

Mirizzi Syndrome: Four Case Reports and a Review of the Literature

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Abstract

Mirizzi syndrome describes obstruction of the common hepatic duct by an impacted stone in the cystic duct or gallbladder infundibulum. We describe four cases, recently diagnosed and managed at our institution. We review the literature and discuss various approaches to the surgical management.

Keywords: Mirizzi syndrome; Gall-stones; Cholelithiasis; Cholecystitis

Introduction

Mirizzi syndrome describes obstruction of the common hepatic duct by an impacted stone in the cystic duct or gallbladder infundibulum [1]. Continued obstruction, inflammation and pressure necrosis can lead to fistula formation between the cystic duct or gallbladder infundibulum and the adjacent bile duct or surrounding structures [2]. It is rare complication of cholelithiasis. We review our recent cluster of four patients with Mirizzi syndrome and discuss the possible options for management.

Case 1

A 61-year old female presented to the ED with a 2-day history of abdominal pain, dark urine and alcoholic stools. Physical exam was unremarkable except for jaundice and right upper quadrant tenderness. Laboratory exam showed AST 99 U/L, ALT 283 U/L, alkaline phosphatase 168 U/L, and a total bilirubin of 9 mg/dL. Abdominal ultrasound revealed a gallbladder with numerous gallstones but no wall thickening. The CBD measured 5.6 mm. A CT scan confirmed extensive gallstones in the gallbladder with intrahepatic and extra hepatic bile duct dilatation. An ERCP showed dilated extra- and intra-hepatic bile ducts, a dilated cystic duct with very distal insertion into the common bile duct. Filling defects were noted in the cystic duct, with external compression of the common hepatic duct by the obstructed cystic duct.

The patient underwent laparoscopic cholecystectomy with intraoperative cholangiogram. Stones were milked out of the cystic duct and a completion cholangiogram confirmed no retained stones. The surgeon noted a very long cystic duct with a very long common wall with the common duct. Cholecystectomy was completed in the usual fashion.

Case 2

A 27-year old obese female presented to the ED with a 2-day history of right-upper quadrant and mid-epigastric pain. On direct questioning, she admitted to a 2-month history of back pain which had worsened over the last 2 days. She noted dark urine and loss of

appetite. Her physical exam was significant for right upper quadrant tenderness and jaundice. Laboratory examinations were notable for the following: AST 325 U/L, ALT 626 U/L, alkaline phosphatase 210 U/L and total bilirubin 6.3 mg/dL. All other laboratory tests were normal. Abdominal ultrasound demonstrated a gallbladder full of stones, including a stone visualized in the gallbladder neck. There was noted to be mild gallbladder wall thickening and a dilated common bile duct measuring 13.9 mm without evidence of intrahepatic ductal dilatation. At ERCP, there were no stones in CBD, but there was extrinsic compression from a stone in the cystic duct. An endoscopic stent was placed in the CBD. The patient underwent subtotal cholecystectomy the next day.

Case 3

A 57-year old man with history of hepatitis C presented to the ED with left lower quadrant pain, diarrhoea and jaundice. He had recently been hospitalized at another facility with jaundice and abdominal pain 3 weeks prior, but improved and had been discharged. Physical exam was notable for right upper quadrant and left lower quadrant tenderness, with the left lower quadrant pain worse than the upper pain. Labs were notable for total bilirubin of 22.7 mg/dL, AST 110, ALT 29 and alkaline phosphatase of 357. A CT scan revealed inflammatory stranding in the distal descending colon and pericolonic fat as well as a partially distended gallbladder without radio-opaque gallstones. The patient underwent ERCP, which revealed a large stone that was thought to be compressing the common hepatic duct. Review of the ERCP images demonstrated faint filling of the gallbladder. A plan for an attempted laparoscopic cholecystectomy with "milking" of the stone back into the gallbladder was made. Intraoperative. However, dense inflammation and duodenal adhesions to the infundibulum of the gall bladder precluded laparoscopic procedure. The procedure was converted to an open procedure and top-down cholecystectomy was performed after opening the gallbladder to assist with orientation. A fistulous opening from the gallbladder to the common hepatic duct was identified as was a large stone impacted in the distal common bile duct. An open common bile duct exploration was performed, T-tube placed, and the remnant gallbladder/cystic duct tissue used to close the fistulous opening in the side of the common duct.

Case 4

A 73-year-old female first presented to our hepatology clinic with a long history of isolated, asymptomatic abnormal alkaline phosphatase. Imaging studies, including an MRCP showed a dilated common bile duct as well as gallbladder stones. An ERCP suggested distal CBD obstruction. A sphincterotomy was performed and a plastic stent placed. The cystic duct and gall bladder were not filled. There appeared to be fusiform dilatation of the mid portion of the CBD. One month later the stent was removed because of pain. Alkaline phosphatase remained elevated.

Approximately 12 months later, she presented *via* the ED with a three-day history of severe right upper quadrant RUQ pain with elevated LFTs including a high bilirubin. AST 48, ALT 24, Alkaline phosphatase 193 and total bilirubin 10.4. MRCP showed cystic duct as well as GB stones, as well as wall thickening. The CBD was dilated to 15 mm and contained stones.

ERCP showed compression of the upper third of the CBD by the distended GB and cystic duct, indicating Mirizzi syndrome. Stones in the CBD were removed by balloon sweep and a stent placed. An open cholecystectomy was performed with intraoperative cholangiogram. A very inflamed GB with frank pus was found containing multiple cholesterol stones. An anomalous insertion of the cystic duct was noted, which predisposed to the Mirizzi syndrome.

Type of MS	Description	Incidence
I	External compression of the common hepatic duct by an impacted stone	11-78%
II	Presence of a cholecysto-biliary fistula involving <1/3 duct wall	15-41%
III	Presence of a cholecysto-biliary fistula involving up to 2/3 duct wall	3-44%
IV	Complete destruction of the wall of the bile duct	1-4%

Table 1: Classification of the severity of Mirizzi syndrome

The first two cases were presumed to have CBD stones. It is typically managed with laparoscopic cholecystectomy and ERCP either preoperatively or postoperatively. The advanced laparoscopic surgeon may elect to perform intraoperative laparoscopic common bile duct exploration CBE in lieu of ERCP.

In patients #1-#3, imaging with ultrasound and CT did not suggest the diagnosis of Mirizzi syndrome, prior to ERCP [11]. In patient 4, the diagnosis was made based on the combination of MRCP and ERTCP. When Mirizzi syndrome is discovered intra-operatively, the rate of conversion to open procedure and duct injury increase dramatically with one group reporting 100% conversion rates [11].

The best surgical approach and treatment of Mirizzi syndrome remains controversial, even with type 1, the least severe. Many suggest that laparoscopic cholecystectomy should not be attempted in the setting of Mirizzi syndrome due to the high rates of duct injury [12]. Current literature supports an attempt at laparoscopic surgery, with low threshold for conversion to open cholecystectomy, if the triangle of Calot is obliterated [12]. However, in the management of Mirizzi syndrome in the setting of complete duct destruction, there is little controversy that this should be managed with biliary-enteric anastomosis. The ideal biliary-enteric reconstruction is up for debate [13].

Discussion

These four cases illustrate the spectrum of severity in the Mirizzi syndrome. Mirizzi syndrome occurs through the external obstruction of the common hepatic duct, due to an impacted stone in the gallbladder infundibulum or cystic duct [1,2]. Associated cholecysto-biliary fistula can arise from pressure necrosis from the impacted stone eroding through the gallbladder or cystic duct and the common hepatic duct.

The reported incidence in the literature ranges from 0.06% to 5.7%. It is more common in women, consistent with the higher frequency of cholelithiasis in women. The incidence is also greater in specific populations with a high incidence of cholelithiasis [3,4].

Our first patient had a long cystic duct running along the common hepatic duct. A fibrous “common channel” between the two ducts likely increased the probability of Mirizzi syndrome [5].

A classification of the severity of Mirizzi syndrome has been proposed based on ERCP findings. Type 1 involves only external obstruction of the common duct, whereas type II involve fistula formation between the two ducts. The classification further grades the severity based on the degree of CBD destruction [6]. As might be expected, the more severe forms of Mirizzi syndrome are less common [7-10] Table 1.

The management of type II MS is much more varied. Some advocate for open cholecystectomy with or without biliary-enteric reconstruction and others advocate for subtotal cholecystectomy alone [14]. Most surgeons who advocate for the laparoscopic surgery will take a multidisciplinary approach when the diagnosis of Mirizzi syndrome is made preoperatively; an ERCP is performed with the placement of a stent followed by sub-total cholecystectomy [15].

Common recommendations for the surgical approach to Mirizzi syndrome include subtotal cholecystectomy with or without CBD exploration and with or without T-tube placement. A major disadvantage is the possibility of retained stones or recurrent stones in the GB remnant. Another approach is cholecystectomy with choledochoplasty over a T-tube, ERCP placed stent or nasobiliary tube. A primary concern with this approach is the possibility of developing biliary stricture. The combination, cholecystectomy with choledochoplasty with remnant gallbladder has also been tried. Finally, biliary-enteric reconstruction with CBDE, include choledochoduodenostomy, serosal patch, and other reconstructions [1,11] have been described.

Complications post treatment of Mirizzi syndrome in a large case series were most commonly post-operative bile leak and retained stones [1,8]. The low overall incidence of Mirizzi syndrome means that most general surgeons will encounter few cases in their careers.

Furthermore, most surgical trainees in the United States have little exposure to major biliary reconstruction and even less to open common bile duct exploration as the skill of our advanced endoscopists and interventional radiologists increases [16,17]. Unfortunately, because the diagnosis of MS is made intra-operatively, preoperative consultation options are often limited. If the diagnosis of MS is suspected preoperatively, especially with a fistula involving the bile duct with an anticipated reconstruction, consultation with a surgeon experienced in biliary reconstruction is recommended [16,18,19].

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