CONGENITAL CYSTIC ADENOMATOID MALFORMATION: CASE REPORT

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Abstract:

Congenital cystic adenomatoid malformation is a rare non hereditary lesion of the lung. In the neonatal period it presents with severe progressive respiratory distress due to expansion of the affected lung. We present a neonate in whom this condition was diagnosed and managed at Omdurman Military Hospital, pediatric surgery unit. In this case the clinical, radiological as well as surgical outcome highlighted.

Key word:

Congenital cystic adenomatoid malformation (CCAM)

Congenital pulmonary air way malformation (CPAM)

Introduction:

CCAM also known as CPAM is congenital, non-hereditary, hamartomatous abnormality of lung tissues with unknown etiology [(1)].The reported incidence of CCAM ranges from 1 in 11,000 to 1 in 35,000 live births [(2)].CCAM accounts for 25% of congenital lung lesions [(3)]. It was first described as a distinct Disease entity by Chi'in and Tang in 1949[(4)].In1977 Stocker et al. classified CCAM into 3 histopathological groups [(2, 3)]:-

Type	Character of cyst	Account for
I	multiple large cysts >2 cm or a single large cyst	55% of cases
II	has multiple small cysts less than 1 cm	40%of cases
III	multiple micro- cysts, measuring less than 0.5 cm	5%of cases

An expanded classification of 5 types was further proposed in 2002[(4, 5)].The prenatal rate of detection of lung cysts at the routine 18–20-week scan is almost 100 %[(6)].It present as respiratory distress in the neonatal period [(7)]. May remain asymptomatic in late childhood or adult life, can be present as recurrent chest infections or malignant transformation also [(2, 6, 8, and 9)]. We aim from reporting this case to highlight the clinical and radiological aspect of this rare disease as well as its successful management despite our limited facilities.

Case presentation:

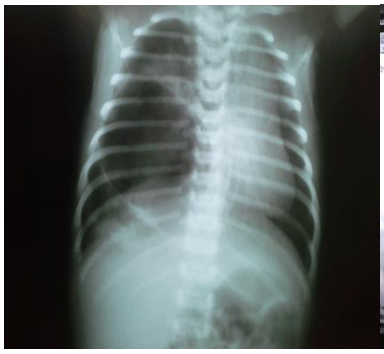
8 days old female neonate, full term, her birth weight was 3.0 kg, born by normal vaginal delivery at hospital .The baby cried soon after birth. Breast fed immediately passed meconium within the first 24hours; antenatal care was supervised and uncomplicated. She presented to a pediatric unit with complaints of poor feeding and shortness of breathing. The baby was looking ill, irritable and a febrile. Respiratory rate was70 breaths per minute, oxygen saturation 85% despite of 100% oxygen mask. Systemic examination was normal except for respiratory system; air entry was decreased on the right side with few creptations.

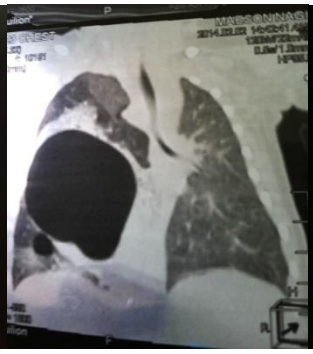
Investigations:

The investigation	The result
CBC	Hb 14,4g/dl TWBCS 13,700mm^3 Granulocyte 56, 7% Lymphocyte 33, 7% Reticulocyte count 1, 5% Platelet 271000mm^3
RFT	Urea 19mg/dl Creatinine 0.8mg/dl Na+ 131mEq/l K+ 4,6mEq/l
LFT	T.bilirubin 7,1 Unconjugated 5.7 T.protein7g/dl albumin. ALT normal. AST normal. ALP normal.
Bleeding Profile	PT 16, 7 control 14se PTT 14 control 25-45se INR 1,1
RBG	79mg/dl

Chest X-ray show (figure-1) air filled cystic like shadow confirmed with CT chest (figure-2) which revealed large air filled cyst. Echocardiography was normal.

The Patient was taken to emergency theatre. Through right thoracotomy, right middle lobectomy was done (Figure-3). Chest drain was fixed. Post-operative vital signs were normal; oxygen saturation was 100% with oxygen mask. 6hours post-operatively the saturation was 95% without Oxygen mask. The Patient followed up for 5days and discharged in a good condition (figure 5, 6, 7, 8). Histopathology showed type I CCAM.





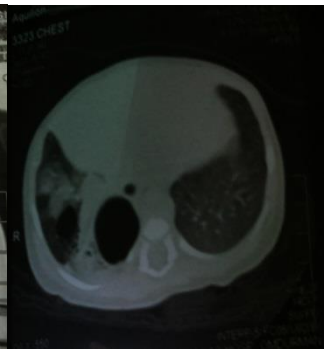


Figure-1(Chest X-ray shows air cyst) figure-2(CT scan)

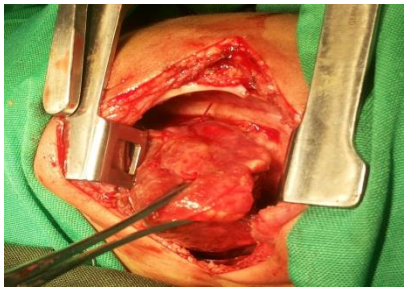
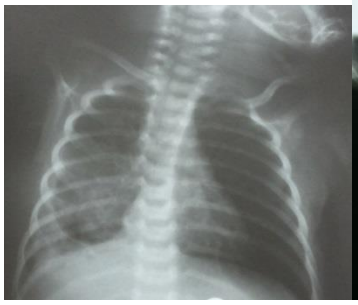


Figure-3middle lobe CCAM.



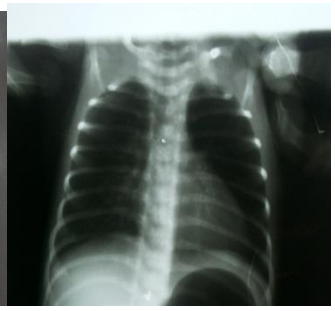


Figure-5 figure -6

(5^{th} day post-operative) (2month post-operative)

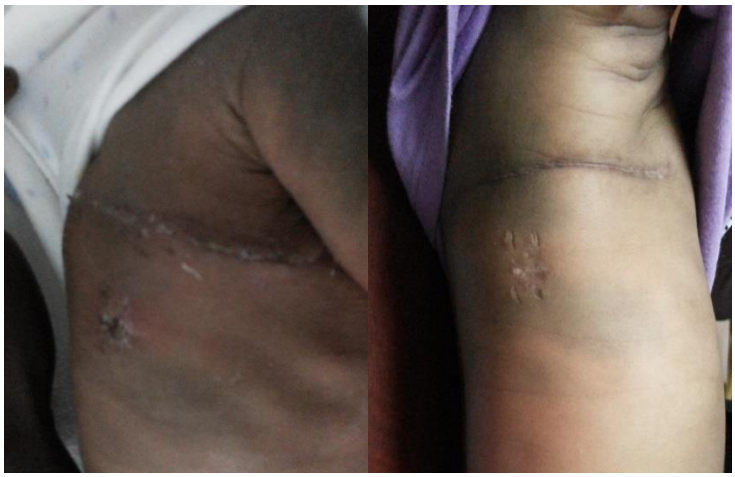


Figure-7(5th day post-operative) figure-8(2 month post-operative)

Discussion:

Congenital adenomatoid malformation can be detected in utero by prenatal scans, sometimes advanced modalities like MRI may be needed to differentiate it from congenital diaphragmatic hernia **(1).** Progressive respiratory distress is the commonest presentation in the neonatal period as had been seen in our patient. Older children present as repeated chest infections **(3).** Type 1 CCAM patients were more likely to remain a symptomatic, but our patient was presented with respiratory distress , however type I carry the best prognosis **(2)** Type 2 CCAM was often associated with other major anomalies**(3,4,10)**. Type (0) is incompatible with life as reported by Sui-Lingliao et al **(5)**

Chest radiograph doesn't differentiate CCAM from other differential diagnoses including lobar sequestration, bronchogenic cyst, congenital lobar emphysema or congenital diaphragmatic hernia **(2, 5).** CT chest is the most sensitive test for diagnosis of CCAM beside identification of its location and it determine any changes in the thoracic position of other lung lobes or in the mediastinum as appear in our work up to diagnose the case . Detection of other congenital gastrointestinal anomalies also can be done by CT chest like renal agenesis, diaphragmatic hernia and bowel atresia **(6, 7, and 8)** in this case cardiac anomaly excluded by echocardiography. MRI is another

modality for diagnosis, which can be used even prenatally to differentiate CCAM from other congenital anomalies [6, 7]. Postnatal management of CCAM has been controversy. Babies with large lesions or those symptomatic at birth should have early evaluation and surgery as was been done in our case, while babies with small asymptomatic lesions, surgical resection could be performed to them at (6-18) months of age as proposed by Bagolan et al. Others suggested that early surgery in asymptomatic cases was associated with low morbidity and mortality [8]. CCAM has more than (95) % survival rate in general in the absence of hydrops and associated congenital anomalies [2,10]. With hydrops mortality rate is more than (95) % [10].

Conclusion:

Early detection of congenital anomalies can be done prenatally by regular prenatal scans. Rapid and early intervention postnatal can improve the outcome markedly. Despite the absence of post operative intensive care unit settings, the post operative period passed uneventful. Nevertheless further studies needed to identify different types of CCAM in order to improve its management guidelines and to prevent development of late complications.

Reference:

1-Wafula .K. C, and Waa.S.Congenital cysticadenomatoidmalformation: case report.East African Medical Journal.2009;86(9): 454- 456.

2-Anna K. and Joshua A.Congenital Cystic Lesions of the Lung: Congenital Cystic Adenomatoid Malformation and Bronchopulmonary SequestrationRev Obstet Gynecol. 2012; 5(2): 85–93.

3-Chikkannaiah P, Kangle R, Hawal M. Congenital cystic adenomatoid malformation of lung: Report of two cases with review of literature. Lung India 2013; 30(2):15-218.

4-Sui-Ling Liao, Shen-Hao Lai1, ChuenHsueh, and Kin-Sun .Comparing Late-onset and Neonatally-diagnosed CongenitalCystic Adenomatoid Malformation of the Lung.Chang Gung Med J.2010; 33(1): 38-43.

5-Sittig .SE1 and Asay.GF.Congenital cystic adenomatoid malformation in the newborn: two case studies and review of the literature.Respir Care. 2000 Oct; 45(10):1188-95.

6-Sood M. Congenital Cystic AdenomatoidMalformation of the Lung. A Case Report. J Nepal PaedtrSoc 2011; 31(1):64-67.

7-Kumar KJ, Anilkumar MG, Shivamurthy YL, Kumar MP.Congenital cystic adenomatoid malformationpresenting as lung abscess in a child.TuberkToraks 2012; 60(4): 389-392.

8-Calvert.J K,Boyd.P A,Chamberlain.P C, and Said.SOutcome of antenatally suspected congenital cysticadenomatoid malformation of the lung: 10 years'experience 1991–2001.Arch Dis Child Fetal Neonatal Ed 2006;91:F26–F28.

9-Shettikeri.A, Padhakrishnan.P.fetaland neonatal out-come of Congenital Cystic Adenomatoid Malformation diagnosed in second trimester of pregnancy. Perinatology 2012 ;12(4):149-152.

10-SL .L,Mary.T,Chan.C,Lee.C. Management and outcome of antenatally diagnosedcongenital cystic adenomatoid malformation of thelung.Hong Kong Med J 2007;13:31-39.