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CASE REPORT | BILIARY

Bilhemia: A Rare Complication of Transjugular Intraheptic Portosytemic Shunt

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Abstract

A 56-year-old woman with cirrhosis due to chronic hepatitis C underwent emergent transjugular intrahepatic portosystemic shunt (TIPS) due to a ruptured esophageal varix during esophagogastroduodenoscopy. Following TIPS, the patient experienced a rapid rise in serum bilirubin with no evidence of biliary obstruction or hepatic injury. She was determined to have bilhemia, a rare but serious complication of TIPS.

Introduction

Transjugular intrahepatic portosystemic shunt (TIPS) is a common procedure used to alleviate the secondary effects of portal hypertension including uncontrolled variceal bleeding, refractory ascites, and hepatic pleural effusion (hydrothorax). There are several well-known complications of TIPS, including portosystemic encephalopathy, hemolytic anemia, hepatic ischemia, and stent thrombosis.[1] Bilhemia is a rare but serious complication of TIPS in which bile escapes into the bloodstream through a fistula between the biliary tree and the hepatic venous system.

Case Report

A 56-year-old woman with cirrhosis due to chronic hepatitis C underwent esophagogastroduodenoscopy (EGD) for evaluation and banding of esophageal varices. During the procedure, a large varix ruptured, requiring an emergent TIPS procedure to control the hemorrhage using a 10 x 10 mm GORE® VIATORR® covered stent (W. L. Gore & Associates, Flagstaff, AZ). Multiple unsuccessful attempts to access the right portal vein were made, requiring repeated repositioning of the cannula. Ultimately, a successful bridge was created between the right hepatic vein and central portal vein, which reduced the portosystemic gradient from 14 mm Hg to 3 mm Hg. The patient was stabilized hemodynamically with a transfusion of 7 units of packed red blood cells and was admitted to the medical intensive care unit.

Three days following the procedure, the patient became jaundiced. Her laboratory profile showed that her total and direct bilirubin levels had increased almost 10-fold, while her transaminases and international normalized ratio (INR) remained stable. Abdominal ultrasound found the stent to be patent with no evidence of biliary obstruction/dilatation. The patient's bilirubin continued to rise while her alkaline phosphatase, transaminases, and INR remained stable. The continued rise in bilirubin prompted an abdominal CT, which showed a patent TIPS and no evidence of hepatic ischemia or biliary duct dilatation.

Fifteen days after the TIPS procedure, an ERCP was performed to evaluate for evidence of a biliary-venous fistula. No evidence of a fistula or bile duct dilatation was found. A common bile duct stent was placed and sphincterotomy was performed in an attempt to lower the biliary pressure in order to reduce the flow of bile

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through a potentially unseen fistula. Despite this procedure, the patient's bilirubin continued to rise, and a venogram to visualize and occlude the biliary-venous fistula was also unremarkable. We determined that the only viable way to reverse the bilhemia was through liver transplantation.

During her hospital course, the patient developed a resistant *Klebsiella* bacteremia. Even with appropriate treatment, she remained persistently bacteremic, possibly due to seeding of her TIPS. She developed sepsis complicated by renal failure and was transferred back to the intensive care unit. Unfortunately, the patient's infection precluded her from undergoing liver transplantation and, after multiple discussions with the patient and her family, she was ultimately discharged home with hospice care.

Discussion

Elevated direct bilirubin levels are typically a sign of hepatocellular or biliary injury/obstruction. After undergoing TIPS placement, the shunting of blood from the portal vein to the systemic circulation can occasionally lead to hepatic ischemia.[1] Clinically, this presents as worsening right upper quadrant abdominal pain, hepatic encephalopathy, elevated aspartate transaminase and alanine transaminase levels, as well as an elevated serum bilirubin.[1-2] Our patient had an isolated hyperbilirubinemia, without transaminase elevation or worsening synthetic liver function. In addition, she had no abdominal pain and demonstrated no signs of encephalopathy. Therefore, it was determined that hepatic ischemia was an unlikely diagnosis.

Worsening cirrhosis was equally unlikely, as the patient demonstrated neither deterioration in her synthetic liver function nor any clinical signs of decompensated cirrhosis such as ascites or encephalopathy. Moreover, worsening cirrhosis post-TIPS is typically associated with TIPS thrombosis or stenosis, and both ultrasound and CT scan showed a patent TIPS.[3] Biliary tree stricture or blockage can also lead to hyperbilirubinemia, but this was unlikely due to our patient's isolated hyperbilirubinemia, normal alkaline phosphatase level, and lack of bile duct dilatation. Finally, post-TIPS hemolysis, a complication that causes an isolated elevation in serum bilirubin, was unlikely as our patient had a stable hemoglobin post-TIPS as well as a normal lactate dehydrogenase.

Bilhemia was the likey diagnosis given the development of *Enterobacteriaceae* bacteremia in the setting of otherwise unchanged liver function tests, absence of hemolysis, and no evidence of biliary obstruction. While no biliary-venous fistula was observed on venogram or ERCP, it is possible that the fistula was too small to be visualized. The difficulty encountered in cannulating the portal vein during the TIPS procedure may explain the creation of a small biliary-venous fistula.

Bilhemia is characterized by a rapid rise in total and direct serum bilirubin without other signs of hepatic dysfunction or biliary obstruction.[4] It is typically a consequence of hepatic trauma, though it has been known to occur as a complication of TIPS procedures.[5] The pathophysiology relates to a pressure gradient between the common bile duct (mean pressure: 12-14 mm Hg) and the hepatic vein (mean pressure: 7 mm Hg), which results in the direct flow of bile into the hepatic vein.[4,6] This is in contrast to a fistulous communication between the higher pressure portal venous system and the biliary tree, which typically results in hemobilia, the flow of blood into the biliary system. One consequence of bilhemia is fat embolism from the passage of large amounts of undissolved bile into the systemic circulation.[6-7] Bacteremia is another consequence, and is thought to be caused by the bilious contamination of the systemic circulation with enteric flora.[8-10] Bilhemia is fatal in approximately 50% of cases.[6]

Treatments for bilhemia aim to decompress the biliary system, commonly with biliary stenting and sphincterotomy.[6-8] Other case studies have shown that percutaneous biliary drainage, endoscopic nasobiliary drainage, and venous balloon occlusion are also viable options.[4-5,7-8] Indeed, in a case series of 20 patients with biliary leak (5 with bilhemia), endoscopic sphincterotomy successfully managed all but 1 patient, who died of bile thromboembolism.[6] In the absence of treatment, spontaneous closure of the biliary-venous fistula within 3 weeks has been observed.[7] However, if these treatments fail or if the patient develops persistent/recurrent bacteremia due to infection of the TIPS, liver transplantation remains the only treatment option. Successful liver transplantation for bilhemia post-TIPS is extremely rare, with only a few documented case reports.[9,4]

Disclosures

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