CASE REPORT

A 64 year-old man presented to our facility with a one-day history of right upper quadrant pain, scleral icterus, and intermittent fever and chills. The patient’s vital signs included a temperature of 97.8, a pulse of 71, a respiration rate of 14, and a blood pressure of 124/80. On physical exam, he was mildly uncomfortable with key physical exam findings of right upper quadrant abdominal pain to palpation and on deep inspiration, scleral icterus, and jaundice. An abdominal ultrasound demonstrated mild intrahepatic biliary ductal dilation with at least one stone visualized in the gallbladder. The wall of the gallbladder was found to be slightly thickened without pericholecystic fluid and the common bile duct was measured at 11mm. A contrast-enhanced CT of the abdomen and pelvis was obtained and revealed prominent intrahepatic ductal dilation and what was interpreted as choledocholithiasis. Notably, the common bile duct had more distension proximally near the stone and less distension distally. GI and surgery consultations were obtained. A plan of preoperative ERCP followed by laparoscopic cholecystectomy was agreed upon.

ERCP revealed intrahepatic ductal dilation and proximal extrahepatic ductal dilation. Initial cholangiogram revealed what appeared to be a single, round 30 mm filling defect consistent with a stone eccentrically obstructing at the level of the CHD (Figure 1). Immediately proximal to the stone, there was visible dilation of the left and right hepatic ducts as well as the intrahepatic ducts. An inflated 15-18mm occlusion balloon could be pulled across the site of the stone without any apparent movement of the stone itself. Of note, the cystic duct was never seen to fill and was presumed to be obstructed. At this point it became apparent that the stone was actually within the cystic duct and was extrinsically compressing the CBD, consistent with a Mirizzi syndrome. An 8.5Fr x 12cm
plastic biliary stent was inserted into the CBD across the site of extrinsic compression, with the proximal end of the stent in the CHD. (Figure 2) The patient improved clinically over the next 24 hours following biliary decompression. The patient was then referred for his cholecystectomy.

At surgery, the gallbladder was noted to be inflamed and adherent to the CBD, with what appeared to be a cholecystocholedochal fistula between the two at the level of the gallbladder neck/proximal cystic duct. Opening of the gallbladder revealed the impacted stone at the level of the fistula, which was then removed. Cholecystectomy was then carried out with subsequent incision of the CHD and removal of the impacted stone. The CHD was felt to have been damaged too severely by the stone/fistula combination for simple T-tube placement. The patient then underwent biliary reconstruction via a Roux-en-Y hepaticojejunostomy, although the distal CBD was left in continuity. The plastic bile duct stent was left in as a guide during surgery. The patient did well postoperatively and without complication.

A follow-up ERCP 8 weeks later demonstrated brisk flow of contrast through the biliary tree as well as a patent hepaticojejunostomy with free flowing contrast and bile into the small bowel. No filling defects were noted, indicating an absence of any residual or recurrent stones. Overall, the patient was felt to have developed a Type IV Mirizzi syndrome.

DISCUSSION

In developed countries, the incidence of gallstone disease has been rising of late with the majority of cases involving cholesterol stones. In the U.S. alone, an estimated 20 – 25 million adults suffer from some form of gallstone disease. Epidemiological studies have found that gallstone disease correlates highly with obesity, diabetes and metabolic syndrome. Though gallstones are understood to be multifactorial from an etiology point of view, one property these risk factors all share seems to be hypercholesterolemia: a condition that can allow supersaturation of bile by cholesterol with subsequent precipitation of gallstones. Gallstones can lead to a variety of conditions including cholangitis, cholecystitis, gallstone pancreatitis, and even gallstone ileus.

Mirizzi Syndrome (MS) is a rare complication of cholelithiasis. Classically, the common hepatic duct (CHD) or the common bile duct (CBD) become obstructed from mechanical compression and surrounding inflammation by gallstone impaction at either the cystic duct or the gallbladder neck. Compression by the impacted stone results in biliary obstruction which can then cause obstructive jaundice and in some patients also lead to cholangitis. From there, this process has the potential to further evolve into an internal biliary fistula, or even complete obliteration of the CHD.

Overall, the incidence of Mirizzi Syndrome is rare, discovered only in 0.7 – 1.4% of patients undergoing cholecystectomy. Management has historically been difficult due to the inadequacy of pre-operative diagnosis as well as the surgical challenge it presents given the
presence of active inflammation, dense adhesions, and
distorted biliary ductal anatomy.5,8

The term Mirizzi Syndrome refers to a collection of
four subtypes of the disease commonly acknowledged
as the Csendes Classification model.9 Type I is the most
dominant form of MS and is characterized by extrinsic
compression of the CBD due to an impacted stone at the
cystic duct or the gallbladder neck.4,10 Types II, III and
IV all include cholecystocholedochal fistulas.11 Type II
describes a fistula where 1/3 of the CBD circumferential
wall has eroded.4,9 Type III refers to a larger fistula
with erosion of 2/3 of the circumferential wall of the
CBD.4,9 Finally, Type IV denotes a fistula that has
cured circumferential damage to the CBD and thus
requires reconstruction of the CBD.4,9

Pathophysiology
Mirizzi Syndrome begins when a gallstone becomes
impacted at the cystic duct or gallbladder neck and
causes an inflammatory response. The offending
stone then applies external pressure on the bile duct,
eventually leading to erosion of the CBD with possible
fistulization.12

Once this situation is established, episodes of biliary
colic cause fibrosis and gallbladder atrophy.12,13 Ongoing
inflammation and proximity allow the gallbladder
and/or the cystic duct and/or the CBD to fuse together.12
An impacted gallstone at this location likely causes
a pressure ulcer which then develops necrosis. The
necrotic tissue can enable partial stone migration into
the CBD and ultimately the production of a chronic
fistula.12 Such cholecystocholedochal fistulas are the
hallmark of Type II, III and IV Mirizzi Syndrome, but
they are still exceedingly rare. In fact, most biliary
fistulas are actually cholecystoduodenal, joining the
gallbladder to the small bowel.11 Despite advanced
imaging, most fistulas are found intra-operatively
during cholecystectomy.4,11

Clinical Presentation
Mirizzi Syndrome is difficult to diagnose clinically
because of its non-specific symptom profile.5,9 Because
MS resembles several other conditions, its differential
diagnosis includes choledocholithiasis, cholecystitis,
cholangitis, gallstone ileus, and gallbladder cancer.5,11,12,14
Clinical presentation is non-specific, but obstructive
jaundice is a common symptom – especially for types
II, III and IV.4 RUQ pain is also frequently seen, along
with a fever. Laboratory values often reflect elevated
liver enzymes, hyperbilirubinemia, leukocytosis, and
a high CA-19-9.12

A reported 28% of MS types II, III and IV cases also
have also been found to have concurrent gallbladder
cancer.11,12 Notably, longstanding gallstones is one of
the primary risk factors for both MS and gallbladder
cancer.12 Average age of onset for MS is around 61
years.9 Most MS patients have had gallstone disease
for a mean of 29 years.12

Diagnosis
Lack of pathognomonic patterns or reliable clinical
or laboratory indicators have made the diagnosis of
MS difficult to distinguish from common
choledocholithiasis.5,15 Optimal management of MS
and avoidance of iatrogenic bile duct injury during
cholecystectomy depends on accurate preoperative
diagnosis, although this cannot be made in all
circumstances.10 Several modalities are available for
biliary imaging including transabdominal ultrasound,
CT, MRCP, endoscopic ultrasound (EUS) and ERCP.12
Transabdominal ultrasound (US) is typically the initial
imaging test ordered for suspected biliary disease
because it is non-invasive, although it’s ability to detect
choledocholithiasis is limited and the test is highly
operator dependent.2 US has high sensitivity to findings
consistent with MS such as gallstones in the gallbladder
neck, acute cholecystitis, a shrunken gallbladder
and a dilated CHD.10 CT scans have the capacity to
demonstrate dilated intra and extrahepatic ducts with
gallstones in the biliary tree, but in general are a poor
test to detect MS or choledocholithiasis.5 EUS is ideal
for detecting choledocholithiasis but may fail to detect
MS if the stone is located too proximally for the EUS
transducer to visualize.16 MRCP (Magnetic Resonance
Cholangiopancreatography) is noninvasive and has a
high sensitivity for finding pericholicystic inflammation
and cholecystobiliary fistulas.11 However, ERCP
(Endoscopic Retrograde Cholangiopancreatography)
is the gold standard for diagnosis of MS.9 The
sensitivity of ERCP for diagnosis of MS approaches
90%.11,12 ERCP is both diagnostic and therapeutic.11
Diagnostically, ERCP identifies ductal abnormalities
and can distinguish with precision the cause, level and
degree of biliary obstruction.10 Therapeutically, ERCP
is used for stone clearance if the stone is endoscopically
accessible.8 It also facilitates biliary decompression
prior to cholecystectomy in patients with MS via
(continued on page 16)
stent placement, as was the case in our patient. If a preoperative diagnosis of MS is not obtained, most experienced surgeons would recognize the entity in the operating room.

Treatment
The goals of treating patients with MS include: removal of the impacted stone and gallbladder, restoration of bile drainage and repair of the anatomical defect. Preoperative ERCP can confirm the presence of MS. It also allows for placement of a temporary stent – providing critical decompression of the obstructed biliary duct. Such decompression alleviates progression of any fistulas. Moreover, the stent will serve as a landmark in subsequent surgery to help minimize the risk of ductal injury. Preoperative ERCP is also a viable alternative for patients who are not candidates for open cholecystectomy (OC).

Removal of the gallbladder and the impacted stone along with repair of the malformed anatomy is performed laparoscopically or by open surgery. OC has historically been the surgical standard of care for MS. However, LC is fast gaining acceptance as a viable alternative to OC. The main advantage conferred by OC is a lower risk of iatrogenic bile duct injury than LC. This is large part due to the feasibility of a fundus first approach enabled by OC. Morbidity from OC has been reported to be 3% for MS type I. However, OC performed on types II and III correlate with a higher morbidity rate (15 – 26%) due to more complicated anatomical deformities. The biggest risk associated with OC is iatrogenic injury to the CBD. Such risk stems from the technical challenge presented by anatomical distortions, cholcystocholedocal fistulas, and adhesions in Calot’s triangle encountered in MS patients undergoing OC. Disadvantages of OC include a larger incision which provides a higher risk of infection and wound dehiscence. Additionally, OC entails a longer recovery.

Laparoscopic Cholecystectomy (LC) for treatment of MS has been somewhat controversial. Indeed, some surgeons still consider MS as a contraindication to laparoscopic cholecystectomy. Nonetheless, LC has become a viable alternative to OC as experience and enhanced instruments continue to improve outcomes with this procedure. LC involves removal of the gallbladder, the impacted stone and suturing the biliary defect. Its advantages are that it is minimally invasive and by extension entails a lower risk of infection, produces less blood loss, and offers a shorter hospital stay. Some disadvantages are that it is technically demanding, requires a high degree of expert skill and thus a longer duration for the procedure.

In general, LC is indicated for the less complicated MS as in Type I. Open surgery is indicated for higher risk cases of MS, difficult dissections (due to anatomical malformations or acute inflammation), and patients who have failed LC. In some cases, higher grades of MS: Type II, III and IV, may require open biliary-enteric bypass to repair extensive destruction of the bile ducts or strictures. Such complete biliary reconstruction is termed a Roux-en-Y hepatico-jejunostomy and involves anastomosis of the jejunum with the fistula to close the eroded duct. Lastly, postoperative ERCP after complete healing of the site removes the temporary stent. The incidence of iatrogenic complications is reported to range between 0 – 14%. Such adverse events can be severe and include cholangitis, bile leakage from perforation of the CBD or perforation of the gallbladder, bile duct strictures from fistula repair, post-ERCP pancreatitis, and sepsis. Regardless of the intervention, a high risk of morbidity exists with treatment of MS.

CONCLUSION
Mirizzi Syndrome is a rare clinical entity that presents a formidable challenge when encountered. The constellation of a contracted gallbladder, obstructive jaundice, gallstones, pain and cholecystitis should arouse suspicion of MS. A preoperative diagnosis is critical to optimal management and is best confirmed by cholangiography during ERCP, although not all patients have the diagnosis made prior to interventions. Once a definitive diagnosis of MS is made, a multidisciplinary team approach including a skilled endoscopist and surgeon offers the best outcome. Treatment for each patient must be individualized, as specific MS types necessitate distinct surgical approaches. Preoperative ERCP with stenting can restore bile flow and mitigate cholangitis, thus enhancing safety of subsequent surgery.

References


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