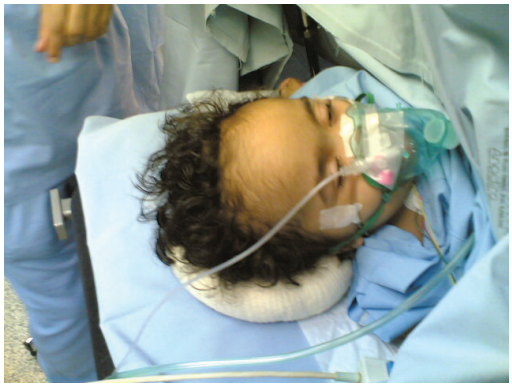
Inhalational induction was done using sevoflurane. Anesthesiologist took long induction before laryngoscopy and tried intubation but found the condition was not proper for intubation, the girl was "fighting", so since she was on sevoflurane inhalation supplemental ketamine was given intravenously in dose 0.5 mg kg^{-1}. Atropine-0.1mg given as well then suxamethonium-5 mg was injected, so the trachea was intubated successfully. The respiration resumed with the tube in place.

Anesthesia maintained on sevoflurane 2% in air: oxygen mixture.

Respiration was spontaneous with occasional ventilatory manual support.

Finally at the one hour procedure, the girl was extubated after she recovered completely and was sent back to NICU on 4L O2 with SpO_2 100%. Later on, she was discharged to primary hospital on 28.8.2007 in good condition.

Fig 1
The patient on operating table before procedure and getting supplementary oxygen



Discussion

The survival rate of extremely preterm infants improved over the last years as a result of a better prenatal and neonatal care mainly due to a greater use of antenatal steroids, appropriate management in the delivery room and in the initial care, surfactant therapy, and better modalities of assisted ventilation. However, this improvement in survival has not been associated with an equal reduction in morbidity. In fact, the frequency of bronchopulmonary displasia, sepsis, poor growth, and neurological disorders in the future may have increased.

Congenital cystic adenomatoid malformation (CCAM).

Pulmonary sequestration, congenital lobar emphysema (CLE) and bronchogenic cysts are all congenital malformations of the lung that present in imaging studies as abnormal air, air/fluid, or fluid-filled cysts. This is a rare developmental, non-hereditary, hamartomatous abnormality of the lung with adenomatoid proliferation of cysts resembling bronchioles. It is usually unilateral with involvement of a single lobe. In case series report from Japan[5]

it stated that: 11 patients under 1 year old showed respiratory distress with mediastinal shift but no episodes of infection. In contrast, 13 of the 19 patients over 1 year old had symptoms of recurrent infection without respiratory distress.

Five patients over 1 year old were entirely asymtomatic from birth.

In patients under 1 year old, cystic lesions were discovered due to manifestation of respiratory distress; and in patients over 1 year old signs of infection were the most important clinical features.

Early recognition of these relatively rare congenital cystic lung lesions would lead to the immediate, proper surgical intervention.

The features of congenital lung cystic malformations are: - Absence of bronchial cartilagen, absence of bronchial tubular glands and presence of tall columnar mucenous epithelium, overproduction of luminal bronchiolar stricture, in the form of cysts of various sizes.

To the naked eye, the affected lobe looks dark, red, solid and bulky with surface bosses corresponding to larger cysts.

CCAM with severe respiratory dysfunction from birth was reported and the anesthetic course was dependent on sevoflurane inhalation and spontaneous respiration. It was 5-day-old-boy with CCAM underwent removal of a large cyst which was present at lower lobe of right lung. Anesthesia was induced slowly and maintained with oxygen and sevoflurane. Severe airway obstruction occurred transiently by the secretion from the lung cyst. Thereafter, the surgery was completed safely and his postoperative course was uneventful[5].

A recent report describing a left pneumonectomy performed on a six week-old female infant suffering from respiratory distress related to cystic changes affecting the entire left lung. Anesthesia was induced with sevoflurane in oxygen and spontaneous ventilation was maintained until intubation of the right main bronchus was secured. the postoperative course was uneventful. The pathological diagnosis was pulmonary interstitial emphysema. In the reported case we are presenting inhalation of sevoflurane took long and the patient did not relax enough for intubation though high concentration (6%) sevoflurane and three minutes has passed. Ketamine supplement and suxamethonium were used to secure the airway[6].

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