Case Report

Congenital cystic adenamatoid malformation: A case report

Sirageldin MK Abdel Rahman (1), Aisha H. Bakhiet(2), Salah A. Ibrahim(3), Amadeldin O. Ahmed (4), SidahmedA.Sidahmed (4), Hana A. Ahmed(4)
(1) Department of Pediatrics, AlzaiemAlazhari University, Khartoum North.
(2) Department of Pediatrics, Faculty of Medicine, Omdurman Islamic University.
(3) Department of Pediatrics, Faculty of Medicine, University of Khartoum.
(4) Department of Pediatrics, Khartoum North Teaching Hospital.

ABSTRACT

We here report a rare case of congenital cystic adenamatoid malformation[CCAM]. This case presented early in the neonatal period with bilateral lung cysts and have favorable outcome. However, the patient continued to be oxygen dependent for more than six weeks.

Key words

Congenital cystic adenomatoid malformations (CCAM), Pulmonary abscess, chronic Lung Disease (CLD). Sudan.

INTRODUCTION

Congenital Cystic Adenomatoid Malformation CCAM is a rare abnormality of lung development [1,2]It accounts for 25% of all congenital lung abnormalities[3] and accounts for 95% of cases of congenital cystic lung disease [4].The incidence of prenatally diagnosed CCAM is 1 : 25,000 – 35,000. CCAM may present in old children and adults as incidental finding secondary to repeated infections [2-8].The mortality rate of prenatally diagnosed cases ranges from 9 to 49%. The risk factors for poor outcome include hydropsfoetalis [5,9], microcystic CCAM [5,10] and the overall size of the lesions[11,12]. The potential for malignant transformation is recognized in CCAM [2,13,14]. The diagnosis of CCAM can be made in utero by prenatal ultrasongraphy [2,9] and postnatally by imaging radiography and MRI. In 1977, Strocker classified CCAM into three typesmacrocystic with excellent prognosis, small cyst with bad outcome and multiple microcysts with also poor prognosis[2,10]. The mainstay of CCAM treatment is surgical excision that prevents complications such as recurrent infections, pneumothorax and malignancy[2,14]. Medical treatment by antibiotics for cases complicated by pneumonia and supportive care range from oxygen supplementation to mechanical ventilation are used when indicated.

We here report an infant with CCAM who behaves like chronic lung disease. To the best of our knowledge, this is the first report from Sudan.

Correspondence to:
Sirageldin Mohamed Kheir,
AlzaiemAlazhari University.
Khartoum North, Sudan
Email: Sirageldin89@hotmail.com
Mobile 0912627429

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CASE REPORT

Our patient is a forty days old male baby born to a consanguineous parents. He was an outcome of normal delivery to aprimiparous healthy mother. The mother was on regular antenatal care but she did not have antenatal ultrasound examination. The baby had an Apgar score of 9 and 10 at one and five minutes. He was discharged home after two hours of delivery. On the third day of life, he presented with shortness of breath, fever and poor suckling. Physical examination showed an ill looking baby with central cyanosis, tachypnea (respiratory rate of 70), .intercostal and subcostal recessions with course right basal crackles. Complete blood count and chest X ray were unremarkable. A working diagnosis of pneumonia was entertained and the patient was admitted to intensive care unit, given humidified oxygen through nasal prongs, intravenous fluids and antibiotics (cefotaxime, ampicillin and cloxcillin).The patient showed minor improvement and accordingly a chest CT was done and showed multiseptate abscesses. Vancomycin was added and all antibiotics were continued for two weeks. A repeat CT chest with virtual bronchoscopy showed numerous cystic lesions seen predominantly at lower lobes with no adjuvant tissue reaction or florid consolidation. Appearance in this age would be in favour of congenital adenomatoid malformation (Figure 1 and 2). The patient showed clinical improvement and was discharged home onoxygen. Ten days later, the patient developed another episode of pneumonia that required hospital admission for a month. He was then successfully weaned from oxygen, experienced fewer attacks of respiratory infections and is currently thriving well.

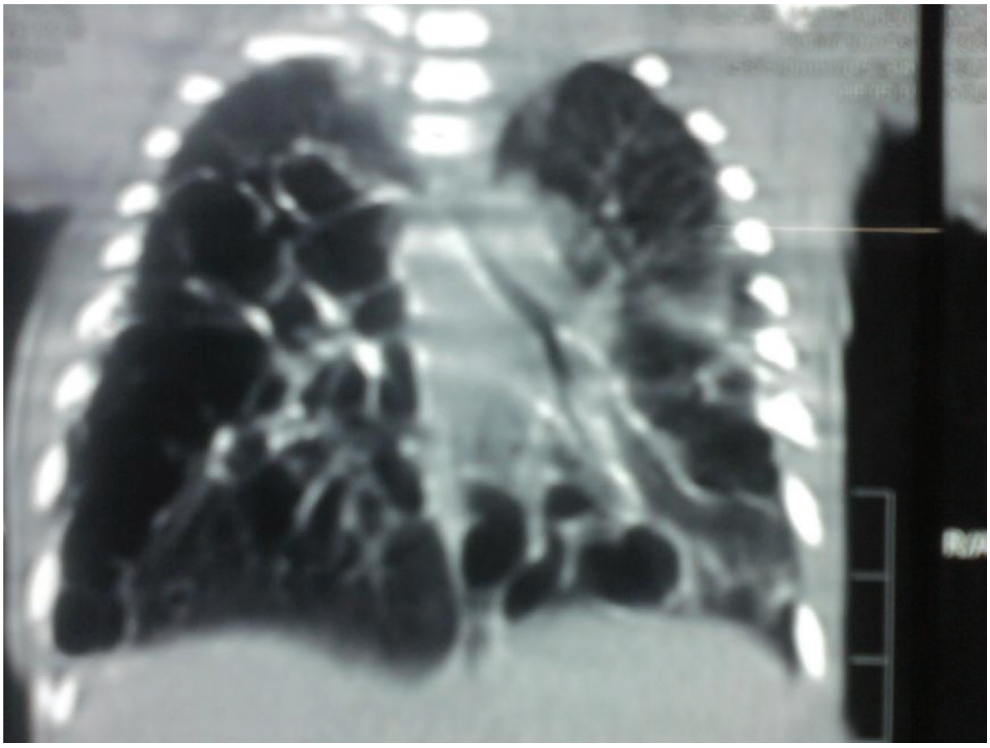

Figure 1 - Shows bilateral cystic lesions mainly in the right side (close PA view)

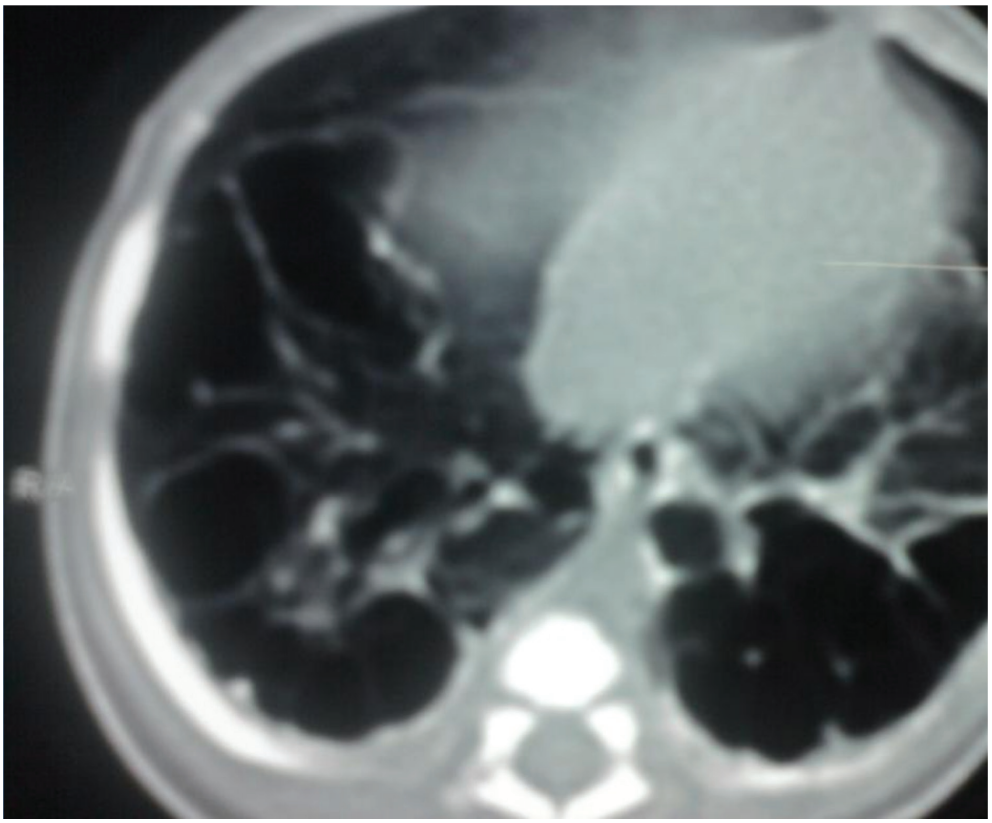

Figure 2 - Shows CT virtual bronchoscopy showing bilateral cystic lesions of CCAM (closecoronal view)

DISCUSSION

CCAM is a rare congenital condition that present postnatally with respiratory distress [5]. Despite the multiple extensive bilateral cystic lesions of our patient, he presented rather late on the third day.

The chest radiograph did not help in the diagnosis. The first CT chest scan report was misleading as it suggested pulmonary septate abscess.

This patient satisfied the definition of Chronic Lung Disease (CLD) which states that infants are considered to have chronic lung disease if they continue to require supplementation of oxygen to maintain adequate oxygentation after 28 days of life and have abnormal appearance of the lung parenchyma on chest radiograph (CXR)[15]..However, this patient does not fit inclusion into any of the four categories mentioned by the author.

It is interesting that our patient improved without surgical intervention despite the extensive multiple bilateral cysts. Although our patient has large cysts which are reported as a good prognostic factor, he has [2,10,] bilateral lesions I which is considered as a bad prognostic sign.

We conclude that this patient is unique as he presented rather late and he had favorable outcome without surgery despite the presence of multiple bilateral cysts that are associated with greater mortality. Finally we recommend to do a routine prenatal ultrasound examination to every pregnant woman in order to detect such rare lesions [5].

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