中文題目：先天性囊腫性腺瘤樣畸形

英文題目：Congenital cystic adenomatoid malformation

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Introduction:

Patients with congenital cystic adenomatoid malformation often present with recurrent respiratory tract infections but can be asymptomatic. With improvement in imaging techniques, most diagnoses are made within first two years of life and cases of CCAM left undiagnosed until adulthood is rare. Diagnosis could be aided by CT scan, but histological findings remained the gold standard. Surgical resection is the treatment of choice in symptomatic patients to make an accurate diagnosis and preclude the potential for malignant transformation. We report a rare case of CCAM in an adult who did not present with typical symptoms of recurrent respiratory tract infections.

Case Presentation:

A 50-year-old woman presented to our hospital with the chief complaint of dyspnea on exertion for several months. She had been smoking 0.3 packs per day for more than 20 years. On physical examination, the patient's height and weight were 164.6 cm and 53 kg, respectively. Her blood pressure was 91/55 mmHg, pulse was 86 bpm, and regular, and chest auscultation showed bilateral clear breathing sounds. Chest radiography (Fig.1A) showed hyperlucency over the left upper lung field. Chest computed tomography (CT) revealed a 2.9 cm lobulated lesion in the left upper lobe with adjacent multilocular cystic lesions (Fig.1B). Pulmonary function tests revealed only mild restrictive ventilatory defects (vital capacity: 76% prediction). She underwent video-assisted thoracoscopic left upper lobe lobectomy(Fig.2A) and the pathological report was compatible with type I congenital cystic adenomatoid malformation (Fig.2B-C). No malignancy was observed in any section. The patient was free from respiratory symptoms six months after the surgery.

Discussion:

Congenital cystic adenomatoid malformation (CCAM) was considered as a lung developmental disease, which characterized by normal pulmonary alveoli replaced with dilated bronchiolar-like airspaces with no cartilage in the wall.(Kim et al., 2011) This disease was first reported in 1949 and classified into three groups based on histological appearance, (Stocker et al., 1977) and currently five main types exist. As in the case of our patient, Type I, which accounts for 70% of cases.

The incidence of CCAM is approximately 1.2 per 10,000 births.(Duncombe et al., 2002). As a developmental disorder, it is most commonly identified in the neonatal period, and with improvement in imaging techniques, most diagnoses are made within the first two years of life.(Morelli et al., 2007) Thus, cases of CCAM left undiagnosed until adulthood is rare. Chest radiographs and CT scans may reveal a lucent, multi-cystic, fluid-filled mass resembling an abscess in the affected pulmonary zone (Kim et al., 2011). CCAM is usually confined to one lobe and occurs equally in the right and left lungs, with a predilection for occurrence in the posterior basal segments of the lower lobes.(Shanmugam, 2005)

Recurrent respiratory tract infection is the most common clinical feature(Kim et al., 2011); other symptoms include spontaneous pneumothorax, hemoptysis, and dyspnea; however, it can be asymptomatic.(Morelli et al., 2007)

A CT scan provides a morphological assessment, especially when a multicystic pattern is present. For a single cystic lesion, the differential diagnosis from other cystic lung diseases, such as an abscess or pulmonary sequestration, relies largely on histological findings.

There has been no standard treatment for CCAM until now; but considering that CCAM has the potential for malignant transformation, (MacSweeney et al., 2003) surgical resection is still the treatment of choice for symptomatic patients and makes an accurate diagnosis.

Conclusion:

Patients with CCAM often present with recurrent respiratory tract infections but can be asymptomatic. Chest radiographs may reveal a lucent lesion, which is easy to be misdiagnosed as a pulmonary cyst or pneumothorax. Diagnosis of CCAM could be aided by CT scan, but histological findings remained the gold standard. Surgical resection is the treatment of choice for CCAM in symptomatic patients to make an accurate diagnosis and preclude the potential for malignant transformation. With the typical radiologic findings and proven diagnosis by histological features, we want to remind clinicians to consider CCAM as a possible diagnosis for adults with cystic lung disease.