Congenital Cystic Adenomatoid Malformations Type 1: Case Report

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Abstract

BACKGROUND: Congenital cystic adenomatoid malformation (CCAM) is a group of multicystic and non-cystic masses due to abnormal lung development. The incidence ranges from 1: 10,000 to 1: 35,000 pregnancies.

CASE REPORT: We report a case of a 3-day male baby presented with worsening respiratory distress, X-ray and chest CT were suggestive of CCAM, and sepsis.

CONCLUSION: The current case adds to the collective clinical and radiological knowledge of this rare congenital lung disorder.

Introduction

Congenital cystic adenomatoid malformation (CCAM) is a congenital abnormality of lung development caused by an overgrowth of abnormal lung tissue that forms cysts, affecting ≥1 lung lobes [1]. CCAM originates from embryological abnormalities of terminal bronchus that occur before 35 weeks of gestation. Nearly 25% of CCAM cases are accompanied by other congenital abnormalities [2]. About 7% of cases show symptoms of severe respiratory distress and 96% can be born alive [3]. Patients with small lesions are usually asymptomatic until they reach middle age when episodes of recurrent airway infections or chest pain occur [4]. CCAM can be diagnosed antenatally by ultrasonography and postnatally by chest CT-scan or MRI [2].

Case Report

A 3-day male baby presented with dyspnea was admitted to the NICU. The baby was delivered by Cesarean section at full term, spontaneously, amniotic fluid was clear and odorless. The APGAR scores were 7 and 9 at one and five minutes. The mother’s prenatal history was unremarkable, but prenatal ultrasound showed the presence of fluid in the fetal lung. The baby showed respiratory distress with decreased breath sound at the right hemithorax. Based on clinical features and laboratory investigations, he was diagnosed with unproven sepsis with increased level of procalcitonin and CRP [5]. The chest X-ray showed a thick-walled multicavity at the left lung and the heart appeared to be pushed to the right (Figure 1). The chest CT showed multi-septate cysts, thin-walled, with the largest size of 3.0 × 4.4 × 3.8 cm almost over the left lung lobes (Figure 2). The baby was on bubble n-CPAP, intravenous fluids, ampicillin, and gentamicin. On the 5th day, the antibiotics were replaced with ceftazidime and amikacin because of no clinical improvement. On the 13th day, the respiratory distress worsened, he was intubated and put on a ventilator, and antibiotics were replaced with meropenem. On the 19th day, he fell in shock. Blood gas analysis revealed a respiratory acidosis. Fluid resuscitation and inotropic medication did not improve the condition, patient passed away on the 20th day of treatment.
Discussion

Stocker et al. divided CCAM into five subtypes; type 0 (rare), consists of microcytic cells throughout the lung and has poor prognosis. Type I accounts for 50–70%; consists of single or multiple large cysts (>2 cm), filled with air or fluid and often cause mediastinal impulse, and has good prognosis. Type II (15–30%), consists of several small cysts (<2 cm), usually associated with other congenital abnormalities, and has poor prognosis. Type III (5–10%), is a mixture of microcytic cells and dense tissue, also has poor prognosis. Type IV originates from distal acini has peripheral large cystic type (>10 cm) [4], [6], [7], [8], [9].

The clinical symptoms depend on the magnitude, degree of expansion, and location of the cyst. Inspiration will trap air inside cystic mass, leading to high pressure in the chest cavity resulting in mediastinal shifting, contralateral atelectasis, pulmonary and diaphragm compression [10], [11].

Total lobectomy is the only treatment for CCAM to prevent sequelae and recurrences. Lobectomy can be performed at 3–6 months old in symptomatic patients and 1–3 months old in asymptomatic patients. Without surgery, the symptomatic CCAM’s mortality rate reaches 100% [7], [10], [11].

CCAM type I has a good prognosis with a total surgical resection and if not accompanied by complications (i.e cardiac shift or septic shock). The current case is type 1 but accompanied with cardiac shift and septic shock.

Based on the CCAM classification proposed by Stocker et al. in 1977, this patient was diagnosed as CCAM type 1. We did not perform lobectomy because of the large number of cysts in almost one left lobe. Respiratory failure that occurs in this patient is of a type of hypercapnic respiratory failure due to airway obstruction (water trapping). CO\textsubscript{2} cannot be released at the time of expiration, it causes the lungs to hyperinflate and increasing the arterial PCO\textsubscript{2} (PaCO\textsubscript{2}) and decreasing pH. As a result of continued excessive production of CO\textsubscript{2}, it will be a failure of the compensation mechanism to maintain gas exchange or oxygen flow is sufficient.

Conclusions

Respiratory distress is the most common symptom in people with CCAM. This can occur due to compression of the normal lung and lung tissue hypoplasia around the cyst. Larger and multiple cysts can cause mediastinal shifting to the contralateral which will disrupt venous return and cardiac output so that it can cause cardiorespiratory distress and respiratory failure. The current case adds to the collective clinical and radiological knowledge of this rare congenital lung disorder.
Declarations

This study has been approved by the ethics committee of Hasanuddin University/DR Wahidin Sudirohusodo Hospital. The patient’s consent to participate and publication has been obtained.

Availability of data and material

The datasets generated during and/or analyzed during the current study are available from the corresponding author on reasonable request.

References