

Portal Biliopathy Causing Recurrent Biliary Obstruction and Hemobilia

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Abstract

A 63-year-old man with extrahepatic portal vein thrombosis presented with biliary obstruction and hemobilia after a liver biopsy. Balloon sweep of the common bile duct removed clotted blood, and cholangiogram showed a common bile duct narrowing, treated with biliary stenting. A percutaneous biliary catheter was later required for recurrent biliary obstruction and hemobilia, and repeat cholangiogram confirmed portal biliopathy—a large peri-biliary varix was compressing the common bile duct, causing biliary obstruction and intermittent portal hypertensive hemobilia. A transjugular intrahepatic portosystemic shunt was inserted, followed by embolization of the peri-biliary varix. Delayed diagnosis of portal biliopathy may lead to significant patient morbidity.

Introduction

Though chronic obstruction of the extrahepatic portal vein in the absence of cirrhosis is frequently associated with bile duct abnormalities due to the formation of prominent collateral veins, symptomatic biliary disease is uncommon. We report a case of extrahepatic portal vein obstruction due to portal vein thrombosis causing recurrent biliary obstruction and hemobilia that was effectively treated with variceal embolization and insertion of a transjugular intrahepatic portosystemic shunt (TIPS).

Case Report

A 63-year-old man presented with 4 weeks of intermittent fever, dark urine, and pruritus, and 2 days of yellowed skin and eyes and abdominal pain, unimproved with empiric antibiotics. Two years prior, he had been diagnosed with extrahepatic portal vein (PV) thrombosis associated with the factor V Leiden mutation, and was receiving chronic anti-coagulation. Two weeks before presentation, abnormal liver tests had prompted serologic testing for infectious and metabolic etiologies of liver disease that were unrevealing. He then underwent a transjugular liver biopsy 1 week before presentation that showed mild non-specific inflammation and no fibrosis. Simultaneous venography revealed patent hepatic veins and a normal hepatovenous pressure gradient of 2 mmHg.

On physical examination, the patient was alert and afebrile, with blood pressure 102/59, heart rate 72, icteric sclerae, jaundice, epigastric abdominal tenderness, and no abdominal distension. Laboratory studies revealed white blood cells 7,300/mL, hemoglobin 14.2 g/dL, platelets 218,000/mL, aspartate aminotransferase (AST) 108 IU/L, alanine aminotransferase (ALT) 115 IU/L, total bilirubin 5.5 mg/dL, alkaline phosphatase 618 U/L, and international normalized ratio 1.8. An abdominal ultrasound showed mild intrahepatic biliary dilation, a 6-mm diameter common bile duct (CBD), and cholelithiasis. Because he had biliary dilation, cholestatic liver test

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abnormalities, and no evidence of intrahepatic disease on serologic testing or liver biopsy, endoscopic retrograde cholangiography (ERC) was pursued. After biliary sphincterotomy, balloon sweep of the CBD removed old blood and clots. Cholangiogram showed an irregularly narrowed mid CBD (Figure 1A, arrow), and a plastic biliary stent was inserted proximally. The etiology of the obstruction was thought to be bleeding into the CBD secondary to the transjugular liver biopsy, but it was unclear what might be causing the cholangiographic abnormality. Symptoms and liver test abnormalities gradually improved, and the biliary stent was uneventfully removed 8 weeks later.

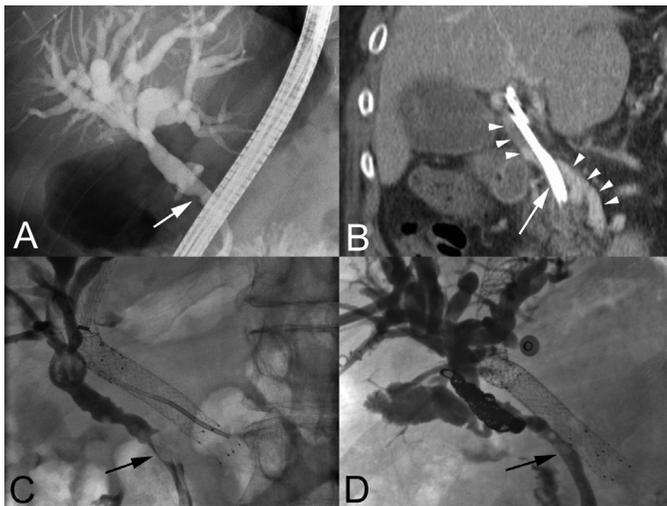


Figure 1. (A) Initial cholangiogram of a narrowed common bile duct (arrow). (B) Computerized tomography of the abdomen obtained after insertion of common bile duct stents revealed a peri-biliary varix (small arrows) adjacent to the stented common bile duct (large arrow). (C) After removal of the biliary stents, cholangiogram demonstrated compression of the common bile duct by the peri-biliary varix (arrow) that resolved after insertion of a (D) transjugular intrahepatic portosystemic shunt (TIPS; arrow).

Four months later, he reported recurrent pruritus and jaundice. The total bilirubin was 6.8 mg/dL and alkaline phosphatase 316 U/L. ERC with balloon sweep of the CBD caused rapid hemobilia that continued despite insertion of side-by-side plastic biliary stents in an attempt to tamponade hemorrhage thought to be occurring from a luminal CBD lesion. A covered metallic stent was considered but not inserted due to brisk bleeding and inadequate visualization. Abdominal computerized tomography revealed the biliary stents (Figure 1B, large arrow) with a 1.2-cm diameter varix coursing adjacent to the CBD without active contrast extravasation, consistent with a diagnosis of portal biliopathy (Figure 1B, small arrows). Chronic superior mesenteric, splenic, and PV thromboses with cavernous transformation were unchanged. Hemobilia resolved after transhepatic PV puncture with main PV stenting and mechanical thrombectomy; a portal venogram showed markedly diminished flow through a large biliary collateral vein thought to be the bleeding culprit. The patient was discharged several days later.

Several weeks later, massive hemobilia recurred during endoscopic removal of the biliary stents, and a percutaneous biliary catheter was inserted. Cholangiogram through the biliary catheter confirmed portal biliopathy—a large varix was compressing the CBD, causing intermittent obstruction and portal hypertensive hemobilia (Figure 1C, arrow). A transjugular intrahepatic portosystemic shunt (TIPS) was inserted, followed by embolization of the large peri-biliary varix, and biliary compression resolved on subsequent cholangiogram (Figure 1D, arrow). Nearly 2 years later, hemobilia and biliary obstruction have not recurred, and his liver tests are within the normal range.

Discussion

Chronic extrahepatic PV obstruction is common in the developing world and rare in Western nations.¹ The most common etiologies of extrahepatic portal vein obstruction are shown in Table 1. Development of collaterals is common and often involves the epicholedochal and pericholedochal venous plexuses. “Portal biliopathy” describes the development of portal hypertensive collaterals impinging on the biliary tract lumen. By cholangiogram, portal biliopathy may be identified in >80% of persons with chronic extrahepatic PV obstruction. However, symptomatic biliary obstruction is uncommon.² Portal biliopathy should be considered in the differential diagnosis of hemobilia and CBD obstruction in any patient with known portal hypertension, especially when chronic PV thrombosis without underlying cirrhosis is recognized.

Table 1. Most Common Etiologies of Extrahepatic Portal Vein Obstruction

Children (N=275) ^a	Adults (N=356) ^b
1. Idiopathic (55%)	1. Idiopathic (54%)
2. Abdominal infections (36%)	2. Abdominal infections (20%)
3. Umbilical catheterization (6%)	3. Trauma/other (15%)
4. Trauma/other (3%)	4. Prothrombotic disorders (9%)
5. Pancreatitis (1%)	5. Pancreatitis (1%)

^aPooled results from 7 studies published from 1962 to 1994.

^bPooled results from 6 studies published from 1979 to 1997.

Table modified from Sarin SK, Agarwal SR. Extrahepatic portal vein obstruction. *Semin Liv Dis.* 2002;22:43-58.

Although endoscopic treatment of choledocholithiasis with biliary sphincterotomy and stenting is indicated when stones are present,³ caution is warranted when portal biliopathy is coincident. Portal hypertensive hemobilia is rare with sphincterotomy alone, despite the frequent presence of adjacent ampullary collaterals, but it may be precipitated by transient pressure elevation in the distal portion of biliary varices during balloon sweeping.⁴ Endoscopic interventions for symptomatic biliary narrowing due to portal biliopathy have limited effectiveness and may cause major hemorrhage. Consequently, we advise a decompressive shunting

procedure such as TIPS if technically feasible, particularly if symptomatic biliary obstruction or hemobilia recur after an initial attempt at endoscopic treatment. As in this case, delays in diagnosis may lead to significant morbidity or even mortality for the patient.

Disclosures

Author contributions: B. Schlansky drafted, critically revised, and edited the article; J.A. Kaufman, G. Bakis, W.E. Naugler critically revised and edited the article; all authors approved the final draft submission. W.E. Naugler is guarantor of this article.

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