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Congenital Cystic Adenomatoid Malformation of Lung- A Rare Case Report

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Abstract: *Congenital cystic adenomatoid malformations (CCAM) of lung are rare congenital cystic lung lesions that arise from excessive proliferation of tubular bronchial structures. CCAM is divided into 5 types. It presents with respiratory distress in neonatal period. We report a case of type 1 CCAM in a female neonate and also describe the pathological features of the various types.*

Keywords: *Congenital cystic adenomatoid malformations lung, cystic lesion, disorganized proliferation, tubular bronchial structures, respiratory distress*

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I. Introduction

Congenital cystic adenomatoid malformation (CCAM) of lung is a rare developmental anomaly with incidence of 1 in 25,000 to 1 in 35,000 pregnancies(1). It represents 25% of congenital lung malformations and 95% of congenital lung lesions. It is commonly seen in male babies(2).

II. Case report

A full term female baby with birth weight 3.0kg was born to a 27 year old G2P1L1 female by normal vaginal delivery at the hospital. The baby cried immediately after birth with APGAR score of 8 at 1 minute and 9 at 5 minutes. However later the neonate developed respiratory distress. On examination heart rate was 138/min and respiratory rate was 76/min. Respiratory examination showed decreased air entry on right side and few crackles were heard over lower part of the chest wall and patient had persistent tachypnea. In view of these clinical findings, radiological investigations were advised. Chest X ray showed dense voluminous right lung with an air-containing cystic area occupying the right upper lung and mediastinal shift towards left. Computed tomography scan of the chest showed gross increased volume of right lung with medisatinal shift towards left causing decrease in left lung volume and mild compression over trachea. There was evidence of large solid lesion in right lung parenchyma with air lucent cavities along the anterior aspect. On CT scan possibility of congenital cystic adenoid malformation was suggested. Patient was initially managed conservatively but in view of persistent tachypnea and mediastinal shift surgical intervention was planned. She underwent thoracotomy and lobectomy and the specimen was sent for histopathological examination.Gross examination revealed a 7. 5 cm X 5 cm X 2 cm mass showing whitish areas with multiple thin-walled small cysts of uniform size (2.5 cm to 3 cm in diameter) as seen in figure. Microscopically, many bronchioles were seen showing cystic dilatation with polypoidal mucosal infoldings lined by cuboidal to pseudostratified ciliated columnar epithelium as seen in figure 2 and figure 3. Few cysts were lined by mucin secreting cells. Solid areas composed of small alveoli filled with mucus material seen. Intervening connective tissue showed sparse lymphocytes and hemorrhagic areas. No necrosis or calcification was seen.

Both histological and radiological findings were consistent with type 1 Congenital cystic adenomatoid malformations (CCAM) of lung. The patient had improvement in breathing and after 3 months, on follow up she had no respiratory distress and also chest X ray showed no midline shift.

III. Discussion

CCAM, an abnormality of the lung development, is defined as a hamartomatous congenital pulmonary airway malformation.(1) Congenital cystic adenomatoid malformation(CCAM) was first described by Chin and Tang in 1949 as a developmental anomaly and termed it as “congenital adenomatoid malformation”.(3)

Stocker et al. first classified CCAM into three subtypes and later in 2002 extended the classification into 5 subtypes. (4)Type 0 consist of microcystic disease throughout the lungs. It is also known as acinar dysplasia and was first described by Rutledge and Jensen as a malformation of the proximal tracheobronchial tree.(5) It is the least common variant and has the worst prognosis. Type 1 is the most common variant. It is characterized by larger cysts measuring more than 2cm and lined by ciliated pseudostratified epithelium. One-third of the cases have cysts lined by mucus secreting cells, these are pathognomic for type 1 CCAM. It involves only a part of 1 lobe and has good prognosis. Type 2 has smaller cysts which are histologically similar to the type 1. Type 3 consists of solid tissue with bronchiole like cystic structure lined with simple cuboidal ciliated epithelium. The prognosis for this type is poor. Type 4 also shows larger cysts but the lining lack mucus cells. It is associated with malignancy occurring later in life. (6) (7)The disorder can be suspected in antenatal period using ultrasonography. After birth chest radiograph and CT thorax constitute important diagnostic modality, but a definitive diagnosis is done only after histopathological examination.Carcinomatous change has been described in patient with congenital pulmonary airway malformation. (8) Therefore early recognition and surgical intervention is important in preventing the consequences of recurrent pulmonary infection and the potential risk of malignancy.(9)The differential diagnosis include bronchogenic cyst, congenital lobar emphysema and cystic bronchiectasis, pulmonary sequestration and pleuropulmonaryblastoma.(10)

Long term outcome is good in surgically managed asymptomatic patients with some studies showing only slight decrease in lung volume. Congenital cystic adenomatoid malformation of lung can cause severe respiratory distress in a neonate but early diagnosis and surgical intervention can improve the condition and prevent death due to respiratory failure.

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Figure 1 Gross image of the lobe of right lung showing whitish solid areas and multiple cysts.

Figure 2 and Figure 3 Low power view showing various cysts lined by pseudostratified columnar epithelium