Pulmonary congenital cystic adenomatoid malformation accompanied with hamartoma in an adult: report of a case

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ABSTRACT

Congenital cystic adenomatoid malformation (CCAM) is mostly recognized in children and adult presentation is rare. Here, a 42-year-old male patient was presented. He had a previous history of pneumothorax, and radiologically there were multiple bullous lesions in the right upper lobe, as well as a nodular lesion of 2 cm. He underwent right upper lobectomy and middle lobe wedge resection which revealed CCAM type I and hamartoma of the lung. Less than 70 adult cases of CCAM have been reported in the literature. Lung cancer accompanying CCAM or malignant transformation in adult are rarely described. But to our knowledge, this is the first adult case of CCAM with hamartoma. CCAM should be considered in the differential diagnosis of multicystic lesions in adults with a history of pneumothorax, and the surgical resection is the treatment of choice.

Keywords: hamartoma, lobectomy, cystic adenomatoid malformation of lung
Introduction

Congenital cystic adenomatoid malformation (CCAM) of the lungs are rare congenital cystic lesions with an incidence of 1 in 25,000 to 35,000 pregnancies [1]. CCAM is firstly defined as a distinct disease by Chin and Tang in 1949 [2]. The disease was classified into 3 groups, then expanded into 5 types with a new name as congenital pulmonary airway malformation [3,4]. Most cases (80 to 85%) are recognized in the first 2 years of life, and adult presentation of CCAM is uncommon [5,6]. CCAM in adults may be complicated by pulmonary bacterial infections and accompanied by adenocarcinoma of the lung [5-7]. Here we described an adult CCAM case accompanied by hamartoma and treated with lobectomy.

Case Report

A 42-year-old non-smoker male patient presented with progressing dyspnea and dry cough. Dyspnea was mostly seen on exertion, and the patient reported that this affected his work and quality of life during the last two years. He had a history of a right pneumothorax treated with chest tube 19 years ago. On admission, the patient had normal vital signs, no fever. The laboratory test results did not show any abnormalities. Pulmonary and cardiac physical examination were unremarkable. Chest X-ray showed increased hyperlucency in the right upper zone. High resolution computed tomography (CT) of the chest demonstrated multiple bullous lesions of several diameters (2 to 7 cm), and a nodular lesion seized 2.0 x 1.4 cm in the right upper lobe (Figures 1a-c). PET-CT scan revealed a SUVmax value of 0.4 for the nodular lesion. Bronchoscopy was negative for any bronchial lesions. Since the patient had a previous history of pneumothorax, and multiple bullous lesions accompanied by a nodule of unknown pathology, a surgical resection was planned. The patient underwent a right upper lobectomy and middle lobe wedge resection without complication via a right posterolateral thoracotomy, since the videothoracoscopic approach was found unsuitable due to dense and heavy adhesions. The patient developed prolonged air leak and discharged with a Heimlich valve on the in the postoperative 9th day. The air leak dissolved and the valve was removed on the 14th postoperative day. The patient had an uneventful follow-up of 10 months without further complaints. He reported that the dyspnea resolved and he could return to his normal daily activities.

Figure 1a-b. Chest computed tomography of the patient demonstrating multiple cystic lesions seized 2 to 7 cm located at the right upper lobe, c. nodular lesion in the same lobe.

The pathological examination of bullous right upper lobe revealed multiple cystic lesions with adenomatous changes. Most cysts were lined by ciliated and some by pseudostratified columnar epithelium. The stroma around the cysts was mostly infiltrated by lymphocytes, and nearby capillaries were dilated which demonstrated chronic inflammation. There was no cartilage in the cyst walls (Figure 2a). These findings were consistent with type I CCAM. The pathological examination of the nodular lesion located nearby the bullous parts of the upper lobe demonstrated hyaline cartilage and foci of adipose tissue surrounded by cleft-like spaces lined by respiratory epithelium, indicating a pulmonary hamartoma (Figure 2b). There was no evidence of malignancy in the resected specimens.

Figure 2a. Photomicrography of the lesions demonstrating multiple varying-sized cysts lined by ciliated and pseudostratified columnar epithelium without the presence of cartilage, b. foci of adipose tissue surrounded by respiratory epithelium, indicating a pulmonary hamartoma (HEx100).

Discussion

We reported an adult case with type I CCAM based on
reported 8 patient with adenocarcinoma in their review consisting of 60 patients. Pulmonary tumors have been reported only once [10]. To our knowledge, our patient is the first case of CCAM accompanied with hamartoma.

The treatment of CCAM in adults, as well as in children, is the complete surgical resection. Lobectomy is the mostly preferred resection type, followed by segmentectomy, pneumonectomy, and wedge resection [5-8]. We performed a right upper lobectomy and middle lobe wedge resection to achieve the complete resection.

In conclusion, CCAM should be considered in the differential diagnosis of multicystic lesions located in one lobe in adults with a history of pneumothorax, although adult cases of CCAM are very rare. The surgical resection is the treatment of choice in all cases of CCAM to perform a histological examination of the lesion and to prevent potential neoplastic transformation.

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