An Uncommon Cause of Biliary Obstruction in a Patient With Budd-Chiari Syndrome

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ABSTRACT

Biliary varices (BVs) are an infrequent complication of chronic portal hypertension. Most cases of BVs are asymptomatic and are likely underdiagnosed. We present a case of a 34-year-old woman with Budd-Chiari syndrome who was found to have BVs caused by a significant inferior vena cava (IVC) stenosis. This case demonstrates that preprocedure imaging for variceal screening should be considered before biliary tract procedures to prevent complications.

INTRODUCTION

Biliary varices (BVs) are an infrequent complication of chronic portal hypertension (PHTN). BVs can be identified as hypoechoic areas surrounding bile ducts on imaging including magnetic resonance cholangiopancreatography, endoscopic retrograde cholangiopancreatography (ERCP), and endoscopic ultrasound. Noninvasive imaging modalities, including traditional ultrasound, computed tomography, and even magnetic resonance imaging, are significantly less sensitive for the detection of BVs. With more invasive techniques, they can be identified as notched defects along the CBD on a cholangiogram. Most cases of BVs are asymptomatic and are likely underdiagnosed. We present the case of obstructive BVs discovered with biliary hemorrhage during ERCP. This is one of few case reports in the literature which demonstrates protrusion of BVs into the lumen of the CBD causing significant biliary obstruction.

CASE REPORT

A 34-year-old woman with polycythemia vera and Budd-Chiari syndrome status post portacaval shunt on chronic warfarin presented with a 1-day history of severe abdominal pain. Her symptoms began with the sudden onset of constant sharp right upper quadrant pain, multiple episodes of nonbloody, nonbilious emesis, tactile fevers, and chills. She had decreased oral intake secondary to pain but denied any jaundice, pale stools, dark urine, black tarry stools, bright red blood per rectum, hematemesis, or changes in bowel habits. She had no recent travel, sick contacts, and did not eat any unusual foods. She did not have a history of tobacco, drug, or alcohol use, and there was no personal or family history of gastrointestinal carcinomas or inflammatory bowel disease. She also had no previous abdominal surgeries and was never diagnosed with human immunodeficiency virus or hepatitis.

On admission to the hospital, she was febrile to 39.9°C and had a soft but distended abdomen with tenderness of the right upper quadrant without evidence of abnormal bowel sounds, icterus, hepatosplenomegaly, palmar erythema, spider angiomas, ascites, encephalopathy, or asterixis. The examination was otherwise unremarkable. Laboratory tests showed a normal hemoglobin/hematocrit without leukocytosis. Her laboratory results were notable for a total bilirubin of 4.1 mg/dL, direct bilirubin of 1.2 mg/dL, aspartate aminotransferase of 79 U/L, alanine aminotransferase of 69 U/L, international normalized ratio of 1.8, alkaline phosphatase of 207 U/L, platelet count of 108 K/µL, creatinine of 0.6 mg/dL, and blood urea nitrogen of 13 mg/dL. Abdominal ultrasound with Doppler demonstrated a patent portacaval shunt, splenomegaly, and stable prominent portosystemic varices causing limited
visualization of the CBD. She was admitted to the hospital and underwent an ERCP for further visualization, given the strong suspicion for choledocholithiasis because of her history of recurrent biliary colic and previous endoscopic ultrasound demonstrating biliary microlithiasis. ERCP demonstrated a filling defect within the middle third of the CBD for which a biliary stent was deployed (Figure 1). Hemorrhage was immediately visualized within the CBD, raising the possibility of an intraductal varix. Hemostasis was achieved with subsequent metal stent placement (Figure 1).

Venography was performed to evaluate for intraductal varices, which displayed multiple infrahepatic IVC collateralization and 2 regions of significant stenosis along the portal-IVC stent and intrahepatic IVC (Figure 2). This resulted in diversion of blood flow through the azygos system and ultimately giving rise to her CBD varices. Balloons were used for venoplasty resulting in successful reduction of the regions of stenosis and improvement of the pressure gradients. After the procedure, the total gradient was noted to decrease from 20 to 9 mm Hg (Table 1).

**DISCUSSION**

The presence of BVs can complicate surgical and endoscopic procedures of the biliary tract. As seen in this case, endoscopic intubation of the duct during ERCP caused unexpected hemobilia. A history of PHTN should increase clinical suspicion for BVs in patients presenting with biliary obstruction. Data on screening for and management of BVs are limited. In the case of acute complication, such as the biliary hemorrhage seen in our patient, recognition of BVs is the first crucial step in management. Although there are no guidelines on management of BV hemorrhage, stent placement can be an effective strategy to achieve hemostasis and temporarily relieve the biliary obstruction. Because most complications of PHTN, such as variceal hemorrhage, ascites, spontaneous bacterial peritonitis, and hepatic encephalopathy, are driven by increasing portal pressures, it is important to estimate these values. Normal portal-cava gradients range from 1 to 5 mm Hg; gradients greater than 5 mm Hg signify a diagnosis of PHTN; however, gradients greater than 10 mm Hg are generally considered clinically significant and suggest a worse prognosis. In our patient, assessing the portal-cava gradients and treating the areas of stricture helped assess and decrease the risk of future BVs.

Assessing the hepatic venous pressure gradient is indicated for diagnosing the degree of PHTN, the risk of developing complications of PHTN, and treatment response. In our patient, venography was conducted to diagnose and treat the IVC stenosis. This ultimately improved pressure gradients in the collateral circulation and should prevent progression of BVs. Based on our patient’s risk factors, a screening venography may have been warranted. It is important to consider preprocedure imaging for variceal screening in patients with a higher clinical suspicion for BVs because this will prevent complications if a biliary tract procedure is required in a patient with asymptomatic BVs.

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**Table 1. Pressure measurements from prevenoplasty and postvenoplasty**

| Site of measurement       | Prevenoplasty measurement (mm Hg) | Postvenoplasty measurement (mm Hg) |
|---------------------------|-----------------------------------|-----------------------------------|
| Proximal shunt            | 31                                | 23                                |
| Mid shunt                 | 29                                | 23                                |
| Distal shunt              | 23                                | 23                                |
| Cardiac side of the IVC   | 11                                | 13                                |
| Intrahepatic IVC near stent | 23                                | 17                                |
| Stent gradient            | 6                                 | 5                                 |
| IVC gradient              | 12                                | 4                                 |
| Total gradient            | 20                                | 9                                 |

IVC, inferior vena cava.
DISCLOSURES
Author contributions: All authors contributed equally to this article. S. Mago is the article guarantor.

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Informed consent was obtained for this case report.

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