CASE REPORT

CONGENITAL BRONCHOBILIARY FISTULA IN A 65-YEAR-OLD WOMAN

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Abstract

Congenital bronchobiliary fistula (CBBF) is quite a rare malformation and the diagnosis is usually made within a few hours or years from birth because of lower respiratory diseases beginning from early infancy. Surgical repair is necessary. Of the 29 cases reported, 4 occurred in adults aged 22-32 years. We detected CBBF incidentally in a 65-year-old woman. During bronchoscopy and thoracic computed tomographic study of the pulmonary nodules, we found an accessory bronchus descending from the carina and composed of a dark green secretion that contained 10% bilirubin. Drip infusion cholangiography revealed air in the left bile duct. Cholescintigraphy showed dilatation of the left bile duct and radiotracer pooling at the top edge of the left hepatic lobe. These findings indicated a narrow fistula between the airway and biliary duct. We attributed the patient’s long survival without major complications to the narrowness of the communication. To our best knowledge, this is the fifth and oldest reported adult diagnosed with CBBF.

Key words: congenital bronchobiliary fistula, lung cancer

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Introduction

Congenital bronchobiliary fistula (CBBF) is uncommon. Only 29 cases have been reported mostly in children (1-11). Because patients with CBBF usually manifest respiratory symptoms from early infancy, the condition is diagnosed at an early age on the basis of bilious sputum and/or recurrent airway infections (2-4). Without surgical treatment, CBBF causes severe respiratory disease and the mortality is high (5). Here, we describe a case in which CBBF was not discovered until the patient was 65 years of age. Such a case is extremely rare.

Case Report

A 65-year-old Japanese woman with bronchiectasis was referred to Yatsushiro General Hospital because of abnormal chest X-ray findings suggestive of lung cancer. She had no smoking history. She complained of persistent cough and low-grade fever over a few weeks even after taking oral antibiotics. She had no history of bilious sputum. Chest X-ray showed right hilar enlargement and multiple nodules (Fig. 1). Physical examination of her chest and abdomen revealed no abnormal findings, and laboratory tests showed no

Figure 1. Chest X-ray film obtained upon admission shows right hilar enlargement (long arrow) and multiple nodular shadows (short arrows).
elevation in total and conjugated serum bilirubin (1.2 and 0.2 mg/dl respectively). γ GTP, Al-phos, AST, ALT and LDH levels were within normal ranges. Thoracic computed tomography (CT) clearly depicted an oval accessory orifice at the carina (Fig. 2a) and the presence of mediastinal air from the carina to the lower part of the mediastinum (Figs. 2b, 2c), suggesting a narrow canal. In addition, the CT scan showed a homogeneous mass, a few nodules (1.5 and 1.0 cm in diameter) in the right lower lobe (Figs. 2a, 2b, respectively) and bronchiectasis in the left lower lobe (Fig. 2c), a small nodule in the left upper lobe, and mediastinal lymph node swelling. However there was no other abnormal finding related to CBBF on chest CT. Histological examination after transbronchial biopsy showed the nodules and homogeneous mass in the right lung to be adenocarcinoma and organized pneumonia, respectively.

Endoscopic examination of the esophagus ruled out tracheoesophageal fistula. We performed bronchoscopy under fluoroscopic guidance and found an accessory orifice at the carina. We inserted the bronchoscope into the orifice, directed air through the bronchoscope to expand the lumen, and found a small number of dark greenish granules just above the left diaphragm that we fluoroscopically determined (Fig. 3). The dark greenish granules were identified as bile granules (10% calcium bilirubinate, 82% cholesterol, and 8% fatty acid calcium).

Figure 2. (a) Thoracic CT revealed a homogeneous mass (white arrow) in the right lung and an oval accessory orifice at the carina (black arrow). (b) It also revealed two nodules (white arrows) in the right lower lobe and mediastinal air (black arrow). (c) It revealed mediastinal air (black arrow) and bronchiectasis in the left lower lobe (white arrows).

Figure 3. Bronchoscopy revealed a small number of dark greenish granules (arrows), which were subsequently identified as bile granules, in the distal part of the fistula.

$^{99m}$Tc-PMT cholescintigraphy revealed markedly dilated bile ducts in the left hepatic lobe and abnormal radiotracer accumulation in the distal area of the left lobe. The area of radiotracer pooling intersected the edge of the left liver (Fig. 4), suggesting that the duct crossed the border of the liver and that the extrahepatic part of the duct continued to the thoracic orifice where the dark greenish granules were observed bronchoscopically. CT with drip infusion cholangiography (DIC) showed no contrast material in the tracheobronchial tree, but it revealed air in the bile duct. Because neither cholescintigraphy nor CT with DIC showed any other biliary tract disease, the air was thought to originate from the airways. Taken together, these findings indi-
cated communication between the carina and left hepatic lobe. Furthermore, the patient had no secondary condition that could have caused bronchobiliary fistula, such as subphrenic abscess, amebic liver abscess, other infection, neoplasm, or trauma to the biliary tracts. Thus, CBBF was diagnosed.

Because one of two nodules in the right lung, the small nodules of the left lung, and the mediastinal lymph nodes were diagnosed as metastases from primary lung cancer in the right lung, surgical therapy was not indicated. The patient therefore underwent chemotherapy for several months, but she finally died of lung cancer 28 months after the initial diagnosis of CBBF. Unfortunately, an autopsy was not performed.

During her hospital days cough and low-grade fever had recurred several times, but the symptoms partly improved with parenteral antibiotics, however the lung cancer had enlarged.

**Discussion**

CBBF is a rare condition, more common in girls than in boys, that is primarily diagnosed in early infancy at the onset of respiratory symptoms and pulmonary infection (2, 4, 6). In addition, medical therapy has proven ineffective; surgical treatment is required. Untreated CBBF, particularly CBBF without surgical treatment, may result in progressive respiratory dysfunction and a high risk of mortality (4). However, if treated correctly, CBBF has a good prognosis with an overall mortality of 25% (4). The patient described herein survived without surgical treatment for this anomaly and was 65 years of age when the CBBF was first diagnosed.

There have been 29 reported cases of CBBF since the first description by Neuhauser in 1952. Of these, only 4 were in adults, aged 22 to 32 years (5, 7-9). The present patient is the fifth and oldest patient with CBBF ever reported. In the 4 previous adult patients, CBBF was clearly visualized by injection of contrast medium into the airways or by cholescintigraphy, which was recently reported to be useful for diagnosing CBBF (10). The present case was unique in that the communication between the airway and bile duct was not visualized by cholescintigraphy. However, bronchoscopy clearly revealed the accessory orifice at the carina, and cholescintigraphy indicated involvement of the left hepatic lobe. These findings were consistent with CBBF, including features reported in the adult cases. The fistula begins at the carina or main bronchus and ends in the left hepatic lobe (4); this is not true of secondary bronchobiliary fistula (12).

It is unclear how an adult patient with CBBF, particularly our patient, could survive for such a long period without surgical treatment. We speculate that the narrow fistula between the airway and the left bile duct in our patient permitted only a small amount of bile to pour into the bronchi and trachea.

Some patients with CBBF who died even after surgery had an associated biliary malformation (4). The reported incidence of a coexisting anomaly of the common bile duct is 36.8% (4). In these cases, the fistula may be the only outlet from the left hepatic lobe. However, if a decompressing communication between the abnormal biliary tract and the normal bile ducts or duodenum exists, a good outcome can be expected. Indeed, in the present case, cholescintigraphy with 99mTc-PMT clearly showed that the abnormally dilated bile duct in the left hepatic lobe continued to the common bile duct, which probably drained into the duodenum.

It is unclear whether the bronchiectasis in our patient resulted from the CBBF. However, we believe that the CBBF may have caused bronchiectasis because the patient had no causative history of bronchiectasis such as pneumonia in infancy, chronic sinusitis and smoking (13). It is also unclear whether the CBBF or lung cancer caused the persistent cough and low-grade fever. We thought the CBBF was predominant cause of these symptoms, because they were improved by parenteral antibiotics while the lung cancer had enlarged even after the anticancer chemotherapy. Again it was also unclear whether the patient’s lung cancer was associated with CBBF, the possibility cannot be excluded because other lung malformation such as tracheal bronchus and congenital lung cyst, coexisting with primary cancer have been reported (14, 15).

In conclusion, we encountered a CBBF in a 65-year-old Japanese woman. This case was unusual with respect to the long-term survival despite absence of surgical therapy. This is the fifth and oldest adult with CBBF ever reported.

**Figure 4.** (a) 99mTc-PMT cholescintigraphy revealed a markedly dilated bile duct in the left hepatic lob continuing proximally to the common bile duct (CB) and (b) abnormal radiotracer accumulation (arrow) in the distal area of the left lobe. GB: gall bladder.
References


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http://www.naika.or.jp/imindex.html