A Rare Case of Pulmonary Bronchogenic Cyst Associated with Bronchial Atresia in the Same Lobe

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Abstract

A rare case of 49-year-old woman having pulmonary bronchogenic cyst associated with bronchial atresia in the same lobe was presented. The diagnosis was confirmed by aortography and operation specimen. Three-dimensional reconstructed images of computed tomography clearly demonstrated the difference between mucoid impaction of bronchial trees in the left S9 not communicating with hilar bronchus and the cyst in the left S10 pressing surrounding vessels.

Key words: bronchogenic cyst, bronchial atresia, mucoid impaction, computed tomography, three-dimensional reconstructions

Introduction

Bronchial atresia is a bronchopulmonary anomaly characterized by mucus-filled bronchocele, blindly-terminating segmental or lobar bronchus, and hyperinflation of the adjacent lung parenchyma (1). Pulmonary bronchogenic cyst is also a congenital anomaly of the bronchial tree, and it is asymptomatic in almost all cases, but if it communicates with the bronchial tree, respiratory symptoms such as sputum and cough appear (2). We report a rare case of pulmonary bronchogenic cyst associated with bronchial atresia in the same lobe, whose anatomical structure was depicted by three-dimensional reconstructed images of computed tomography (CT) scans.

Case Report

A 39-year-old, nonsmoking woman first visited our hospital in 1991 because of a mass shadow in the left lower lobe which had been found by chest roentgenogram in an annual health check. We first suspected that this patient had pulmonary sequestration but aortography showed no aberrant arteries. Therefore she was followed up for 10 years under a diagnosis of pulmonary cyst filled with fluid, as determined by magnetic resonance imaging (MRI). In 2001, she often complained of a large amount of brown sputum and the size and shape of the pulmonary cyst in the left S10 had been different in each examination. She was admitted again to our hospital for a newly appearing irregular shadow in the left S9 field on July 17, 2001.

On admission she was 157 cm tall and weighed 59 kg. Her radial pulse rate was 80/min and regular. There were no abnormalities in her physiological findings including auscultation of the lung. Her white blood cell count was 9,100/mm³ and the C-reactive protein concentration was 0.51 mg/dl. Serum tumor markers, such as carcinoembryonic antigen, alpha-fetoprotein, and cytokeratin 19 fragment were all within normal limits. Arterial blood gas study in room air showed PaO₂ 94.4 Torr, PaCO₂ 39.8 Torr, and pH 7.415. There was no abnormal finding in respiratory functions. Chest roentgenogram revealed a cyst in the left S10 area and an irregular shaped shadow in the left S9 field (Fig. 1). CT scan showed the left S9 to have a bronchial cast-like tree opacity, measuring about 1 cm in diameter, surrounded by segmental emphysema (Fig. 2), and this opacity had high-intensity on T1-weighted images and iso-intensity on T2-weighted images in MRI. CT scans were obtained using a multi-detector-row CT (Aquilion-multi (4PAS), Toshiba, Japan); 0.5 seconds rotations with X-ray beam width of 1 mm and a couthptop volume pitch of 0.75 at a single breath-hold. Axial images (1 mm collimation) were reconstructed to...
Figure 1. Chest X-ray film on admission shows a mass in the left S10 (arrow heads) and an irregular shaped shadow is disclosed in the left S9 (arrow).

Figure 2. Chest computed tomography scan on admission demonstrates a sharply defined mass 5 cm in diameter in the left S10 and a bronchial tree-like opacity about 1 cm in diameter surrounded by hyperlucent lung in the left S9.

Figure 3. Three-dimensional reconstructed images show that a structure like bronchial trees does not communicate with hilar bronchus of the left B9 and that the bronchogenic cyst in the left S9 oppresses the adjacent normal bronchi and vessels.

Figure 4. Macroscopic findings of cut surface of the resected specimens.
Bronchogenic Cyst with Bronchial Atresia

Figure 5. The histological findings show that the cyst surface is lined by ciliated columnar epithelium and the wall consists of submucosal lymphocytes proliferation and cartilage, associated with lymphocyte infiltration.

Figure 6. Microscopic findings of the atresia in the left S' area show that the bronchiectatic lesion is filled with mucous, surrounding emphysematous lung parenchyma.

Figure 7. Abnormal vessels (arrow) in the hilar area of the left S' lung field.

we suspected she had bronchial atresia of S' and a pulmonary bronchogenic cyst in S"10, and it was speculated that the large amount of brown sputum might result from a transient communication of the pulmonary cyst in S"10 with bronchus. She underwent left lower lobectomy and histological examination of the resected specimens revealed a cyst, measuring 45×30×50 mm, filling with brown fluid in the left S"10 area (Fig. 4). Its luminal surface was lined by ciliated epithelium and the wall contained cartilage, seromucinous glands, and submucosal lymphocytic proliferation (Fig. 5). On the other hand, histological findings in the S' area showed ectatic bronchus not communicating with any other central bronchi of the left lower lobe, surrounded by localized emphysema (Fig. 6). Furthermore, in the hilar area of the left S', winding vessels that were unidentifiable pathologically as arteries or veins existed (Fig. 7). From these results a diagnosis of pulmonary bronchogenic cyst associated with bronchial atresia in the same left lower lobe was made.

Discussion

Bronchogenic cyst and bronchial atresia co-existed in the same lobe in this case. Three-dimensional reconstructed images of CT scans easily revealed the anatomical differences between these two lesions; the bronchogenic cyst oppressed adjacent normal bronchi and vessels, and the atresia lesion was completely separated from normal bronchi. Since there were no abnormalities from order 1 to 4 bronchi in left B"b confirmed by fiberscopic bronchoscope, the interruption might occur at more distal point. It has been reported that bronchiectasis or a bronchogenic cyst sometimes appear in the distal area of the obstructive bronchus in cases of bronchial atresia (3), however, in this case the cyst and the atresia existed independently. To our knowledge, there are only 4 previous reports of bronchogenic cyst associated with but existing independently of bronchial atresia in the same lobe.
Bronchogenic cysts are believed to represent localized portions of the tracheobronchial tree that become separated from normal airways during the branching process and do not undergo further development (8). If they occur at an early stage, when there is little tissue surrounding the developing airways, the anomalous bud is likely to remain in the mediastinum. On the other hand if they occur at a later stage of bodily development, the abnormal branch is more apt to be contained within lung tissue already present and they thus have an intrapulmonary location, as in the present case (9). Histologically the cyst wall is lined by a pseudostratified, ciliated epithelium, often with a focal area of metaplastic squamous or attenuated cuboidal epithelium. In the present case we did not detect any communication between the cyst and any bronchi, but the patient sometimes complained of repeated brown sputum and the size of the cyst had changed at each examination. Therefore we speculated that this cyst temporally communicated with the normal bronchial tree.

Bronchial atresia consists of atresia or stenosis of a lobar, segmental, or subsegmental bronchus near its point of origin. Two pathogenetic theories have been proposed to explain this condition as follows: the first one is that an island of multiplying cells at the tip of a bronchial bud loses its connection with the bud itself but continues to branch independently, resulting in a normal distal bronchial branch pattern without a connection between distal and central airways; and the second is a localized intrauterine interruption of the bronchial artery blood supply resulting in bronchial wall ischemia and secondary luminal obliteration (10). The present case showed winding vessels, which were unidentifiable pathologically as arteries or veins. Although interruption of blood supply might be one of the causes of atresia, we could not confirm it from our results. Chest CT scan revealed mucous plugging in a branching pattern with associated segmental hyperinflation (11, 12). The hyperlucency was produced as a result of effective collateral ventilation, which maintained aeration of the lung tissue that did not communicate directly with central airways (13). In case where atresia or stenosis of a lobar, segmental, or subsegmental bronchus happens in adults, hyperlucency is not seen as much because each of the proposed pathways for collateral ventilation has already been completed. In children, on the other hand, distal hyperlucency accompanied with atresia is seen more often than mucoid impaction. Therefore, it was suggested that the atresia and localized emphysema in this case might have been completed during childhood.

The present case was a rare case of pulmonary bronchogenic cyst associated with bronchial atresia in the same lobe; this is the fourth case report in the English language literature. Three-dimensional reconstructed images were able to help to recognize the anatomical structures more clearly.

References