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Case Report

Congenital cystic adenomatoid malformation: a rare serial case report

Dedy C. Hariyono[1]*, Muhammad Kartika[1], Prima K. Hayuningrat[2], Darmawan Ismail[2]

[1]Surgery Resident of Sebelas Maret University, Surakarta, Indonesia
[2]Department of Surgery, Cardiothoracic Division, Dr. Moewardi General Hospital, Surakarta, Indonesia

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***Correspondence:**
Dr. Dedy C. Hariyono,
E-mail: dedychandramd@gmail.com

ABSTRACT

Congenital cystic adenomatoid malformation (CCAM) is a rare congenital lung malformation which a part of the lung becomes polycystic. CCAM accounts for 25% of congenital lung malformations and 95% of lung lesions. Case 1 was a 5-month-old female infant who was diagnosed with pneumothorax, with multiple cysts in the right lung, using chest computed tomography (CT). Thoracotomy lung resection was performed. Case 2 was one-day-old newborn infant who had respiratory distress with Downe score 3. Multiple cystic lesions with septations in left lung was observed on chest CT. Lobectomy inferior lobes of left lung was performed. Both patients were diagnosed as CCAM type 1 pathologically. CCAM can be detected in the gestation by ultrasonography or after delivery through the appearance of respiratory distress signs.

Keywords: CCAM, Congenital, Lung malformation

INTRODUCTION

Congenital cystic adenomatoid malformation (CCAM), also called congenital pulmonary airway malformation (CPAM), is an uncommon development abnormality of the lung that accounts for 25% of all congenital lung malformations and 95% of congenital lung lesions.[1] CCAM is considered as a hamartomatous lung anomaly, characterized by a formation of many cysts in the lung because of adenomatous hyperplasia in bronchiolar epithelium.[2] Firstly in 1977, CCAM is classified into three subtypes, and into five types with a new name as CPAM by Stocker in 2002. Both classification is based on histologic patterns.[3]

The presence of respiratory distress is a helpful indicator to identify the CCAM in neonatus, although it can also be found in older children or adults with recurrent infection.[4] A routine prenatal exam might improve the chances of CCAM detection. Abnormal chest radiographs or more definitive computer tomography scans can confirm CCAM lesions that are exclusively present at birth and cause respiratory problems.[5] We present two cases of CCAM, which was cured by open resection surgery.

CASE SERIES

Case 1

A 5-month-old female infant referred to the hospital from another facility with complaints of pneumothorax. The past medical history of patient shows that one week ago, the patient experienced shortness of breath then admitted to the pediatric intensive care unit (PICU) for further evaluation. In PICU, the patient was diagnosed with pneumothorax based on X-ray chest and performed water sealed drainage (WSD). WSD was kept *in situ* even when the patient was referred. She had no documented history of stay in hospital previously.

X-ray chest PA shows opacity in left center zone, increased vascularity of left lung, and right hyperlucent lung with WSD on it (Figure 1). Chest computed tomography (CT) revealed multiple large sized cystic lesions with septations in superior-median lobes and inferior lobes of right lung, also tracheal and cardiac structure deviation. CT chest was described by radiologist as CCAM Type 1 (Figure 2). Patient was referred to

cardiothoracic surgical side and was further managed there with thoracotomy lung reresection under general anesthesia and removal of cystic lesion (Figure 3 and 4).

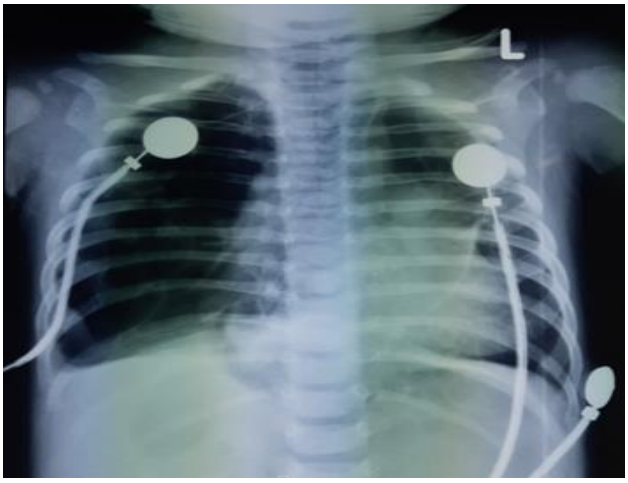

Figure 1: Posteroranterior (PA) chest X-ray before surgery.

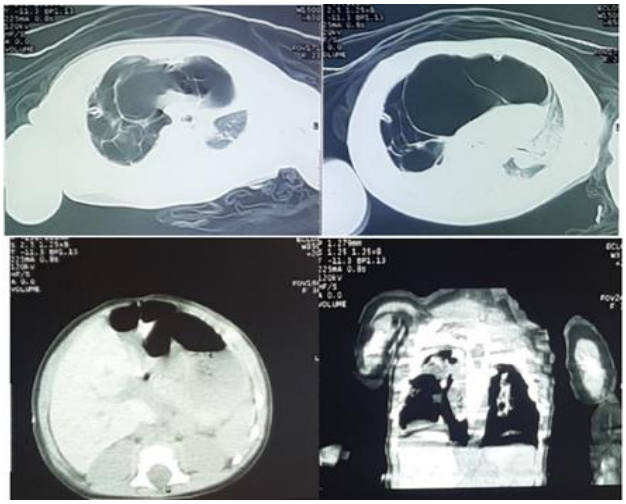

Figure 2: Chest CT-scan.

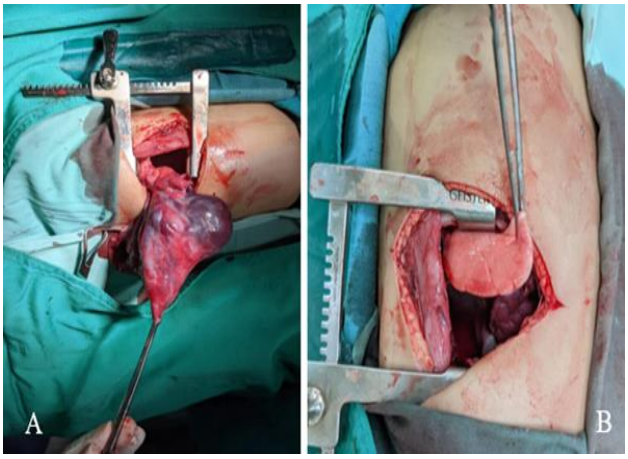

Figure 3 (A and B): Before lung resection and after lung resection.

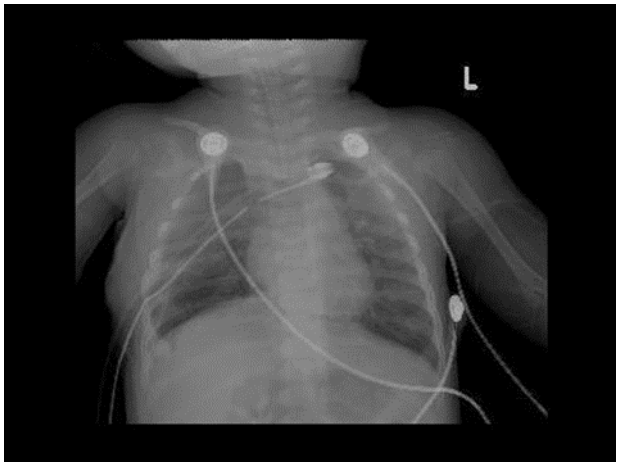

Figure 4: PA chest x-ray after surgery.

Case 2

One-day-old newborn infant, female, was born to G2P1 mother. Sectio caesaria (SC) aterm labor with indication mayor congenital malformation (CCAM) occurred at 38 weeks of gestation. The APGAR scores were 7-8-9. The birth weight was 3400 grams. Patient's mother had a history of premature rupture of membrane (PROM) in 20 hours before come to the hospital and previous SC in 20 months ago. A routine prenatal ultrasound, revealed cystic lesions within the left lung of the baby. The patient was referred to fetomaternal unit and ultrasound was performed and revealed echogenic cyst mass with lobes within left hemithorax. The mass wasn't appeared to receive any arterial blood supply and measured 4×3 cm (Figure 5).

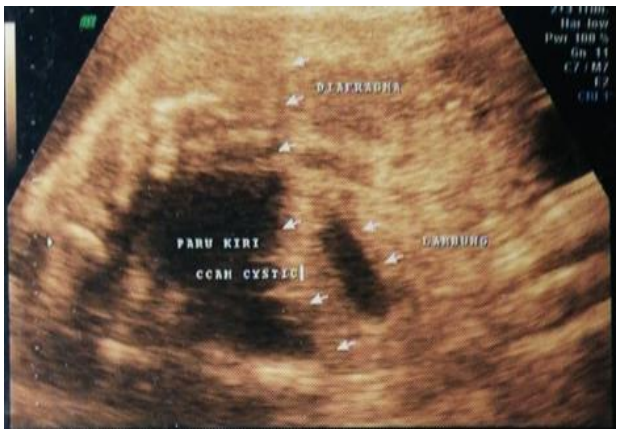

Figure 5: Fetomaternal USG.

The infant was admitted to surgical emergency unit for further evaluation. On examination, baby had respiratory distress. Inspection resulted asymmetry of anterior and posterior chest, decreased left chest expansion, and mild intercostal retractions. Percussion note was dull in lower zone of left lung and resonant in right lung. The Downe score was 3 based on increased respiratory rate, decreased breath sound, and mild restrictions. The diagnose was

mild shortness of breath et causa suspect CCAM type-1. Then patient was admitted to Neonatal Intensive Care Unit (NICU). Babygram was done and showed multiple cystic lesions measured >2 cm with thin walls in left lung suggested of CCAM type 1. There was a mediastinal shift of the heart to the right hemithorax (Figure 6). A computed tomography scan of chest revealed multiple cystic lesions with septations in left lung, some of the cysts were filled as air fluid level (+). The largest individual cyst measured 5 cm. The lesions induced left lung collapse and caused mediastinal shift appearance (Figure 7).

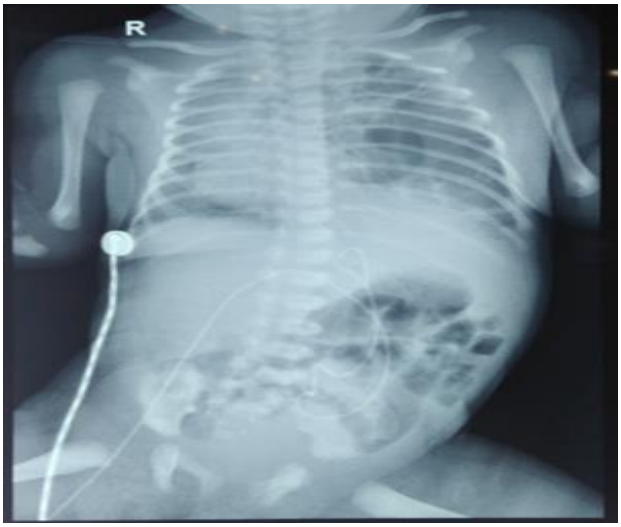

Figure 6: Babygram was suggestive of CCAM type 1. Gastric tube and umbilical catheter were *in situ.*

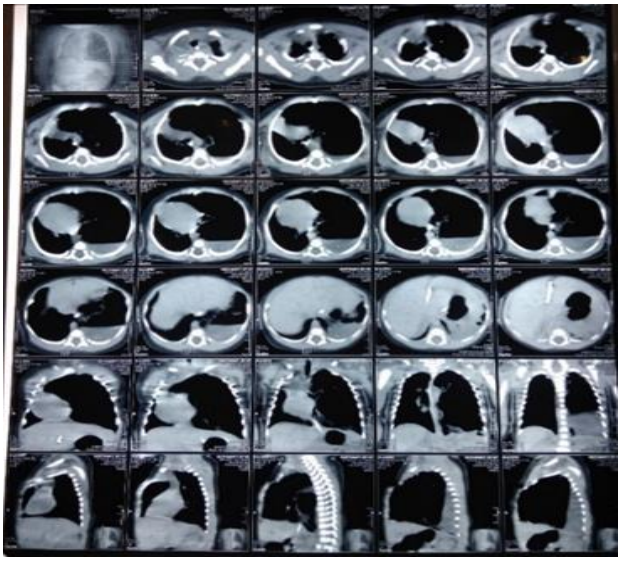

Figure 7: Chest CT-scan.

The patient underwent surgery for 90 minutes. Lobectomy inferior lobes of left lung was done under general anesthesia. Postoperatively, the infant was treated in NICU for several days. The pathology from the fetal excision demonstrated pulmonary tissue with multi size cystic lesions. The cyst wall lined with ciliated pseudo-scollumnar epithelium, CCAM-like changes (Figure 8). Repeat X-ray chest after surgery was different to previous X-rays, showed progression of left lung (Figure 9) and patient went home.

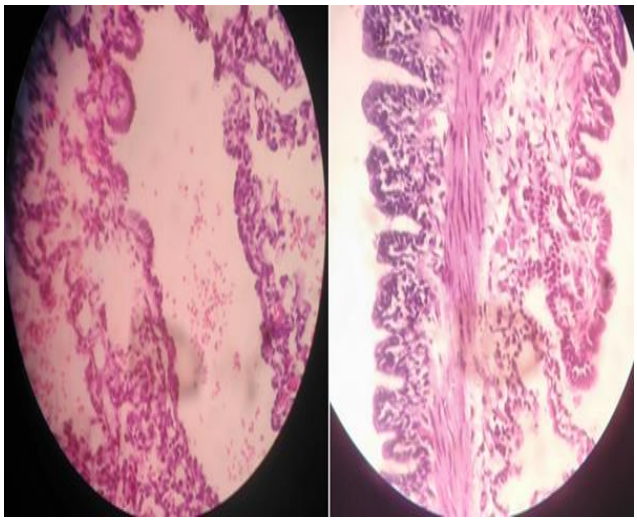

Figure 8: Histopathology of left lung lobe.

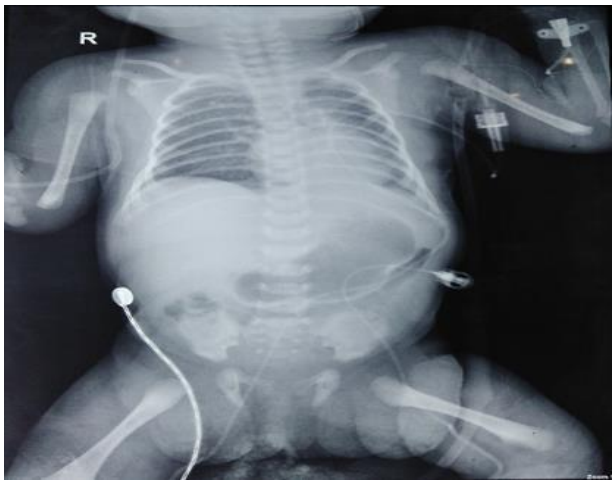

Figure 9: Chest X-ray after surgery.

DISCUSSION

CCAM is a rare developmental lung abnormality that occurs early in fetal lung development, between the fifth and eighth weeks of gestation. In the majority of cases, CCAM presents in one lung only.[5] The terminal bronchiole abnormally proliferates, inhibiting alveolar development and generating intercommunicating cysts. Reduced pulmonary growth causes pulmonary hypoplasia, which can result in postnatal respiratory distress, mediastinal shift, pleural effusion, and spontaneous pneumothorax.[6] Approximately 60% of CCAM cases are associated with other findings such as cardiac anomalies, renal agenesis/dysgenesis, gastrointestinal atresia, and skeletal anomalies.[7] Type 1 CCAM is the most prevalent, accounting for 50-70

percent of cases and originating in the distal bronchus or proximal bronchiole. Because these CCAMs may be big, they may have a substantial mass effect, which might cause hydrops.[8]

Cases are generally identified prenatally by regular ultrasound screening. The majority of postnatally identified cases appear during the neonatal period. CCAM presents on ultrasound as a cystic, solid or hyperechoic intrathoracic, space-occupying mass.[6] The cyst growing and squeezing its surrounding tissues causes acute respiratory distress, which is the most prevalent manner of presentation.[4] CCAM can be detected in the gestation by ultrasonography or after delivery through the appearance of respiratory distress signs.[5] The size and type of the tumor, as well as the presence of mediastinal shift and hydropic alteration on ultrasonography, have all been linked to a bad prognosis.[9] Postnatally, computed tomography (CT) is the gold standard for diagnosis in pre-operative because of its excellent spatial resolution and fast acquisition periods. However, due to overlapping characteristics and uneven terminology use, diagnosis can be difficult.[10]

The therapy of CCAM is determined on the size of the tumor and if it is symptomatic.[6] CCAM is treated with lobectomy. Depending on the clinical situation, this may have to be done as an emergency. If the patient is asymptomatic and the cyst is resolving, treatment might be postponed.[5] Most literatures suggest resection of all antenatally detected CCAMs, however surgery can be postponed for several months after delivery.[7] Pulmonary resection in infancy is linked with low morbidity and death rates and may avoid late problems such as infection and hidden malignant transformation. Thoracotomy and delivery of the hyperinflated lobe into the wound relieves the ventilatory and circulation issues immediately. Generally, lobectomy is required, however segmental resection is sometimes possible.[4]

CONCLUSION

CCAM is a rare case. Some babies with CCAM born with respiratory distress so early detection and treatment could remove the risk of further complications. A routine prenatal ultrasound examination is recommended to detect such rare lesions. Open surgery with pulmonary resection gives a good result in CCAM patients.

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