Research Paper Medical Science

Congenital Cystic Adenomatiod Malformation Type-1

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ABSTRACT

Congenital cystic adenomatoid malformation (CCAM), a rare type of developmental anomaly of the lung was first acknowledged as a separate entity and introduced into English literature by Chin and Tang in 1949(1). The disorder is often referred to by another name, congenital pulmonary airway malformation (CPAM). Several patterns of clinical presentation have been observed-

(i)still birth or neonatal death frequently associated with fetal hydrops

(ii) acute progressive respiratory distress in a newborn

(iii) an indolent course characterized by recurrent pulmonary infections

(iv) rarely pneumothorax(2)

(v) even more rarely as a prenatal diagnosis on maternal ultrasound or polyhydramnios(3).

We are reporting here a case of CCAM came with respiratory distress.

Keywords : Congenital cystic adenomatoid malformation,CT scan, pneumenectomy

CASE REPORT-

A 2 yr. male child(fig:3) was admitted with complain of cough(nonproductive) and fever(High grade and continuos) for 1 week and difficulty in breathing from 3-4 days. Patient had multiple similar episodes in past and was treated as respiratory tract infection but never hospitalized . His birth was uneventful and antenatal USG was not done.

O/E pt. had growth failure, halitosis and clubbing. At the time of admission pt. was in respiratory distress with dullness & decrease air entry over right side of chest, and B/L crepitations. Clinical impression was considered right sided collapse/ consolidation or right sided pleural effusion and started parentral antibiotics with oxygen support. Chest roentogeogram(fig:1) showed multiple right sided heterogenous opacities with contra lateral mediastinal shift. Hemogram showed PMN leucocytosis . CECT thorax(fig:2) showed right lung hyperinflation with multiple air filled cystic lesions of varying sizes occupying entire right lung. Cyst were separated from each other by opaque pulmonary tissue strands.Medaistinum was shifted to left sided. Left lung appeared normal . As per CT finding(fig:2) pt. was diagnosed as Congenital cystic adenomatoid malformation(**TYPE 1)** involving right lung and pt. advised pneumenectomy.

Fig-1 x-ray chest PA view showing multiple cystic lesion with mediastinum shift

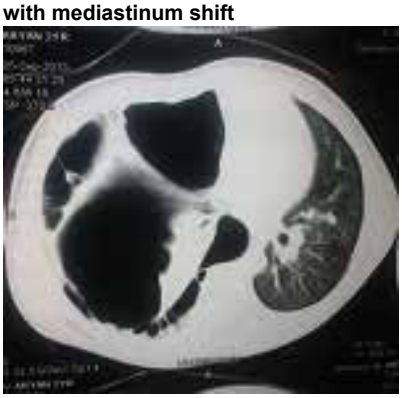

fig-2 CT SCAN THORAX showing cysts in right lung seprated from each other by opaque pulmonary tissue strands

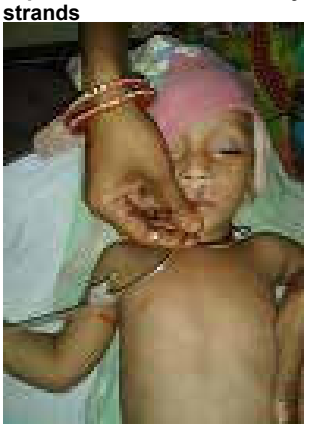

Fig-3

DISSCUSSION-

Congenital cystic adenomatoid mal-formation (CCAM) of the lung is caused by anomalous fetal development of terminal respiratory structures, resulting in adenomatoid proliferation of bronchiolar elements and cyst formation leading to enlargement of the affected lobe. The clinical spectrum varies depending on the extent of malformation in the lung and associated conditions.

Some authors have observed predilection of the right lung over the left for this anomaly(4)as seen in our case. Involvement of an entire lung is distinctly rare. Single lobe involvement is the most common. Out of 153 cases reported till 1993 only 27 had multiple lobes affected(5). In our case we also having entire right lung involvement. Since the cysts are conglomerated into a well defined expansive mass, the space occupying lesion produces compression on surrounding tissues and pushes the mediastinum. A less common presentation is without mediastinal shift which is true about type II CCAM *vis-a-vis* type I(6). There was no associated pulmonary hypoplasia which is a well described associated anomaly(7). High frequency of other anomalies has been seen in type II CCAM(6).

CCAM can be differentiated from other cystic lesions of the lung namely pulmonary sequestration, bronchogenic cyst, congenital lobar emphysema or diaphragmatic hernia and cystic bronchiectasis. In bronchiectasis, CT scan shows bronchi with thick walls sometimes looking like a string of pearls when aligned. In contrast, thin-walled cystic spaces are seen in CCAM(8).

Table 1. Pathologic Features of Congenital Cystic Adenomatoid Malformation lung (according to Stocker) (6)

Classification	Type 0	Type 1	Type 2	Type 3	Type 4
Frequency (%)	1-3	>65	20-25	8	10
Cyst size (maxi-mum)	0.5 cm	0.5-10 cm	0.5-2 cm	0.3-0.5 cm	7 cm
Epithelial lining	Ciliated, pseu-dostratified columnar	Cuboidalcells, flattened, co-lumnar	Ciliated, cuboidal, ciliated pseudostrati-fied	Ciliated, cuboi-dal, columnar	Type 1&2 al-veolar lining cells
Muscular wall thickness (mm)	100-500	100-300	50-100	0-50	25-100
Mucus cells	Present in all cases	Present (33%of cases)	Absent	Absent	Absent
Cartilage	Present in all cases	Present(5-10%of cases)	Absent	Absent	Rare
Skeletal muscle	Absent	Absent	Present (5%of cases)	Absent	Absent

The definitive treatment of CCAM is surgery. However, there is a controversy whether or not all cases be subjected to surgery. Surgery is indicated for the following reasons: (i) definite histological diagnosis, (ii) history of recurrent infections, and (iii) risk of malignancies which have been rarely reported.

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Disclosure

The authors report no conflicts of interest in this work.

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