Case report

Bronchobiliary fistula complicating oriental cholangiohepatitis

SF Howman¹, TL Feng², RS Chamberlain³, JS Groeger⁴ and LH Blumgart⁴

¹Department of Anesthesia and Critical Care, ²Department of Radiology, ³Department of Surgery, ⁴Department of Surgery,
Enid A Haupt Chair in Surgery, Memorial Sloan-Kettering Cancer Center, New York, NY, USA

Abstract

Bronchobiliary fistula in adults is a rare event defined by the passage of bile into the bronchus and the sputum (biloptysis). Typically these lesions occur in the congenital form, as a result of thoracoabdominal trauma, or in rare instances as a result of iatrogenic injury or long-standing biliary tract disease and obstruction. In this paper, we report a novel case of a fatal bronchobiliary fistula that developed in a 67-year-old Chinese male with Oriental cholangiohepatitis. To our knowledge, this is the first case report of a bronchobiliary fistula complicating the clinical management of a patient with this disease.

Keywords
Liver abscess, empyema, fistula, cholangitis

Abbreviations
ERCP = Endoscopic retrograde cholangiopancreatography, MRI = Magnetic resonance imaging

Introduction

Oriental cholangiohepatitis, also known as recurrent pyogenic cholangiohepatitis, or primary intrahepatic stone disease, is characterized by recurrent attacks of cholangitis and intrahepatic pigmented stone formation [1, 2]. The pathogenesis of Oriental cholangiohepatitis is uncertain, but chronic infestation of the biliary tree by Clonorchis sinensis and Ascaris lumbricoides, pyogenic infections, biliary stasis, and nutritional deficiency have been implicated [1, 2]. The disease is endemic to Southeast Asia, but has been encountered with increased frequency in western countries due to immigration patterns [1, 2]. We report a case of a patient with Oriental cholangiohepatitis who developed fatal pulmonary complications due to a bronchobiliary fistula.

Case Report

A 67 year-old Chinese man with known oriental cholangiohepatitis was admitted to the hospital for management of progressive ascites and nutritional support in preparation for elective resection of an hepatic abscess. His extensive surgical history consisted of cholecystectomy (1963), common bile duct exploration with stone extraction (1967), partial right hepatectomy for liver abscess (1978), and choledochoduodenostomy (1987). Eighteen months prior to admission the patient began experiencing multiple episodes of cholangitis. Endoscopic retrograde cholangiopancreatography (ERCP) revealed mucoid obstruction of the intra- and extrahepatic bile ducts, multiple intrahepatic stones, and a stricture of the left hepatic duct. Multiple percutaneous transhepatic and endoscopic attempts at stone extraction and biliary stenting were performed.

Two months prior to admission, endoscopic evaluation demonstrated a bronchobiliary fistula in which the right posterior sectoral duct communicated directly with the pulmonary parenchyma and right mainstem bronchus via a subdiaphragmatic/intrahepatic abscess cavity. Figures 1 and 2 are fat suppressed T2 – weighted sagittal and coronal magnetic resonance imaging (MRI) images obtained on admission. They demonstrated a heterogeneous mass involving both the liver and the pleural space, communicating via a broad-based tract in the middle of the right hemidiaphragm. The hepatic component consisted of markedly dilated intrahepatic ducts, a subcapsular collection, low signal areas consistent with stones or debris, and
complex loculations consistent with abscesses. Intra-thoracic extension of this lesion resulted in right lower lobe consolidation. Contiguity with the collapsed lung, while not diagnostic, suggested the possibility of extension of this abscess into the lung parenchyma.

At presentation, the patient complained of two weeks of increasing abdominal girth, fever, chills, nausea, vomiting, and cough productive of yellow sputum streaked with black. On hospital day 2, he developed acute respiratory distress, hypoxemia, and hypotension requiring mechanical ventilation and vasopressors. Chest radiography revealed a large right pleural effusion. Tube thoracostomy yielded 1.2 L of serosanguinous fluid. Upon analysis it revealed a pH 7.44, lactate dehydrogenase 93 U/L, total protein of 45 g/L, glucose of 7.94 mmol/L, bilirubin 6.84 µmol/L, amylase 13 U/L, white blood cells $154 \times 10^9$/L, and red blood cells $964 \times 10^{12}$/L. Simultaneous comparison serum levels were lactate dehydrogenase 117 U/L, total protein 82 g/L, glucose 11.66 mmol/L, bilirubin 20.5 µmol/L. Cultures of the pleural fluid, sputum, ascitic fluid, stool, and urine were negative. Blood cultures grew Streptococcus viridans. Computed tomography (CT) guided drainage of the hepatic abscess was performed on hospital day 9. Cultures of the abscess grew Candida albicans, Enterococcus fecalis, and Pseudomonas aeruginosa. Despite appropriate antibiotic therapy and intensive care support, the patient expired of progressive sepsis.

**Discussion**

Oriental cholangiohepatitis is encountered with increasing frequency in the Western Hemisphere [2]. While the pathogenesis of this disorder is unknown, chronic parasitic infestation of the biliary tree has been implicated. The resulting inflammatory and fibrotic changes of the biliary tract and hepatic parenchyma lead to stricture, stone formation, and bile stasis, which in turn, predispose to pyogenic cholangitis [1]. Bacterial deconjugation of bilirubin leads to its precipitation and repeated cycles of cholangitis and stone formation may ensue [3]. Complications of Oriental cholangiohepatitis include cirrhosis, biliary sepsis, hepatic abscesses, and cholangiocarcinoma in up to five percent of patients [2].

Non-invasive radiologic evaluation of Oriental cholangiohepatitis typically reveals dilatation of the extrahepatic bile ducts disproportionate to intrahepatic bile duct dilatation. Intra- or extrahepatic stones, segmental atrophy, and increased periportal echogenicity may be also be present. Multiplanar MRI imaging can be helpful to evaluate extrahepatic extension. Although direct cholangiography and
cholangioscopic investigations can be used to confirm these findings, due to the risk of infection and the adequacy of noninvasive imaging, these procedures are usually employed when intervention such as biliary decompression, hepatic abscess drainage, stricture dilatation or stenting is necessary.

We report the first case of a bronchobiliary fistula developing as a complication of Oriental cholangiohepatitis. This fistula may have resulted from an iatrogenic etiology, disease progression, or both. Although iatrogenic bronchobiliary fistulae are typically the result of hepatic surgery or penetrating thoracoabdominal trauma, multiple reports of bronchobiliary fistula resulting from aggressive endoscopic decompression, long-term nasobiliary decompression, and inadvertent crossing of the diaphragm during percutaneous transhepatic biliary decompression exist [4, 5]. In this case, the patient's prior hepatic resection may have caused subclinical diaphragmatic injury and predisposed him to develop this condition. Similarly, his previous percutaneous and endoscopic procedures may have resulted in direct extension of the intrahepatic abscess to the pleural space. If not iatrogenic, the bronchobiliary fistula may have developed spontaneously, secondary to infection (subphrenic or intrahepatic abscesses) or obstruction (stones and strictures). Gugenheim et al., reported sixteen cases of non-traumatic bronchobiliary fistulas with similar presentations to our patient [4]. Moreira reported 26 cases of bronchobiliary fistulae resulting from biliary lithiasis [5].

All patients evidenced bilioptysis, and 42% had an associated subphrenic or intrathoracic abscess. In this series, bronchobiliary fistula development resulted in death of half of the patients, the mechanism of which was sepsis and pulmonary complications such as diffuse airspace disease and parapneumonic effusions. Successful management of this disease depends upon early recognition and a high level of clinical suspicion. Definitive treatment consists of adequate drainage and surgical resection of the affected areas of the liver together with appropriate antimicrobial and intensive care support [5].

To our knowledge this is the first report of pulmonary complications resulting from Oriental cholangiohepatitis.

References