INTERNAL MEDICINE

□ CASE REPORT □

Spontaneous External Biliary Fistula: A Rare Complication of Cholangiocarcinoma

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Abstract

A 68-year-old woman presented with yellowish discharge oozing from a fistula opening in the upper epigastric area that had persisted for one month prior to her visit. The patient had undergone a left lateral segmentectomy of the liver ten years prior for treatment of intrahepatic duct (IHD) stones. An abdominal computed tomography (CT) scan showed focal stricture and proximal dilatation of remnant IHD and a 1 cm-sized rim-enhancing lesion located under the surgical bed of the abdominal wall surrounding the dilated remnant IHD. Despite conservative management including nasobiliary drainage, no further improvement was anticipated. Partial hepatectomy and fistulectomy were performed for pathologic diagnosis and treatment of the enhancing lesion. Histopathology revealed adenocarcinoma. In this case, cholangiocarcinoma might have arisen in association with IHD stones and then developed a choledocho-cutaneous fistula as a clinical manifestation.

Key words: choledocho-cutaneous fistula, cholangiocarcinoma

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Introduction

Biliary fistulae can be divided into two types; internal and external. Fistulous connections may occur between the biliary tract and various structures including the bowel, bronchial tree, skin and vessels. They may develop as a complication of chronic cholelithiasis, infection, peptic ulcer, or malignancy, and result from abdominal or operative trauma.

External biliary fistulae can be further subdivided based on etiology into spontaneous, therapeutic, traumatic and iatrogenic one. Iatrogenic fistula complicating biliary tract surgery is the most common cause of external biliary fistulae. Spontaneous fistulae are becoming even less common due to prompt diagnosis and expedient surgical intervention (1). Only one case of spontaneous choledocho-cutaneous fistula due to cholangiocarcinoma has been reported (2). Herein, we report a rare case of choledocho-cutaneous fistula associated with cholangiocarcinoma.

Case Report

A 68-year-old woman presented with yellowish discharge oozing from a fistula opening in the upper epigastric area that had persisted for one month prior to her visit. She complained of general weakness without fever or pain. The patient had undergone left lateral segmentectomy of the liver ten years prior for treatment of intrahepatic duct (IHD) stones. There was neither dysplastic nor carcinomatous change in the resected specimen. She stated no history of trauma. On examination, she was thin and not jaundiced. Culture and cytology of the drained fluid were negative. Blood chemistry was normal except for a mildly elevated gamma-glutamyl transpeptidase level of 138 U/L (normal range: 8-39 U/L). CT scans showed focal stricture and proximal dilatation of remnant IHD and a 1 cm-sized rim enhancing lesion under the surgical bed of the abdominal wall surrounding the dilated remnant IHD (Fig. 1a and 1b). A cholangiogram showed no leakage of contrast from the remnant IHD into the peritoneal cavity or fistula tract

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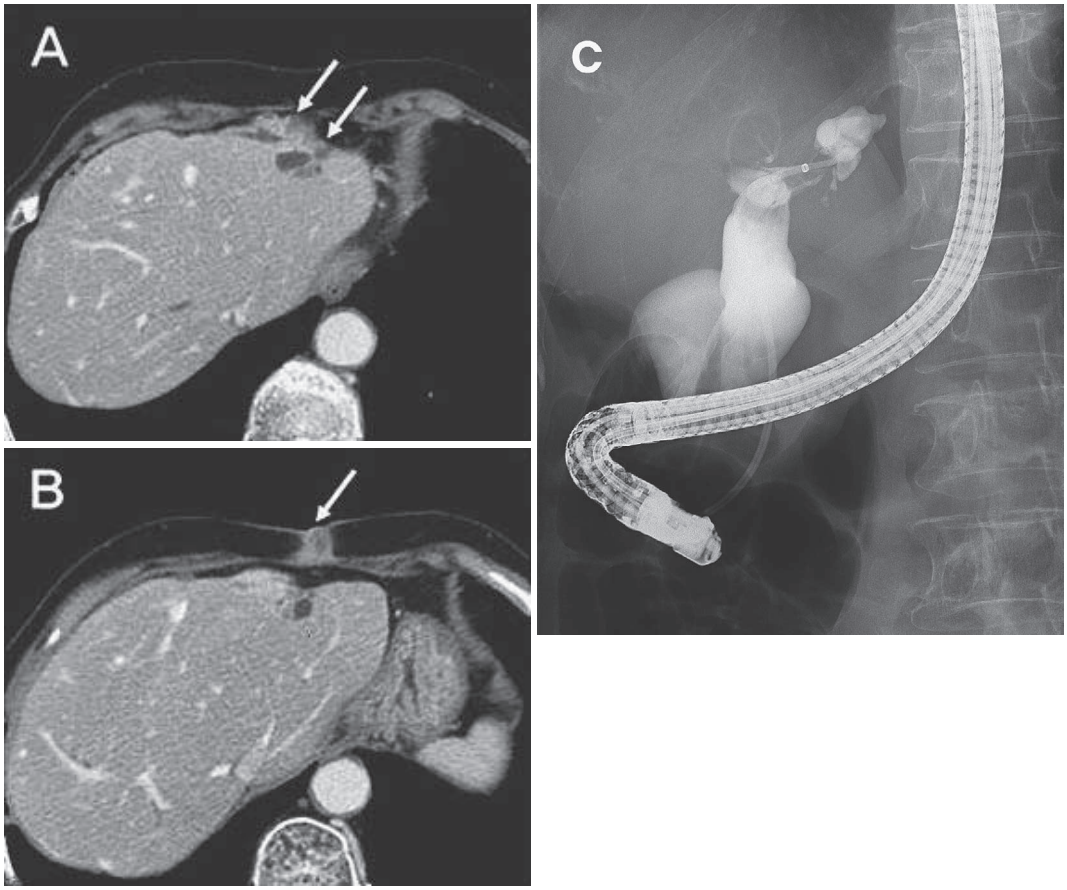

Figure 1. **A, B: Radiologic findings. (A) (B) CT scan showed focal stenosis and proximal dilatation of the remnant intrahepatic duct and a 1 cm-sized rim enhancing lesion (a, arrows) in the bed of the cutaneous fistula (b, arrow). (C) Cholangiography showed no leakage of contrast from the remnant IHD into the peritoneal cavity or fistula tract.**

(Fig. 1c). Both brush cytology at the stricture site of the remnant IHD and bile cytology were negative. Despite conservative management including nasobiliary drainage from the remnant branch of left IHD, no further improvement was anticipated. Therefore, surgery was performed for pathologic diagnosis and treatment of the enhancing lesion. On exploration, an infiltrating mass was observed on the serosal surface of the left hepatic lobe and the fistula tract between the cutaneous opening and the tumor was documented by probe insertion. Neither adhesion nor enlarged lymph nodes was detected around the omentum. Partial hepatectomy and fistulectomy were performed. Surgical pathology revealed that atypical cells forming a glandular structure, consistent with adenocarcinoma, were scattered along the fistula tract and also near the fistula opening in the abdominal wall (Fig. 2). The patient has been followed-up without relapse for eight months.

Discussion

Most spontaneous biliary fistulae are internal (cholecystoduodenal). External biliary fistula is a rare problem that usually results from a bile duct injury, undetected distal biliary obstruction, or breakdown of a biliary anastomosis (3, 4). Typically, the fistulae arise from extrahepatic ducts and rarely communicate with the IHD. Spontaneous external biliary fistulae, secondary to gallstones perforating through the abdominal wall, were common in the past (5). Since the advent of surgical treatment for hepatobiliary disease, the incidence of external biliary fistulae has markedly decreased (6).

Spontaneous external biliary fistulae are more frequent in women than men, with a ratio of 3 : 1, and they occur most commonly in the fifth to seventh decade of life (7). They are usually a complication of acute suppurative cholecystitis associated with gallstones. The suppurative process leads to necrosis and perforation, and the subsequent fistula usually moves into the duodenum (77%), colon (15%), or very rarely, through the abdominal wall. The fistula may discharge stones, pus, or bile (8).

Spontaneous external biliary fistula arising from IHD is very rare condition, and neoplasms such as cholangiocarcinoma are rare causes of such fistulae. In a retrospective review of 210 cases of external biliary fistulae over a 22-year period, only one case was due to a spontaneous external biliary fistula. This external fistula resulted from spontaneous discharge of gallbladder empyema (9). Spontaneous external biliary fistulae are easy to diagnose if the fistula is

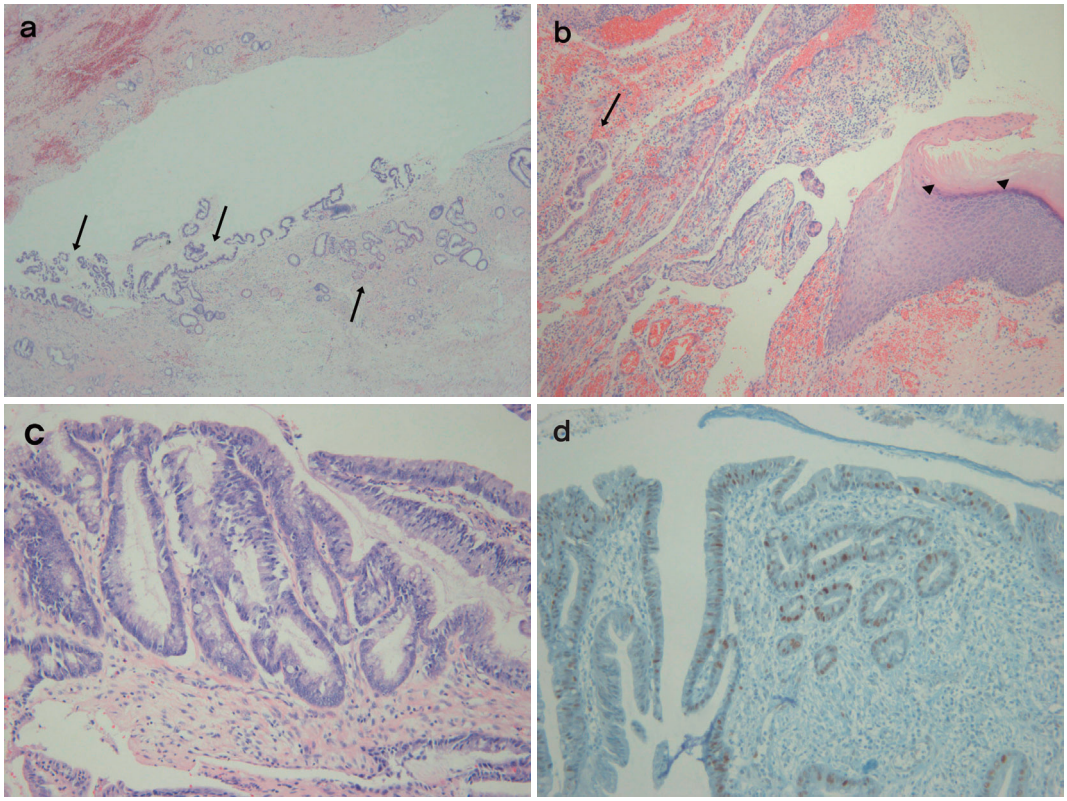

Figure 2. **a-d: Histologic findings of the resected specimen. (a) Hyperchromatic cells were arranged in irregular glandular patterns along the fistula tract, consistent with adenocarcinoma (Hematoxylin and Eosin staining, ×40). (b) Cancer cell nest (arrow) was also observed near the fistula opening in the abdominal wall that was indicated by squamous cells (arrowhead) (Hematoxylin and Eosin staining, ×100). (c) (d) High power microscopic view of carcinomatous change (Hematoxylin and Eosin staining, ×200), which was also strongly positive for p53 immunostaining (×200).**

close to the gallbladder with leakage of bile or calculi. Differential diagnosis is difficult when the fistula is more distant, and a fistulography and surgery are often needed (10). According to the literature search, there has been only 1 report of such a fistula as the presenting symptom of cholangiocarcinoma (2). In that case, hilar cholangiocarcinoma resulted in marked dilatation of the peripheral IHD and bile stasis. Then, liver abscess developed and extended into the abdominal wall, and finally induced a cutaneous fistula (2). In the present patient, cancer developed in the subcapsular portion of the remnant IHD, extended into the abdominal wall, and finally resulted in a cutaneous fistula by direct invasion.

Insertion of an endoprosthesis or nasobiliary catheter relieves pressure on the bile duct and most benign choledocho-cutaneous fistulas close within a few days (11-13). However, in the present case, the fistula leak persisted even after sphincterotomy and nasobiliary catheter insertion. The response to biliary diversion may be also useful for the differential diagnosis of fistulae. In conclusion, this case highlights the fact that cholangiocarcinoma might arise in a patient who has a past history of IHD stones and may induce a choledocho-cutaneous fistula as the clinical manifestation.

The authors state that they have no Conflict of Interest (COI).

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