Original Research Paper

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Neonatology

CONGENITAL CYSTIC ADENOMATOID MALFORMATION (CCAM) MASQUERADING AS PNEUMOTHORAX

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KEYWORDS :

Introduction:
Congenital cystic adenomatoid malformation (CCAM) is a rare developmental malformation of lung(1). CCAM is an uncommon fetal lung anomaly involving cystic changes of the terminal bronchioles(2). It is a part of a spectrum of disorders known as congenital pulmonary airway malformation (CPAM) which is described as hamartomatous lesion of the lung which results from abnormal branching of the distal airway(3). It is a rare lesion with incidence of 1 in 25,000 to 1 in 35,000 pregnancies(1). We present a rare case of CCAM in a 25 day old baby boy.

Case Report:
A full term 25 day old male neonate was admitted with severe respiratory distress. The baby was born to a registered and immunized primigravida mother whose antenatal scans were reportedly normal. Mother was delivered by lower segment caesarean section done in view of meconium stained liquor and fetal distress. Baby cried immediately after birth and weighed 3 kg. The baby was asymptomatic till 15 days after birth when he developed difficulty in breathing. Child was admitted in private hospital where chest X-ray was done showing left sided radiolucency. He was erroneously diagnosed as left pneumothorax and Intercostal drain (ICD) was put. Child was started on antibiotics and oxygen therapy. CT scan thorax was done which was suggestive of large thin walled air filled cystic lesion with internal thin septations in mid and lower lung fields on left side, largest 5.3*3.8 cm with moderate pneumothorax with collapse of left side with mediastinal shift to right side. Child underwent left lower lobectomy on day of life 32. Patient had an uneventful post-operative period before being discharged on day of life 44.

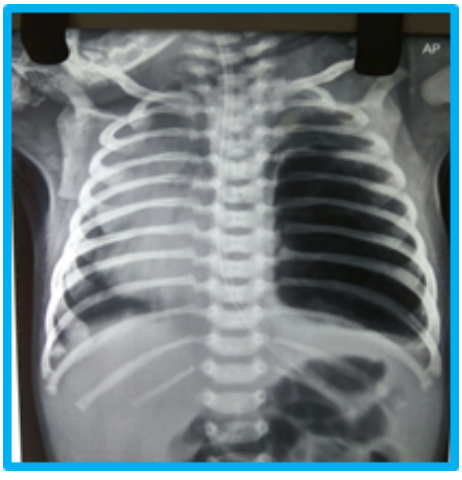

Fig no 1. Chest x ray reveals left side CCAM

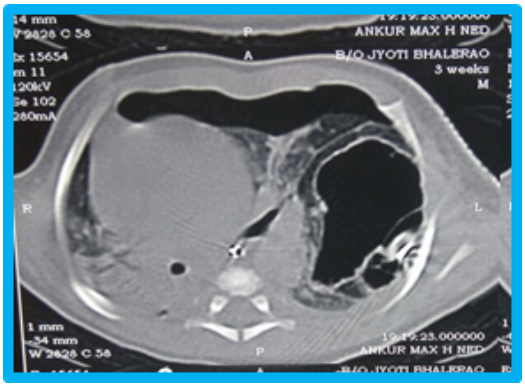

Fig no 2. CT thorax detected left lower lobe CCAM, Pneumothorax with ICD in situ

Discussion:
Congenital pulmonary airway malformation (CPAM) is a developmental anomaly of lung that may give rise to a spectrum of clinical presentations. This spectrum comprises of Bronchopulmonary sequestration (BPS) accounting for 14% of the cases, Congenital lobar emphysema (CLE) and CCAM which are 60% of the cases and a 'hybrid' of CCAM and BPS, accounts for the remaining 26% of the cases (4). The incidence of CCAM has been estimated at 1 case per 25,000 to 35,000 pregnancies (5). Males are more frequently affected than females and are not associated with race, age, maternal exposure to any given factor, or genetic factor(6).

Etiologies such as overgrowth, hyperplasia and hamartoma in the bronchioles have been implicated in the pathogenesis of CCAM. Arrest during the pseudoglandular period of airway development, which is characterised by rapid growth of conducting airways, results in development of CCAM types 1 to 3. CCAM type 4 is a result of this arrest occurring later, between 22 – 36 weeks of gestation. Genetically, the development of CCAM is attributable to the mutation in gene HOXB5 which is expressed during early lung development. Increased expression of platelet derived growth factor (PDGF-B) gene is responsible for rapidly progressive CCAM (7).

Historically, CCAM has been divided by Stocker into 3 types depending on size and consistency of lesion as type 1 to type 3 (8), however the newer expanded Stocker classification is from types 0 to 4 and are now named as congenital pulmonary airway malformations (CPAM).

Type 0 occurs in <3% of cases. It arises from the trachea and either does not have cysts or cysts are <0.5 cm. It involves all lobes and is

incompatible with life. Type 1 CPAM occurs in 60–70% which are cystic lesions >2 cm in size. It arises from distal bronchial or proximal bronchiolar region and is characterized by single or multiple cysts. Type 2 occurs in 15–20%, cases and arises from terminal bronchioles and the cysts are <2 cm in size. Type 3 has occurrence of 5–10%. The origin is from acini. It is predominantly solid with cyst size <0.5 cm.

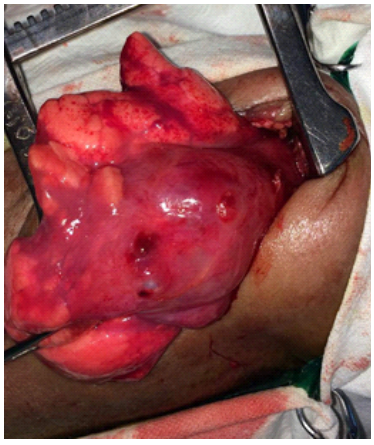

Fig no 3. Intra-operative lung cyst

Type 4 is seen in 10–15% of cases and has alveolar or distal acinar origins. It contains large cysts >2 cm in size, similar to type-1 CPAM. Type-2 and 3 CPAM are usually associated with other congenital malformations such as tetralogy of Fallot, renal agenesis, intestinal atresia, and diaphragmatic hernia. In type-4 CPAM, a sudden respiratory distress occurs due to tension pneumothorax (9). Our patient was a case of CCAM stocker type-1.

Clinical presentation of CCAM depends upon the size, location, type of lesion and its communication with the airway or the gastrointestinal tract and can vary from an asymptomatic infant to those with respiratory failure. Neonates with large CCAMs may be symptomatic on the first day of life itself (10). They present with predominant respiratory symptoms such as tachypnea, grunting, chest retractions and cyanosis (1). Infants may present with respiratory distress and reduced local air entry, and require supplemental oxygen therapy. Pulmonary hypoplasia in these infants is a risk factor for pulmonary hypertension (8). A CCAM may become symptomatic if associated with an intercurrent chest infection or with a pneumothorax (10).

Antenatally, CCAM can be diagnosed by ultrasound with Doppler studies and fetal magnetic resonance imaging (MRI). MRI improves the accuracy of diagnosis as well as helps in distinguishing a CCAM from a diaphragmatic hernia. The appearance of fetal CCAM varies from predominantly solid to purely cystic masses which may be macrocystic or microcystic (7). Postnatal diagnosis is often made with the help of chest skiagram and CT scan. CT scan helps in better delineation of CCAM size and site (11). The diagnosis of CCAM in our patient was not detected on antenatal ultrasound scans and was diagnosed in postnatal period after becoming symptomatic which was confirmed on chest X-ray and CT scans.

Complications of CCAM in neonates include cardiovascular compromise secondary to compression of mediastinal structures, pneumonia, lung abscess, fungal infections, haemoptysis, air embolism, intralobar sequestration and bronchogenic carcinoma (1).

Infants and older children with symptomatic CCAMs require surgery at the earliest. Infants requiring mechanical ventilation for large CCAMs need an urgent surgery. Treatment in asymptomatic neonates and infants with CCAM detected on imaging is controversial. Most authors favor an early surgery in contrast to a delayed one in infants. Regardless of size, it should be removed. Advantages of an early surgery are to facilitate optimal lung growth and minimise the risk of potential local morbidity, including infection and pneumothorax and to minimise the risk of subsequent malignancy. So continued review is appropriate before considering surgery (7). Although asymptomatic for initial few days of life, our patient developed respiratory distress and iatrogenic pneumothorax 15 days after birth and hence, had to undergo lobectomy.

In utero management techniques include open maternal-fetal therapy with fetal thoracotomy and lobectomy, thoracoamniotic shunting of macrocystic CCAM's and third trimester ex utero intrapartum therapy (EXIT) delivery with fetal thoracotomy, and lobectomy on maternal placental bypass (7). The survival rate after such treatment is around 50%. Steroid treatment can prevent progression of CCAM (12).

Depending upon the Stocker type, prognosis varies. Type 1 CCAM patients have overall good prognosis. The prognosis of type 2 depends upon the associated anomalies. Stocker type 3 carries the bad prognosis owing to its large size and cardiovascular compromise. Prognosis is poor in bilateral involvement, patients with hydrops and associated congenital anomalies (1).

Conclusion:

With improving antenatal imaging techniques, Congenital Pulmonary Airway Malformations (CPAM) can easily be detected and such neonates require close follow up with postnatal X-ray and CT scan. It is recommended to do elective resection in CCAM in early infancy to minimise morbidity and mortality.

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References:

1. Chikkannaiah P, Kangle R, Hawal M. Congenital cystic adenomatoid malformation of lung: Report of two cases with review of literature. Lung India [Internet]. 2013;30(3):215–8. Available from: http://www. pubmedcentral. nih.gov/ articlerender .fcgi ? artid =3775204&tool=pmcentrez&rendertype=abstract
2. Norris M. Congenital cystic adenomatoid malformation. Australasian journal of ultrasound in medicine. 2009 Aug 1; 12(3):39-41.
3. Onkar D. Congenital pulmonary airway malformation. Journal of the Anatomical Society of India. 2013 Jun 1; 62(1):90-2.4. Laberge JM, Puligandla P, Flageole H. Asymptomatic congenital lung malformations. Semin Pediatr Surg. 2005;14(1):16–33.
4. Farrugia MK, Raza SA, Gould S, Lakhoo K. Congenital lung lesions: Classification and concordance of radiological appearance and surgical pathology. Pediatr Surg Int. 2008;24(9):987–91.
5. Laberge JM, Puligandla P, Flageole H. Asymptomatic congenital lung malformations. Semin Pediatr Surg. 2005;14(1):16–33.
6. Herrero Y, Pinilla I, Torres I, Nistal M, Pardo M, Gómez N. Cystic adenomatoid malformation of the lung presenting in adulthood. Ann Thorac Surg. 2005;79(1):326–9.
7. Di Prima FAF, Bellia A, Inclimona G, Grasso F, Teresa M, Cassaro MN. Antenatally diagnosed congenital cystic adenomatoid malformations (CCAM): Research Review. J Prenat Med. 2012;6(2):22–30.
8. Shanti CM, Klein MD. Cystic lung disease. In Seminars in pediatric surgery 2008 Feb 29 (Vol. 17, No. 1, pp. 2-8). WB Saunders.
9. Stocker JT. Congenital pulmonary airway malformation: a new name and an expanded classification of congenital cystic malformation of the lung. Histopathology 2002; 41(Suppl2):424–31.
10. Fitzgerald DA. Congenital cyst adenomatoid malformations: resect some and observe all? Paediatr Respir Rev. 2007;8(1):67–76.
11. Calvert JK, Lakhoo K. Antenatally suspected congenital cystic adenomatoid malformation of the lung: postnatal investigation and timing of surgery. J Pediatr Surg. 2007; 42(2):411–4.
12. Adzick NS. Management of Fetal Lung Lesions. Clin Perinatol. Elsevier Ltd; 2009;36(2):363–76.