CASE REPORT

Transjugular intrahepatic portosystemic shunt for the treatment of portal hypertensive biliopathy with cavernous transformation of the portal vein: a case report

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Abstract

Background: Portal hypertensive biliopathy (PHB) was caused by anatomical and functional abnormalities in the intrahepatic and extrahepatic bile ducts secondary to portal hypertension. Currently, there is no consensus regarding to the optimal treatment for PHB. Transjugular intrahepatic portosystemic shunt (TIPS) is the treatment choice for the management of symptomatic PHB, however, it could be very difficult in patients with PHB and cavernous transformation of portal vein.

Case presentation: We report a case of PHB, successfully managed with TIPS. A 23-year-old man with liver cirrhosis presented with jaundice. Magnetic resonance cholangiopancreatography (MRCP) showed multiple tortuous hepatopetal collateral vessels compressing the common bile duct (CBD) and leading to the dilated proximal bile duct. He was diagnosed with PHB and treated with TIPS. A guidewire was inserted into the appropriate collateral vessel through transsplenic approach to guide intrahepatic puncture and TIPS was performed successfully. After the operation, portal vein pressure decreased and the symptoms of biliary obstruction were relieved significantly. In addition, the patient showed no jaundice at a follow-up of one year.

Conclusions: For PHB patients presenting for cavernous transformation of the portal vein, which precludes the technical feasibility of TIPS, a combined transjugular/transsplenic approach could be an alternative option.

Keywords: Portal hypertensive biliopathy, Transjugular intrahepatic portosystemic shunt, Cavernous transformation of the portal vein

Background

Generally, portal hypertensive biliopathy (PHB) was caused by abnormalities in the intra- or extrahepatic bile ducts secondary to portal hypertension and commonly accompanied with extrahepatic portal venous obstruction (EHPVO). Only 5–30% of patients show symptoms of fever, abdominal pain, jaundice, and skin itching [1–4].

There is no consensus regarding to the optimal treatment for PHB. Endoscopic, surgical and interventional treatment are the treatment options for patients with symptomatic PHB [5–7]. Individualized treatment maybe decided case-by-case.

Some studies have reported that transjugular intrahepatic portosystemic shunt (TIPS) could be used for symptomatic PHB, but it is very challenging in patients with cavernous transformation of portal vein [8–11]. In this report, we present a case of EHPVO-related PHB
that was alleviated by TIPS via a combined transjugular/transsplenic approach.

**Case presentation**

A 23-year-old man with liver cirrhosis presented with jaundice for one month. The initial laboratory test indicated total bilirubin (TB) of 4.81 mg/dL, direct bilirubin (DB) of 4.44 mg/dL, alanine aminotransferase of 268 U/L, aspartate aminotransferase of 128 U/L, alkaline phosphatase (ALP) of 1105 U/L, and gamma-glutamyl transferase (GGT) of 1018 U/L. Child–Pugh's classification was graded as A. Gastroscopy showed moderate esophageal and gastric varices. Magnetic resonance cholangiopancreatography (MRCP) showed multiple tortuous hepatopetal collateral vessels compressing the common bile duct (CBD) and leading to the dilated proximal bile duct (Fig. 1a). TIPS was considered as an alternative approach to decrease portal venous pressure and alleviate bile obstruction. The main trunk and intrahepatic branches of portal vein were completely occluded and replaced by collaterals. Most collaterals were small, tortuous and not suitable for TIPS placement.

Traditional TIPS based on cross-sectional images may not guarantee that the suitable collateral was punctured. Therefore, a guidewire was inserted into the appropriate collateral vessel through transsplenic approach to guide intrahepatic puncture to ensure a linear intrahepatic shunt (Fig. 1b). Once the collateral vein was accessed successfully, indirect portography was performed. The intrahepatic tract was dilated using a balloon catheter to allow the implantation of an 8 mm × 60 mm expanded polytetrafluoroethylene covered stent. The portosystemic pressure gradient decreased from 24 to 13 mmHg and an 6 mm coil was used to embolize the splenic access after withdrawal of the sheath (Fig. 1c). Three months after the operation, TB dropped to 2.23 mg/dL, DB to 1.39 mg/dL, ALP to 504 U/L and GGT to 670 U/L. A follow-up MRCP revealed that the biliary obstruction was alleviated (Fig. 1d). In addition, the patient showed no jaundice.

**Discussion and conclusions**

Although the pathogenesis of PHB is not fully elucidated, it has been postulated that external pressure caused by dilated porto-porto collaterals and/or
ischaemic strictures of the bile duct may the may rea-
on. In the present case, biliary obstruction was caused
by compression of the collaterals due to EHPVO. Sur-
gical shunt, which was gradually replaced by TIPS, has
proven to feasible and effective in patients with EHPVO
and complications of portal hypertension. However, it
is very difficult since the intrahepatic portal branch was
totally occluded. During TIPS procedure, it is vital to
identify the appropriate collateral to insure the good
outflow in the stent. Transsplenic access and retrograde
catheterization would make it easier. Habib et al. [12]
demonstrated the feasibility of transsplenic TIPS in
11 patients with chronic portal vein thrombosis with
a success rate of 100%. We also emphasized the trans-
splenic tract to avoid the risk of perisplenic hemorrhage.

Endoscopic treatment is preferred in patients with
CBD stones, cholangitis or patients with dominant bili-
ary stricture, but without a shuntable vein. It includes
endoscopic sphincterotomy, stone extraction, and bili-
ary stricture dilatation with or without stent or naso-
biliary drain placement [6, 9, 13]. In this case, MRCP
showed dominant biliary stricture without CBD stones
and cholangitis. It may be risky if endoscopic treat-
ment such as biliary stricture dilatation with or without
stent in the presence of collaterals in the region. The
patient presented with jaundice and moderate esopha-
geal and gastric varices and the shuntable vein was
present. Therefore, TIPS was considered as alternative
approach to decrease portal venous pressure and allevi-
ate bile obstruction and performed successfully. After
the operation, portal vein pressure decreased and the
symptoms of biliary obstruction were relieved signifi-
cantly. Therefore, for PHB patients with cirrhosis, pre-
senting for cavernous transformation of the portal vein,
which precludes the application of TIPS, the combined
transjugular/transsplenic approach can be used as an
alternative treatment.

Abbreviations
PHB: Portal hypertensive biliopathy; TIPS: Transjugular intrahepatic portosys-
temic shunt; EHPVO: Extrahepatic portal venous obstruction; TB: Total bilirubin;
DB: Direct bilirubin; ALP: Alkaline phosphatase; GGT: Gamma-glutamyl trans-
ferase; MRCP: Magnetic resonance cholangiopancreatography; CBD: Common
bile duct.

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Authors’ contributions
LXF and WQZ performed the operation. ZM drafted the original manuscript
and LBX contributed to data collection. All authors read and approved the
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Availability of data and materials
Data sharing is not applicable to this article as no datasets were generated
or analysed during the current study.

Declarations
Ethics approval and consent to participate
Written informed consent was obtained from the patient for publication
of clinical data, including all images in this case report. This study was
approved by the Ethics Committee of the West China Hospital (Chengdu,
China).

Consent for publication
Consent for publication was obtained from the patient.

Competing interests
The authors declare that they have no competing interests.

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References
1. Dilawari JB, Chawla YK. Pseudosclerosing cholangitis in extrahepatic
portal venous obstruction. Gut. 1992;33:272–6.
2. Khuroo MS, Yattoo GN, Zargar SA, et al. Biliary abnormalities associ-
ated with extrahepatic portal venous obstruction. Hepatology. 1993;17:807–13.
3. Condat B, Vilgrain V, Asselah T, et al. Portal cavernoma-associated
cholangiopathy: a clinical and MR cholangiography coupled with MR
portography imaging study. Hepatology. 2003;37:1302–8.
4. Sezgin O, Oğuz D, Altintaş E, et al. Endoscopic management of biliary
obstruction caused by cavernous transformation of the portal vein.
Gastrointest Endosc. 2003;58:602–8.
5. Wallner BK, Schumacher KA, Weidenmaier W, et al. Dilated biliary tract:evaluation with mr cholangiography with a t2-weighted contrast-
enhanced fast sequence. Radiology. 1991;181:805–8.
6. Chattopadhyay S, Nundy S. Portal biliopathy. World J Gastroenterol.
2012;18:6177–82.
7. Suarez V, Puerta A, Santos LF, et al. Portal hypertensive biliopathy: a
single center experience and literature review. World J Hepatol.
2013;5:137–44.
8. Bayraktar Y, Öztürk MA, Egesel T, et al. Disappearance of “pseudochol-
gangioma sign” in a patient with portal hypertension due to
complete thrombosis of left portal vein and main portal vein web after
web dilatation and transjugular intrahepatic portosystemic shunt. J
Clin Gastroenterol. 2000;31:328–32.
9. Oo YH, Olliff S, Haydon G, et al. Symptomatic portal biliopathy: a
single centre experience from the UK. Eur J Gastroenterol Hepatol.
2009;21:206–13.
10. Cellich PP, Crawford M, Kaffes AJ, et al. Portal biliopathy: multidis-
ciplinary management and outcomes of treatment. ANZ J Surg.
2015;85:561–6.
11. Gorgül A, Kayhan B, Dogan I, et al. Disappearance of the pseudochol-
gangioma sign after TIPSS. Am. J. Gastroenterol 1996;150–154
12. Habib A, Desai K, Hickey R, et al. Portal vein recanalization-transjugu-
larintrahepatic portosystemic shunt using the transluminal approach
to achieve transplant candidacy in patients with chronic portal vein
thrombosis. J Vasc Interv Radiol. 2015;26:499–506.
13. Dhiman RK, Behera A, Chawla YK, et al. Portal hypertensive biliopathy. Gut.
2007;56:1001–8.
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