

## Incidence and Surgical Strategies in Management of Mirizzi Syndrome

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**Abstract:** Mirizzi syndrome (MS) is a rare complication of prolonged gallstone disease, characterized by narrowing of the common hepatic duct (CHD) due to mechanical compression and/or various grade of inflammation due to biliary calculus impacted in the neck of the gallbladder or in the cystic duct, presentation sometimes varies from obstructive jaundice associated with extrinsic compression or, when the stones migrate through the cystic duct, to the presence of cholecystobiliary fistula. From February 2008 to July 2011, 400, patients with a diagnosis of cholelithiasis were included in a retrospective review of case notes. According to the observations in the operation, 12, patients with the final diagnosis of MS, who were managed in our surgical department, were retrospectively evaluated. The patients who presented with an initial diagnosis of MS and were later found to suffer from a malignancy such as Klatskin tumor or gallbladder carcinoma were excluded from the study. Patient's demographic variables, clinical presentation, laboratory findings, diagnostic modalities, presence of choledocholithiasis, therapeutic procedures and postoperative complications were evaluated. During the study period, 400 cases underwent cholecystectomy. Incidence of MS diagnosis among the patients was (3 %). There were (33.3 %) male and (66.6 %) female, with a mean age of 46.5 years (range: 30-70years). According to Csendes classification (41.6 %) were classified as type I; 6 (50 %) as type II, and 1 (8.3 %) as type III no patient was determined as type IV. In conclusion, preoperative diagnosis is essential, the only risk for MS is gallstones in the patients with cholelithiasis, the diagnosis of MS should be definitive before operation and therefore detailed evaluation must be performed. In addition, good outcome can be achieved with an appropriate surgical procedure, although open surgical procedure is safer and preferred, laparoscopic procedure is still the gold standard in the management of MS, especially for type I variety.

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**Key words:** Mirizzi Syndrome, cholecystocholedochal fistula, Roux en-Y hepaticojejunostomy.

### 1. Introduction

Mirizzi syndrome (MS) is a rare complication of prolonged gallstone disease, characterized by narrowing of the common hepatic duct (CHD) due to mechanical compression and/or various grade of inflammation due to biliary calculus impacted in the neck of the gallbladder or in the cystic duct, presentation sometimes varies from obstructive jaundice associated with extrinsic compression or, when the stones migrate through the cystic duct, to the presence of cholecystobiliary fistula. In 1948, Pablo Mirizzi first described a patient with partial obstruction of the CHD as "functional hepatic syndrome" and this presentation became known as Mirizzi syndrome<sup>(1)</sup>.

In 1982, McSherry *et al.*<sup>(2)</sup> initially classified Mirizzi syndrome into two types. Type I involves the external compression of the common hepatic duct due to a stone impacted in the neck of the gallbladder or the cystic duct. Type II refers to cholecystocholedochal fistula and stone migration into the common hepatic duct. A further modification of this classification was suggested in 1989 by Csendes *et al.*<sup>(3)</sup> in which, type II is an obstruction that involves less than one-third of the bile duct,

Type III is an obstruction involving up to two-thirds of the duct, and Type IV is complete obstruction of the bile duct<sup>(4)</sup>. The prevalence of this rare complication varies from 0.3 to 3%<sup>(5)</sup>.

In clinical practice, the diagnosis is rarely made preoperatively. The diagnostic approach usually begins with ultrasonography followed by cholangiography via direct cholangiography, endoscopic retrograde cholangiopancreatography, or magnetic resonance cholangiography. Endoscopic retrograde cholangiopancreatography (ERCP) is the procedure of choice to establish the diagnosis and to classify the lesion<sup>(4)</sup>.

The surgical management of MS has more difficulties compared to the cholelithiasis without any complication. Especially dangerous anatomic alterations make the surgical intervention more complex and risky. We therefore emphasize the importance of preoperative determination of surgical strategies and optimal surgical intervention when diagnosis is done at the time of operation, particularly for those whose classification was high grade<sup>(6)</sup>.

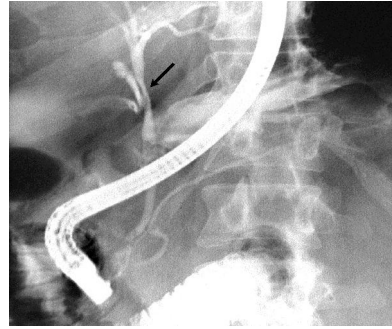
### 2. Material and Methods

From February 2008 to July 2011, 400 patients with a diagnosis of cholelithiasis were included in a retrospective review of case notes. According to the observations in the operation, 12 patients with the final diagnosis of MS, who were managed in our Surgical Department, were retrospectively evaluated. The patients who presented with an initial diagnosis of MS and were later found to suffer from a malignancy such as Klatskin tumor or gallbladder carcinoma were excluded from the study. Patient's demographic variables, clinical presentation, laboratory findings, diagnostic modalities, presence of choledocholithiasis, therapeutic procedures and postoperative complications were evaluated. To determine the surgical procedure, Csendes classification was applied prospectively to the patients, generally considering preoperative and/or intraoperative observations.

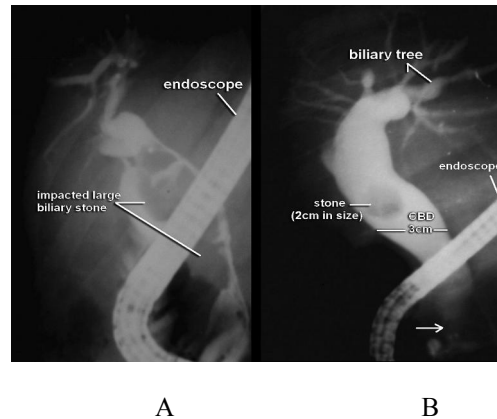
### 3. Results

During the study period, 400 cases underwent cholecystectomy. Incidence of MS diagnosis among the patients was (3 %), 12 patients. There were 4 male (33.3 %) and 8 female patients (66.6 %), with a mean age of 46.5 years (range: 30-70years). Jaundice and itching were the major symptoms, followed by abdominal pain. 8 patients (66.6 %) presented with jaundice and 4 of these patients (50%) had associated cholangitis. The most frequent signs and symptoms are jaundice, itching, abdominal pain, vomiting, nausea and fever. Only 4 patients (33.3%) experienced recurrent symptoms of epigastric or right hypochondrial pain during the last year prior to presentation. Mean total bilirubin in the patients with jaundice was 3.8 mg/dl (range: 1.7-7.7 mg/dl), SGPT (range: 40-450 u/l), SGOT (range: 10-600 u/l) and ALP (range: 70-950 u/l).

All the patients subjected to ultrasonography initially, which revealed the gallstones in both the common bile duct (CBD) and gallbladder-associated dilatation of the intrahepatic bile ducts in 7 patients (58.3 %), whereas presence of gallstones only in the gallbladder was reported for 3 patients (24.9%). The remaining 2 patients were aggressively investigated using CT and MRCP. The other diagnostic modality, endoscopic retrograde cholangiopancreatography (ERCP), was performed in 9 patients (74.9%), and MS was suspected in 6 patients, preoperatively (figure,1&2). The remaining 3 patients who underwent ERCP could be diagnosed intraoperatively.



Figure(1):Image obtained during endoscopic retrograde cholangiopancreatography shows smooth narrowing of the bile duct (arrow) at the site of insertion of the cystic duct (Mirizzi syndrome). Note the small calculus in the cystic duct.



Figure(2):A) A dilated CBD due to impaction of a huge biliary stone; cholangiography was performed through a fistula tract just above the papilla. B) obstruction of the distal CBD (white arrow) and proximal bile duct dilatation, which also included a biliary stone.

According to Csendes classification 5 patients( 41.6 %) were classified as type I; 6 ( 50 %) as type II, and 1 ( 8.3 %) as type III no patient was determined as type IV. Open cholecystectomy without additive surgical procedure was performed in 4 (33.3 %) patients with type I, and laparoscopic cholecystectomy was attempted in (1) of these patients and converted to the open technique because of dense adhesions and distorted anatomy due to edematous tissue and inflammatory process.

The open technique, fundus-first cholecystectomy, was applied to this case. 4 patients with type II, underwent cholecystectomy, choledochotomy and insertion of T-tube for biliary drainage through a separate choledochotomy (33.3 %). The patient with type III and the remaining

2 patients (16.6 %) with type II underwent cholecystectomy with excision of the external bile ducts and reconstruction with Roux en-Y hepaticojejunostomy.

The mean hospital stay was 8.3 days (range: 4-18 days). There were no complications including biliary problems in the early postoperative period. However, in the late postoperative period, 2 patient (16.6 %) one type I and one type II were diagnosed with benign biliary stricture, in the twelve postoperative month, and was managed with Roux en-Y hepaticojejunostomy, because recurrence is common after balloon dilatation.

#### 4. Discussion

Mirizzi syndrome is a rare complication (frequency about 1%) of chronic cholecystitis and prolonged cholelithiasis, which consists of inflammatory process of gallbladder wall and direct compression (Mirizzi syndrome type I) or erosion (Mirizzi syndrome type II, III, IV) of the common bile duct and subsequent fistulous formation<sup>(7)</sup>.

After Mirizzi<sup>(1)</sup> and McSherry *et al.*<sup>(2)</sup> classification, the most recent and predominant classification was made by Csendes *et al.*<sup>(3)</sup>. According to it, Mirizzi syndrome is divided in four types depending on the size of the destruction of the common bile duct<sup>(8)</sup>. A wide communication that included the entire circumference of the CBD (Over 66% of the common bile duct diameter) classifies this case as Mirizzi syndrome type IV<sup>(9)</sup>.

MS, which is a rare condition, has remained a mystery for preoperative confirmation of the diagnosis, which is the cornerstone in determining the surgical procedure to be used. The mechanism of the pathology includes two possible explanations: (a) Chronic and/or acute inflammatory changes due to impacted gallstone causes stenosis of the CHD, or (b) the impact of the gallstones leads to cholecystocholedochal fistula formation associated with necrosis of the adjacent ductal walls<sup>(10)</sup>.

There are also anatomical predispositions that are comprised of the presence of a long cystic duct in parallel with the CHD or a low insertion of the cystic duct into the CBD<sup>(11)</sup>.

In a large study (219 patients), Csendes *et al.* reported that 11% of their patients with MS had type I lesions, 41% had type II, 44% had type III, and 4% had type IV<sup>(3)</sup>. In other study, no type IV lesions were detected, and most of the patients had type II. The incidence of MS was approximately 0.3-3% of all patients undergoing cholecystectomy and in 0.1% of all patients with gallstone disease<sup>(12)</sup>.

In this study, no type IV lesions were detected, and most of the patients 6 patients had type II. The incidence of MS was approximately 3% of all

patients, the patients had type I was 5 and type II was 1 patient.

Clinical manifestation of this syndrome includes recurrent cholangitis, jaundice, right upper quadrant pain, and abnormal liver biochemistry. It is remarkable that this patient was an icteric<sup>(13)</sup>. The most frequent signs and symptoms in our study are jaundice, itching, abdominal pain, vomiting, nausea and fever. Mirizzi syndrome is a rare complication of cholelithiasis that represents a dangerous alteration of the anatomy and bears the potential to lead to significant morbidity and biliary injury, particularly in the laparoscopic era. In this condition the cystic duct usually runs parallel to the CBD (10% of cholangiograms)<sup>(14)</sup>.

Preoperative recognition of this variation is important to avoid inadvertent ligation or severance of the bile duct. An attempt to expose Calot's triangle may lead to severe bile duct injury such as: i) iatrogenic communication between the gallbladder and CBD, ii) complete transection of CBD after dissection of the gallbladder neck, iii) tear of CBD. Additionally, a high coincidence of Mirizzi syndrome and gallbladder cancer has been reported by Prasad *et al.*<sup>(15)</sup>.

The intraoperative confirmation of Mirizzi syndrome was made after the extraction of the large gallstone. The contracted gallbladder, the inflammatory wall and the subsequent adherence with the common bile duct (CBD) resulted to the wide communication (fistulas diameter: = 3.5 cm). After temporary stenting of the CBD through the fistula opening we classified the condition as Mirizzi syndrome type IV which represents the most uncommon type<sup>(3)</sup>.

The surgical technique depends on the type of MS. If the type of MS has not been classified preoperatively, the best way to determine the operative procedure is "fundus-first" technique<sup>(16)</sup>, which relieves the fistula formation via permitting the reflux of the bile as an indicator of it. Most inflammatory strictures return to normal when the inflammatory process resolves. Otherwise, retrograde dissection is contraindicated due to risks of injury to the Callot's triangle in the presence of inflammation resulting in adhesions and distorted anatomy. In addition to observation, examination of the intraoperative cholangiography helps to detect not only CBD stones but also presence of the fistula and its size<sup>(17)</sup>.

The surgical treatment of type I MS generally involves minimal interventions such as partial or total (open or laparoscopic) cholecystectomy. The reported incidence rate of conversion to open cholecystectomy was remarkably high, with a range of 37-78%<sup>(18)</sup>, and the incidence of 100 % in our

study was well correlated with the literature. However, some authors consider this a contraindication for laparoscopic cholecystectomy<sup>(19)</sup>.

In our study, the type I cases underwent only cholecystectomy, while type II needed T-tube insertion with choledochotomy after cholecystectomy for temporary decompression. Except 2 cases needed reconstruction with Roux en-Y hepaticojejunostomy with type III case.

Type II defects can usually be treated successfully with either complete or partial cholecystectomy followed by closure of the fistula with T-tube placement in CBD. However, isolated cholecystectomy in the patients with type II should be avoided to prevent likely postoperative biliary complications<sup>(20)</sup>.

In the present study, most of the patients 50% with type II were submitted to cholecystectomy with choledochotomy and insertion of T-tube for drainage. The remaining cases with type II and of the type III MS cases underwent cholecystectomy with excision of the external bile ducts and reconstruction with Roux-en-Y hepaticojejunostomy. There were no postoperative complications including biliary leak. However, in the late postoperative period, 2 patient (16.6%) one type I and one type II were diagnosed with benign biliary stricture, in the MS patients and was managed with Roux en-Y hepaticojejunostomy.

The cholecystoenteric fistula usually develops insidiously, and an association with gallstones is always present. Cholecystoenteric fistula appears in 1.2 to 5% of patients with acute and chronic cholecystitis in a large series of cholecystectomies. About 50% of diagnosis for biliary-enteric fistulas is preoperative. ERCP is the most important diagnostic tool in order to identify the presence of biliary-intestinal fistula. Two other significant diagnostic methods MRI and MRCP set up a high index of suspicion revealing the fistulous tract<sup>(21)</sup>. In this case, colonoscopy was unable to detect the fistulous tract, but the notification of the inflammatory area of the hepatic flexure was very helpful in the overall diagnosis. A causative correlation between Crohn's disease and cholecystocolic fistula formation was also precluded in this case, by colonoscopy<sup>(22)</sup>. The most common type of biliary-enteric fistula is cholecystoduodenal (75%); cholecystocolic is next common (10–20%), with a variety of other types being less frequent (15%)<sup>(23)</sup>.

### Conclusion

In conclusion, to determine the best surgical procedure in order to well-manage the condition, preoperative diagnosis is essential. Because the only

risk for MS is gallstones in the patients with cholelithiasis, the diagnosis of MS should be definitive before operation and therefore detailed evaluation must be performed. Good outcome can be achieved with an appropriate surgical procedure. Although open surgical procedure is safer and preferred, laparoscopic procedure is still the gold standard in the management of MS, especially for type I variety. In the operation, which is a challenge for the surgeons, bile duct injury, which can occur easily, can be avoided with a judicious approach during dissection of Callot's triangle and early recognition of its presence. In the current study, the operative procedure of choice in the patients with type I MS without a fistula was cholecystectomy.

In the cases with type II-IV varieties, who require more complex surgical approaches, T-tube insertion or bilioenteric anastomosis, especially Roux-en-Y hepaticojejunostomy, following cholecystectomy is a more preferable technique because it is safer and provides good long-term results with low morbidity and mortality rates. The major factor for successful treatment is primarily a good preoperative evaluation to determine the best surgical procedure considering the type.

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