

Unraveling Mirizzi Syndrome – Uncommon Cause of Biliary Obstruction: A Case Report

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Abstract

Mirizzi syndrome is an unusual complication of chronic cholelithiasis due to impacted gallstone in the cystic duct or neck of the gallbladder causing Common Bile Duct (CBD) obstruction. Here, we report a case of middle aged female presenting with acute abdominal pain and jaundice. Endoscopic Retrograde Cholangio Pancreatography (ERCP) revealed a large gallstone in the cystic duct compressing the common bile duct suggestive of Mirizzi syndrome which was managed surgically.

Keywords: Cholelithiasis; Jaundice; Endoscopic Retrograde Cholangio Pancreatography (ERCP)

Abbreviations: ERCP: Endoscopic Retrograde Cholangio Pancreatography; CBD: Common Bile Duct; MRCP: Magnetic Resonance Cholangio Pancreatography; USG: Ultrasonography; CT: Computed Tomography

Introduction

Mirizzi syndrome is a condition characterized by gallstones causing obstruction and erosion of CBD. In extreme cases, it can lead onto cholecystocholedochal fistula. It is of immense importance to identify the condition prior to cholecystectomy in order to avoid postoperative complications and it also has coincident association with gallbladder carcinoma [1-3].

Case Report

A 36 year old Hispanic female presented to the emergency room with acute epigastric pain and non-bloody vomiting of 2 days duration. Her past medical history was unremarkable. On examination, she was febrile (101.2 F), icteric and had a positive Murphy's sign. Her liver function test showed a total bilirubin of 5.1 mg/dl (normal, 0.2-1.0 mg/dl), aspartate aminotransferase of 359 IU/L (normal, 10-40 IU/L), alanine aminotransferase of 1115 IU/L (normal, 10-40 IU/L) and alkaline phosphatase of 281 IU/L (normal: 40-160 IU/L).

Ultrasound (USG) of the abdomen revealed a large gallstone in the gallbladder. Subsequently, she had a Magnetic resonance cholangio pancreatography (MRCP) showing 1.5 cm gallstone in the distal cystic duct and dilated intrahepatic biliary duct suggestive of Mirizzi syndrome (Figure 1). ERCP showed dilated common bile

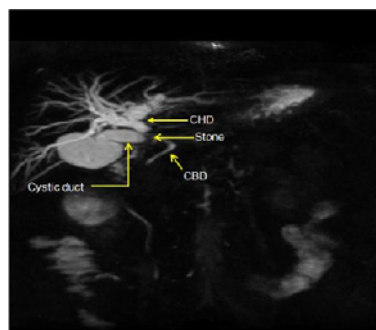


Figure 1: Magnetic resonance cholangio pancreatography (MRCP) showing 1.5 cm gallstone in the distal cystic duct and a dilated intrahepatic biliary duct suggestive of Mirizzi syndrome.

duct (CBD) and common hepatic duct with a 1.5 cm impacted stone in the cystic duct compressing CBD confirming the diagnosis of Mirizzi syndrome (Figure 2). A biliary sphincterotomy was also performed. Subsequently she had a laparoscopic cholecystectomy with stone removal. Intraoperative cholangiogram showed no obvious cholecystocholedochal fistula or stones in CBD. At the time of discharge, her liver function tests were within normal limits and she was pain free. The pathological examination revealed chronic cholecystitis with a large gallstone within the cystic duct.

Discussion

Pablo Luis Mirizzi first described Mirizzi syndrome in 1948 as a condition with obstructive jaundice due to gallstone/s impacted in the cystic duct or Hartmann's pouch, compressing the common hepatic duct [4]. Its incidence is about 0.2%-1.5% among patients with calculus of the gallbladder [2,5,6]. These gallstones can produce

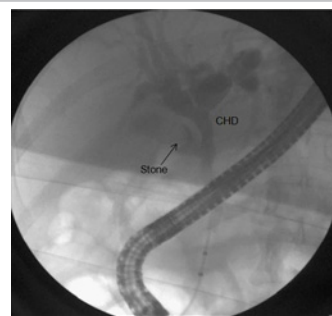


Figure 2: Endoscopic retrograde cholangio pancreatography (ERCP) reveals a dilated common bile duct (CBD) and common hepatic duct with 1.5 cm impacted gallstone in the cystic duct compressing CBD confirming the diagnosis of Mirizzi syndrome.

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CBD obstruction by two mechanisms: (1) mechanical obstruction of the hepatic duct because of the proximity of the cystic duct and the common hepatic duct, and (2) secondary inflammation with frequent episodes of cholangitis that leads to gallbladder contraction and consolidation with the common hepatic duct, or development of a fistula from pressure necrosis caused by the impacted stone. The mean age of presentation is 44 to 62 years [2]. Obstructive jaundice, fever and acute abdominal pain are the most common symptoms in these patients [2,5]. Elevated bilirubin and alkaline phosphatase levels are most often present. Associated gallbladder carcinoma can be found in about 11-28% of them [1].

McSherry and Virshup classified Mirizzi syndrome into 2 types—type I is compression of common hepatic duct or CBD by stone impacted in cystic duct or Hartmann's pouch and type II is erosion of calculus into common hepatic duct or CBD producing cholecysto-choledochal fistula [7]. Csendes et al. [8] further simplified the classification based upon the extent of fistula formation which is described as follows,

Type 1: Extrinsic compression of CBD without a cholecysto-choledochal fistula and has either an intact (IA) or obliterated (IB) cystic duct

Type 2: A cholecysto-choledochal fistula involving less than one-third of the circumference of CBD

Type 3: A cholecysto-choledochal fistula involving at least two-thirds of the circumference of CBD

Type 4: A cholecysto-choledochal fistula involving the entire biliary wall of CBD

Adequate delineation of the surgical anatomy by preoperative imaging of this condition has been shown to reduce postoperative complication rates [3,8]. The characteristic feature of Mirizzi syndrome in USG is stone impaction in gallbladder neck with dilatation of proximal biliary system. Although USG is the initial imaging modality, it has limited sensitivity of 8% to 48% in the diagnosis of Mirizzi syndrome [2,9]. Computed topography (CT) scan can identify dilated intrahepatic and extrahepatic ducts with stones in biliary tree but has poor sensitivity of about 40% to 50% [2,10]. CT scan helps in ruling out any associated gallbladder cancer, any mass lesion in the porta hepatis that cannot be detected by cholangiography.

Although ERCP has a variable sensitivity of 50% to 100% in the diagnosis of Mirizzi syndrome, it is the most accurate imaging technique to identify the cholecysto-choledochal fistula and offers therapeutic drainage of biliary tract [2,3,9]. Increasingly, MRCP has been used for the diagnosis of Mirizzi syndrome. A recent study showed that MRCP has excellent sensitivity of 92% in identifying gall stones but has poor ability to delineate cholecysto biliary fistula [9,10]. Addition of MRCP to CT increases the overall sensitivity (42% to 96%) and diagnostic accuracy (85% to 94%) for Mirizzi syndrome, respectively [10].

Surgical management remains to be the mainstay of treatment but often is associated with high complication rates (around 16%) that are mainly due to postoperative biliary leak and retained gallstone [3]. Surgical management is based on the type of Mirizzi syndrome and degree of inflammation. An intraoperative frozen section can be useful in ruling out coincident gallbladder carcinoma [1]. In general, laparoscopic or open cholecystectomy is often reserved only for type 1 Mirizzi syndrome while the presence of cholecystobiliary fistula (type 2, 3 and 4 Mirizzi syndromes) can be managed by: surgical repair of the cholecystocholedochal fistula, choledochoplasty with the gallbladder

remnant, end-to-end anastomosis over a T-tube and biliary enteric anastomosis. In type 2 Mirizzi syndrome, subtotal cholecystectomy, followed by choledochoplasty with the gallbladder remnant or end-to-end anastomosis over a T-tube can be attempted in the setting of limited inflammation. In cases of type 2 Mirizzi syndrome with severe inflammation, bilioenteric anastomosis is indicated as both choledochoplasty with the gallbladder remnant and end-to-end anastomosis over a T-tube can be complicated by the development of biliary stricture [11-14]. Bilioenteric anastomosis to the duodenum or a Roux-en-Y hepaticojejunostomy are the only two options in cases of type 3 Mirizzi syndrome [13,15]. Roux-en-Y hepaticojejunostomy is preferred in type 4 Mirizzi syndrome due to low morbidity and mortality rates on long term basis [11-13].

Conclusion

In conclusion, Mirizzi syndrome is a rare cause of obstructive jaundice that requires adequate delineation of anatomy by imaging for diagnosis and appropriate surgical treatment.

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