Case report

Lobar bronchial atresia demonstrating a cystic lesion without overinflation

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Abstract

Congenital bronchial atresia (CBA) is an infrequent pulmonary anomaly characterized, in general, by a blindly terminating bronchus, mucoid impaction, and hyperinflation of the peripheral pulmonary parenchyma. We herein report an adult case of lobar bronchial atresia who showed no hyperinflation of the peripheral pulmonary parenchyma.

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1. Introduction

Congenital bronchial atresia (CBA) is a rare pulmonary anomaly characterized by a blindly terminating bronchus, mucoid impaction, and hyperinflation of the obstructed segment of the lung [1,2]. There have been many reports of atresia of a segmental bronchus with hyperinflation of the pulmonary parenchyma. However, here we report an adult case of congenital lobar bronchial atresia who showed a simple cystic lesion with no hyperinflation of the peripheral pulmonary parenchyma.

2. Case report

A 61-year-old woman exhibited a chest X-ray abnormality from her childhood with no history of any treatment. She had a history of repeated respiratory infections. A chest roentgenogram showed a homogeneous radio-opacity in the right hemithorax (Fig. 1A). She did not have any complaints or symptoms at admission. Computed tomography (CT) revealed a homogeneous cystic lesion near the pulmonary hilum showing no contrast enhancement, and no evidence of emphysema (Fig. 1B). Furthermore, the CT images also suggested the absence of the right upper lobe bronchus. Bronchoscopy revealed that the orifice of the right upper bronchus was missing, and stenosis of the right lower lobe bronchus. There was no evidence of tumors, foreign bodies, or inflammatory changes. Magnetic resonance angiography revealed no aberrant arteries from the systemic circulation. Based on these findings, we made a preoperative diagnosis of CBA.

A right thoracotomy was performed. An elastic mass lesion, soft in consistency, was found to communicate with the right pulmonary hilum, but it was clearly separated from the other two normal pulmonary lobes. The lesion received two small arterial branches directly from the right main pulmonary artery, and showed three normal-sized vessels draining into the superior pulmonary vein, and communicated with the right main bronchus by a band of fibrous connective tissue (Fig. 2). The cystic lesion was resected after dividing the above-mentioned vessels.

The cyst measured 70 mm × 65 mm × 52 mm in size, had a septal membrane and contained a brown-green jelly-like fluid. The jelly-like fluid showed a high amylase concentration (29,000 IU/L), and bacterial culture was negative. The internal wall of the cyst was lined by pseudostratified ciliated epithelium, cartilage, bronchial glands, and alveolar tissues, which showed no aeration.

3. Discussion

CBA belongs to the category of bronchopulmonary anomalies. The first case of CBA was reported in 1953 [1]. Jedertinic et al. [2] described the clinical characteristics of CBA based on the findings in 86 cases. The most common sites of CBA have been reported to be the left upper pulmonary lobe (64%), followed by the left lower lobe (14%) and the right upper lobe (12%). Although there have been many reports of
CBA, cases with congenital lobar bronchial atresia remain infrequent. Only a few cases with a lobar obstruction, including fetal dysplasia or hypoplasia, have been reported [2,3].

In our case, preoperative plain roentgenograms and CT revealed the presence of a simple cystic lesion near the right pulmonary hilum. Careful evaluation of the CT images also suggested absence of the right upper lobe bronchus, which was confirmed by bronchoscopy. Furthermore, the cystic lesion received two arterial branches directly from the pulmonary artery and drained via vessels entering the superior pulmonary vein, which completely coincided with the blood supply distribution of the normal upper lobe of the right lung. Based on these findings, the patient was diagnosed to have a congenital anomaly.

Regarding the differential diagnosis of CBA, pulmonary sequestration and bronchogenic cyst may be considered. In this case, preoperative magnetic resonance angiography ruled out pulmonary sequestration. Histopathological examination revealed the presence of alveolar tissue in the lesion, which ruled out the possibility of bronchogenic cyst. Therefore, the condition was diagnosed as congenital lobar bronchial atresia.

CBA is considered to result from an insufficient blood supply to the bronchial bud during development [4—6]. Interference with the development of the bronchial bud results in blind termination of the bronchus. In contrast, the distal pulmonary parenchyma develops normally because of the normal vascular supply. After parturition, the distal pulmonary parenchyma in CBA is often connected with other parts of the normal lung tissue. Therefore, hyperinflation of the distal pulmonary parenchyma frequently occurs because of collateral ventilation through the pores of Kohn and air trapping. Furthermore, a sufficient blood supply causes the glandular cells to secrete mucinous products. As a result, emphysematous changes and mucoid impaction frequently occur in the atretic pulmonary region. Thus, lobar bronchial atresia could be characterized by a variety of CT findings, including a cystic lesion due to mucoid impaction and hyperinflation of the distal pulmonary parenchyma.

However, hyperinflation of the pulmonary parenchyma is not an essential feature in lobar bronchial atresia, according to the lobation of the lung. In the presented case, the lesion in the right upper lobe was completely separated from the other pulmonary lobes, preventing collateral ventilation from the adjacent lung. Thus, the case exhibited only a simple cystic lesion with no evidence of hyperinflation.

Although this cystic lesion had no bronchial or parenchymal communications at all, the patient had a history of repeated respiratory infections. Preoperative bronchoscopy revealed that the right lower bronchus was stenotic, which was considered to be due to a bronchial kink because of deviation of the pulmonary lobes. Therefore, the patient might have had repeated pneumonitis of the right lower lobe. In fact, there was no evidence of infection in the cystic lesion.

Consequently, we consider proof of the presence of an atretic bronchus to be most important for the diagnosis of CBA. Recently, reconstruction of CT images or high resolution CT has been shown to be clinically useful for detecting atretic bronchi, in addition to bronchoscopy or bronchography [2,3,5,7,8]. Furthermore, thoracic surgeons should bear in mind that not only bronchogenic cysts but also lobar

Fig. 1. (A) Chest X-ray shows a well-defined tumor shadow. (B) Chest CT shows a homogeneous cystic lesion at the right hemithorax without a hyperinflation.

Fig. 2. The atretic right upper lobe completely separated from the other lobes of the lung. The arrow head indicates the arterial branch directly caused from the main pulmonary artery. The arrow indicates drainage veins.
bronchial atresia could appear as a simple cystic lesion communicating with the pulmonary hilum at surgery. Simultaneous dissection of the pulmonary vessels using auto-suture devices could result in postoperative pulmonary arterio-venous fistula. Therefore, complete evaluation of the airways and vessels, including CT, bronchoscopy, and angiography, should be performed preoperatively whenever CBA is suspected.

References


