Congenital Cystic Adenomatoid Malformation: A Sixteen-year Old Case of a Rare Clinical Entity

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Abstract
A 16- year old male was referred to a chest diseases clinic with fatigue, cough and weight loss of 10 kg within two months. His radiologic evaluation showed multiple parenchymal consolidations with air-fluid levels on the right lower lobe and he had no clinical sign of infection. The patient underwent right posterolateral thoracotomy and right lower lobectomy was performed. The diagnosis after pathological examination was congenital cyctic adenomatoid malformation (type II). This case presenting with fatigue, cough and weight loss but without any lung infections or respiratory distress is a rare form of presentation of this rare entitity.

Keywords: Congenital cystic adenomatoid malformation, lung, congenital diseases

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INTRODUCTION

Congenital cystic adenomatoid malformations (CCAMs) are lesions which have solid and cystic components with an overgrowth of terminal bronchiolar type tubular structures and a lack of mature alveoli [1]. A predominantly solid mass is usually found in the stillborn or premature infant. The combined solid-cystic lesion may produce respiratory distress in the near-term infant at birth. The primarily cystic lesion is usually found in the older infant, child or adult because of an associated unresolving or recurrent pneumonia [1]. We present a case of a 16 year old male without any pulmonary infection history who was found to have CCAM type II.

CASE REPORT

A 16- year old male was referred to the chest diseases clinic with fatigue, cough and weight loss of 10 kg within two months. His cough and associated fatigue started three weeks before and continued despite oral antibiotherapy prescribed by his prior doctor. His past medical and family history had nothing significant to note. He was not a smoker.

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On physical examination he was cachectic.Turgor and tonus were diminished. There was no peripheral lymphadenopathy, cyanosis or edema. Oropharynx was normal. His minute ventilation rate was 20. Lung sounds were coarse on the right lower chest on auscultation. No rhonchi was heard. His cardiac, abdominal and neurological examinations were within normal limits.

Laboratory findings were as follows: Leucocytes 6.33 K/uL, sedimentation rate 5 mm/h, C – reactive protein 1.4 mg/L. The sputum investigations showed no growth of a pathogenous bacteria and three consecutive direct microscopic evaluation revealed no acid resistant bacteria (ARB).

Chest X-ray showed multipl, the largest being 4 cm in diameter, parenchymal consolidations with air-fluid levels on the right lower lobe. Further investigation with a computed tomography (CT) scan of the chest had cystic formations with air-fluid levels, largest being 2.7 cm in diameter, and parenchymal infiltration of the neighbouring area in the right lower lobe (Fig. 1).

Concerning the physical examination , laboratory and radiology findings there was no clear sign of infection. The sputum investigations for ARB being negative, the patient was started on nonspesific parenteral antibiotherapy and consultated with the thoracic surgery department. The most probable differential diagnosis was a long-standing bronchogenic cyst partially controlled by prior oral antibiotherapies so surgical removal was planned. The patient underwent right posterolateral thoracotomy. A subpleural cystic lesion was seen and the polycystic mass was palpated in the lower lobe. Right lower lobectomy was performed. His postoperative period was uncomplicated and his control chest X-ray showed total clearance of the cystic lesions with good aeration.

The pathological analysis of the resected lesion revealed many cysts, the largest being 3 cm in diameter, filled with with mucus and yellow-stained material, macroscopically. Microscopically; lesion with multiple cysts, largest being 2 cm in diameter, lined by respiratory epithelial cells were detected. Some cysts showed ulceration of the superficial

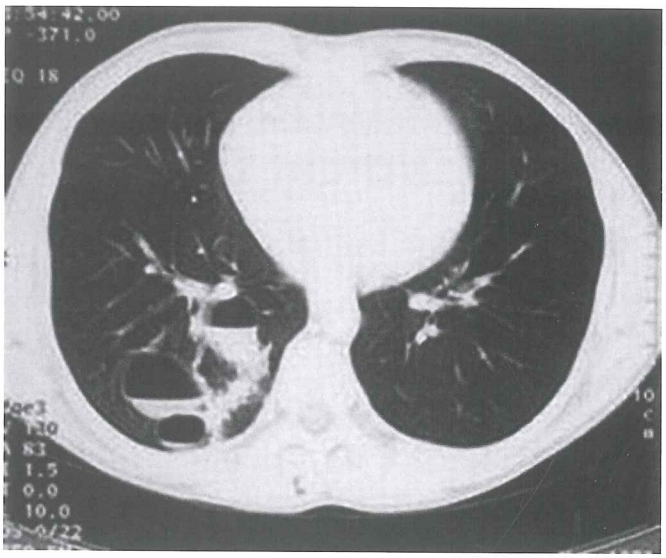

Figure 1. Thorax CT showed cystic formations with air-fluid levels, largest being 2,7 cm and parenchymal infiltration of the neighboring area in the right lower lobe.

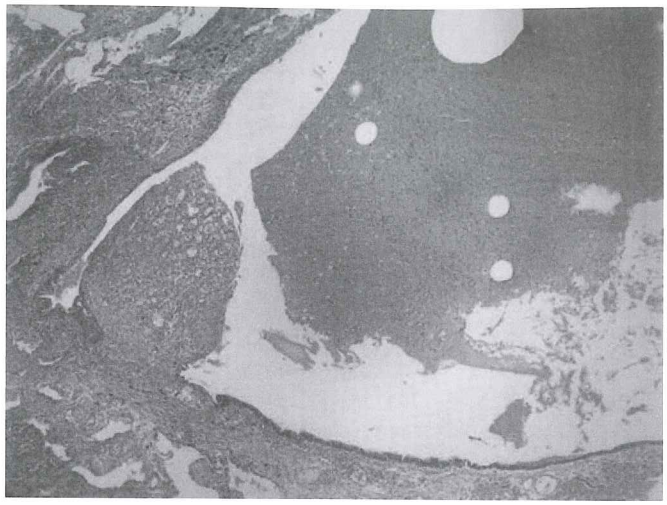

Figure 2. Cystic lesion with respiratory type epithelial lining and secretions, inflammatory granulation tissue arising from the ulseration of the cyst wall (H&E x 40).

epithelium but these areas were covered with inflammatory granulation tissue (Fig. 2). One cyst had inspissed neutrophils within. Granulation tissue plugs were apparent in the alveolar and bronchiolar sites near the cysts. These findings were consistent with CCAM type II.

DISCUSSION

The differential diagnosis of a cystic mass of the bronchopulmonary system includes bronchogenic cyst, lung abscess, bronchiectasis, herniation, atelectasis, pneumonia, pneumatocele and CCAM. .Most of these entities require signs of acute or recurrent infection with fever, cough, sputum production and associated laboratory findings. However our patient presented mainly with fatigue and weight loss. Since bronchogenic cysts tend to appear as an incidental mass in either the mediastinum or the lung and are usually asymptomatic in adults, our primary differential diagnosis happened to be a possible bronchogenic cyst. However the macroscopic and microscopic evaluation of the lung specimen revealed CCAM type II.

CCAM is a developmental hamartomatous abnormality of the lung with adenomatoid proliferation of cysts resembling bronchioles. It is believed to result from a rest in the fetal lung development where intrapulmonary bronchial system is lacking. Pathologically it is an intralobar mass of disorganized lung tissue characterized with overgrowth of bronchioles. Lesions are often cystic sometimes solid and communications with the bronchial tree is possible [1,2].

CCAM is subdivided into 3 major types: In type I, accounting for 65% of cases, there are few cysts that can reach up to 10 cm in diameter. Cyst walls contain muscle, elastic, or fibrous tissue and are frequently lined by pseudostratified columnar epithelial cells, which occasionally produce mucin, that is unique to this subtype. In type II, accounting for 25% of cases, numerous small, uniformly shaped cysts resembling bronchioles will be found with diameters ranging from 0.5 to 2 cm. Cysts are lined by cuboid-to-columnar epithelium and have a thin fibromuscular wall. Type III, accounting for 10% of cases, involves an essentially solid mass formed of very small cysts less than 0.5 cm in diameter [2,3].

CCAM is differentiated from other congenital cystic diseases by: absence of bronchial cartilage (unless it is trapped within the lesion); absence of bronchial tubular glands; presence of tall columnar mucinous epithelium; overproduction of terminal bronchiolar structures without alveolar differentiation, except in the subpleural areas; and massive enlargement of the affected lobe that displaces other thoracic structures [4].

Most of the patients present within 2 years of life. The prognosis primarily depends on the size of the lesion. Larger lesions have a higher incidence of mediastinal shift, vascular compromise, polyhydramnios, pulmonary hypoplasia, and hydrops, which may lead to intrauterine fetal demise or neonatal death [1,2,4]. If it occurs in later years of life, it is almost always associated with recurrent pulmonary infections [5-10]. The ultimate treatment remains as the resection of the effected lung tissue.

CONCLUSION

We believe that around 40 cases of late-onset CCAM cases have been reported, mostly with a history of recurrent pneumonia or lung abscess and CCAM type I being the most common histological type [5-10]. Our case presenting with fatigue, cough and weight loss but without any

lung infections or respiratory distress who was found out to have CCAM type II, is a rare form of presentation of this rare clinical entity.

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