CASE REPORT

Congenital Cystic Adenomatoid Malformation of the Lung (CCAM): Report of 2 Atypical Cases

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ABSTRACT Congenital cystic adenomatoid malformation (CCAM) presents mainly in neonates, rarely in children beyond infancy, and has been reported in adults. Two females (aged 26 months and 34 days) who had CCAM in the right and left lower lobe, respectively, are reported. One of them presented with recurrent respiratory infection and the other as newborn with respiratory distress secondary to mediastinal displacement and pulmonary compression as a result of expanding cystic lesions. Both cases had different lesions from typical neonatal cases reported in the literature. Single cyst was shown in the first case, but multiple cysts in other. The young child survived, but the baby died. All lesions had lining varied from pseudostratified columnar to cuboidal epithelium. Cartilage plates was found in the second case but not in the first. The absence of inflammation is typical in neonates' lesions, by contrast, all of our two patients had clinical and pathologic evidence of chronic inflammation. CCAM may be clinically silent in infancy and may present as pneumonia associated with cystic lesion on chest X-ray in childhood or later in life. To support the diagnosis, CT scanning is needed. **[Paediatr Indones 1999; 39:229-236]**

Introduction

Congenital cystic adenomatoid malformation (CCAM) is a rare congenital lesion of the lung. It was first described by Ch'in and Tang in 1949 as an original/typical case.[1] Thereafter several cases were reported in the literature and some of them were described as atypical cases.[2] The distinction between congenital and acquired lung lesion

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is a problem. The various report in literature are based on either clinical and histologic criteria. In practice a histologic distinction between congenital and post-infectious acquired occasionally cannot be made. A better distinguishing sign is clinical assessment, namely the presence or absence of current or chronic pulmonary infections.[3] The most difficult differential diagnosis in order infants or children with recurrent infections is between CCAM and lung abscess or necrotizing pneumonia, therefore a CT scan may be needed to resolve uncertain cases.[2] These are only 2 cases which was found in our division during almost the last 15 years and reported here. One of them was found in neonatal period and other in a young child past the age of infancy.

Case 1

D, a-26 month-old malnourished girl, was admitted to hospital after previous recurrent pulmonary infections, referred for accidentally finding of the right lung mass on chest X-ray, diagnosed elsewhere. She has been on recurrent pulmonary infections since almost 6 months but never had respiratory distress. Physical examination supporting with chest X-ray and pleural puncture showed encapsulated empyema or an infected lung cyst (Figure 1). CT scan showed a thick-walled infected right lung cyst (Figure 2). Antibiotic injection was administered but chest tube drainage was not performed. There was still no change on chest X-rays after given an antibiotic. A right thoracotomy was planned and an uneventful right lower lobectomy was performed. With removal of the malformation, lung expansion was shown on chest X-ray and her general condition was good.

The malformed lower lobe weight 75 gram, measuring 9x7x5 cm. The cyst was 6 cm in diameter, located in inferior lobe filled with red liquid, no systemic arterial branch was seen. Medial and superior lobe was normal. Histology (Dr. L Silitonga): cyst wall of ciliary cuboidal epithelium with sub-epithelium focally thrown into papillary fold to the lumen, but no cartilage plates was seen. On cut sections it was lined by stratified cuboidal epithelium; an area of acute and chronic suppurative inflammation were also seen (Figure 3).

Case 2

ASA, a-34 day-old girl, was admitted for persisting dyspnea after receiving medical care for 6 days in other hospital which produced no improvement and then transferred. She was born in good clinical condition from an uneventful pregnancy, 2800 gram birth weight and no respiratory distress after delivery. Dyspnea was first shown when she was 26 days old. The physical examination showed an acutely ill, febrile baby, 3200 gram body weight, tachypnoe, retraction but no cyanosis. No specific

findings were found on percussion and auscultation except crackles on both lung fields. The chest x-ray showed multiple cystic lesion on left hemithorax, tracheal and mediastinal shift to the right (Figure 4). The peripheral white blood cell count on admission was 30 100/mm^3 with shift to the left. A lung ultrasonography (USG) showed left lung cyst and a CT scan also revealed an infected left lung cyst (Figure 5). Because the general condition became worse, respiratory difficulty progressed rapidly and cyanosis was noted and the serial chest X-ray showed progressive enlargement of the cyst and shift of the mediastinum despite of parenteral cefotaxime, ampicillin, gentamicin and assisted ventilator, then she underwent an emergency thoracotomy. During operation a broken cyst filled with yellowish pus was found in the inferior lobe left lung, so a left lower lobectomy was performed. Decortication was also done for pleural thickening in superior lobe. No lung expansion was shown in serial chest X-ray for several hours after the operation. The baby died 16 hours after the operation.

Histology (DR. B Hernowo): the specimens was a 5x5x1.5 mass of the lung, most of it was solid, firm. A cross sectional cutting showed a browned-yellowish solid mass with a space of 0.25 in diameter in one part. Microscopic findings showed normal bronchial and bronchiolar structures and abnormally alveolar collapse. There were many small cysts accounting most of the lesion demonstrated pseudostratified cuboidal epithelium lining and cartilage plates. The supporting stroma consisted of bundles of smooth muscle and was infiltrated by lymphocytes and also demonstrated small likely abscess formation (Figure 6).

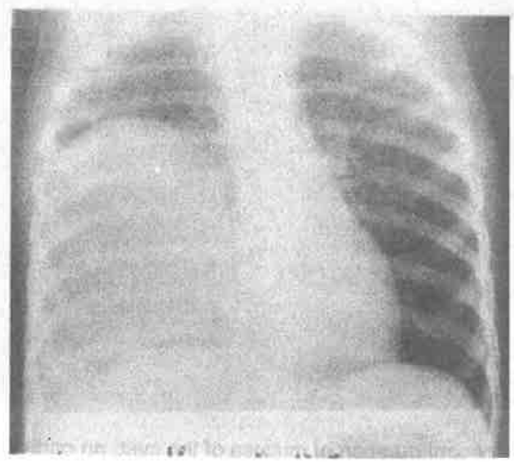

Figure 1. Chest X-ray showed right located empyema or lung cyst

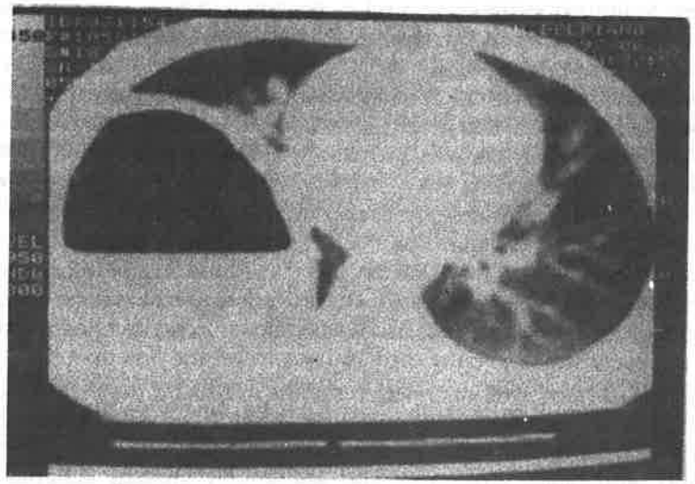

Figure 2. CT scan showed a thick-walled infected right lung cyst

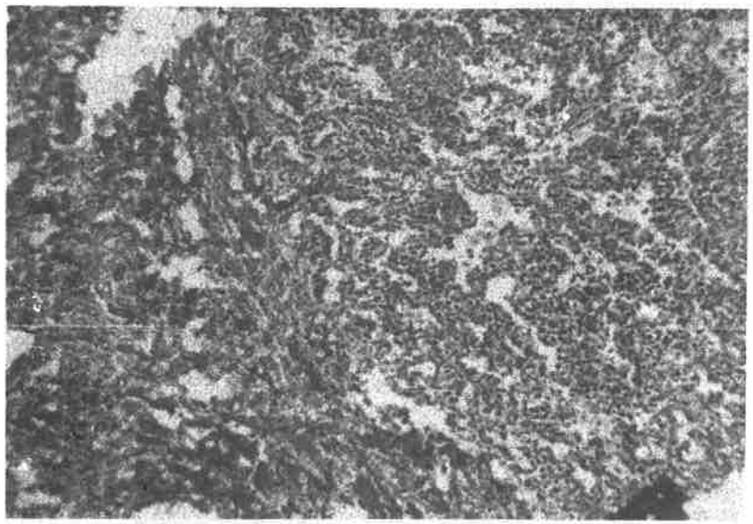

Figure 3. Papillary configuration of mucosa of the cyst, no cartilage, but inflammatory cells was shown

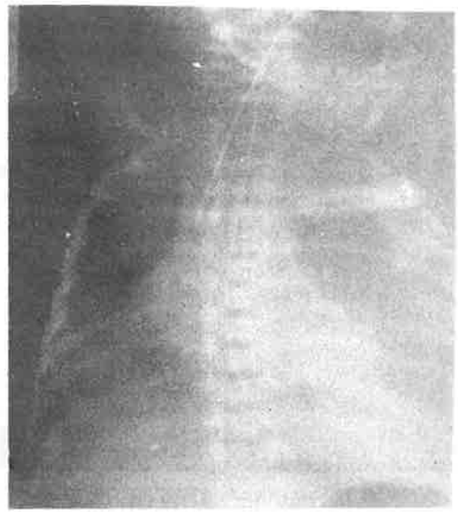

Figure 4. Chest X-ray in case no. 2 showed multiple cysts on the left hemithorax, tracheal and mediastinal shifting to the right hemithorax

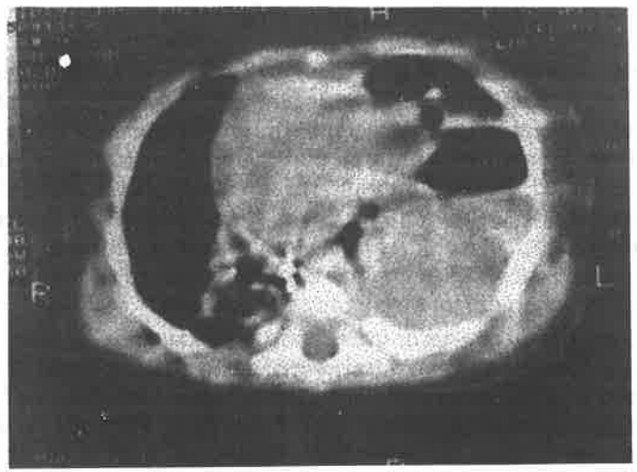

Figure 5. CT scan case no. 2 showed multiple cysts on the left lung

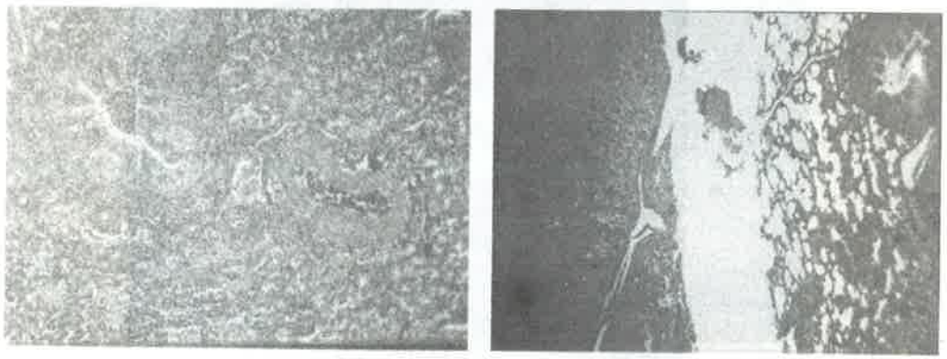

Figure 6. Histologic findings showed small cyst with cartilage plates

Discussion

Congenital cystic adenomatoid malformation of the lung presents mainly in neonates and infants.[1,4-7] rarely in children and has been reported in 2 adults[8] can produce respiratory difficulty by tension and infection. Cases described by Ch'in and Tang were original cases with premature, anasarca and maternal hydramnion; the affected lobe was a greatly enlarged, heavy tumorlike mass with multiple small cysts or slit like spaces. The cysts and spaces resembled bronchioles and the intervening tissue resembled very immature fetal "alveoli".[1,3] No inflammation were found in the original cases. Many reported cases thereafter showed variable clinical presentation and pathologic findings which have often differed from the original cases.[4,7,8]

The atypical cases have been term infants without anasarca, usually survived lobectomy, and whose specimens were often soften and aerated instead of solid mass. Microscopy revealed that the cysts had similar bronchiolar-like walls, but the intervening tissue was composed of mature alveoli.[3] The commonest postnatal presentation is progressive respiratory distress in a newborn associated with large cystic pulmonary mass on chest X-ray.[9] Children older than 3 months had infected cysts;[10] usually with repeated pulmonary infection later during infancy or childhood,[9,11-13] or even relatively asymptomatic[7] and found accidentally. One of our patients (Case 1) with recurrent

pulmonary infections but never had respiratory distress were initially thought to have encapsulated empyema which was very difficult to differentiate with infected lung cyst through clinical and physical examination as well as chest X-ray. But because an infected right lung cyst was shown from thoracic CT scan, right chest tube drainage was not done. Chest tube drainage was not done in encapsulated empyema.[14] An infected congenital cyst and encapsulated empyema may look alike; in many patients with congenital cyst, chest drainage has been performed on the basis of a diagnosis of empyema, but characteristically unlike empyema, obliteration of the infected cyst does not occur in the presence of adequate drainage.[9] Depending on the configuration of the structural deformity and on volume of parenchyma replaced, malformations of very limited dimensions may not become clinically manifest until late childhood or even in adulthood.[4]

Clinical findings found in case no. 2 might be due to progressive tension. The clinical pathogenesis in this case may derived from the pores of Kohn, with free access on inspiration and obstruction during expiration so an acute or chronic distention of the cyst leads to progressive increase in intrathoracic tension. If cyst drainage is poor, suppuration develops.[9] This patient died after undergoing thoracotomy. The majority of surviving children have had surgical resection soon after the diagnosis of CCAM was made.[6,7,9] Delaying surgery to permit the infant to gain weight is not recommended. The large size of the malformation will continue to interfere with pulmonary function and development. A pathological classification describing three types of CCAM has been used in predicting outcome; small cysts as in Case 2 give a bad prognosis.[5] The microscopic findings in the two cases here shown differently with descriptions of CCAM in the literature. It may be summarized as follows: 1. No increased amounts of elastic tissue and abundant smooth muscle in the walls of the cystic portions lined with bronchial-type epithelium (Case 2). 2. Absence of cartilage plates in the cystic parenchyma (Case 1). 3. There were inflammation in both cases.

ACKNOWLEDGMENTS

The authors wish to thank to dr. Bobby Akbar and dr. Suhartinah from the Department of Radiology for their cooperation and assistance in the preparation of this report.

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