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Mirizzi Syndrome with a Biliobiliary Fistula: a case report

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ABSTRACT

Mirizzi syndrome is a rare complication of cholelithiasis, which is defined as a common bile duct obstruction due to stones impacted in Hartman's pouch or the cystic duct of the gallbladder. The impacted stones and surrounding inflammation can lead to a biliobiliary fistula. We herein present the case of a 73-year-old Japanese man with a biliobiliary fistula that was diagnosed by peroral cholangiography (POCS). We performed partial cholecystectomy and choledochoplasty as the stone had eroded almost the entire circumference of the bile duct. Postoperative complications included a minor bile leak from the repaired common bile duct apparent on postoperative day 1, which was managed conservatively. The patient was discharged on postoperative day 9. Based on this experience, POCS is useful for detecting the existence of a biliobiliary fistula in cases of Mirizzi syndrome. Once a biliobiliary fistula is confirmed, it is important to select an appropriate surgical procedure based on the extent of common bile duct involvement in the inflammatory process.

Key words: *Mirizzi syndrome, Biliobiliary fistula, Peroral cholangiography*

BACKGROUND

Mirizzi syndrome is defined as obstruction of the extrahepatic bile duct by stones impacted in Hartman's pouch or the cystic duct. An impacted stone and inflammation sometimes lead to thinning of the wall between the gallbladder and the extrahepatic bile duct, pressure necrosis of the common wall, and finally formation of a fistula. This rare complication of Mirizzi syndrome is known as a biliobiliary fistula. This report describes the case of a patient with a biliobiliary fistula who underwent choledochoplasty using the posterior wall of the gallbladder neck.

CASE PRESENTATION

A 73-year-old Japanese man presented with pruritus and dark urine for one month and was admitted to our hospital. He had a history of appendicitis, gout, and diabetes. At the time of his initial hospitalization, physical examination revealed no remarkable findings except for jaundice. His vital signs were: heart rate, 72 beats/min; temperature, 35.8; blood pressure, 102/68 mmHg; and SpO2, 100% (room air). The laboratory data showed a cholestatic pattern including 3.4 mg/dl total bilirubin, 89 IU/liter aspartate aminotransferase, 108 IU/liter alanine aminotransferase, 813 IU/liter alkaline phosphatase, 227 IU/liter leucyl aminopeptidase, and 492 IU/liter gamma glutamyl transpeptidase (Table 1). Abdominal computed tomography (CT) with contrast showed a hypertrophied gallbladder (GB) wall, gallstones, and dilated intrahepatic bile ducts (Fig. 1). Endoscopic retrograde cholangiography (ERC) demonstrated a smooth defect on the right side wall of the common bile duct (CBD) causing obstruction. ERC also detected a CBD stone; neither the cystic duct nor the gallbladder was visualized (Fig. 2). Intraductal ultrasound (IDUS) demonstrated extrinsic compression of the upper biliary tract (Fig. 3). Biliobiliary fistula was confirmed by subsequent peroral cholangioscopy (POCS). POCS demonstrated the impacted GB stone compressing the bile duct at the level of the obstruction (Fig. 4). While the perihilar and intrahepatic bile ducts could not be ob-

Abbreviation:
CT: computed tomography, GB: gallbladder, ERC: Endoscopic retrograde cholangiography, CBD: common bile duct, IDUS: Intraductal ultrasound, POCS: peroral cholangioscopy, ENBD: endoscopic nasobiliary drainage
ERBD: endoscopic retrograde biliary drainage, MRC: Magnet resonance cholangiography

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Table 1. The laboratory data showed a cholestatic pattern

Complete Blood Count			ALP	813	IU/liter	Na	140	mEq/liter
WBC	5690	/mm^3	LAP	227	IU/liter	K	4.1	mEq/liter
RBC	365×10^4	/mm^3	γ-GTP	492	IU/liter	Cl	102	mEq/liter
Hgb	11.5	g/dl	CHE	206	IU/liter	CRP	1.11	mg/dl
PLT	17.7×104	/mm^3	LDH	188	IU/liter	Hb-A1c	7.3	%
			P-Amy	159	IU/liter			
Blood chemistry			TP	6.9	g/dl	HBs Ag	(-)	
T.Bil	3.4	mg/dl	Alb	3.9	g/dl	HCV Ab	(-)	
D.Bil	2.0	mg/dl	T.Chol	208	mg/dl			
AST	89	IU/liter	BUN	15.3	mg/dl	Tumor marker		
ALT	108	IU/liter	Cre	0.89	mg/dl	CEA	2.3	ng/ml
						CA19-9	9	U/ml

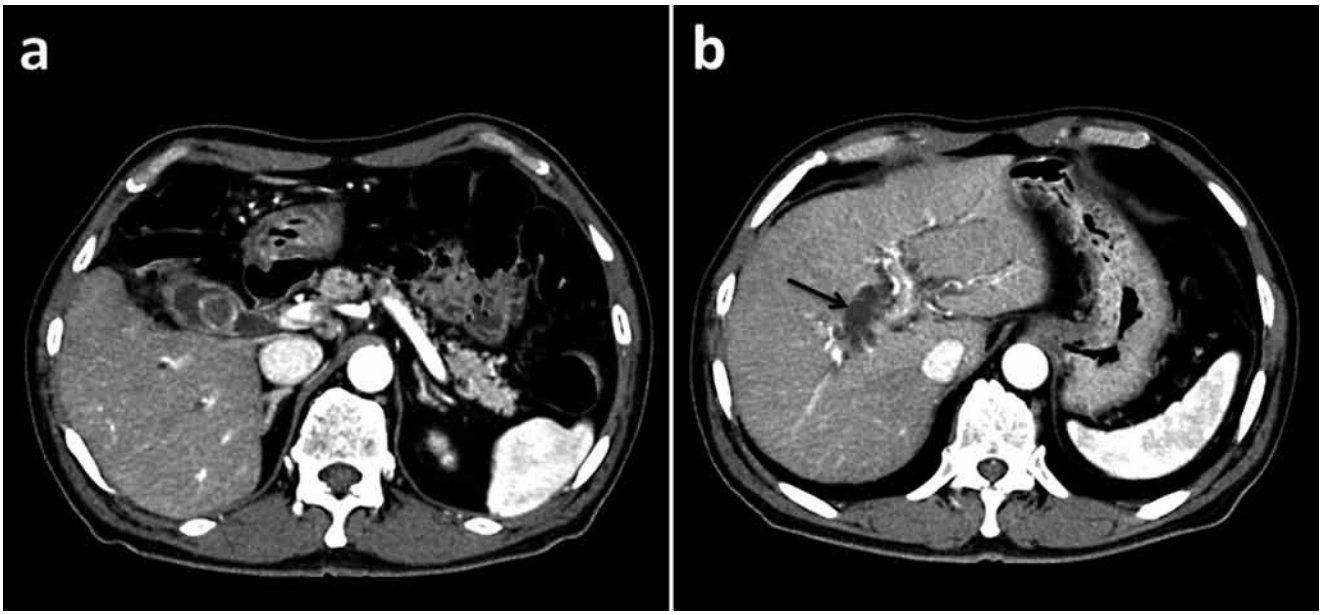

Fig. 1. Abdominal computed tomography (CT) with contrast showed (a) hypertrophied gallbladder wall and gallstones, and (b) dilated intrahepatic bile duct (black arrow).

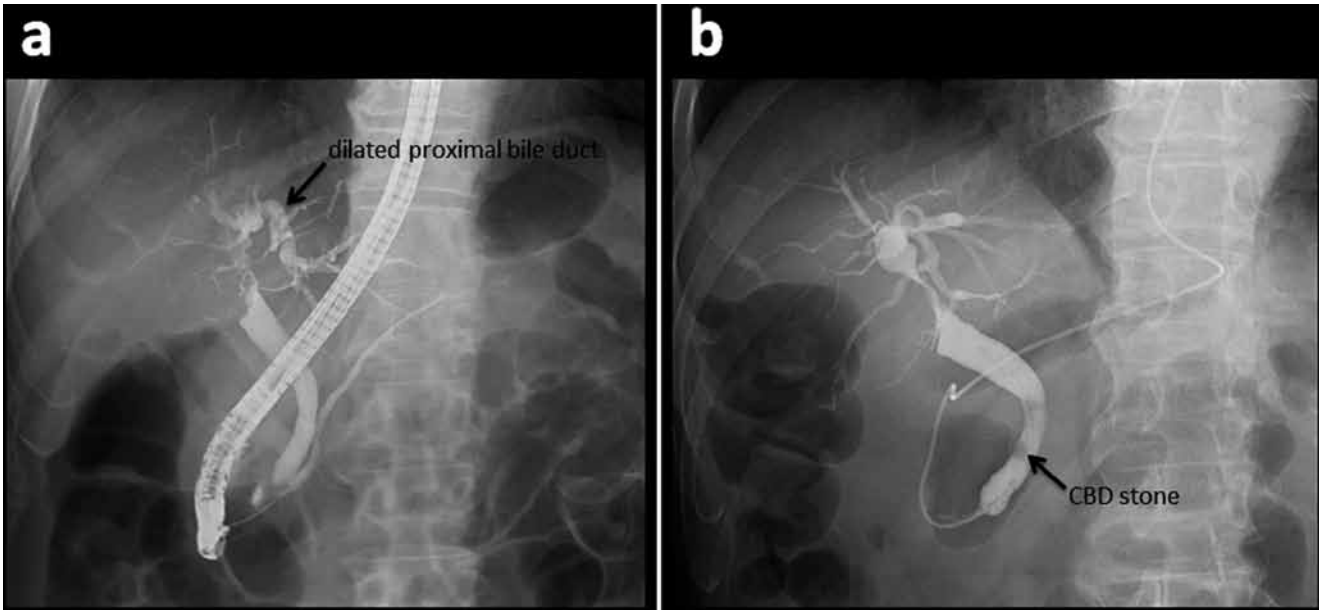

Fig. 2. Endoscopic retrograde cholangiography (ERC) showed (a) smooth defect on the right side wall of the common bile duct (CBD), and stricture of the upper biliary tract with a dilated proximal bile duct (black arrow). The gallbladder and cystic duct were not visualized. (b) ERC also showed a CBD stone in the dilated distal CBD (black arrow).

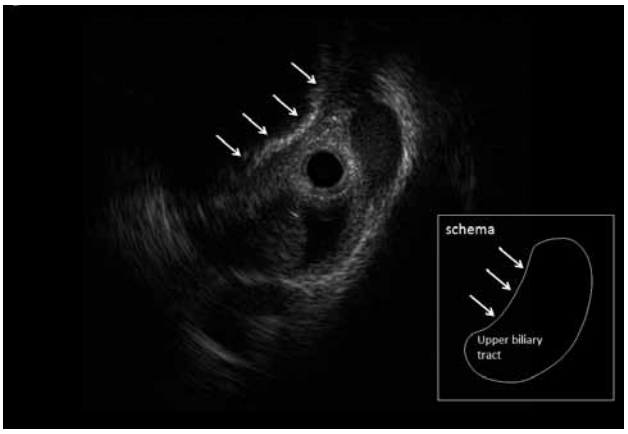

Fig. 3. Intraductal ultrasound (IDUS) showed extrinsic compression of the upper biliary tract (white arrows).

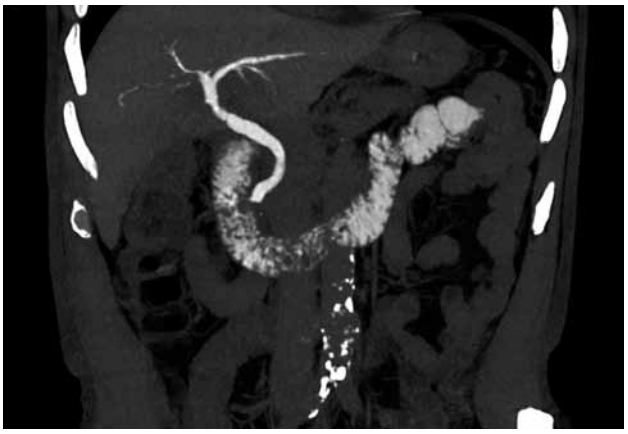

Fig. 5. Drip infusion cholangiography-computed tomography (DIC-CT) revealed no obstruction in the common bile duct.

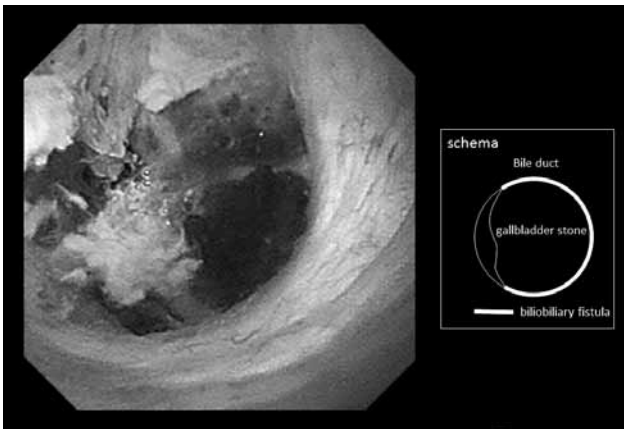

Fig. 4. Peroral cholangioscopy (POCS) showed the impacted gallbladder stone exposed to the bile duct at the level of obstruction.

served by POCS due to the obstruction, there was no evidence of an intraductal mass in the visualized portion. At this stage, an endoscopic nasobiliary drainage (ENBD) catheter was inserted to treat the choledocholithiasis. No malignant cells were found on the cytology of the bile or from brush cytology of the bile duct. After improvement of the jaundice, the ENBD was exchanged for endoscopic retrograde biliary drainage (ERBD). Based on these findings, the diagnosis of Mirizzi syndrome with a biliobiliary fistula was confirmed.

The patient subsequently underwent partial cholecystectomy and choledochoplasty. Laparotomy revealed that the GB was slightly atrophied and its wall was thickened, although there was no suspicion of malignancy microscopically. The GB was then dissected using an antegrade approach. Since the dissection of the GB neck could not be completed due to severe adhesions, the GB was opened and two 2 cm-sized stones were removed. The stone that was impacted in the cystic duct involved up to two-thirds of the circumference of the CBD and caused a biliobiliary fistula. Partial cholecystectomy was performed followed by ERBD tube removal, and the fistula was closed by continuous suture using the remaining wall of the GB neck with 4-0 Vicryl (Ethicon Inc.). At the end of surgical procedure, intraoperative cholangiography was performed showing no leakage and a patent CBD. A 19 French closed suction tube drain was placed in the subhepatic space near the bile duct. Although there was a small amount of bile in the drain on postoperative day 1, it improved by conservative management alone and the drain was removed on postoperative day 7. This was considered a Grade II complication based on the Clavain-Dindo classification[7]. On the day of discharge, cholangiographic-computed tomography revealed a wide open CBD (Fig. 5). The patient was discharged on postoperative day 9, and has been doing well without any complications for over 3 months.

DISCUSSION

Mirizzi syndrome is defined as a bile duct obstruction due to GB stones impacted in Hartman's pouch or the cystic duct, with an incidence of 0.7 to 1.4% of patients undergoing cholecystectomy[2,4,12]. Furthermore, these impacted stones and surrounding inflammation may erode into the bile duct wall causing a biliobiliary fistula. Csendes et al[4] classified biliobiliary fistula into four groups according to the extent of circumferential erosion of the bile duct. Type I is obstruction of the CBD by stone(s) impacted in Hartman's pouch or the cystic duct (11%). Type II is a biliobiliary fistula with erosion of less than 1/3 of the circumference of the CBD (41%). Type III is erosion of up to 2/3 of the circumference of the CBD (44%). Type IV is complete destruction of the CBD wall (4%). In our case, the defect in the CBD was more than 2/3 of the circumference, a Csendes type III.

Common symptoms of Mirizzi syndrome include

abdominal pain, fever, and jaundice. Some prior reports cite the presence of Charcot's biliary triad in 44 - 71% of patients with Mirizzi syndrome[5,9]. While biochemical parameters of liver function often show a cholestatic pattern, no one specific clinical or laboratory presentation has been identified in patients with Mirizzi syndrome. ERC and percutaneous transhepatic cholangiography are common and reliable methods for the preoperative diagnosis of Mirizzi syndrome. The typical findings on cholangiography are a smooth defect on the lateral wall of the CBD at the level of the cystic duct or GB neck and intra- and extrahepatic bile duct dilation proximal to the obstruction, with a normal caliber distally[15]. IDUS is an effective modality for diagnosis that commonly shows compression of the CBD from outside the wall[14]. Magnet resonance cholangiography (MRC) is almost equivalent to ERC for delineating the bile duct anatomy, and is less invasive[10]. Thus, while several modalities have been utilized to diagnose Mirizzi syndrome, it is still difficult to distinguish this entity from other causes of obstructive jaundice including GB cancer, cholangiocarcinoma, or metastatic tumor[1]. Furthermore, biliobiliary fistula due to Mirizzi syndrome is rarely diagnosed preoperatively. Hamada et al[8] reported that the preoperative diagnosis rate of biliobiliary fistula is only 8.3%, and only a few reports have demonstrated the usefulness of percutaneous transhepatic cholangioscopy for preoperative diagnosis[13]. However, it is important to detect the existence of a biliobiliary fistula preoperatively as the surgery to treat this is different than that used to treat a simple Mirizzi syndrome (cholecystectomy alone). In our case, biliobiliary fistula with Mirizzi syndrome was identified by POCS. POCS allows for examination of bile duct mucosa and, testing with brush cytology and biopsy, useful for detecting malignancy. Therefore, based on our experience, POCS should be considered to identify the existence of a biliobiliary fistula when Mirizzi syndrome is suspected.

The treatment of a biliobiliary fistula is by surgery. Mirizzi syndrome is frequently accompanied by severe pericholecystic adhesions, a shrunken and sessile GB, and an obliterated Calot's triangle. Therefore, intraoperative cholangiography should be performed in the early part of surgery to help understand the biliary anatomy[3]. A partial cholecystectomy with an antegrade approach is recommended when a biliobiliary fistula is suspected or confirmed[3]. Opening the fundus of the GB followed by removal of the impacted stones makes it possible to visualize the presence of a biliobiliary fistula. In the case of a biliobiliary fistula, complete cholecystectomy is difficult, and forcible dissection of the cystic duct may cause common bile duct injuries. Csendes et al[4], who classified Mirizzi syndrome into four types based on the extent of CBD circumference erosion, recommended that a different procedure should be applied to each type. Namely, Type I - partial cholecystectomy; type II - suture closure of the fistula or choledochoplasty; type III - choledochoplasty, and type IV - bilioenteric anastomosis. Since the present case corresponded to a Csendes type III with severe inflammatory adhesions around the GB neck, we elected to perform a partial cholecystectomy, leaving the wall of the GB neck to avoid bile duct injuries. Subsequently, choledochoplasty using the posterior wall of the GB neck was completed. In this case, a suture closure of the fistula was not attempted from the first, since it was considered to cause a bile duct stricture postoperatively. Regarding a bilioenteric anastomosis, it should be required when choledochoplasty is impossible. The bile drainage catheter was not left indwelling because intraoperative cholangiography showed no stricture or leakage at the site of repair. However, minor bile leakage appeared after the operation, which resolved without intervention. Based on this experience, perioperative ENBD, which enables pre and postoperative bile drainage and cholangiography, might be useful to prevent postoperative bile duct obstruction[6, 11]. Although the usefulness of T-tube drainage is reported, we consider ENBD to be preferable to a T-tube because of the shorter time needed to remove it[3,4].

CONCLUSION

In conclusion, the presence of a biliobiliary fistula should always be considered in the case of Mirizzi syndrome. POCS is useful to identify the existence of a biliobiliary fistula in cases of Mirizzi syndrome. When a biliobiliary fistula is confirmed before or at the time of surgery, operative strategy is selected according to the extent of erosion of the CBD.

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