Mirizzi Syndrome in a Cirrhotic Patient After TIPS Resolved by Technetium$^{99}$m Mebrofenin Hepatobiliary Scan

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ABSTRACT

Cholestatic pattern on the hepatic panel is common and can be caused by a broad array of etiologies. Although rare, with a prevalence as low as 0.06%, it is imperative to keep Mirizzi syndrome in the differential diagnosis when evaluating cholestasis. Due to the nonspecific presentation and inconsistent radiologic features, a high index of suspicion is needed to diagnose Mirizzi Syndrome. We present an unusual case of a 51-year-old man with worsening cholestatic laboratory tests and a normal ultrasound and abdominal computerized tomography. A technetium$^{99}$m mebrofenin hepatobiliary acid scan suggested the diagnosis of Mirizzi syndrome that was later confirmed during an open cholecystectomy.

INTRODUCTION

Mirizzi syndrome (MS) refers to the extrinsic compression of the common hepatic duct by a stone in the cystic duct, gall bladder neck, or Hartman’s pouch.$^{1,2}$ There is a female predominance and the prevalence varies from 0.06% to 2.9% among the patients undergoing cholecystectomies, with lower incidence reported in the western countries.$^{1,3}$ Mirizzi syndrome is classified as Type I (common hepatic duct obstruction by the stone in cystic duct or Hartman’s pouch) and Type II (erosion of calculus into common hepatic duct or common bile duct producing a cholecystocholedochal fistula).$^{1,4}$ The classification is further defined based on the extent of common bile duct damage from cholecystocholedochal fistula and presence of cholecystoenteric fistula.$^2$ The determination of the type of MS helps choose the appropriate surgical approach.

CASE REPORT

A 51-year-old man with alcoholic cirrhosis (Model for End-Stage Liver Diseas [MELD] 9, Child Pugh Class B) was referred to our tertiary care center for refractory ascites. He had been abstinent from alcohol for >6 months but required weekly paracentesis. A standard hepatic panel was completely normal. He underwent transjugular intrahepatic portosystemic shunt (TIPS) placement. At 1-month follow-up, he was requiring paracentesis less frequently and was otherwise symptom free. However, there was interval development of cholestasis. An ultrasound showed a patent TIPS, multiple stones in the gallbladder without evidence of cholecystitis, and no biliary duct dilation. New medications (ranitidine and a vitamin supplement) were discontinued, but his bilirubin continued to rise. Abdominal computerized tomography showed cholelithiasis and patent TIPS, with no evidence of choledocholithiasis or intra- or extrahepatic biliary dilation. Endoscopic retrograde cholangio-pancreatography was deferred as he remained asymptomatic.

A transjugular liver biopsy showed cirrhosis, mild cholestasis, and ductular proliferation, suggestive of a large duct obstruction (Figure 1). Because of concern for image degradation from the TIPS, magnetic resonance...
cholangiopancreatography was not performed. Instead, technetium\textsuperscript{99m}-labeled mebrofenin hepatobiliary scan was obtained, which demonstrated prompt uptake and clearance of the tracer by the liver and excretion into the duodenum, but no visualization of the gallbladder on initial or delayed images (Figure 2), suggesting MS. During open cholecystectomy (OC), pericholecystic inflammation with fibrosis around the gallbladder neck and cystic duct were seen. An intraoperative cholangiogram was performed with extraction of multiple stones from the cystic duct.

Figure 1. Liver biopsy showing (A) bile stasis and (B) portal bile ductular proliferation and chronic inflammation, and (C) CK7 stain showing bile duct proliferation.

Figure 2. Technetium\textsuperscript{99m}-labeled mebrofenin hepatobiliary scan without gallbladder filling on initial or delayed images.
The microscopic examination of the resected gall bladder showed chronic cholecystitis without evidence of malignancy. Postoperatively, his cholestasis resolved.

DISCUSSION

There are no pathognomonic signs of MS, but it typically presents with right upper quadrant or epigastric abdominal pain (54%–100%) and jaundice (24%–100%) and, less frequently, nausea, vomiting (22%–31%), anorexia (11%–27%), fever (19%), pruritus, hepatomegaly, and acute cholecystitis (up to one-third of the cases). Among the imaging tests, the clinicians should consider a reevaluation of hepatic panel.

Various imaging tests may assist in the diagnosis of MS, although the results are inconsistent. Sensitivities of radiologic testing are: ultrasonography (23%–48%), computerized tomography (43%), and endoscopic retrograde cholangiopancreatography (50%–100%).2 Abdominal ultrasound is usually the first screening test that may show contracted gall bladder with cholelithiasis, dilated intrahepatic, and common hepatic duct with abrupt change in diameter distal to choledocholithiasis.2 Endoscopic ultrasound may reveal similar findings.2 Computed tomography can be a useful if there is a suspicion of malignancy.1 Magnetic resonance cholangiopancreatography has the highest preoperative cost effectiveness.2 Endoscopic retrograde cholangiopancreatography has the additional advantage of endoscopic treatment with stents or prosthesis, although it may have a significant morbidity (10%) and mortality (0.4%).2,4,6 When endoscopic interventions are unsuccessful, percutaneous transhepatic cholangiography may be used. Despite the various diagnostic modalities that are widely available, the diagnosis of MS may be challenging, as these imaging tests may fail to illustrate an underlying cause or there may be relative contraindications to perform them. This is illustrated in our case, in which we diagnosed this unusual cause of cholestasis in an atypical manner; that is the preoperative diagnosis of MS was important to decrease the chances of intraoperative complications.5

Open cholecystectomy is the treatment of choice for MS, especially in Type II, because the altered anatomy from dense adhesions causes a high rate of conversion from laparoscopic cholecystectomy to OC.4,5 Intraoperative cholangiography can be useful to locate the ducal stones and fistula, particularly if the diagnosis of MS is made during the surgery.6 Laparoscopic cholecystectomy may be used in certain cases of Type I and even Type II MS.7 However, laparoscopic cholecystectomy can be a surgical challenge, which warrants a low threshold to convert to OC to minimize the risk of bile duct injury.1,2

Despite its low prevalence, physicians should be aware of MS as a potential cause of cholestasis even in asymptomatic patients, especially if they have a prior history of cholelithiasis. The variable sensitivities of various imaging tests emphasize a high index of suspicion when the initial radiological evaluation is noncontributory. Rarely, nuclear scans can be helpful and prevent a more invasive liver biopsy.

DISCLOSURES

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