Title:
Cholestasis after TIPS placement in a patient with primary sclerosing cholangitis: an uncommon complication

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DOI: 10.17235/reed.2017.5159/2017
Link: PubMed (Epub ahead of print)

Please cite this article as:

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INTRODUCTION
Transjugular intrahepatic portosystemic shunt (TIPS) consists of the establishment of a low-resistance communication between the portal vein circulation and the systemic venous area in order to reduce portal blood pressure. It is primarily indicated for variceal bleeding refractory to medical and endoscopic management but is also useful for other complications related with portal hypertension (PHT) (1). The most common complication is hepatic encephalopathy (HE), but others should not be overlooked due to their potential severity. Hyperbilirubinemia after TIPS placement may result due to hemolysis or hepatocellular insufficiency (HI), and cholestasis is an uncommon complication. We report the case of a patient with primary sclerosing cholangitis (PSC) who had a rapidly evolving cholestatic syndrome with severe hyperbilirubinemia following TIPS placement. No similar cases have ever been reported in the literature to date. In addition, experience with TIPS in patients with PSC is limited.

Key words: Cholangitis. PSC. TIPS.
CASE REPORT
A 39-year-old male was admitted for TIPS placement. The patient was diagnosed with PSC following a cholangitis episode at the age of 32 which subsequently evolved to liver cirrhosis with clinically significant PHT in the form of esophageal varices (EVs). The patient received primary prophylaxis with beta-blockers and refractory ascites. He required frequent large-volume paracentesis for evacuation. He was subsequently referred to our center, where he was placed on the waiting list for liver transplantation (LT) and scheduled for TIPS placement as a bridge therapy. His clinical history was also remarkable except for a penicillin allergy, type-1 diabetes mellitus (DM) and IgA nephropathy.

On admission, liver function was Child B7 and Meld 9. While no complications developed during TIPS placement, a right portal branch puncture during the procedure was difficult due to a stony liver. The puncture tract was dilated using an 8 mm balloon, and a 10 mm wide by 7 + 2 mm long VIATORR endoprosthesis was placed, which was also dilated using a balloon of the same caliber. A control portogram revealed an appropriate flow (Fig. 1) with a final portacaval gradient of 9 mmHg (14 mmHg previously); 48 hours after TIPS placement, the patient developed a rapidly evolving cholestatic syndrome with hyperbilirubinemia levels reaching 29 mg/dl on the seventh day, and there was no evidence suggestive of concomitant hemolysis. This was accompanied by progressive hepatic panel impairment (AP 1046, GGT 283, GPT 165 and GOT 106) and worsening plasma creatinine levels (3.2 mg/dl), while the latter had been normal previously. No other LF signs developed and the level of consciousness, albumin and coagulation parameters remained normal. An urgent abdominal doppler scan was performed which revealed evidence of cirrhosis with PHT, moderate ascites and a normal-functioning TIPS (Fig. 2) with adequate permeability of the suprahepatic veins and the inferior vena cava area. The bile tree was not dilated and empiric broad-spectrum antibiotic therapy was started after blood culture. The patient was prioritized on the LT waiting list and an ERCP was performed. Retrograde cholangiography showed no contrast leakage. A consistent stricture was identified in the common hepatic duct which was approximately 15 mm long and 3 mm in diameter. The suprastenotic duct was slightly dilated with a beading pattern and there was no material inside (Fig. 3). The stricture was brushed for cytology (no malignant cells were found) and finally, a plastic stent was inserted. The patient course was satisfactory, with a gradual decrease in bilirubinemia.
levels and an improved renal function back to baseline values. Four weeks later, the patient underwent liver transplantation and, currently, he has no complications and undergoes regular monitoring.

DISCUSSION
TIPS is used for the management of severe PHT complications such as bleeding from esophageal varices or ascites (1). The main complication is hepatic encephalopathy in up to 35% of cases. Hyperbilirubinemia after TIPS occurs in up to 15% of patients and results from hemolysis within the stent in most cases. Liver failure may also cause jaundice after TIPS placement. In this case, hyperbilirubinemia could not be accounted for by these conditions, therefore other rarer causes of post-TIPS hyperbilirubinemia such as those secondary to bile duct conditions were investigated.

The patient had obstructive jaundice following the placement of the stent with compression of a previously stenotic common hepatic duct due to PSC and an associated surrounding fibrotic parenchyma that limited stent distension. The condition improved rapidly with a biliary stent, therefore extrinsic compression by the TIPS endoprosthesis was considered as the most likely cause. This is an uncommon complication which, in reported cases, was satisfactorily treated with biliary drainage (2), as in this case. Biloma resulting from an accidental bile duct rupture can also lead to a similar clinical presentation (3), which was not present in this case. Another potential cause of cholestasis that usually responds to a stent is a vascular origin. Biliary obstruction from variceal veins, known as portal biliopathy, is common in patients with extrahepatic PHT and portal cavernomatosis. Decreased portal pressure is a primary pillar of therapy (4) but does not account for the outcome in this case. A clot may develop that blocks the bile duct in patients with accidental biliary fistula, which is managed by removal via ERCP (5). There was no evidence of biliary sepsis or positive blood cultures, which are the typical presentation of a biliary fistula (6).

Portal puncture can be difficult due to the stony consistency of the liver and this PSC case also had an anatomically abnormal biliary tree. While no data exist about TIPS in these individuals, there is evidence of a higher frequency of biliary or vascular complications in the presence of an atrophic or anatomically altered liver. In conclusion, due to the development of hyperbilirubinemia following TIPS, a precise differential diagnosis should include a
potential biliary origin as identifying the etiology is crucial for early treatment.

REFERENCES
Fig. 1. Follow-up portogram after TIPS placement that shows a normal flow through the vascular tree and stent.

Fig. 2. High-speed hepatopetal portal flow with normal TIPS functioning data on the doppler sonogram.
Fig. 3. An endoscopic retrograde cholangiogram that shows a stricture in the common hepatic duct.