Adult-Onset Congenital Cystic Adenomatoid Malformation in Combination with Intralobar Bronchopulmonary Sequestration: a case report

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Congenital cystic adenomatoid malformation (CCAM) and bronchopulmonary sequestration are congenital lung lesions. They are classified as separate entities. Each of them has distinct embryology, pathology, and natural history. However there are some commonalities between them. CCAM may have a pulmonary artery supply, or be supplied like a sequestration from the aorta, and histological features of both lesions may coexist [1]. CCAM is usually diagnosed before the age of 2. It is rarely encountered in adult [2]. We present an adult case of congenital cystic adenomatoid malformation in combination with intralobar bronchopulmonary sequestration.

Keywords: Congenital cystic adenomatoid malformation; Bronchopulmonary sequestration

Cystic adenomatoid malformation (CCAM) of the lung is a rare developmental abnormality of the lung and it has no sex predilection. CCAM receives its blood supply from the pulmonary circulation and is not sequestrated from tracheobronchial tree. CCAM can be classified into three major types. Type II and III lesions can occasionally coexist with bronchopulmonary sequestration [3]. In such cases, they may receive systemic arterial supply. Most cases of CCAM are diagnosed in stillborn fetus or newborn infants, but some are not detected until adult life. We present an adult case of CCAM in combination with intralobar bronchopulmonary sequestration.

CASE REPORT

A 20-year-old male came to the Chest Clinic of our hospital because of long-term productive cough. He described suffering from chronic cough with yellowish sputum and one episode of hemoptysis. He denied smoking history and other systemic disease. On physical examination, breathing sounds were decreased with mild crackle over the right lower chest. No cardiac murmur was heard. His pulmonary function test is normal. Laboratory signs of infection were absent. Findings from chest radiograph (Fig. 1) showed cystic lesions with some infiltrations at the right lower lung field. Thoracic computed tomography (Fig. 2 and Fig. 3) revealed multiple cystic lesions of variable size, ground-glass opacity and honeycombing over the right lower lobe. There was no bronchiectasis, neither systemic artery supplying these lesions. Magnetic resonance angiography (Fig. 4) revealed feeding artery from descending aorta at the level of T10-11 and draining vein into the right inferior pulmonary vein. Under the impression of bronchopulmonary sequestration, the patient was submitted to a right lower lobectomy via thoracotomy.

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Macroscopic examination revealed a solid tumor sized 5 \times 4 \times 4cm with multicystic appearance. Histologically the tumor was consisted of uniformly distributed cysts lined by columnar ciliated epithelium, foci of papillary epithelial proliferation, inflammatory cell infiltrations, and thick-walled vessels. CCAM in combination with intralobar bronchopulmonary sequestration was the pathologic diagnosis. The post-operative course of the patient was uneventful.

**DISCUSSION**

The term CCAM was first introduced by Chin’ in and Tang in 1949 [4]. CCAM of the lung is a rare lesion with incidence about 1/25,000-35,000 [5]. It is considered bronchopulmonary foregut malformations resulting from focal arrest in fetal lung development between 4th and 7th gestational week to a variety of pulmonary insults [6]. Depending on the time and type of insult, 4-26% of cases can be associated with other congenital abnormalities.

CCAM can be classified into three major types. Type I, the most common, is characterized by single or multiple large cysts measuring more than 2cm in diameter. Type II is composed of multiple small cysts not exceeding 2cm in diameter. Type III consists of large solid-appearing lesions containing microscopic adenomatoid cysts.

CCAM usually presents in children. The advent of prenatal ultrasound scanning also allows its detection in utero. Occasionally CCAM is discovered in adult, usually as a result of chronic or recurrent pulmonary infection [7].

Bronchopulmonary sequestration is also a rare congenital lung disease and has an estimated incidence of 0.15% to 1.7% [8]. It consists of aberrant lung tissue that has no normal bronchial communication and receives blood supply from one or more systemic arteries. It is subdivided into extralobar sequestration (ELS) and intralobar sequestration (ILS). ELS has its own pleural envelope. ILS is contained within the substance of the lung.
CCAM is usually diagnosed in infancy, though it has been rarely described in adults.

Patients usually present with chronic cough or recurrent pulmonary infections. The diagnosis should be suggested on CT findings of a thin-walled lower lobe complex cystic mass. According to Samuel et al. [9], CCAM occasionally coexists with bronchopulmonary sequestration. The diagnosis of CCAM in combination with bronchopulmonary sequestration should be always kept in mind. A careful search for systemic anomalous arterial supply in patients with CCAM should be performed by Doppler ultrasound or MRI. Surgical excision of this congenital lesion is recommended to prevent complications such as pulmonary infection and potential malignancy [10].

Figure 4. Magnetic resonance angiography showed feeding artery (a) of the bronchopulmonary sequestration from the aorta and draining vein (v) into the right inferior pulmonary vein.

REFERENCE

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成年發現的先天性囊泡腺瘤畸形合併肺葉內游離肺：病例報告

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「先天性囊泡腺瘤畸形」與「肺葉內游離肺」是先天性的肺部病灶。二者分屬不同的疾病，有著不同的胚胎學由來、病理學構造、與自然病史。雖是如此，二者之間還是有某些共通的特性。「先天性囊泡腺瘤畸形」的動脈供應，可以來自肺動脈，也可以像「肺葉內游離肺」一樣來自主動脈，且二者在組織學上的特質可以共存。「先天性囊泡腺瘤畸形」通常在二歲之前就會獲得診斷，故在成年人身上的鮮少發現。我們報告一成年病例同時併有「先天性囊泡腺瘤畸形」與「肺葉內游離肺」。

關鍵詞：先天性囊泡腺瘤畸形；游離肺