CASE REPORT

Endoscopic Biliary Plastic Stenting and Successful Intentional Stent Retrieval in a Benign Biliary Stricture with Mural Spherical Calcification and Porcelain Gallbladder

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Abstract

We report a very rare case of benign biliary stricture with calcification and porcelain gallbladder, causing difficulty in differential diagnosis. A 64-year-old man was referred for further examination of jaundice. Computed tomography showed calcifications in the gallbladder wall and the common bile duct. Endoscopic retrograde cholangiopancreatography revealed narrowing and a filling defect in the distal common bile duct. Percutaneous cholangioscopy showed a protruded lesion and stricture, and pathological examinations revealed no evidence of malignancy. The stricture was resolved after temporary insertion of progressively larger of plastic stents. Patients with benign biliary stricture and/or porcelain gallbladder should be followed carefully, because malignancy can occur as a complication, although infrequent.

Key words: benign biliary stricture, porcelain gallbladder, mural calcification of the distal common bile duct, endoscopic biliary stenting, gallbladder carcinoma

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Introduction

Benign biliary strictures are caused by various etiologies and involve various sites (1-3). The benign nature and etiology of the stricture should be confirmed in order to ensure appropriate therapy. Surgical repair has been the traditionally preferred approach. With recent advances in therapeutic endoscopy, there is increased opportunity for endoscopic management.

Porcelain gallbladder (calcification of the gallbladder wall) is a rare condition with prevalence in cholecystectomy specimens ranging from 0.06 to 0.8% (4). Although various hypotheses regarding the mechanism of porcelain gallbladder have been reported, the etiology is unclear. It has been reported that porcelain gallbladder is often complicated by gallbladder carcinoma (4-10), but another study suggests a lower incidence of gallbladder carcinoma in porcelain gallbladder than previously estimated (11). Therefore, controversy remains as to whether patients with porcelain gallbladder carry a risk of gallbladder carcinoma.

We report a very rare case of benign biliary stricture of the distal common bile duct (CBD) and porcelain gallbladder, accompanied by mural spherical calcification in the distal CBD causing much difficulty in differential diagnosis from choledocholithiasis and malignancy. Endoscopic biliary stenting and intentional stent retrieval completely resolved the symptoms and abnormal laboratory data.

Case Report

A 64-year-old Japanese man with diabetes mellitus (DM) had been consulting a family physician. During medicinal treatment for DM, he became aware of general fatigue, appetite loss, jaundice, and pruritus. Blood chemistry showed increased levels of liver transaminases, conjugated bilirubin, and biliary enzymes. Because abdominal ultrasonography revealed dilatation of the intrahepatic bile ducts (IHBD), he
was referred and admitted to our hospital for further examination in February 2008. He had undergone glossectomy for lingual cancer at the age of 50, but had no past medical history of abdominal operations or trauma. He had a habit of drinking and consumed alcohol every day (25 g of alcohol/day for more than 30 years). He was well-nourished with a temperature of 36.4°C, had no tenderness or masses in the abdomen, but looked icteric.

Upon admission, blood chemistry results were as follows: total bilirubin 22.9 mg/dL, direct bilirubin 17.5 mg/dL, aspartate aminotransferase 67 IU/L, alanine aminotransferase 124 IU/L, alkaline phosphatase 1,136 IU/L, \( \gamma \)-glutamyltranspeptidase 432 IU/L, serum amylase 105 IU/L, and C-reactive protein 1.3 mg/dL. Glucose tolerance was not impaired with HbA1c level of 5.6%. Serum levels of CA19-9 and CEA were 38 U/mL (normal; 0-37 U/mL) and 2.7 ng/mL (normal; 0.5 ng/mL), respectively. Serum \( \gamma \)-globulin, IgG, and IgG4 concentrations were not elevated: 19.2% (1.34 g/dL, normal; 10.9-20.7%), 1,482 mg/dL (normal; 870-1,700 mg/dL), and 18.7 mg/dL (normal; 4.8-105 mg/dL), respectively. Serum antinuclear antibody was negative. Serum level of rheumatoid factor was 49 IU/mL (normal; 0-18 IU/mL). Calcium metabolism was not impaired with serum calcium and intact parathyroid hormone levels of 9.1 mg/dL (normal; 8.5-10.3 mg/dL) and 29.6 pg/mL (normal; 10.3-65.9 pg/mL).

Abdominal computed tomography (CT) showed flecks of mural calcification in the gallbladder wall (Fig. 1a) and another calcified lesion in the distal CBD area (Fig. 1b), which was suggestive of choledocholithiasis. Magnetic resonance cholangiopancreatography (MRCP) showed narrowing of the distal common bile duct with dilatation of the proximal bile ducts. Arrows indicate a periampullary duodenal diverticulum.

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Abdominal computed tomography (CT) showed flecks of mural calcification in the gallbladder wall (Fig. 1a) and another calcified lesion in the distal CBD area (Fig. 1b), which was suggestive of choledocholithiasis. Magnetic resonance cholangiopancreatography (MRCP) showed narrowing of the distal CBD with marked dilatation of the IHBD and proximal CBD (Fig. 2). Endoscopic retrograde cholangiopancreatography (ERCP) revealed not only narrowing but also a rounded filling defect in the distal CBD (Fig. 3). There was a periampullary duodenal diverticulum, but no evidence of pancræaticobiliary malunion, no typical signs of sclerosing cholangitis, no gallstones in the gallbladder lumen, and no abnormal findings in the main pancreatic duct. To remove the rounded defect, which resembled a choledocholith, endoscopic sphincterotomy was performed, but the filling defect did not move at all in the distal CBD. Endoscopic naso-biliary drainage (ENBD) was performed, and treatment with ursodeoxycholic acid was started. His symptoms and abnormal blood chemistries were gradually improved. Repeated post-ENBD cytological examinations of bile were negative for malignancy, and cultures were negative for bacteria. One week after placement, the ENBD catheter (7.5 Fr) was replaced by a biliary plastic stent (8.5 Fr). The stent was replaced by a larger one (10 Fr) 10 weeks after the first stenting. Concurrently with the second stenting, peroral cholangioscopy (POCS; CHF-B260, 3.4 mm diameter; Olympus Co., Tokyo, Japan) showed a whitish protruded lesion located within the portion of reddened irregular stricture in the distal CBD (Fig. 4) (which still allowed passage of the cholangioscope) and smooth nonstenosed lumen in the proximal bile ducts. POCS-directed biopsies of the stricture and whitish protruded sites revealed hyperplasia of the bile duct epithelium and no evidence of malignancy (Fig. 5a). Calcification was observed only in the whitish protruded site (Fig. 5b). Following 10 more weeks (21 total weeks of biliary tract drainage), the stent was removed, and therapy with ursodeoxycholic acid was stopped. Posttreatment MRCP suggested a certain level of improvement of biliary stricture (Fig. 6). For the 7 months following the writing of this article, the patient has been under obser-
Figure 3. Endoscopic retrograde cholangiopancreatography (ERCP). (a) ERCP revealed a rounded filling defect (arrow) in the distal common bile duct (CBD). Flecked calcifications of the gallbladder wall (arrowheads) were also observed. (b) ERCP revealed a rounded filling defect (arrow) and narrowing (arrowheads) in the distal CBD.

Figure 4. Peroral cholangioscopy showed a whitish protruded lesion located within the portion of reddened irregular stricture in the distal common bile duct.

Figure 5. Peroral cholangioscopy-directed biopsies revealed hyperplasia of the bile duct epithelium (a) with calcification (b) and no evidence of malignancy. Arrows indicate calcified deposit. (Hematoxylin and Eosin staining, original magnification ×200)
cholangitis, including abdominal pain, fever, jaundice, and elevations of biliary enzymes. Because benign biliary stricture is a rare disease, and the etiology of the stricture is diverse (1-3). Common causes include intraoperative injury, chronic pancreatitis, primary sclerosing cholangitis, recurrent cholangitis, abdominal trauma, ischemic injury, chemotherapy, radiation therapy, Mirizzi syndrome, and papillary stenosis. The clinical manifestations and findings of laboratory examinations are associated with cholangitis, including abdominal pain, fever, jaundice, and elevations of biliary enzymes. Because benign biliary stricture sometimes causes difficulty in differential diagnosis from malignancy, efforts should be made to improve accuracy of pretreatment diagnosis.

There are no pathognomonic symptoms or characteristic images of benign biliary stricture, and it appears particularly difficult to differentiate between a localized fibrotic change and malignancy using only imaging studies. Additionally, the present patient had a peculiar mural spherical calcification within the biliary stricture, closely resembling choledocholithiasis. In such cases, cholangioscopy and direct-vision biopsy are occasionally useful (12, 13). Because a hyperplastic lesion and carcinoma tissue of the bile ducts frequently exist adjacent to each other, it is often difficult to establish the diagnosis even by direct-vision biopsies (14, 15). In this patient, POCs revealed a whitish protruded lesion located within the portion of reddened irregular stricture in the distal CBD. Although POCs-directed biopsies revealed hyperplasia of the bile duct epithelium with calcification and no evidence of malignancy, the possibility of malignancy could not be completely excluded. Therefore, we must strictly follow the clinical course by laboratory examinations and imaging studies at least every 6 months in order to prevent delaying the timing of surgery.

Because the present patient also has porcelain gallbladder, the benign biliary stricture might have been caused by spreading of the inflammation of his gallbladder. However, the patient had not experienced symptoms of cholecystitis. It is uncertain whether the porcelain gallbladder was related to the benign biliary stricture and calcification described here. Although the patient had a habit of drinking and consumed alcohol every day, he did not present symptoms of, nor did clinical tests suggest, chronic pancreatitis. Thus, the cause of the biliary stricture in this patient remains obscure.

If the possibility of malignancy is excluded, benign biliary stricture may be treated noninvasively, for instance endoscopically by constructing internal fistula by insertion of a biliary stent (1-3). Although there have been no controlled trials comparing surgery with endoscopic treatment, there is growing evidence (2, 16) suggesting a long-term benefit of endoscopic treatment. Of the two types of biliary stents available, the plastic stent and the expandable metallic stent (EMS), we generally use a biliary plastic stent for the treatment of benign stricture. Although we consider that the use of EMS is contraindicated in benign biliary stricture, it is reported to be effective in some cases (17). Following a study by Hoffman et al (18), the plastic stent in the present patient was exchanged 10 weeks after insertion of the previous stent to avoid clogging and resulting cholangitis. Additionally, the stent diameter was increased with each replacement in order to gradually expand the stricture. For fear of delaying the timing of surgery, we completely removed the stent 21 weeks after insertion of the ENBD catheter. As it is difficult to differentiate benign from malignant stricture as described above, surgical resection should be considered whenever malignancy is suspected.

Porcelain gallbladder as a form of chronic gallbladder disease is rare. It is most commonly found in the sixth decade of life, with a higher prevalence in women (by about 4-5 to 1) (19). The etiology of porcelain gallbladder is still unclear. There are hypotheses that this disease is a rare manifestation of chronic cholecystitis with mural hemorrhage and subsequent calcification of the gallbladder wall (10, 20). Others hypothesize pathology in calcium metabolism (11). A possibility of a concurrence with Giardiasis has also been examined (21). Another possible sequel of chronic cholecystitis is a xanthogranulomatous inflammation (22).

The symptoms of porcelain gallbladder depend on the presence or absence of cholelithiasis and its manifestation, on the extent of wall calcification and thus the gallbladder function derangement, and on the extent of chronic inflammatory infiltration of the surrounding tissue. The last point might be relevant to the stricture and mural calcification of the distal CBD in the present patient.

In conclusion, no other report has described a case of benign biliary stricture with mural calcification and porcelain.

Figure 6. Magnetic resonance cholangiopancreatography (MRCP) after endoscopic treatment. Posttreatment MRCP suggested a certain level of improvement of biliary stricture. Arrows indicate a periampullary duodenal diverticulum.
gallbladder. Although it is not clear that porcelain gallbladder has a direct relationship with benign biliary stricture, it is possible that porcelain gallbladder is a precursor of benign biliary stricture or vice versa. If the possibility of malignancy is excluded, benign biliary stricture may be treated noninvasively by temporary insertion of a biliary plastic stent. Nonetheless, patients with benign biliary stricture and/or porcelain gallbladder should be followed carefully, because malignancy does occur, although infrequent.

References