CASE REPORT

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Portal cavernography during endoscopic retrograde cholangiopancreatography: from bilhemia to hemobilia

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Portobiliary fistulas are rare but may lead to life-threatening complications. Biliary plastic stent-induced portobiliary fistulas during endoscopic retrograde cholangiopancreatography have been described. Herein, we present a case of portal cavernography and recurrent hemobilia after endoscopic retrograde cholangiopancreatography in which a portobiliary fistula was detected in a patient with portal biliopathy. This likely indicates a change in clinical presentation (from bilhemia to hemobilia) after biliary drainage that was successfully treated by placement of a fully covered, self-expandable metallic stent.

Keywords: Endoscopic retrograde cholangiopancreatography; Hemobilia; Portal cavernography; Portobiliary fistula

INTRODUCTION

Internal biliary fistulas are rare and occur in 0.9% to 3.2% of patients with hepatobiliary disease.[1] This term generally refers to the communication of any part of the biliary tree with the gastrointestinal lumen, whereas fistula formation between the biliary tract and the portal vein is rare. The clinical manifestations of this type of fistula depend on both the etiology and type of fistula. Herein, we report a case of hemobilia due to a portobiliary fistula (PBF) in a patient with portal cavernoma. In addition, we report the results of a literature review that identified available evidence from previously reported cases.

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CASE REPORT

A 36-year-old male patient with polycythemia vera-induced portal vein thrombosis and cavernoma was referred to our hospital for endoscopic retrograde cholangiopancreatography (ERCP) due to recent onset of obstructive jaundice (bilirubin, 29 mg/dL). Cholestasis was mild (alkaline phosphatase, 160 U/L; gamma-glutamyl transferase, 52 U/L). He had no cholangitis (C-reactive protein, 4.2 mg/L), hemoglobin was 12.9 g/dL, and platelet count was 508,000/mm^3. Abdominal magnetic resonance angiography demonstrated a portal cavernoma compressing the common bile duct (Fig. 1). ERCP with biliary sphincterotomy was performed, followed by 4-mm balloon dilation of a fibrotic stenosis associated with the biliary varices and placement of a straight plastic stent (10 cm, 10 French [Fr]). Two days later, the patient presented with melena, fever (38.4 °C), and a hemoglobin drop to 8.5 g/dL. Urgent endoscopy revealed fresh blood and clots in the duodenum. A second ERCP was performed and, after removing the stent, fresh blood from the papilla was observed (Fig. 2).

After contrast injection, the portal venous system was opac-

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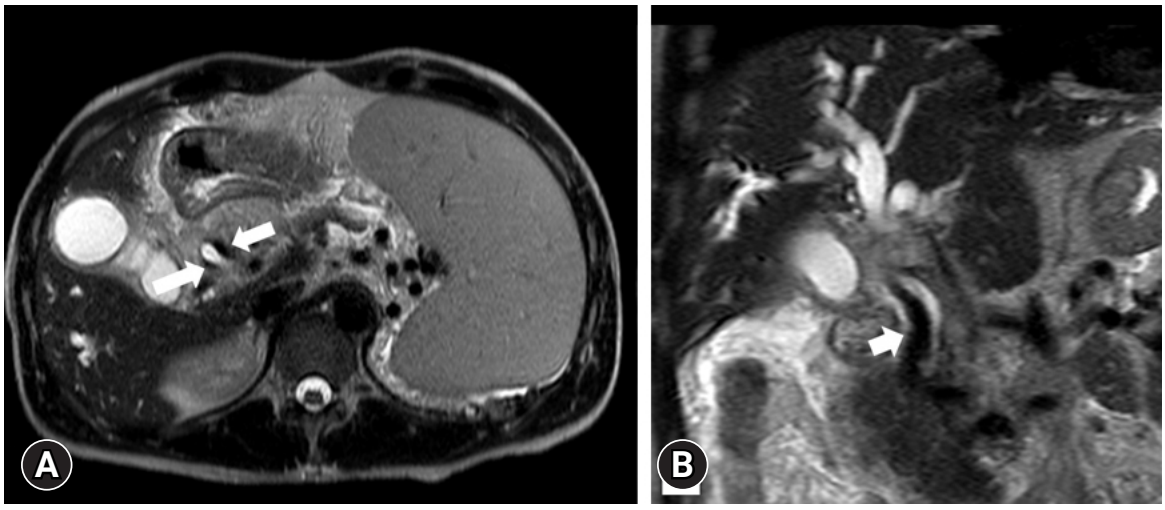

Fig. 1. (A) T2-weighted image (axial plane) depicting compression of the common bile duct between two venous structures (arrows). (B) T2-weighted image (coronal plane) showing a vessel as a low signal tubular structure (arrow) parallel to the common bile duct.

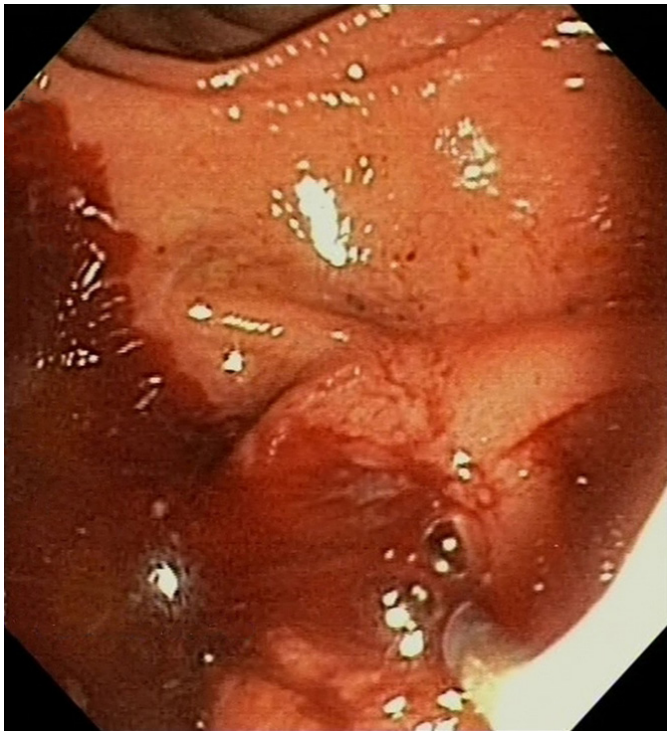

Fig. 2. Endoscopic image showing active bleeding from the papillary orifice (hemobilia).

ified (portal cavernography) (Fig. 3A). Two straight plastic stents (9 cm, 8.5 Fr) were placed to bridge the stenosis and the PBF, while IV somatostatin was initiated. The patient remained stable and was discharged five days later, without further endoscopic control, on oral antibiotics and beta-blockers.

Two months later, the patient presented again with jaundice (bilirubin 9.9 mg/dL), melena (hemoglobin 9.6 g/dL), and chills. Abdominal computed tomography scan showed intrahepatic biliary duct dilation. During ERCP, after removal of the two occluded plastic stents, active bleeding was again observed and controlled after insertion of three biliary plastic stents (8.5 Fr). Unfortunately, 12 hours later, the patient developed hypovolemic shock, requiring intensive care unit admission. Another urgent ERCP determined again an opacification of the portal venous system (Fig. 3B) and a 60×10 mm fully covered, self-expandable metallic stent (FCSEMS) (Wallflex; Boston Scientific) was inserted (Fig. 4). The patient was discharged 2 weeks later with no bleeding recurrence or jaundice. Owing to the patient's hemodynamic stability and favorable clinical condition, a second-look upper gastrointestinal endoscopy was not deemed necessary.

After the patient had been doing well for the next 6 months, the FCSEMS was replaced owing to occlusion with sludge, and no residual PBF was observed (Fig. 5A). Nine months later, the stent was removed without replacement, after balloon extraction of sludge and check for proper calibration of the stricture (Fig. 5B). Five months later, the patient was still in follow-up without relapse of bleeding, cholangitis, or jaundice.

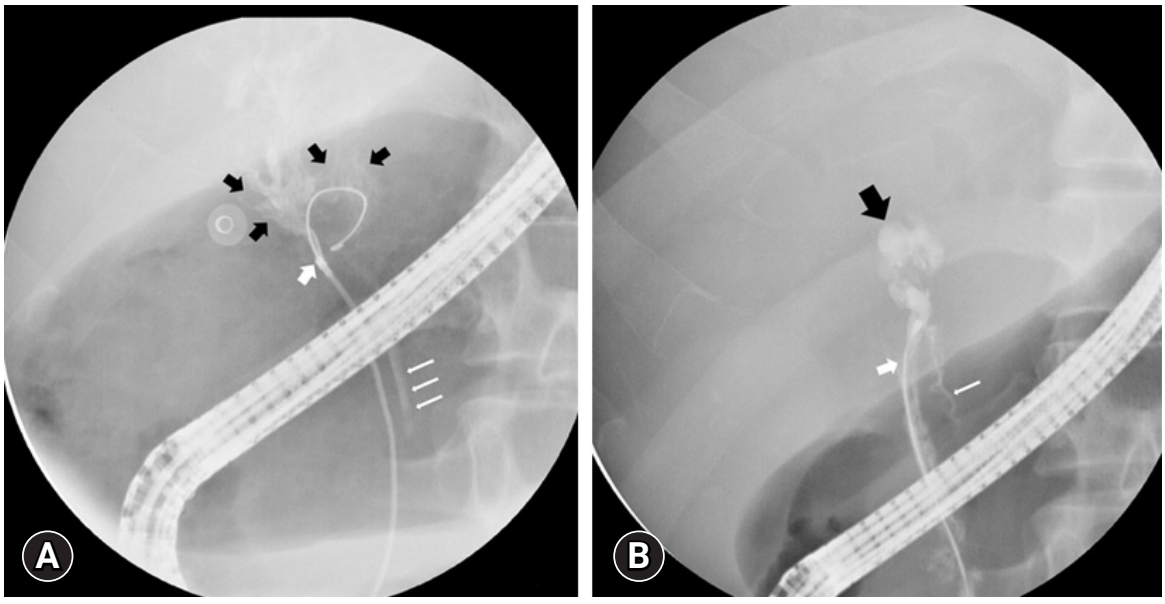

Fig. 3. (A, B) Cholangiogram showing opacification of a venous structure compatible with the known cavernoma (black arrows) and contrast dye in a venous branch parallel to the bile duct (long white arrow) with possible orifice of portobiliary fistula (short white arrow).

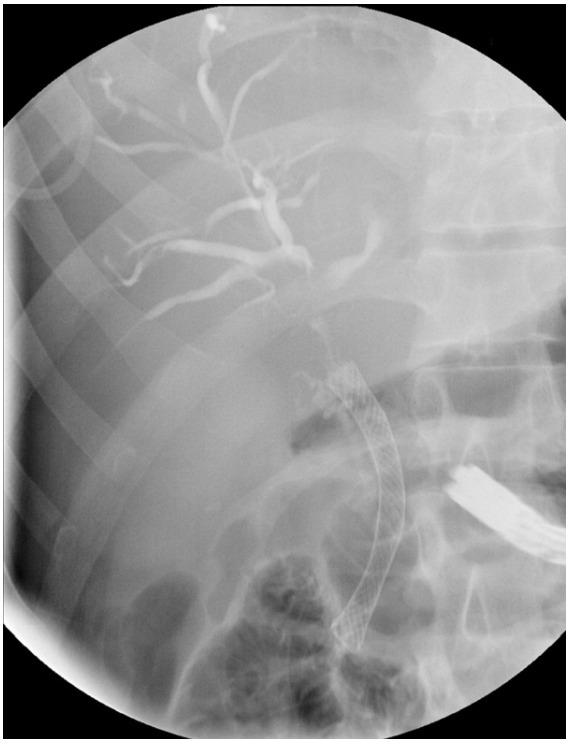

Fig. 4. Radiography showing the fully covered self-expandable metal stent deployed in the main bile duct.

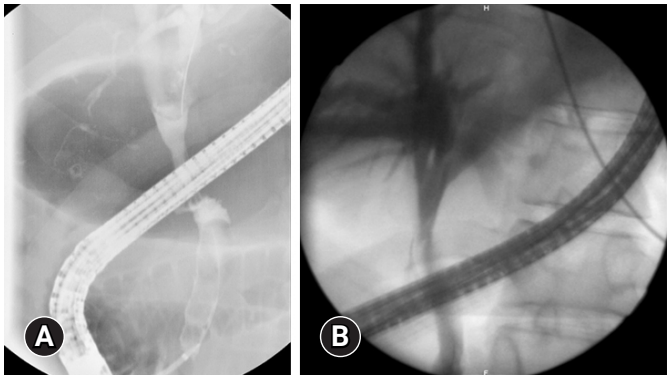

Fig. 5. (A) Radiography showing opacification of the common bile duct with no residual fistula during the endoscopic retrograde cholangiopancreatography performed to replace the initially placed fully covered self-expandable metal stent. (B) Radiography showing opacification of the common bile duct with no residual fistula or stenosis after definitive removal of the fully covered self-expandable metal stent.

DISCUSSION

PBF is a rare condition, and only a few case reports have been described in the literature. The first report of a PBF was in 1559,[2] describing gallstones found in the portal vein of a patient who underwent autopsy. In our case, we believe that the fistula resulted from a bile duct wall injury that occurred during the index

Table 1. Published case reports of ERCP-related portobiliary fistula

Study	No. of patients	Probable mechanism	Direct clinical manifestation	Treatment	Follow-up	Outcome
Huibregtse et al. (1988)[3]	1	Regular papillotomy	Minor bleeding	10 Fr BPS placement	6 wk	Uneventful
Ricci et al. (1992)[4]	1	Papillary fistulotomy	None	10 Fr BPS placement	7 day	Uneventful
Tighe and Jacobson (1996)[5]	1	Balloon dilation (4 mm and 6 mm)	Spontaneously resolving bleeding upon deflation of the 6 mm balloon	12 Fr straight BPS placement	24 hr	Uneventful
Kennedy et al. (1997)[6]	1	Biliary sphincterotomy 11.5 mm balloon and basket sweeping	Bradycardia and hypoxia followed by cardiac arrest	None	-	Death due to air embolism
Mutignani et al. (2002)[7]	2	Biliary sphincterotomy and two BPS (10 Fr)	Hemobilia after stent removal one month later	FCSEMS	4 yr	Partial inward stent migration with recurrent cholangitis treated with NBD and multiple stent in stent placement
		Biliary sphincterotomy	Spontaneously resolving hemobilia immediately after balloon-occlusion cholangiogram	3 BPS placement without proximal side-flaps (2×10 Fr, 1×11.5 Fr) and NBD for 48 hr	1 yr	One month later massive hemobilia after BPS removal. Persistent PBF after one week of endoscopic balloon tamponade and NBD requiring surgical splenorenal shunt
Espinel et al. (2007)[8]	1	Needle-knife sphincterotomy	None	Procedure termination	3 wk	Fistula healing
Layec et al. (2009)[9]	1	Biliary FCSEMS placement	Massive bleeding through the papilla upon stent removal	Placement of FCSEMS	18 mo	Uneventful
Furuzono et al. (2009)[10]	1	Catheter and guidewire manipulation Biliary sphincterotomy	None	Endoscopic NBD tube withdrawal in operating room	NA	No subsequent complications
Kawakami et al. (2011)[11]	1	Guidewire induced	Minor hemobilia	Rapid withdrawal of catheter and guide wire	Regular interval (not specified)	Fistula healing
Kalaitzakis et al. (2011)[12]	1	Guidewire induced	None	Procedure termination	4 day	Uneventful
Dawwas et al. (2013)[13]	1	Biliary sphincterotomy, balloon sweeping and BPS placement	Minor hemobilia upon stent removal	Procedure termination	At least 2 mo	Healing of all portal vein abnormalities
So et al. (2015)[14]	1	BPS placement	Exacerbation of hemobilia upon stent removal	FCSEMS	NA	Patient recovered from bacteremia and hemobilia

ERCP, endoscopic retrograde cholangiopancreatography; Fr, French; BPS, biliary plastic stent; FCSEMS, fully covered self-expandable metal stent; NBD, nasobiliary drainage; PBF, portobiliary fistula; NA, not applicable.

ERCP with balloon dilation. Other possible ERCP-related causes of trauma have also been reported, including biliary sphincterotomy, guidewire insertion, and stent placement (Table 1).[3-14] Four of the 13 reported cases of ERCP-related PBF occurred in the setting of portal cavernoma.[5,7,9]

The major message to be taken from our experience is that in the case of PBF identified during ERCP, the optimal treatment should be the placement of a FCSEMS covering the PBF, at least if it is located in the middle or distal part of the common bile duct (to avoid the possibility of crossing the hilum with the risk of contralateral biliary obstruction). It is probable that the compression induced by FCSEMS favors thrombosis of biliary varices, a feature that cannot be achieved with plastic stents.

Other possible causes of PBF include blunt liver trauma, bile duct stone, liver abscess, malignancy, percutaneous transhepatic cholangiography, biliary surgery, transjugular intrahepatic por-

tosystemic stent shunt, and percutaneous or transjugular liver biopsy.

PBF may result either in hemobilia, where blood enters the biliary tract, or more commonly, in the absence of portal hypertension and/or in case of untreated biliary stricture, in bilhemia, where bile enters the bloodstream. The pressure gradient between the biliary tree (10–15 mmHg) and the portal vein system (5–10 mmHg) explains the bilhemia. In our case, it is possible that bilhemia was already present before the index ERCP and that both the presence of portal hypertension and the treatment of biliary obstruction modified the clinical presentation towards hemobilia.

As previously illustrated, hemobilia associated with PBF may be a life-threatening condition. FCSEMS may be an acceptable and effective treatment option for both bleeding control and stricture calibration, as was seen in our case. Only three cases of PBF-induced hemobilia treated with FCSEMS have been described to date.[7,9,14] In one of these cases, FCSEMS was complicated by recurrent cholangitis due to partial proximal stent migration.[7] Other treatments that have been offered to control hemobilia in ERCP-related PBF include balloon tamponade, coil embolization, percutaneous or transjugular portosystemic stent placement, and surgical repair. However, transarterial embolization is not a good treatment option due to the venous origin of the bleeding.

In summary, this report describes a case of PBF that was identified during an ERCP performed for the treatment of a benign biliary stricture. Initial balloon dilation and plastic stenting treated the cholestasis (and possibly preexisting bilhemia) and induced recurrent and severe hemobilia, which was ultimately resolved by placement of a FCSEMS. This clinical presentation and possible treatment option should be kept in mind, especially for patients with portal hypertension presenting with elevated bilirubin levels and limited cholestasis.

Conflicts of Interest

The authors have no potential conflicts of interest.

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Author Contributions

Conceptualization: RAY, PG, JD, DB; Data curation: all authors; Project administration: RAY, PG, JD, DB; Supervision: all authors; Writing–original draft: RY, PG, JD, DB; Writing–review & editing: all authors.

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