Case Report: Unusual Presentation of Congenital Cystic Adenomatoid Malformation in A Toddler.

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ABSTRACT

Congenital cystic adenomatoid malformation of the lung is a rare lesion that typically manifests as severe progressive respiratory distress in the neonate secondary to expansion of the affected lung. We present a 2.5 year old female child diagnosed to have congenital cystadenomatoid malformation

Introduction:
Congenital cystadenomatoid malformation (CCAM) also named congenital pulmonary airway malformation(CPAM), 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). Congenital cystic adenomatoid malformation (CCAM) is defined as a mass of abnormal solid or cystic pulmonary tissue in which there is a proliferation of bronchial structures at the expense of alveolar development.(2)
Seventy per cent present in first month of life (3).

Case Report:
A 2.5 year old female child,fifth born to a third degree consanginous marriage was admitted with complaints of cough and fever since 20 days which responded poorly to antibiotics. Cough was non productive and gradually progressive. Neonatal period was uneventful and there was no history of similar complaints in the past and no history of contact with tuberculosis. Right hemithorax was dull to percussion and had features of volume loss with crackles all over. Clinical impression was right sided collapse/ fibrosis. Chest X-ray showed right sided heterogenous opacity with ipsilateral mediastinal shift. Ultrasound chest revealed right basal lung consolidation with air bronchogram.Flexible fiberoptic bronchoscopy did not show any intra bronchial obstruction. Hemogram showed anemia with polymorphonuclear leucocytosis.CT chest showed a large thin walled cavity in the right upper and middle lobe measuring 80 x 54 mm. Few septae were seen with in the cavity. Adjacent lung was collapsed due compression by the large cavity within the right lung.

Fig 1: Photograph of chest X-ray showing heterogenous opacity right lung with mediastinal push.

Discussion:
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. This congenital pulmonary disorder occurs in ~1-4/100,000 births. There is a predilection of the right lung over the left for this anomaly(4). Single lobe involvement is the most common.CCAM usually presents with respiratory distress in the neonatal period. 80% of the cases are diagnosed by one month of age.

The Features unique in our case is the late age of presentation and the involvement of both upper and middle lobes.

Prenatally, the challenge is mostly to differentiate this condition from congenital diaphragmatic hernia. To do this, the diaphragm would have to be visualised and for this reason, obstetric MRI is sometimes required.Other conditions that need to be differentiated from CCAM are pulmonary sequestration, bronchogenic cyst, congenital lobar emphysema and cystic bronchiectasis. Large lesions may compromise normal development of lungs in utero leading to stillbirth or neonatal death. CCAM is a recognised cause of non-immune foetal hydrops.

Three histologic patterns have been described. Type 1 (50%) is macrocystic and consists of a single or several large (>2 cm in diameter) cysts lined with ciliated pseudostratified epithelium.

Fig 2: CT scan showing cystic lesion of right lung.
The wall of the cyst contains smooth muscle cells and elastic tissue. One third of cases have mucus-secreting cells. Cartilage is rarely seen in the wall of the cyst. This type has a good prognosis for survival. Type 2 (40%) is microcystic and consists of multiple small cysts with histology similar to that of the type 1 lesion. Type 2 is associated with other congenital anomalies and carries a poor prognosis. In type 3 (<10%), the lesion is solid with bronchiolo-like structures lined with cuboidal ciliated epithelium and separated by areas of nonciliated cuboidal epithelium. This lesion carries the poorest prognosis and can be fatal.

The definitive treatment of CCAM is surgery. The outcome of lobectomy/pneumonectomy is good in children. In lobectomy the remaining lung grows and expands well enough so that total lung volume and pulmonary function tests return to normal(11). This response is most vigorous in the very young because new acini and alveoli form up to 5 years age(12). Post-resection majority of patients have an excellent result. Younger groups have lower ratio of residual volume to total lung capacity and higher maximum breathing capacity. This suggests that hyperplasia rather than over distension occurs in the remaining lung(12). If left untreated there is a higher chance for malignant transformation.

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