Congenital Cystic Adenomatoid Malformation of Lung, a Rare Case Report

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Abstract: Congenital cystic adenomatoid malformations (CCAM) are rare developmental anomalies of the lung occurring in 1:4-100,000 live birth. It arises from excessive disorganized proliferation of tubular bronchial structures. It is a disease of infancy with most of the cases diagnosed within first 2 years of life. We report a case of congenital cystic adenomatoid malformation (CCAM) type 1 in a 5-yr-old male child with features of respiratory distress.

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

I. Introduction

Congenital Cystic Adenomatoid Malformations are rare abnormalities of lung development in which there is adenomatoid proliferation of bronchioles that form cysts at the expense of normal alveoli. The etiology remains unknown, but a disturbed interaction between mesodermal and ectodermal components of the lung during embryonic development has been suggested(1). It represents 25% of congenital lung malformations(2). 80% of the lesions are discovered during neonatal period due to respiratory distress however may be discovered in older children or adults occasionally. In 1977 Stocker et al. originally described the findings and classified CCAM into 3 subtypes(3). In 2002 it was expanded into five types and renamed as congenital pulmonary airway malformation(4).

II. Case History

A 5 year male child presented with complaints of intermittent fever, recurrent chest infection and respiratory distress for one month. On general examination there was no cyanosis, clubbing or lymphadenopathy. Chest examination revealed rhonchi and crepitations predominantly on right side. His cardiovascular and abdominal examination revealed no abnormality. His hemogram showed anemia with polymorpho-nuclear leukocytosis. Sputum and blood culture were non-contributory. Chest X-ray revealed multiple cystic lucencies in right lower lung field. Patient received oxygen and antibiotics therapy but continued to have decreased breath sound on right side, so advised for CECT chest which revealed Cystic lesion suggesting possibility of CCAM type 1(Figure 1&2). Antibiotics was continued for 2 weeks and patient was referred for surgical consideration. Lobectomy was done and specimen(Figure 3) sent for histopathological examination. Histopathological examination of the excised mass showed many normal-sized to dilated alveoli lined by flattened or low-cuboidal epithelium and containing blood and alveolar macrophages(Figure 4).

III. Discussion

CCAM lung are rare developmental anomaly of lung characterized by overgrowth of terminal bronchioles with reduction in alveoli(5). Incidence of CCAM is 1:25,000 to 1:35,000 with male and female equally affected(5). They are usually unilateral and restricted to a single lobe(6). Extra-pulmonary abnormalities can be found in 10% of the cases such as renal, central nervous system and cardiac defects(7). Most of the patients can present with respiratory distress or a history of recurrent chest infections while some cases are asymptomatic and incidentally found on radiography. Differential diagnosis of CCAM includes diaphragmatic hernia, bronchogenic cyst, cystic fibrosis, congenital lobar emphysema, and pulmonary sequestration(2). CCAM will have connections to tracheobronchial tree and derives its blood supply from pulmonary circulation. In contrast, pulmonary sequestration contains immature lung tissue without connection to tracheobronchial tree and derives its blood supply from aberrant systemic blood vessel(8). Congenital lobar emphysema should be
considered as close differential diagnosis because it can present in a similar way. Based on the anatomical changes development of the vertebrate lung has been sub-divided into five distinct periods: Embryonic (3-7 weeks), Pseudoglandular (7-17 weeks), Canalicular (17-29 weeks), Saccular (24-36 weeks), and Alveolar (36 weeks to maturity). The CCAM develops during the pseudoglandular and saccular period(8). In 1977 Stocker et. al subdivided CCAM into three subtypes based on site of origin of the malformation and later added two more sub-types in 2002,[5]: (i) type 0 – acinar dysplasia, (ii) type I – multiple large cysts or a single dominate cyst, (iii) type II – multiple evenly spaced cysts, (iv) type III – bulky firm mass, (v) type IV – peripheral cyst type. In neonatal period the main complication of CCAM is compression of the mediastinal structure. In adult patient, CCAM is a nidus for pneumonia, abscess formation, spontaneous pneumothorax, hemoptysis, intralobar sequestration, and development of bronchogenic carcinoma(9). Stocker type, type I lesions carry overall good prognosis. Type III lesions carry bad prognosis as they are usually large in size and presents with cardiovascular compromise. Overall bilateral involvement and associated congenital anomalies carry poor prognosis(10).

References

Legends
1. Figure 1 coronal section of CECT chest revealing Cystic lesion suggestive of CCAM type I
2. Figure 2 Mediastinal window of CECT CHEST showing CCAM type 1 in right lung lower lobe

Figure 3 Lobectomy specimen
**Figure 4** Showing many normal-sized to dilated alveoli lined by flattened or low-cuboidal epithelium and containing blood and alveolar macrophages.