CASE REPORT

Mirizzi Syndrome: A Rare Cause of Obstructive Jaundice

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ABSTRACT

Mirizzi syndrome was first described by Pablo Luis Mirrizi. The incidence of this syndrome ranges from 0.05% to 4%. This rare condition is caused by the obstruction of common hepatic duct or common bile duct due to compression caused by several impacted stones or a single large impacted gallstone in Hartmann's pouch. The clinical presentations may vary from no symptoms to severe cholangitis. It is a rare cause of obstructive jaundice and hence we are reporting a case to emphasize that while evaluating a case of obstructive jaundice, one must consider Mirizzi syndrome as a differential diagnosis.

Keywords: Obstructive jaundice, Mirizzi syndrome, common bile duct, cholecystoenteric fistula

Mirizzi syndrome was first described by Pablo Luis Mirizzi. The incidence of this syndrome ranges from 0.05% to 4%. It is a rare condition wherein a gallstone is impacted in the cystic duct or neck of gallbladder, which compresses the common hepatic duct. This leads to obstruction and jaundice. Obstructive jaundice can occur due to direct compression from the stone or due to fibrosis, which develops as a result of chronic cholecystitis. The clinical presentations may vary from no symptoms to severe cholangitis. There may be recurrent episodes of jaundice and cholangitis. It can be associated with cholecystitis. There can be development of a fistula between gallbladder and the common duct, with passage of stone into the common duct. It is a rare cause of obstructive jaundice and hence, we are reporting a case in order to emphasize that while evaluating a case of obstructive jaundice, one must consider Mirizzi syndrome as a differential diagnosis.

CASE REPORT

A 49-year-old male presented to us with chief complaints of low-grade fever, yellowish discoloration of urine, itching in body and right upper abdominal pain for 5 days. The patient was a known case of type 2 diabetes mellitus for the past 8 years and was on oral hypoglycemic agents. He had history of jaundice 20 years back. On examination, the patient was conscious, cooperative and oriented to time, place and person. His vital signs were stable and temperature was 99.6°F (oral). He had no pallor, cyanosis, clubbing, lymphadenopathy or edema, but icterus was present. On systemic examination, liver was palpable but spleen and gallbladder were not palpable. We kept the diagnosis as febrile illness, possibly due to malaria, dengue fever, scrub typhus or acute cholangitis.

Patient was further evaluated and his blood biochemistry showed that renal function tests were normal (blood urea - 16 mg/dL, serum creatinine - 0.70 mg/dL). Liver function tests were deranged (total bilirubin - 5.7 mg/dL, direct bilirubin - 2.3 mg/dL, serum glutamic oxaloacetic transaminase [SGOT] - 245 U/L, serum glutamic pyruvic transaminase [SGPT] - 513 U/L, alkaline phosphatase [ALP] - 341 U/L, total protein - 7.5 g/dL, serum albumin - 3.8 g/dL). His lipid profile was also deranged (serum cholesterol - 195 mg/dL, serum triglyceride - 262 mg/dL, high-density lipoprotein cholesterol [HDL-C] - 18 mg/dL, low-density lipoprotein cholesterol [LDL-C] - 130 mg/dL). Complete hemogram showed hemoglobin - 11.5 mg/dL, mean corpuscular volume (MCV) - 60.1 fL, total leukocyte count (TLC) - 9,940/mm^3, platelet count - 3.51 × 10^3/mm^3. His fever profile and viral markers were negative (Malarial parasite quantitative buffy coat [MPQBC], Dengue immunoglobulin M/immunoglobulin G [IgM/IgG],

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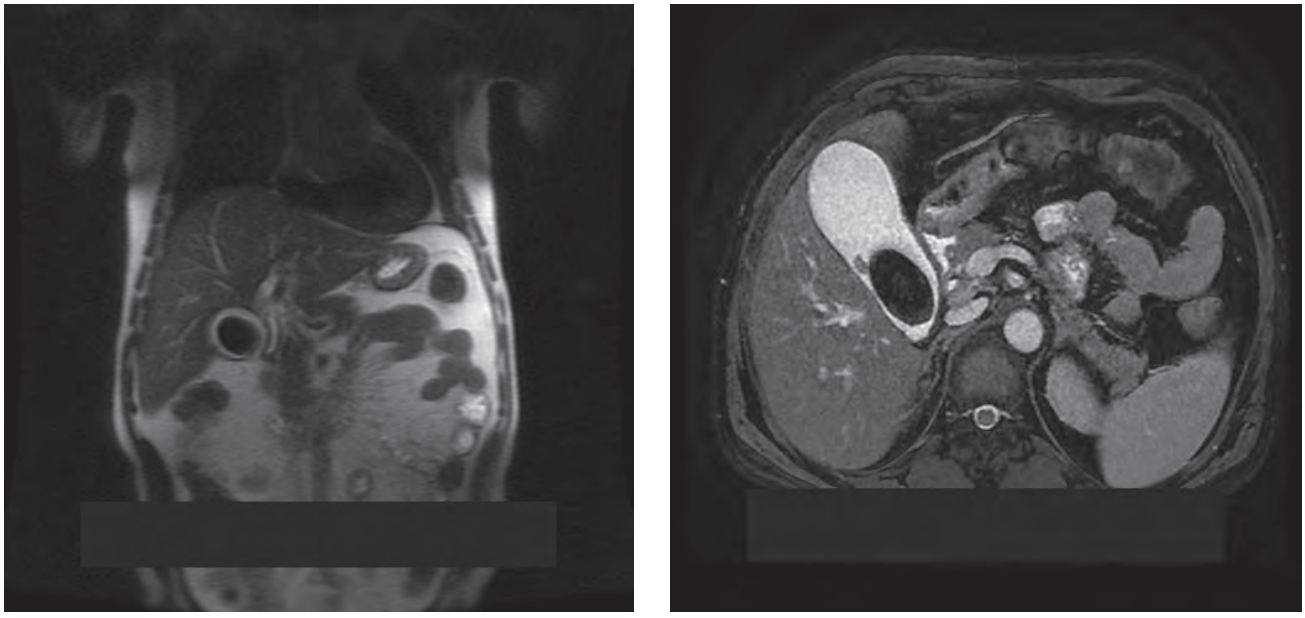

Figure 1. MRCP of the patient in which a 42 mm calculus is seen in gallbladder neck compressing the CBD, suggestive of Mirizzi syndrome.

scrub typhus IgM capture ELISA test, hepatitis B surface antigen [HBsAg], anti-hepatitis C virus [HCV], IgM anti-hepatitis A virus [HAV], IgM anti-hepatitis E virus [HEV], human immunodeficiency virus [HIV]). Ultrasonography (USG) showed fatty liver, cholelithiasis with ill-defined hypoechoic area in gallbladder neck region resulting in distended gallbladder with dilated intrahepatic vascular and biliary radicles, possibly indicating cholecystitis.

On further investigating, contrast-enhanced computed tomography (CECT) abdomen showed evidence of calcified calculus of size 4.4 × 2.56 cm in gallbladder lumen in body and in neck region, associated with dilatation of central (right hepatic duct [RHD] - 7 mm and left hepatic duct [LHD] - 8 mm) and peripheral intrahepatic biliary radical (IHBR). Gallbladder appeared significantly overdistended; however, wall thickness appeared normal. Dilatation of central and peripheral IHBR with overdistended gallbladder likely due to gallbladder neck calculus impact on coronary heart disease or CHD (suggestive of Mirizzi syndrome). Magnetic resonance cholangiopancreatography (MRCP) showed a 42 mm calculus noted in gallbladder neck causing compression over common bile duct (CBD). Mild IHBR dilatation was seen. These findings were suggestive of Mirizzi syndrome with cholelithiasis, as shown in the Figure 1. Hence, the diagnosis of Mirizzi syndrome was confirmed. Patient was advised surgical treatment for this, and therefore, was transferred to the surgical ward for further management.

DISCUSSION

Mirizzi syndrome is a rare condition known to occur as a result of obstruction of the common hepatic duct or CBD due to compression from multiple impacted stones or a single large impacted gallstone in the Hartmann's pouch. The syndrome is named after Pablo Luis Mirizzi. The first published paper on this syndrome was in 1940. It is relatively uncommon and mere 0.1% of patients with gallstones have been reported to develop this condition. About 0.7% to 25% of patients who underwent cholecystectomies were found to have this condition. While it may be more common in older populations, no particular inclination has been noted for either male or female patients with gallstones. The exact pathophysiology of this syndrome is not clearly known but it appears to be associated with floppy Hartmann's pouch with multiple stones or a single large impacted stone, which causes inflammation and fistula formation. Most patients present with repeated bouts of fever, pain and jaundice.

McSherry et al proposed a classification that divides Mirizzi syndrome into 2 types based on endoscopic retrograde cholangiopancreaticography (ERCP) findings. In type 1, the hepatic duct is compressed by a large stone impacted in the cystic duct or Hartmann pouch. Associated inflammation may contribute to obstruction and formation of a stricture in the central section of the extrahepatic bile duct. In type 2, the calculus erodes the common hepatic duct to produce a cholecystocholedochal fistula.

Csendes et al modified McSherry's classification in 1989, and classified Mirizzi syndrome into 4 types:

- Type I: Extrinsic compression of the CBD by an impacted gallstone.
- Type II: Cholecystobiliary fistula present, involving one-third of the circumference of the CBD.
- Type III: Cholecystobiliary fistula present, involving two-third of the circumference of the CBD.
- Type IV: Cholecystobiliary fistula present, involving the whole circumference of the CBD.

Csendes, in 2007, added a new type (Type V) to his classification of Mirizzi syndrome, and it was validated by Beltran et al. This type corresponds to any type of Mirizzi syndrome associated with cholecystoenteric fistula with (Vb) or without (Va) gallstone ileus.

There is an increased risk of developing gallbladder cancer after Mirizzi's syndrome. USG reveals gallstones with a contracted gallbladder and intrahepatic ductal dilatation. The typical findings of MRCP and ERCP are a dilated intrahepatic biliary tract with a normal-sized bile duct, secondary to obstruction at the level of cystic duct insertion into the common hepatic duct.

A cholecystobiliary or cholecystoenteric fistula due to persistent inflammation is the most common complication of Mirizzi syndrome. Other complications include cutaneous fistula and secondary biliary cirrhosis. This syndrome may be confused with Klatskin tumor because of the appearance of obstruction and surrounding inflammation.

Management of Type I Mirizzi syndrome includes cholecystectomy with or without bile duct exploration. Types II and III can be treated with partial cholecystectomy, removal of calculus and choledochoplasty, as needed. Roux-en-Y hepaticojejunostomy is required to repair a large defect as seen in Type IV Mirizzi syndrome. In elderly patients with comorbidities and high risk of surgical complications, nonoperative methods should be considered to minimize morbidity associated with the surgery. Exploration of the CBD should be undertaken due to the occurrence of choledocholithiasis in Mirizzi syndrome. Performing a frozen section biopsy from the removed gallbladder is advisable, since there is association between Mirizzi syndrome and gallbladder cancer.

CONCLUSION

Mirizzi syndrome can present to us in various forms, including obstructive jaundice. It is a rare cause of obstructive jaundice. So, while evaluating a case of obstructive jaundice, one must consider Mirizzi syndrome as one of the differential diagnoses.

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