Congenital Cystic Adenomatoid Malformation of Lung

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Introduction

Congenital cystic adenomatoid malformation (CCAM) is a rare entity and is considered as a focal pulmonary dysplasia. Rarity of this lesion can be assessed from the fact that in a teaching 600 bedded hospital with a clientele of 3,50,000 there were only 12 cases reported over a period of 16 years [1]. In another study, the incidence was 1:25000 to 1:35000 pregnancies [2]. It is defined as a multicystic mass of a pulmonary tissue in which there is proliferation of immature bronchial structures at the expense of alveolar development [1]. It represents 25% of congenital lung malformations and 95% of congenital lung lesions [3]. This lesion occurs more often in males with a ratio of 1.8:1, and is primarily unilateral, but may occur bilaterally [4]. Associated anomalies are rare. Some CCAM lesions present only at birth with respiratory distress symptoms but are confirmed by an abnormal chest radiograph or computed tomography scan. We present a case of CCAM in a child who presented with features of pneumothorax.

Case Report

A two year old female child presented with complaints of sudden onset of breathlessness and pain in the left side of chest. Clinically there was no air entry on the left side. An urgent radiograph chest revealed a large pneumothorax with collapsed left lung. Few cystic lesions were noted in the collapsed portion of the left lung. No significant mediastinal shift was noted and there was no herniation of the affected lung to contralateral side. To relieve the breathlessness, a needle was put into the left hemithorax and after initial drainage of air it was followed up with ICD drainage. On follow up radiograph there was complete opacification of the left hemithorax with few cystic lesions in the left lower zone. A provisional diagnosis of CCAM Type I was offered. Blood and biochemical parameters did not reveal any significant finding. Computed tomography (CT) scan showed hydro-pneumothorax on the left side with multiple large cystic

lesions including the lingular lobe of left lung. The largest cyst measured 36x24mm. There was subpleural cystic lesion seen on left side. The child underwent a lobectomy at a tertiary care hospital. Histopathology proved the diagnosis of congenital cystic adenomatoid malformation. At present the child is completely asymptomatic and on regular follow up.

Discussion

Histological classification of CCAM was first described in 1977 [5]. The Stocker classification system divides the lesions into three groups. The Stocker Type I, accounts for 50-70% of diagnosed cases. It is composed of single or multiple large cysts (3 to 10 cm), confined to one lobe and filled with air or fluid. Only 11% of these lesions are complicated with associated anomalies and majority of these patients survive. Our case fits into this group. CCAM Stocker Type II lesions found in 18-40% of cases, consist of multiple evenly distributed, medium sized cysts (<2.0 cm) that resemble terminal bronchioles. They are associated with a higher incidence (50%) of other congenital anomalies. CCAM Stocker Type III lesions represent 10% of diagnosed cases and are usually large, bulky lesions with evenly distributed small cysts. These cysts may occupy an individual lobe or the entire lung. Mediastinal displacement is extremely common. The condition is usually isolated. CCAM III has been reported in association with grossly elevated amniotic fluid alpha fetoprotein [6,7]. The essential problem in congenital cystic adenomatoid malformation is excessive overgrowth of the terminal bronchioles leading to enlargement of the lobe, while the development of the alveoli is completely suppressed except at the periphery.

The differential diagnosis for fetal lung space occupying lesions includes diaphragmatic hernia, pulmonary sequestration, bronchogenic cysts, bronchiectasis, cystic fibrosis and congenital lobar emphysema [3,6]. Prognosis in CCAM depends upon

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Fig. 1 : Chest radiograph AP view shows large left sided pneumothorax with compression of the collapsed lung. Also seen are few cystic lesion in the collapsed left lung.

Fig. 2 : Following needle aspiration there was accumulation of fluid giving the appearance of consolidation of left lung. Still well defined cystic lesions are seen in left lower zone.

Fig. 3 & 4 : CT scan in mediastinal and lung window shows left sided pneumothorax with parenchymal cystic lesion

the size of the lung mass and the pathophysiologic involvement [7]. Early identification of patients with CCAM helps in prenatal counseling, potential fetal interventions and birth planning at a tertiary care facility. The mortality in patients who present with fetal hydrops is much higher (up to 68%) as compared to those without (30%) [8,9]. Those infants with fluid filled cysts may benefit from the placement of a thoracoamniotic shunt under ultrasound guidance. Foetal shunt placement has been associated with a mortality rate of 12.5% and with obstructions and dislocations in 29% of the cases [3]. Additional risks of these procedures include premature rupture of membranes, premature birth, haemorrhage, infection and injury to the fetus. It has been documented that a percentage (15-60%) of these lesions may regress spontaneously in utero [7,10]. For those greater than 32 weeks without foetal hydrops, early delivery may be considered.

The commonest presentation of CCAM in post natal period is progressive respiratory distress including tachypnoea, grunting, retractions and cyanosis [3]. A chest radiograph may show displacement of the mediastinum away from the affected side with loculated translucencies in the affected side which represent the CCAM as in our case. CT scan shows multilocular cystic mass with thin walls. In case of an infected lesion CT shows mixture of solid and cystic lesion with variable definition of thick walls. On magnetic resonance imaging, the appearance of CCAM is determined by the size of the lesion as well as the number and size of the cysts. In contrast to pulmonary sequestrations which have a homogeneous appearance, CCAM demonstrates inhomogeneity because of the multiple cysts. Severe respiratory distress warrants surgical resection. When symptoms are present at birth, surgical resection is almost always required.

Conflicts of Interest

None identified

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