CASE REPORT

Congenital Cystic Adenomatoid Malformation Type I in a Newborn with Sepsis: A Case Report

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ABSTRACT

Congenital cystic adenomatoid malformation (CCAM) is a rare bronchopulmonary tissue proliferation disease. Although CCAM type I (CCAM I) has a good prognosis, respiratory distress and sepsis comorbid can worsen surgical outcomes. This case aims to explain the intensive care of a newborn with respiratory distress and sepsis before lobectomy. In this case, a female newborn with severe respiratory distress due to CCAM I faced two dilemmas of sepsis comorbid and early lobectomy consideration. We decided to increase the sensitive antibiotics dose of Ampicillin-Sulbactam to 100 mg/kg/12hours (standard dose: 50 mg/kg/12hours) and Amoxicillin to 90 mg/kg/day (standard dose: 45 mg/kg/12hours). This decision successfully showed negative blood culture before surgical consideration. A left inferior lobe lobectomy was then performed, despite the newborn respiratory function had not been well improved. Eight days after lobectomy, the left lung developed well without any complications. Hence, immediate lobectomy is recommended to restore lung function and prevent recurrent infections and pneumothorax.

Keywords: Congenital cystic adenomatoid malformation, Cystic adenomatoid malformation of lung, Newborn, Sepsis

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INTRODUCTION

Congenital cystic adenomatoid malformation (CCAM) is a rare birth defect consisting of bronchopulmonary cystic lesions. This occurs as multilobar or bilateral, but the commonest type is located in the left lung(1). The estimated incidence is approximately 1:25,000 live births with male predominance. CCAM is often misdiagnosed as pneumonia due to respiratory distress, a recurrent lung infection sign(2). It can cause polyhydramnios or hydrops fetalis, and pneumothorax.

We reported a female newborn that suffered respiratory distress syndrome and sepsis. CCAM type I (CCAM I) diagnosis was obtained from the x-ray and from computerized tomography (CT) scan. It commonly has a good prognosis, but respiratory distress and sepsis are certainly clinical challenges in CCAM I. Early lobectomy was indicated because of continued pulmonary function deterioration. Meanwhile, sepsis tended to worsen the operative outcomes.

After making the clinical decision to add sensitive antibiotics, the patient no longer had sepsis. Consequently, an early lobectomy was performed to restore normal pulmonary function and prevent further complications. This report aims to explain CCAM I pre-operative, operative, and post-operative care in a newborn with respiratory distress and sepsis.

CASE REPORT

The female baby was born at 38 weeks of gestation by caesarian section (C-section) due to premature rupture of membrane 20 hours before delivery. The APGAR score was 7-8-9, and 3.4kg of birth weight. Physical examination showed Downe's score 4 (grunting, flaring, mild left intercostal retraction, respiratory rate 70x/minute), SpO2 91-93%, temperature 37.1 C. The newborn was treated in the neonatal intensive care unit (NICU) with respiratory distress and sepsis symptoms.

Treatment included 0.5lpm nasal oxygen cannula, 10% Dextrose infusion, 1 mg intramuscular Vitamin K injection and 0.5 ml Hepatitis B immunization, and Ampicillin-Sulbactam standard dose 50 mg/kg/12hours,Amoxicillin 45 mg/kg/day,warmth care maintenance, incubators care, and umbilical cord care. Moreover, fluid and calories intake were targeted to 80 ml/kg/day and 100 kcal/kg/day (breastfeeding8 x 10-20 ml/day). Laboratory studies included septic workup, blood sugar, and babygram. These showed 23,500/µL leukocyte (normal: 4,500-11,000/µL), Enterococcus sp. on blood culture,

and 50 mg/dL glucose level (normal: 40-50 mg/dL). The initial babygram (Fig. 1) showed >2 cm thin-walled multiple cysts in the left lung accompanied by right side mediastinum and heart deviation. The initial diagnoses were CCAM, congenital pneumonia, pneumatocele, and bronchogenic cyst.

The newborn was further treated in the NICU with Ampicillin-Sulbactam and Amoxicillin as sensitive antibiotics while monitoring general condition every hour and fluid balance every 8 hours (Table I). The chest contrast CT scan was performed after a common condition assessment was stable on the fifth day of hospitalization. This showed multiple cystic lesions having several features, partly filled with water with an air-fluid level. The largest cyst was 5 cm in the left inferior lobe, which showed contrast enhancement at its septation. The lesions caused left lung collapse and right mediastinum deviation. The CT scan interpretation was supported by CCAM I diagnosis.

The surgery was carried out for 1 hour and 30 minutes on May 1, 2018. A left posterolateral thoracotomy incision was performed, and the entire inferior lobe was covered with bullae. The bullae covering the operational field were broken, and they looked like bubbles. A left inferior lobectomy was performed. Afterwards, chest tube drainage, gastric tube, and peripherally inserted central catheter were inserted.

In the NICU, a pressure control mechanical ventilator was installed with 5 cmH2O positive end-expiratory pressure, 11 cmH2O peak inspiratory pressure, 40x/minute respiratory rate, 60% FiO2, and inspiration/expiration ratio 1:1.5. Additional therapies were injection of 12.5 mg/day Gentamicin, 30 mg/8 hours Paracetamol alternating with 30 mg/8 hours Metamizole, 75 µg Fentanyl plus Normal Saline up to 12 ml. Blood gas analysis showed pH 7.42 (normal: 7.35-7.45), base excess -1.9 (normal: -2 to +2), pCO2 3.4 (normal: 4.5-6.0), HCO3 22.1 (normal: 22-26), SpO2 100%, and lactate 1.7. Meanwhile, the FiO2 was planned to decrease to 35%. The third babygram showed left inferior lobe post-lobectomy pneumothorax, lucent avascular area and visceral pleural line in left inferior hemithorax, and right-side heart deviation (Fig. 1).

On May 7, 2018, the clinical condition improved with Downe's score 0. Moreover, sepsis and respiratory distress were absent. The left lung was well developed in the last babygram (Fig. 1). The CCAM I tissue size was 9 x 5 x 2 cm. Its histopathology result showed cystic chambers with ciliated pseudocolumnar epithelium and glandular structure. These features established the CCAM I diagnosis (Fig. 2). The patient was discharged on May 9, 2018, after the general medical conditions

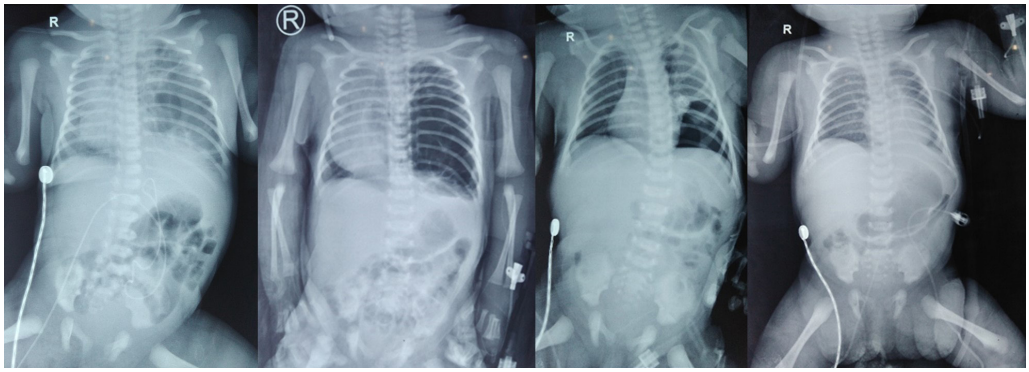

Figure 1: Left to right: the first pre-operative babygram, second pre-operative babygram, third post-operative babygram, and fourth post-operative babygram consecutively

Table I: Important medical progress of the patient before the surgical procedure

Dates	Clinical findings	Clinical Decision
April 20, 2018	C-section due to PROM indication. Downe's score 4, respiratory distress and sepsis symptoms. Leukocyte 23,500/µL. *Enterococcus sp.* on blood culture. Multiple cystic appearances on the first pre-operative babygram.	Treatment and routine monitoring in NICU. Oxygen therapy, Dextrose, Vitamin K, Hepatitis B immunization, Ampicillin-Sulbactam. Maintenance of warmth and incubator care. Fluid therapy and calorie intake management.
April 21-23, 2018	Downe's score 5, respiratory distress, and sepsis symptom.	Treatment and monitoring in NICU.
April 24, 2018	Downe's score 5, respiratory distress, and sepsis symptoms. CT-scan confirmed CCAM I in the left inferior lobe.	Treatment and monitoring in NICU. Lobectomy risk and benefit assessment.
April 25-29, 2018	Downe's score 5, respiratory distress and sepsis symptom.	Lobectomy was planned to restore normal respiratory function. The Ampicillin-Sulbactam and Amoxicillin dose was increased to 100 mg/kg/12 hours and 90 mg/kg/day consecutively.
April 30, 2018	Downe's score 3, respiratory distress. No organisms were grown in blood culture. Multiple cystic appearances on the second pre-operative babygram.	Treatment and monitoring in NICU (one day prior to lobectomy).

CCAM I, congenital cystic adenomatoid malformation type I; C-section, caesarian section; CT, computed tomography; NICU, neonatal intensive care unit; PROM: premature rupture of membrane

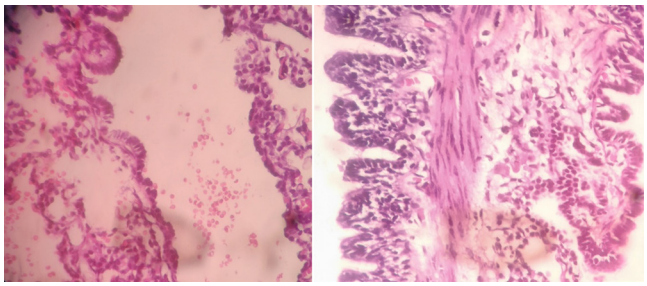

Figure 2: The histopathology examination of the post-surgical tissue sample

were stable.

DISCUSSION

CCAM prevents normal healthy lung tissue development, and CCAM I is the commonest type of CCAM. The distal bronchial and proximal bronchioles are the frequent locations of the CCAM lesions. CCAM can cause breathing difficulties, recurrent infections, and pneumothorax complications(1). CCAM is also associated with lung cancer, such as bronchoalveolar carcinoma or pleuropulmonary blastoma. In some cases, CCAM appears asymptomatic.

In this case, early surgery was considered because of the respiratory distress indication. Lobectomy needs to be performed before ten months of age for the optimal outcome (3). The study also recommended that blood culture have to be negative by adding sensitive antibiotic doses (4). Therefore, the pre-operative care must ensure haemodynamic and pulmonary function improvement before the lobectomy.

The CCAM resection needs to be clear from the remnant to facilitate normal lung development (3). Moreover, post-operative mechanical ventilation must be monitored continuously because newborns' lungs were very much susceptible to mechanical injury and oxygen(5). The limitation of this case report is the inadequate evidence-based medicine references of neonatal sepsis care before major surgery.

CONCLUSION

Early CCAM lobectomy needs to be considered to restore optimal lung function. However, sepsis comorbid require intensive care and more dose of sensitive antibiotics before lobectomy. CCAM is acknowledged as a rare disease. More evidence-based medicine of CCAM cases with respiratory distress and sepsis is necessary before lobectomy.

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