Case Report

Cystic Adenomatoid Malformation In A Young Male Adult With Recurrent Respiratory Tract Infections

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Abstract: We present a case of 26-year-old male with congenital cystic adenomatoid malformation. His medical history started after birth as recurrent respiratory tract infections and breathlessness during his infancy which persisted even after continued medical treatment. Cystic bronchiectasis of right lower lobe was suspected. On cross-sectional imaging at our hospital, congenital cystic adenomatoid malformation of right lower lobe was diagnosed. The clinical features, radiological characteristics, differential diagnosis and prognosis of the disease are discussed.

Introduction

Cystic disease of the lung, in various forms, has been well-described for over a century. Congenital cystic adenomatoid malformation (CCAM) of the lung was recognized as a distinct entity of cystic lung disease by Staerk in 1897,[1] but remains a rare disorder. It is considered a developmental disorder and most of the cases reported have been described in premature or stillborn infants.[1-4] This disorder has also been described in older children and, rarely, in adults in whom the disease has been localized to one lobe or at most one lung.[5-12]

History

A 26 years old young shopkeeper presented to our hospital in Dec. 2012 with complaints of recurrent respiratory tract infections and occasional breathless in association. His chronic cough was productive of greenish sputum along intermittent pleuritic chest pain. No obvious triggering factors for his respiratory symptoms were found. He had a good appetite, and no progressive weight loss or any other significant systemic symptoms, such as fever associated with night sweats. He was a non-smoker and non-alcoholic. There was neither a history of atopy nor a familial history of respiratory disease. There was no history of significant dust or chemical exposure in his workplace.

On physical examination, he was a heavy built male besides normal vitals; His chest was normal on inspection, but revealed prominent inspiratory and expiratory wheezing over right lower chest. The apex beat was palpable and the heart sounds were normal. The examination of central nervous, cardiovascular as well as gastrointestinal system was unremarkable.

Preliminary clinical diagnosis was cystic bronchiectasis. Patient underwent radiological imaging through plain X-ray followed by cross-sectional imaging as a part of work up to confirm diagnosis.

A frontal chest radiograph showed multi-cystic change in the right lower zone besides normal cardiac size and contour. The multiple cysts were thin walled without any obvious air-fluid levels. Right upper as

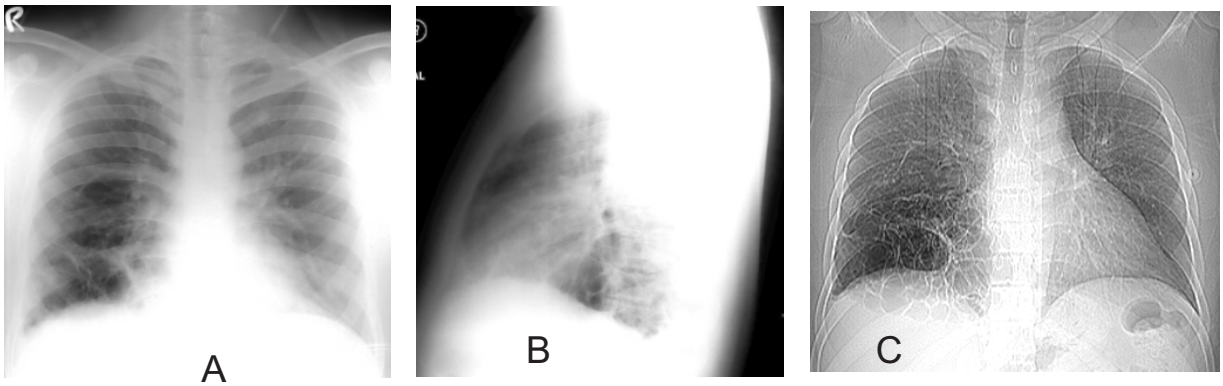

Fig-1: A frontal (a) and lateral (c) chest radiographs along with scanogram (c) showing thin walled multicystic changes in the right lower lobe (blue and white arrows) without any obvious air-fluid levels. Note that right upper as well as left hemithoraces are unremarkable for any obvious pathology.

well as left hemithoraces were unremarkable for any obvious pathological pattern. Both costo-phrenic angles were clearly outlined. Lateral chest projection confirmed the cystic changes in the right lower lobe being confined to the retrocardiac region.

High resolution computed tomography (HRCT) of the lung was ordered as a next modality for investigation. The patient was scanned from the lung apices down to the lung bases with 2 mm high resolution slices. The bronchi throughout right lower lobe showed an unusual, dilated, variable sized cystic appearance extending all the way to the periphery. These multiple cysts were thin walled without any significant intra-cystic fluid. No lung parenchyma was appreciated in the right lower lobe, however, right middle and upper lobes as well as left lung revealed normally aerated lung parenchyma. The trachea, main bronchi and cardiomediastinum were centered normally with normal anteroposterior diameter of chest.

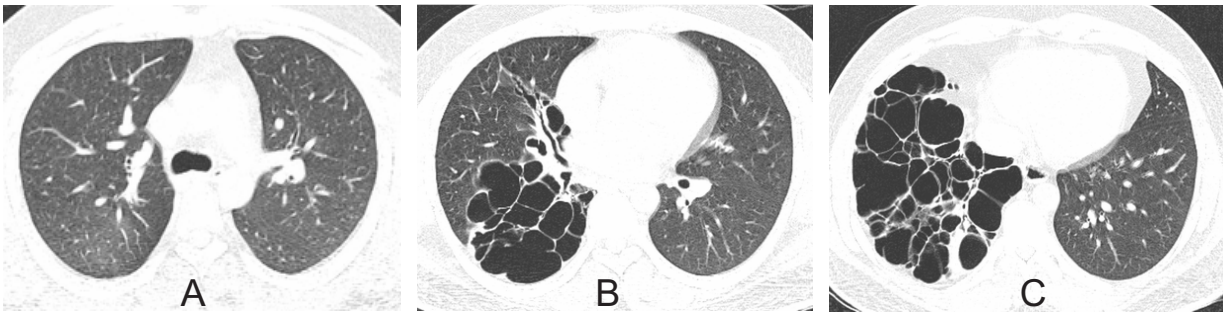

Fig-2: Axial HRCT cuts at the level of carina (a), left pulmonary trunk (b), and left ventricle (c) showing thin walled multiple cysts only involving right lower lobe. Note that right upper lobe (a) is completely spared.

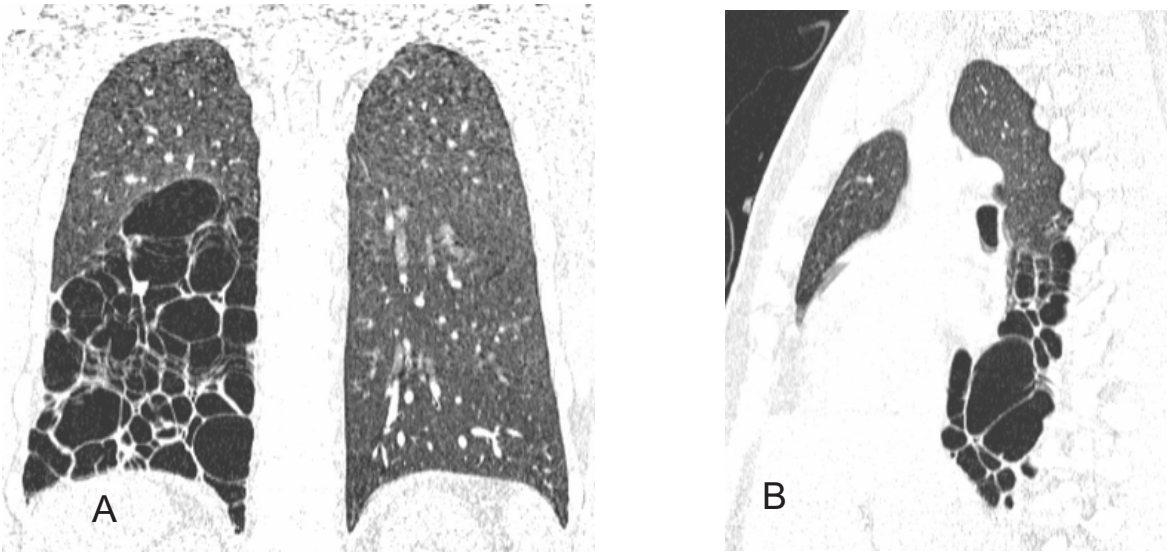

Fig-3: Coronal (a) and sagittal (b) reformatted HRCT cuts confirming multi-cysts only involving right lower lobe. Note that right upper lobe and apex is completely well preserved.

Features demonstrated by HRCT were considered compatible with a diagnosis of type I congenital cystic adenomatoid malformation (CCAM Type-I).

Discussion

Cystic lung disease in adults broadly included bronchiectasis, post-inflammatory pneumatoceles, bullous disease and cavitating lung infection. Congenital lesions, such as sequestration, bronchopulmonary- foregut anomalies and bronchogenic cysts are also encountered.[7]

The characteristic features of CCAM include "adenomatoid" increase of terminal respiratory structures, manifested by various-sized cysts, variably lined by cuboidal to ciliated pseudostratified columnar (bronchial type) epithelium or a single-layered cuboidal epithelium; polypoid configuration of the mucosa and increased amounts of elastic tissue

In the walls of the cystic portions, lined with bronchial-type epithelium; absence of cartilaginous plates in the cystic parenchyma, except as constituents of nondeformed bronchial structures entrapped within the diseased lung; the presence of a group of mucogenic cells lining the cyst wall or alveolar-like structures; and absence of inflammation.

On rare occasions, cystic dilatation and polypoid formation may be absent, adenomatoid structure being the only feature of the malformation.[2] Another rare variant is characterized by abundant cartilage on the walls of malformed bronchioles.[11]

CCAM is classified into three main types-3. Type I is composed of a single or number of large cysts with smooth muscle and elastic tissue wall; size of the cysts bein more than 20 mm. Type II variants contain numerous smaller cysts (<10 mm in diameter), with a thin muscular coat beneath the ciliated columnar epithelium; the area between the cyst is occupied by large alveolar-like structures; the lesion blends with the normal parenchyma. Type III variants occupy the entire lobe or lobes and are composed of regularly spaced bronchiole-like structures, separated by masses of cuboidal epithelium-lined alveolar-like structures. CCAM has been associated with various other congenital abnormalities, including renal agenesis, Potters' syndrome, polyhydramnios,[13] and bile duct hypoplasia.[14]

Adult cases have been diagnosed incidentally, presenting as cystic lesions on chest radiographs 10, 15. Cases have also been recognized after presenting with recurrent infection[5,11] pneumothoraces,[16] haemoptysis,[17] mycetoma,[16] and bronchioloalveolar carcinoma.[18,19]

Our report emphasizes the importance of HRCT with cystic disease, as the pathognomonic cystic bronchial wall abnormalities were not readily apparent from the plain radiograph. Due to the paucity of adult cases reported in the literature, it has not been possible to formulate any treatment guidelines. However, the appropriate treatment of complicating lower respiratory tract infection constitutes primary therapy. Lobectomy should be considered for localized disease associated with recurrent infection and haemoptysis, but it can also be recommended that this procedure should be undertaken in asymptomatic patients in view of the relatively high incidence of malignant change, as reported previously.[18,19]

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