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Completely Air Filled Bronchogenic Cyst: A Rare Case

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INTRODUCTION

Bronchogenic cysts are formed due to abnormal budding of bronchial tree. They are lined by respiratory epithelium. They are not air filled as they do not commonly communicate with the bronchial tree. They are filled with fluid containing proteins, blood products and calcium oxalate.

One third are located in the lung parenchyma and two third are located in the mediastinum. 40 to 50% of all congenital cysts in the thorax are bronchogenic cysts. On CT scans they appear as round mass with attenuation similar to soft tissue or water.

Symptoms arise only when the cyst compresses surrounding structures. Most cysts are asymptomatic.

We present a case of a young lady who presented with an incidentally detected cavitory lesion on routine health checkup. Bronchogenic cysts which are completely air filled are rare and need treatment if very large or causing symptoms.

Keywords: *Bronchogenic cyst, respiratory epithelium, intra-parenchymal, intra- pulmonary.*

CASE REPORT

A 16-year-old asymptomatic lady presented for evaluation of an incidentally detected chest radiographic abnormality while undergoing routine health checkup for abnormal menstrual cycles with dysfunctional uterine bleeding. There was no history of chest trauma or any other symptom suggesting recurrent lung infections in the past. She is a non-smoker. There is no history of tubercular disease in the past. Previous chest radiographs and clinical records were not available for verification.

General physical examination and systemic examination were normal. Chest examination revealed diminished air entry in left lower lung fields. Examination of other systems was normal.

Routine blood and urine investigations did not reveal significant abnormality. The chest radiography revealed a large thin-walled cavity occupying the left middle and lower lung fields. There was no significant mediastinal shift.

She eventually underwent surgery for excision of the cyst and a left lower lobectomy was performed to excise the cyst.

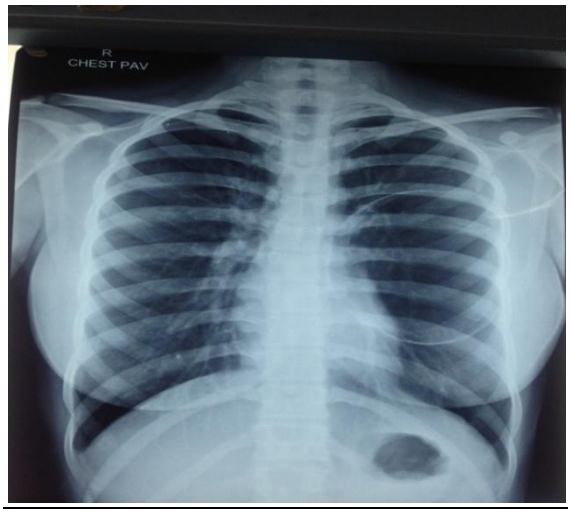

Fig 1: chest x-ray showing large air filled cavity in left middle lung field.

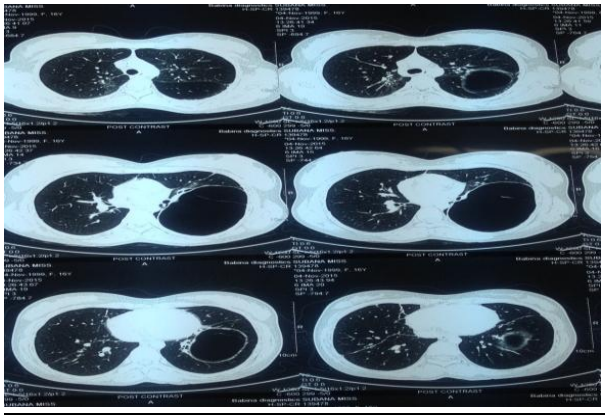

Fig 2: CT scan showing the cyst cavity completely air filled.

DISCUSSION

The earliest reports date from the late 1890s when a cystic disease process was identified in non-surviving infants.[1]In 1949 Chi'in and Tang used the term congenital adenomatoid malformation to describe these cystic lung disease.[2]

In 1973, Van Dijk described three varieties: cystic, intermediate, and solid.[3] Stocker proposed a classification in 1997 on the basis of cyst size.[4]CCAMhave a reported incidence between 1/8,000 and 1/35,000. [5, 6.]

CCAM is most commonly found in the neonatal period and up to 90% of diagnoses are made within the first two years of life. [7, 8.]

CCAMs are thought to originate from different levels of the trachea-bronchial tree and at different stages of lung development.[9] CCAM is distinguished from normal lung and other lesions by five main criteria. a) polypoid projections of the mucosa, b) an increase in smooth muscle and elastic tissue within the cyst walls, c) an absence of cartilage in the cystic parenchyma, d) the presence of mucous secreting cells, and e) the absence of inflammation.

The CCAMs do not participate in normal gas exchange although there are connections to the tracheobronchial tree. It can lead to air-trapping and respiratory distress in the newborn period.

The mechanism resulting in CCAM is thought to include an imbalance between cell proliferation and apoptosis (i.e., increased cell proliferation and decreased apoptosis).

CCAM can affect all lobes and equally distributed on the right and left sides. The blood supply is from the normal pulmonary vasculature.

Stocker's classification currently has 5 types based on the size of the cyst and the cellular characteristics.[10, 11, 12.]

The initial classification included Type 1, which include large cysts composed of primarily bronchial cell type characteristics; Type 2, intermediate cysts with bronchiolar type cells; and Type 3,small cysts with bronchiolar/alveolar duct cells. Types 0 and 4 were added later, based on the site of origin of the malformation.

Rosenberg summarizes – "Type 0 CCAMs are the least common, account for less than 3% of all cases and are thought to have a tracheal origin. The cysts are very small with a maximum diameter of <0.5 cm and are lined with ciliated pseudo-stratified epithelium. Mucous and cartilage are present but skeletal muscle is absent.

Type 1 is the most common and accounts for over 65% of CCAMs. This type is thought to be of distal bronchial or proximal bronchiolar origin. The cysts are 2–10 cm in diameter and may be single or multi-loculated. These cysts are lined with ciliated pseudo stratified columnar epithelium and contain both smooth muscle and elastic tissue. In a small number of cases, there are occasional islands of abnormal cartilage. Mucus producing cells are present in approximately one third of cases. Adjacent tissue

tends to be relatively normal but the cysts can cause significant compression of normal lung tissue and mediastinal shift.

Type 2 accounts for 20–25% CCAMs and are composed of multiple smaller cysts 0.5–2 cm in diameter. The cysts are primarily bronchiolar in origin and are lined with ciliated cuboidal and columnar epithelium. Mucus secreting cells and cartilage are absent. There is usually little mass effect on surrounding normal lung. Interestingly, Stocker noted that type 2 CCAMs were associated with other congenital anomalies in up to 60% of cases. These included esophageal atresia, renal agenesis, intestinal atresias, and other defects.

Type 3 CCAMs are usually very large and can affect the entire lobe or multiple lobes. This type arises from alveolar duct cells and accounts for less than 10% CCAMs. They consist of either a combination of cystic and solid elements or appear completely solid. Because of their large size and poor differentiation, these CCAMs are thought to develop in early gestation, before 4 weeks. The cysts are <0.5 cm and lined by nonciliated cuboidal epithelium. Mucus secreting cells and cartilage are absent.

Type 4 CCAM is rare, accounting for only 2–4% of CAMs.12 The cysts can be as large as 7 cm and consist of nonciliated, flattened, alveolar lining cells lacking mucus and skeletal muscle cells. These CCAMs are thought to have an alveolar or distal acinar origin."[9]

The diagnosis of CPAM in adults requires a high degree of clinical suspicion. CPAM diagnosed in adults are result of an earlier misdiagnosis, partial excision in childhood or nonoperative observant management of a detected lesion.[13]

Most cases of Types 1 and 3 can be diagnosed during routine antenatal ultrasonography. Sonologically, Type 1 lesions appear like a mass with mediastinal shift and shrinkage in cyst size is noticed with advancement in pregnancy, whereas Type 3 lesion appears non-cystic and does not collapse as pregnancy progresses.[14, 15.]

The findings on ultrasound ranges from an incidental finding of a cystic lesion to massive pulmonary involvement with the development of hydrops.[16]

Type 1 lesion is the most commonly identified variant in chest radiograph and single large cyst is the most unusual of all presentations whereas Type 2 lesion may not be visible in chest radiograph.[14, 17.]

CT scan is the investigation of choice for the diagnosis of cystic lesions of lung as it identifies the cysts that are not detected in chest radiographs, delineates the nature and extent of cysts and also detects any other associated congenital anomaly.[18]

The radiological differentials in this case include post-infectious pneumatocele, giant bulla, pulmonary sequestration, congenital lobar emphysema and bronchogenic cyst.

The large size of the cyst, absence of preceding lung infection and healthy surrounding parenchyma exclude the possibility of a pneumatocele.

The age of the patient, non-smoker status, absence of emphysema or abnormality suggesting a previous tuberculosis insult lessens the possibility of a bulla. Cystic bronchiectasis is unlikely considering the solitary nature, lower lobe location and lack of recurrent lung infections.

Treatment for both symptomatic and asymptomatic patients is surgical resection. Surgical resection is the treatment of choice in all patients including those who are asymptomatic considering the risk of recurrent infections. If CCAM is detected antenatally or at birth, surgical resection is done between 1 and 6 months of age as the scope for compensatory lung growth after resection is much better in infants compared to adults.[9]

The standard therapy has been a posterolateral thoracotomy or median-sternotomy with intrapulmonary cysts requiring segmental or lobar resection.[9]

Histopathological Examination in our patient revealed hyalinised collagen tissue devoid of any lining epithelium except a localized area having squamous epithelium and syncytium with multiple nuclei.The underlying lung parenchyma showed dilated bronchioles, normal alveolar spaces and focal lymphocytic infiltration.

CONCLUSION

Outcome for CCAM depend largely on the type of lesion. Type 1 lesion has excellent outcome if operated in the neonatal period. Type 4 also has a favorable outcome. Type 2 and 3 have less favorable outcome.

It is rare to come across a completely air filled bronchogenic cyst in adults and this condition has to be differentiated from other lesions especially from tubercular cavity and bulla.

Surgery is the mainstay of treatment. Complete resection is necessary to avoid recurrence and the possibility of missing malignancy. The excised cyst has to be examined for malignancy.

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