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Adult Presentation of Congenital Cystic Adenomatoid Malformation of the lung : A Case Report

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Adult presentation of congenital cystic adenomatoid malformation(CCAM) of the lung is so rare that only 5 cases have been reported in the literature to date. We report the case of a 19-year-old female with CCAM in the left lower lobe. Computed tomography showed a multilobulated cystic lesion with multiple air-fluid levels and also showed focal enhancement of the solid component in the eccentrical portion of the lesion. Thoracic aortogram and selected bronchial arteriograms show a slightly enlarged and tortuous bronchial artery feeding the multilobulated cystic lesion.
We present the clinical and radiological features of our case with a brief review of the literature.

Key Words : *Cystic adenomatoid malformation lung, Lung abnormalities, Lung cysts, Lung computed tomography, Lung angiography*

INTRODUCTION

Congenital cystic adenomatoid malformation of the lung(CCAM) is a developmental anomaly characterized by proliferation of terminal bronchiole-like interconnecting structures with formation of cysts of varying sizes. CCAM accounts for approximately 25% of congenital lung lesions(Avitabile et al., 1984). The vast majority of the patients with CCAM present with signs of respiratory distress in the immediate neonatal period, but antenatal diagnosis is often possible and late presentation in older children and adults sometimes occurs(Stocker et al., 1988). Late presentation of CCAM usually involves patients with history of recurrent pneumonia(Hulnick et al., 1984 ; Pulpeiro et al., 1987)

In our review of the English literature, 5 cases in which the patients were older than 15-years at the time of diagnosis were reported(Avitabile et al., 1984 ; Hulnick et al., 1984 ; Pulpeiro et al., 1987). We report a 19–year-old female patient with CCAM which was confirmed by operation.

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CASE REPORT

A previously healthy 19-year-old female was admitted to the hospital for evaluation of a round soft tissue mass with an air-fluid level in the left lower lobe of the lung. This lesion was first detected 3 weeks previously. The patient had suffered from upper respiratory infection intermittently for the previous 2 years.

On admission, the patient had cough, sputum production, and mild fever. The physical examination was unremarkable. The routine laboratory examination revealed leukocytosis. The culture and smear of the sputum for acid-fast bacilli failed to reveal any Mycobacterium tuberculosis. The bronchoscopic examination was unremarkable. Pulmonary function tests showed a mild obstructive and

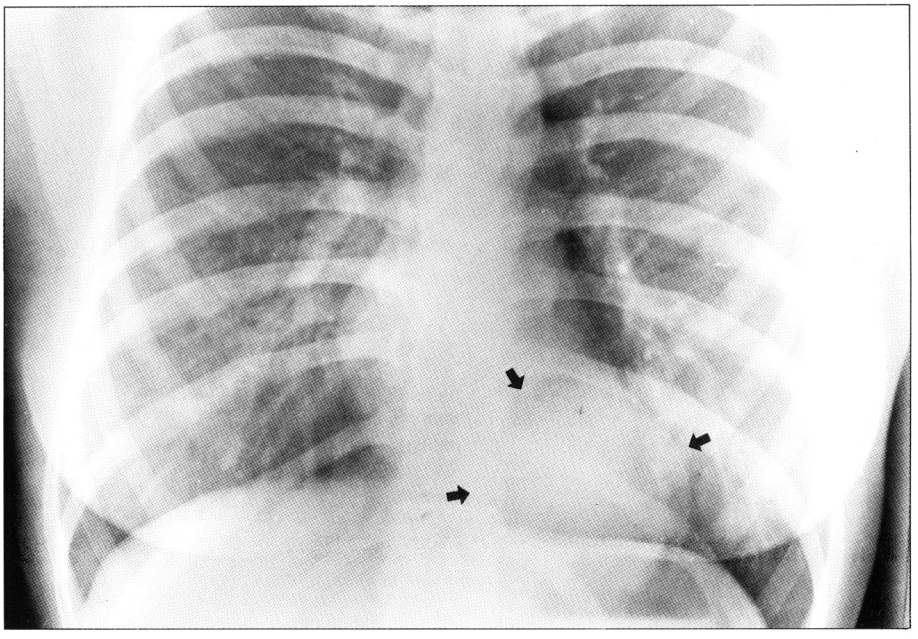

Fig. 1. Radiograph obtained 3 weeks prior to admission demonstrates a homogeneous mass density in the left lower lobe(arrows).

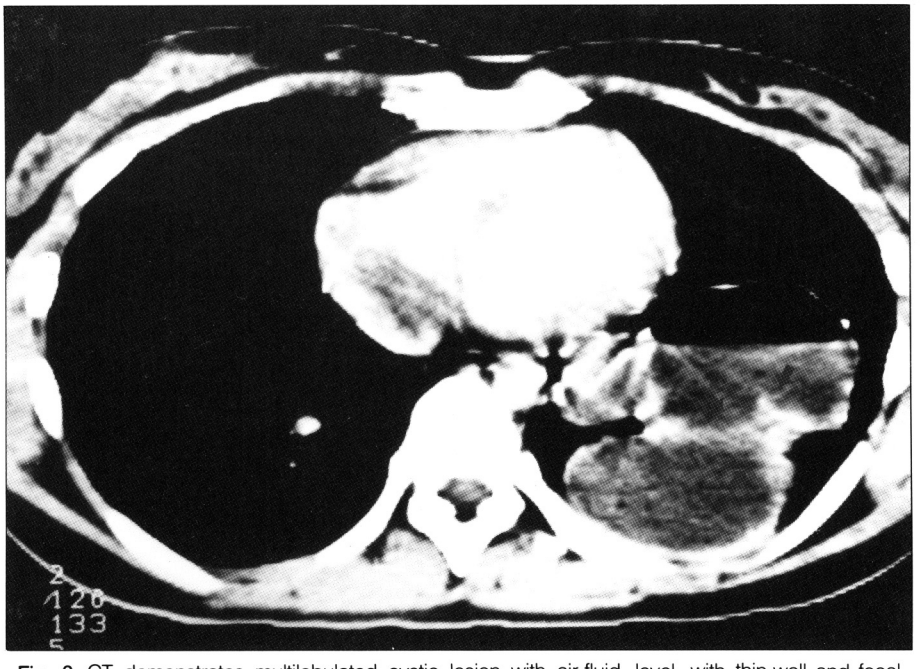

Fig. 2. CT demonstrates multilobulated cystic lesion with air-fluid level, with thin-wall, and focal enhancing solid component(arrows).

restrictive pattern.

Chest radiographs showed a round, soft tissue mass with an air-fluid level in the basal segment of the left lower lobe. The round soft tissue mass had been demonstrated on the chest radiographs taken at another hospital 3 weeks before, bur air-fluid level was not present at that time(Fig. 1). She was treated by antibiotics(Ceftezole 2.0g IV q 12hr., tobramycin 80mg IM q 12hr., clindamycin 1.2g) for 11days. Eleven days after admission, the mass changed to a cystic lesion having multiple air-fluid levels. After postural drainage, the air-fluid levels disappeared. Computed tomography(Somatom Hi-Q, Simens, Germany) taken 4 days after admission showed a multilobulated cystic lesion having multiple air-fluid levels with thin walls which were enhanced after injection of contrast media. A focal enhancement of the solid component in the eccentrical portion of the cyst was noted(Fig. 2). There was no evidence of lymphadenopathy in the thorax. Thoracic aortogram and selected bronchial arteriograms showed a slightly enlarged and tortuous bronchial artery feeding the multilobulated cystic lesion(Fig. 3).

The contrast media drained into the left atrium via the left superior pulmonary vein. A pulmonary angiogram showed displacement and stretching of the pulmonary arteries or vessels due to the mass effect of multilobulated cystic lesion.

During surgery, multilobulated, two cystic lesions were found in the left lower lobe, and severe adhesive changes in the diaphragmatic and posterior aspect of the left lower lobe were noted. An abnormal arterial supply to the lesion was not indentified during the operation. A left lower lobectomy was performed.

Grossly, the mass was composed of two large cysts(6 cm X 5 cm and 5 cm X 5 cm in diameter) occupying the resected left lobe(20% of left lower lobe) and innumerable cysts with 0.1 to 0.7 cm diameter around the large cysts. The cysts were expanded by air and there was no bronchial communication within the cysts. The walls of the large cysts and the surrounding solid areas were yellowish, fibrotic and intermixed with inflammatory exudates. Histologically, the greater part of the epithelial lining of the large cysts was denuded, but the remaining portions revealed pseudostratified ciliated columnar epithelial lining. Sections from the solid areas adjacent to the large cyst showed a network

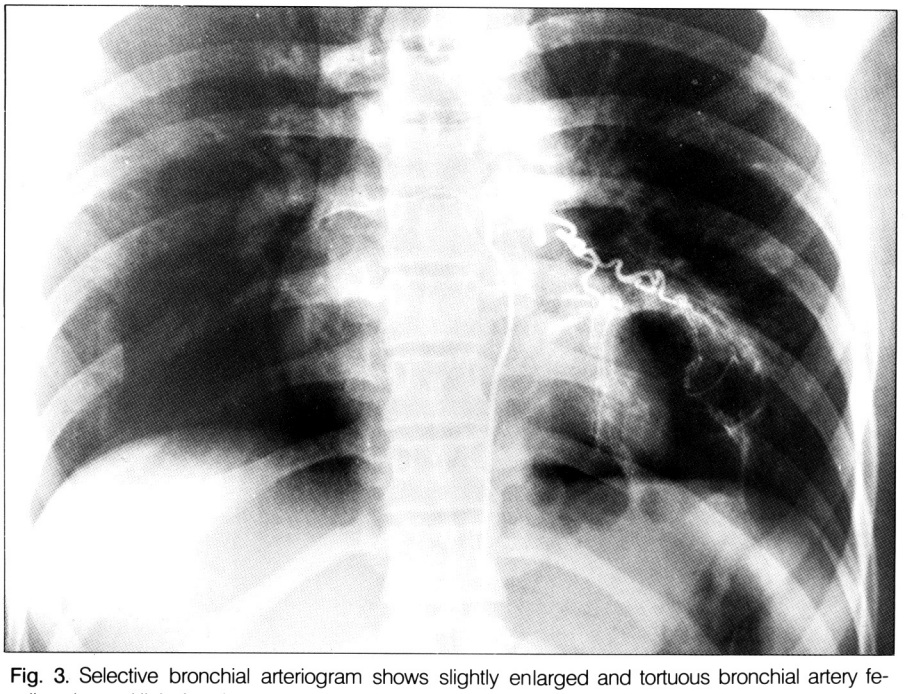

Fig. 3. Selective bronchial arteriogram shows slightly enlarged and tortuous bronchial artery feeding the multilobulated cystic lesion.

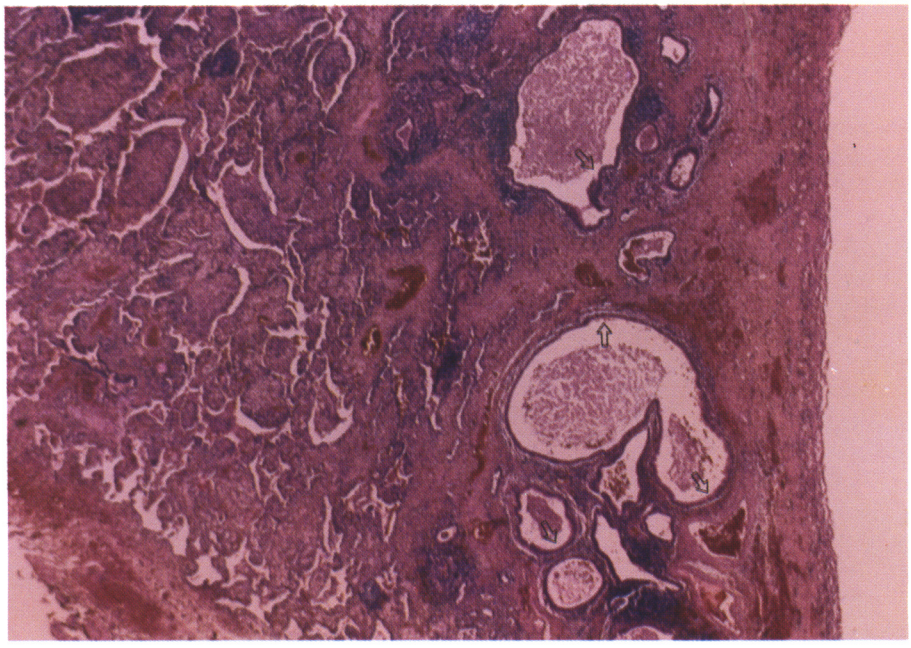

Fig. 4. Microphotograph of CCAM showing multiple irregular variable sized cystic structures with bronchial epithelial lining(arrows). (H-E stain, ×20)

of interconnecting, irregular, variable sized bronchiole-like spaces lined by cuboidal to pseudostratified ciliated columnar epithelium. The supporting stroma was composed of loose connective tissue containing numerous bundles of smooth muscle and thick walled blood vessels, and was infiltrated by exuberant histiocytes and lymphoplasma cells. No cartilage plates or mucous cells were present in any part of the lesions(Fig. 4). The final pathologic diagnosis was Stocker's type I CCAM of the lung.

DISCUSSION

Congenital cystic adenomatoid malformation is characterized by anomalous fetal development of terminal respiratory structures, resulting in an adenomatoid proliferation of bronchiolar elements and cyst formation(Stocker et al., 1988). CCAM typically is restricted to all or part of one lobe, without preference for either side or any lobe. The right and left lung are affected almost equally, with lower lobe involvement slightly more prominent(Stocker et al., 1988). The lesion affects males slightly more frequently than females and has no racial predilection-(Stocker et al., Hulnick et al., 1984 ; Pulpeiro et al., 1987). In generally, communication with the normal bronchial tree and arterial supply and venous drainage of CCAM are normal, although anomalous arterial supplies and venous drainage have been reported(Rashad et al., 1988 ; Hutchin et al., 1971).

Fluid accumulation may transform air-filled cystic spaces into areas of homogeneous density, and conversely, reestablishment of bronchial pathways or progressive air trapping may transform solid-appearing areas into air-filled cysts(Tucker et al., 1977). In our case, the initial homogeneous density transformed into a multilobulated cystic lesion with air-fluid level, and then into a multilobulated air-filled cyst after postural drainage with the prone position. It may be presumably due to reestablishment of bronchial communications between the malformation and the normal adjacent lung after antibiotic treatment. Increased intrathoracic pressure reestablishes bronchial pathways between cysts and the normal adjacent lung and collateral ventilation through the pores of Kohn(Madewell et al., 1975).

The radiographical findings in CCAM may include a multicystic appearance, a dominant cyst in a multicystic background, or a solid homogeneous mass(Madewell JE et al., 1984). Radiologic findings

of CCAM were altered by the presence of superimposed infection. CT may show a multicystic lesion with multiple air-fluid levels and focal enhancement of the solid component in the eccentrical portion of the cyst(Shackelford et al., 1989). The arteriography is often performed preoperatively in order to rule out sequestration, which can show the aberrant blood supply. The blood supply to the involved lobe is usually derived from the pulmonary artery in CCAM(Hutchin et al., 1971 ; Tucker et al., 1977). In pulmonary sequestration, supplying vessels are most frequently derived from the abdominal aorta, and rarely from the aortic arch, the intercostal artery, or one of the branches of the celiac artery(Felker et al., 1990). To the best of our knowledge, pulmonary sequestrations supplied by the bronchial artery have not been reported. The area of chronic or recurrent pulmonary infection usually receives its blood supply from the bronchial or intercostal arteries(Takahashi et al., 1975). The presence of anomalous systemic arterial supply to the lower lobe lesion favors the diagnosis of pulmonary sequestration but does not exclude CCAM as CCAM also can have a systemic blood supply in rare cases(Rashad et al., 1988 ; Hutchin et al., 1971). In equivocal cases the diagnosis can only be established by pathologic examination of resected tissue(Shackelford et al., 1989). Type I CCAM, the large-cyst type, contains one or more cysts varying from 2 to 10cm in diameter, surrounded by multiple smaller cysts. The larger cysts are lined with ciliated pseudostratified columnar epithelium and often display a papillary or polypoid appearance because of the presence of elastic tissue beneath the epithelium.

In addition to elastic tissue, the cyst wall contains thin to broad bands of smooth muscle and fibrovascular connective tissue. The smaller cysts resemble dilated bronchioles and are lined by cuboidal to columnar epithelium overlying a thin fibromuscular wall. Alveolar ducts, alveolar saccules, and alveoli are interspersed among the cysts. A unique findings of the type I lesion is the presence of clusters of mucogenic cells amid the lining of the large cysts or within alveoli adjacent to the cysts. In intralobar sequestration, the cut section reveals a consolidated parenchyma that frequently contains a single or multiple cysts varying in size from a few millimeters to 5 or more cm in diameter and filled with thin to viscoid yellow-white fluid or gelatinous material. Microscopically, chronic inflammation and fibrosis replace the normal pulmonary parenchyma. The cysts noted grossly are lined by cuboidal, columnar, or rarely, squamous epithelium and are filled with amorphous eosinophilic debris and/or foamy macrophages. Remnants of bronchi and bronchioles are surrounded by fibrous connective tissue infiltrated by lymphocytes, plasma cells, and macrophages. Remnants of alveolar ducts and alveoli are present as cuboidal epithelial-lined structures amid loose-to-dense connective tissue infiltrated by inflammatory cells. The finding of irregular proliferation of bronchiole-like structures, a feature that is routinely found in CCAM, is not noticed a intralobar sequestration(Stocker et al., 1988).

In summary of our case, radiological studies showed a multicystic lesion with multiple air-fluid levels and focal enhancement of the solid component in the eccentrical portion of the cystic lesion which was supplied by the bronchial artery. The patient had a clinical history of recurrent pneumonia with changing contour and volume in the same location of the lung.

Late presentation of CCAM may be quite similar to the pulmonary sequestration clinically and radiologically.

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