Delayed Diagnosis of Mirizzi Syndrome

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Abstract

Keywords

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- pseudoaneurysm
- cholangiocarcinoma
- gallstone
- Mirizzi syndrome
- interventional radiology

Biliary obstruction occurs in a wide variety of malignant and benign conditions. The following is a unique case of biliary obstruction caused by external compression of the hepatic duct by a gallstone (Mirizzi syndrome). Owing to unusual imaging characteristics of the stone, the mass was initially mistaken for a malignancy or hepatic pseudoaneurysm. It was not until the patient developed gallstone ileus that the correct diagnosis was made.

Objectives: Upon completion of this article, the reader will be able to discuss the pathogenesis, classification, diagnosis, and treatment of Mirizzi syndrome.

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Interventional radiologists commonly treat biliary diseases that are not amenable to surgical intervention. The National Health and Nutrition Examination Survey estimated that 6.3 million males and 14.2 million females between the ages of 20 and 74 years in the United States have gallbladder disease.¹ Females are especially prone to developing gallstones, as studies have demonstrated a 4.5:1 female-to-male ratio.² Gallstone predominance in females is likely secondary to estrogenic effects on the enterohepatic circulation.^{3,4} It follows, then, that biliary obstruction secondary to cholelithiasis/choledocholithiasis is more frequently encountered in

this population.⁴ Less common benign causes of biliary obstruction include infection, post-cholecystectomy stricturing, post-pancreatitis stricturing, primary sclerosing cholangitis, and Mirizzi syndrome.^{5,6} Mirizzi syndrome is often not recognized at initial presentation, which can result in morbidity and biliary injury.

Case Description

A 74-year-old woman with systemic sarcoidosis presented to an outside hospital with progressive jaundice and 7 days of epigastric pain, nausea, and bilious vomiting. The initial contrast-enhanced abdominal computed tomographic (CT) scan showed an abnormal low-attenuation lesion in the porta hepatis (► **Fig. 1**). The patient underwent endoscopic cholangiopancreatography with brushings which were negative for neoplasia. A percutaneous transhepatic biliary drain was placed for decompression. She was deemed a poor surgical candidate and was transferred to our institution for further workup. Diagnostic workup at our institution included tumor markers which were also found to be negative, as well as a repeat magnetic resonance cholangiopancreatography which showed intrahepatic biliary ductal dilatation and the hilar mass. On this study, a discrete spherical area of low signal was seen reportedly separate from the mass, which suggested a flow void and possible pseudoaneurysm (► **Fig. 2**). Repeat contrast-enhanced CT showed marked thickening of the

Fig. 1 Axial view of initial contrast CT scan. Note the mass-like density in the subhepatic region near the biliary confluence (arrow).

gallbladder wall and soft tissue filling the hepatic hilum which was thought to represent tumor. A small focus of high density within the mass was suggestive of hemorrhage or pseudoaneurysm (►Fig. 3) left-sided biliary ductal dilation was also noted. Subsequent abdominal ultrasound demonstrated Doppler signal within the mass with a to-and-fro waveform (yin-yang phenomenon) suggestive of a hepatic artery pseudoaneurysm near the liver hilum (►Fig. 4a, b). A conventional catheter angiogram failed to support these findings (►Fig. 5). Percutaneous transhepatic cholangiography showed extrinsic compression of the common hepatic duct with dilation of the intrahepatic ducts (►Fig. 6). The patient was asymptomatic and over the course of almost 1 year underwent routine biliary drain exchanges.

Eight months later, the patient was transferred to our institution from an outside hospital with jaundice, epigastric pain, nausea, and intractable vomiting. Follow-up CT revealed that the previously reported mass at the hepatic hilum had migrated to the distal small bowel, suggestive of a gallstone ileus (►Fig. 7a–c). She underwent laparoscopic enterolithotomy for removal of the impacted gallstone from the proximal ileum. Operative findings included a dilated bowel proximal to the stone and collapsed bowel distal to the stone. The extracted stone measured 2.5 cm in diameter. The patient recovered from surgery without complications. Repeat cholangiogram showed no residual obstruction and the biliary drain was removed.

Discussion

Mirizzi syndrome is a rare cause of biliary obstruction caused by extrinsic compression of the common hepatic duct by a gallstone in the cystic duct or Hartmann pouch of the gallbladder. It occurs in less than 2% of cholecystectomies.⁷ Patients typically present with jaundice, fever, and abdominal pain. Initial imaging evaluation usually consists of sonography, which may reveal biliary dilation above the gallbladder neck, an impacted stone, and an abrupt change to a normal width of the common duct below the stone.⁸ CT scanning can

Fig. 2 T2-weighted MRI showing low signal mass in the hepatic hilum (arrow).

be helpful to establish whether malignancy is present, but may not adequately show the gallstone as the source of biliary obstruction.⁹ Beltran's simplified classification of the different stages of Mirizzi syndrome is included in ►Fig. 8.^{10,11}

In this case, given the unusual appearance of the gallstone on MRI and ultrasound, the gallstone was mistaken for a mass. Coincidentally, Mirizzi syndrome has been associated with an increased frequency of gallbladder carcinoma. Between 6 and 28% of patients with a preoperative diagnosis of Mirizzi syndrome were later found to have cholangiocarcinoma¹² and this co-occurrence is mainly found in Mirizzi type II or higher.¹³ The underlying factor for both cholangiocarcinoma and Mirizzi syndrome appears to be long-standing inflammation from gallstone disease with stasis.¹² For this reason, some may classify Mirizzi syndrome as a risk factor for cholangiocarcinoma.¹³ In our patient, all tumor markers, including CA-19-9, were within normal limits. CA-19-9 may be moderately elevated in patients with Mirizzi syndrome but would be strikingly elevated, to over 800 U/mL, in patients with cholangiocarcinoma.¹³

Fig. 3 Repeat contrast-enhanced CT scan at the authors' institution showed marked thickening of the gallbladder wall with associated soft tissue filling the hepatic hilum (not shown). A high-density focus which was thought to represent a pseudoaneurysm is noted (white arrow). Black arrow indicates a dilated left-sided intrahepatic biliary duct.

Fig. 4 (a) Doppler ultrasound of the hepatic hilum showing vascular abnormality with to-and-fro flow (yin-yang phenomenon) suggestive of a pseudoaneurysm. In retrospect this was likely due aliasing artifact with a normal left portal vein and hepatic artery. (b) Echogenic foci with acoustic shadowing, indicative of gallstones. This was separate from the vascular abnormality noted at the hilum.

Gallstone ileus is a rare cause of small bowel obstruction, but should always be included in the differential for a patient with a known history of gallstones who presents with symptoms of small bowel obstruction.¹⁵ In gallstone ileus, a cholecystoenteric fistula provides an escape route for a previously impacted gallstone.¹⁶ Using Beltran's simplified

Fig. 5 No pseudoaneurysm was identified on digital subtraction angiography.

Fig. 6 Percutaneous transhepatic cholangiogram demonstrating extrinsic compression of the common hepatic duct (arrow) with dilation of the intrahepatic ducts.

classification system for Mirizzi syndrome, this patient had Mirizzi type IIIb. The first stage begins with extrinsic compression of the hepatic duct. The continuous inflammatory process in the gallbladder results in obliteration of the cystic duct, while the gallstones produce pressure ulcers over the gallbladder wall to the point of erosion into the bile duct (cholecystobiliary fistula) and duodenum (cholecystoenteric fistula) eventually leading to gallstone ileus.¹⁷ A recent study examining the relationship of Mirizzi syndrome and cholecystoenteric fistula found that increased age was associated with Mirizzi syndrome and cholecystoenteric fistula, and that female gender was also significantly associated with Mirizzi syndrome.¹⁷

Surgery is the primary treatment modality for Mirizzi syndrome. Distorted biliary anatomy with increased risk of bile duct injury poses a daunting surgical challenge, given that preoperative diagnosis is uncommon.¹⁸ Dissection of Calot triangle (cystohepatic triangle) proves especially difficult during surgical extraction of the stone. In patients who are not surgical candidates, endoscopic or percutaneous biliary decompression may be the initial treatment.¹⁹ The surgical approach for Mirizzi syndrome type IIIb is controversial, but it is generally suggested that the gallstone ileus is treated first. After the patient has recovered from surgery, the gallbladder should be approached based on the presence or absence of external hepatic duct compression and cholecystobiliary fistula.^{16,18} In this case, the patient underwent laparoscopic enterolithotomy almost immediately after the gallstone ileus was revealed on imaging studies. Interestingly, current literature does not recommend laparoscopic

Fig. 7 Noncontrast CT abdomen performed during most recent hospitalization. (a) Axial image with no significant intra- or extrahepatic biliary ductal dilatation, an improvement from prior CT studies. The previously visualized hypodense ring-enhancing mass at the hepatic hilum is no longer noted. (b and c) Axial and coronal images showing significant proximal bowel dilatation to the level of the distal ileum, with a gallstone at the proximal ileum (white arrows).

Fig. 8 Three stages of Mirizzi syndrome according to Beltran's 2012 simplified classification scheme.¹³

cholecystectomy in patients with Mirizzi type II or higher, given the greater technical complexity and increased risk for bile duct injury.²⁰

In summary, Mirizzi syndrome refers to common hepatic duct obstruction caused by extrinsic compression from an impacted stone in the cystic duct. Associated inflammatory changes may mimic a mass and Mirizzi syndrome has been associated with a high frequency of gallbladder cancer. In this case, the diagnosis of Mirizzi syndrome was delayed because the gallstone was mistaken for a mass or pseudoaneurysm. It was not until the patient developed gallstone ileus and the "mass" was shown to migrate that the correct diagnosis was made.

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