MIRIZZI SYNDROME:
A DIAGNOSTIC AND SURGICAL PROBLEM

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Abstract

This study was conducted at King Saud Hospital (350 beds), Al-Qassim Region, Saudia Arabia; between December 2000 till May 2005. The aim of this work is to describe a series of 13 patients presented with obstructive jaundice and proved to have Mirizzi syndrome, at a single institution, submitted to surgical treatment and to comment on their aspects with emphasis on the diagnosis and treatment. The following items were evaluated: clinical presentation, laboratory results, preoperative evaluation, operative findings, type of Mirizzi syndrome according to the classification of Csendes et al, 1989, choice of operative procedures, and complications.

The study comprised 13 patients (5 males & 8 females with mean age 58.2±9.3 years) with MS detected out of 1834 patients (0.7%) treated for cholelithiasis during the period of the study. Preoperative radiological examination succeeded in the diagnosis of MS in 6 cases; 2 cases by ultrasonography (15.4%), 4 cases by ERCP (30.8%), one out of 4 cases by CT (25%) and one out of 4 cases by MRCP (25%) and failed to diagnose 7 cases with a success rate of 46.2%. Surgical exploration through a right subcostal incision detected the presence of impacted stone in the infundibulum of the gallbladder or in the cystic duct of the all patients; there were 4 patients (30.8%) with MS type I, 3 patients (23.1%) had MS type II, 2 patients (15.4%) had MS type III and 4 patients (30.8%) had MS type IV. The surgical procedure done was cholecystectomy for patients with type I MS, Cholecystectomy, primary closure of the cholecysto-biliary fistula and T-tube drainage of CHD for patients with MS type II. Patients with MS type III underwent cholecystectomy and choledocho-
duodenostomy, while cholecystectomy and Roux-en-Y hepatico-jejunostomy were done for patients with MS type IV. Liver function tests returned to normal values in all patients within 43.8±20.7 days (range: 30-70 days postoperatively) and the mean duration of post-operative follow-up was 20.7±12.8 months (range: 6-48 months) with no post-operative major procedure-related complications or mortality.

It could be concluded that MS is an uncommon form of benign obstructive jaundice identified with a frequency of 0.7% of patients with cholelithiasis. The preoperative diagnosis of Mirizzi syndrome is difficult and an awarded suspicion is necessary to avoid injuries of the biliary tree. The problem may only become evident during the operation due to firm adhesions around Calot’s triangle. The success of the treatment is related to a precocious recognition of the condition during surgery, and adapting the management according to the individual characteristics of each case.

**Introduction**

In 1948, P. L. Mirizzi described an unusual presentation of gallstones which, when lodged in either the cystic duct or the Hartmann pouch of the gallbladder, externally compressed the common hepatic duct (CHD), causing symptoms of obstructive jaundice (Mirizzi, 1948). Impaction of a large gallstone (or multiple small gallstones) in the Hartmann pouch or cystic duct results in the Mirizzi syndrome in 2 ways: (1) Chronic and/or acute inflammatory changes lead to contraction of the gallbladder, which then fuses with and causes secondary stenosis of the CHD, or (2) large impacted stones lead to cholecystocholedochal fistula formation secondary to direct pressure necrosis of the adjacent duct walls. (Pemberton and Wells, 1997 and Hazzan et al, 1999).

The presence of a long cystic duct in parallel with the CHD or a low insertion of the cystic duct into the common bile duct (CBD) also increases the likelihood of this syndrome. Mirizzi syndrome occurs in approximately 0.7-1.4% of all patients undergoing cholecystectomy and in 0.1% of all patients with gallstone disease (Pemberton and Wells, 1997 and Hazzan et al, 1999). Extensive ad-
hesions may make visualization of the biliary anatomy exceptionally difficult, especially within the hepatoduodenal ligament. Consequently, the CBD may be mistaken for the cystic duct, and ligation or permanent injury may occur during surgery (Becker et al, 1984). Postoperative bile leakage may occur if a fistula is not recognized; rarely, this may result in bile peritonitis (Schafer et al, 2003).

CSENDES et al;1989, classified Mirizzi syndrome (MS) into four types: type 1 represented by the presence of extrinsic compression of the common biliary duct by calculus in the area of the infundibula of the gallbladder or in the cystic duct; type II, characterized by cholecystobiliary fistula with the diameter of the orifice less than 1/3 of the circumference of the common biliary duct; in type III, the orifice of the cholecystobiliary fistula has a diameter up to 2/3 of the circumference of the common biliary duct; and in type IV, the cholecystobiliary fistula involves the entire circumference of the common biliary duct wall with the inflammatory process (Fig.1 and Table 1)

The objective of this study was to describe a series of 13 patients with Mirizzi syndrome, submitted to surgical treatment, and to comment on aspects of the etiopathogenesis and clinical presentation, with emphasis on the diagnosis and treatment of this complication of biliary lithiasis.

Patients & Methods
This study was conducted at King Saud Hospital (350 beds), Al-Qassim Region, Saudia Arabia; since December 2000 till May 2005. The inclusion criteria was characterization of MS by the presence of cholecystolithiasis and dilatation of the common hepatic duct, above the level of biliary obstruction caused by impacted calculus in the cystic duct or in the gallbladder infundibula and/or inflammatory process due to the presence of biliary stones with a normal common duct diameter below the obstruction with or without cholecysto-biliary fistula. Patients enrolled in the study underwent clinical evaluation; estimation of serum total and direct bilirubin, aspartate transferase
(AST), alanine transferase (ALT) and alkaline phosphatase (ALP). Abdominal ultrasonography (US) and endoscopic retrograde cholangio-pancreatography (ERCP) were the initial imaging studies for all patients of the study. CT and magnetic resonance cholangiopancreatography (MRCP) were done for 4 patients for whom ERCP was done and CBD stones were extracted but jaundice persisted after ERCP.

All patients underwent surgical exploration for assuring the diagnosis provided by the preoperative diagnostic modalities and appropriate operative procedure was undertaken. All specimens excised were sent for histopathology. Postoperative complications and liver functions tests were recorded.

**Results**

The study comprised 13 patients (0.7%) with Mirizzi syndrome detected out of 1834 patients treated for cholelithiasis during same period. There were 5 males (38.5%) and 8 females (61.5%) with mean age 58.2±9.3 years (range: 45-77 years). The presenting symptom was jaundice. Eleven patients presented with painless jaundice but with history of biliary colic and 2 patients presented with cholangitis that resolved with antibiotic therapy. Liver function tests (LFT) showed elevated levels of total and direct bilirubin, alkaline phosphatase (ALP), AST and ALT. The mean value for total bilirubin was 140.7umol/L (range from 71 to 204 umol/L), for ALP it was 1115.5 U/L (range from 920 to 1380 U/L), for AST it was 150.5 U/L (range from 70 to 230 U/L) and for ALT it was 213.3 U/L (range from 76 to 310 U/L) (Table 2).

Ultrasonography showed evidence of chronic calculic cholecystitis, dilated intrahepatic and extrahepatic biliary ducts in all patients. In two patients, the dilatation could be traced to its beginning at the level of the CHD, where an impacted stone was seen in the infundibulum of the gallbladder suggestive of Mirizzi syndrome (Fig.2), (Table 3). ERCP confirmed the diagnosis of Mirizzi syndrome that was suggested by ultrasonography and also diagnosed another two cases (total 4
cases) as Mirizzi syndrome (Figs.4,5 and 6). In the remaining 9 cases (Table 4), the cause of obstructive jaundice as diagnosed by ERCP was stone CBD. Extraction of CBD stones succeeded in 4 cases, however, serum bilirubin was increasing in those 4 cases after stone extraction, so they were subjected to CT and MRCP. CT diagnosed one case as Mirizzi syndrome (1 out of 4) (Fig. 3), and MRCP reached the diagnosis of Mirizzi syndrome in another case (1 out of 4) (Figs. 7 and 8), (Table 3).

All cases were reported as type 1 Mirizzi syndrome. So pre-operative diagnosis of Mirizzi syndrome was reached in 6 cases (46.2%), 2 cases by ultrasonography (15.4%), 4 cases by ERCP (30.8%), one out of four cases by CT (25%), and one out of four cases by MRCP (25%).

Surgical exploration was undertaken through a right subcostal incision in all patients; intraoperative exploration confirmed the presence of impacted stone in the infundibulua of the gallbladder or in the cystic duct of all the patients included in the study and were diagnosed as having Mirizzi syndrome. According to intraoperative findings, MS type 1 was found in 4 cases (30.8%), type 11 in 3 cases (23.1%), type 111 in 2 cases (15.4%) and type IV in 4 cases (30.8%), (Table 5).

In type 1 MS, cholecystectomy, as an isolated surgical procedure, was performed in 3 patients, where the cystic duct could be isolated and intra-operative cholangiography (IOC) was done and showed that the biliary ducts were free of stones. Partial cholecystectomy was done for the 4th case and IOC was performed by puncture of CHD by fine needle and biliary ducts were free of stones.

For patients with MS type 11, cholecystectomy was done and the fistula was closed with interrupted vicryl 3/0 sutures. A T-tube was put in common hepatic duct above the fistula.

For patients with MS type 111, cholecystectomy and choledocho-duodenostomy were done, while cholecystectomy and Roux-en-Y hepatico-jejunostomy were done
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for patients with MS type 1V, (Table 5).

The main difficulty encountered during surgery was the dense inflammatory adhesions in Calot's triangle, so dissection was done with extreme care in order to avoid injury of the main bile ducts. The fundus first technique for cholecystectomy was done for all patients. The gallbladder was opened at its fundus and gallstone was removed. A cholecysto-biliary fistula was suspected if a gush of bile into the gallbladder occurred, so IOC was done by inserting a Foley's catheter into the neck of the gallbladder to confirm the presence of the fistula and to check the biliary tree (Fig.9). All specimens were sent for histopathology and no carcinoma of gallbladder was detected for all cases.

There were no major procedure-related complications. All patients were monitored with serial liver function tests. Liver function tests returned to normal values in all patients within 30 to 70 days postoperatively. The mean duration of post-operative follow-up was 20.7 months (range: 6 to 48 months).

Table (1): Classification of Mirizzi syndrome, (Csendes et al., 1989)

<table>
<thead>
<tr>
<th>Csendes type</th>
<th>Characteristics</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I</td>
<td>Extrinsic compression in the common hepatic duct by stone generally impacted in the cystic duct or in the infundibula of the gall bladder.</td>
</tr>
<tr>
<td>Type II</td>
<td>Presence of cholecystobiliary fistula with a diameter one-third of circumference of the common hepatic duct wall</td>
</tr>
<tr>
<td>Type III</td>
<td>Presence of cholecystobiliary fistula with a diameter two-thirds of circumference of the common hepatic duct wall</td>
</tr>
<tr>
<td>Type IV</td>
<td>Presence of cholecystobiliary fistula that involves the entire circumference of the common hepatic duct wall</td>
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</table>

Table (2): pre-operative liver function tests

<table>
<thead>
<tr>
<th>Test</th>
<th>Mean±SD</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Alkaline phosphatase (U/L)</td>
<td>1115.5</td>
<td>920-1380</td>
</tr>
<tr>
<td>AST (U/L)</td>
<td>150.5</td>
<td>70-230</td>
</tr>
<tr>
<td>ALT (U/L)</td>
<td>213.3</td>
<td>76-310</td>
</tr>
<tr>
<td>Total bilirubin (umol/L)</td>
<td>140.7</td>
<td>71-204</td>
</tr>
</tbody>
</table>
Table (3): Preoperative radiological diagnosis of MS

<table>
<thead>
<tr>
<th>Radiologic Study</th>
<th>MS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ultra sonography</td>
<td>2/13 (15.4%)</td>
</tr>
<tr>
<td>C.T</td>
<td>1/4 (25%)</td>
</tr>
<tr>
<td>MRCP</td>
<td>1/4 (25%)</td>
</tr>
</tbody>
</table>

Table (4): ERCP Findings:

<table>
<thead>
<tr>
<th>Findings</th>
<th>No.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mirizzi syndrome</td>
<td>4 (30.8%)</td>
</tr>
<tr>
<td>CBD Stones</td>
<td>9</td>
</tr>
<tr>
<td>CBD Stone Extraction</td>
<td>4</td>
</tr>
<tr>
<td>Failed stone extraction</td>
<td>5</td>
</tr>
</tbody>
</table>

Table (5): Operative procedures done for patients with Mirizzi syndrome

<table>
<thead>
<tr>
<th>Csendes type</th>
<th>Operative procedure</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I</td>
<td>Cholecystectomy</td>
<td>3 (23.1%)</td>
</tr>
<tr>
<td></td>
<td>Partial cholecystectomy</td>
<td>1 (7.7%)</td>
</tr>
<tr>
<td>Type II</td>
<td>Cholecystectomy and fistula closure with a T-tube drainage of common hepatic duct</td>
<td>3 (23.1%)</td>
</tr>
<tr>
<td>Type III</td>
<td>Cholecystectomy &amp; choledochoduodenostomy</td>
<td>2 (15.4%)</td>
</tr>
<tr>
<td>Type IV</td>
<td>cholecystectomy and Roux-en-Y hepatico-jejunostomy</td>
<td>4 (30.8%)</td>
</tr>
</tbody>
</table>

Total: 13 (100%)
FIGURE 2: Ultrasonography of biliary tract. Impacted stone in the infundibula of the gallbladder (large arrow) and dilatation of the common hepatic duct (small arrow).

Figure 3: CT Showing a stone in the cystic duct "arrowed" with compression of the CHD with intrahepatic biliary ducts dilatation.
Fig. 4 ERCP showing a big stone at cysto-hepatic junction and two smaller stones are impacted in the cystic duct.

Figs 5 and 6 demonstrate stone at the cysto-hepatic junction.
Fig. 7 MRCP showing a 1.2-cm stone (arrow) resulting in biliary ductal dilatation. Gallbladder calculi are also seen (arrowheads).

Fig. 8 MRCP obtained 5mm anterior to Fig 5 shows two calculi in the dilated cystic duct (arrowheads) which parallels the extrahepatic bile duct. The inferior stone (arrow) corresponds to the calculus seen in Fig 5; this stone eroded through the wall of the cystic duct in to the extrahepatic bile duct, bridged the two structures, and resulted in obstruction of the bile duct (Mirizzi syndrome).

Fig. 9. Intra-operative cholangiogram with Foley's catheter at the neck of gall bladder demonstrating a fistula. Note absence of cystic duct and direct opposition of the neck of the gall bladder to the CHD with filling of the biliary tree.
Discussion

In 1948, the Argentinean surgeon PABLO MIRIZZI described a patient with partial obstruction of the common hepatic duct secondary to impacted biliary stone in the cystic duct or in the infundibula of the gallbladder, associated to an inflammatory response involving the cystic duct and the common hepatic duct. This presentation became known as Mirizzi syndrome.

Mirizzi syndrome is a rare complication of prolonged cholelithiasis. It presents a spectrum that varies from extrinsic compression of the common hepatic duct to the presence of cholecystobiliary fistula. For this reason, the disease represents a dangerous alteration in the anatomy during the performance of cholecystectomy, by predisposing the patients to the risk of an inadvertent lesion of the common hepatic duct (Martin and Rossi, 1994 and Nunes et al, 2000)

Throughout a period of about 5 years, 1834 patients were admitted to and treated for cholelithiasis; out of these patients 13 patients were found to have Mirizzi syndrome with a frequency rate of 0.7%. This reported frequency supported the fact that Mirizzi syndrome is a rare condition and goes in hand with Chan et al., (2003), who identified 18 cases of MS through a period of 5 years and with Yeh et al., (2003) who reported a frequency of 0.24% of MS among their series. Also, Tan et al., (2004), reported 24 MS cases among a total of 1881 cholecystectomies through a period of 5 years. In the present study, all patients were jaundiced; This mode of presentation coincided with that previously reported by Johnson et al., (2001) and Penkov, (2003).

In the present study, the percentage of MS type 1 was 30.8%, type II, 23.1%, type III, 15.4% and type IV, 30.8%. There is great discrepancy in the percentage of
types of MS in different studies. Csendes et al, 1989, in their study that included 219 patients with MS reported that the majority of cases were of MS type 111 (44%), MS type 11 (41%), MS type 1 (11%) and MS type 1V (4%). Tan et al, 2004 reported that MS type 1 represented the majority of cases (19 out of 24 i.e. 79.2%).

The diagnosis can be suggested when US or abdominal CT features reveal biliary stones in the junction of the cystic and common hepatic ducts and associated with dilatation of the proximal biliary tree (Hilger et al,1988). However, a dilated cystic duct can be confused with the common hepatic duct of normal diameter (Schreiber et al,1988). thereby hindering the diagnosis of this condition. MRCP is a useful method for diagnosis of Mirizzi syndrome. MRCP can demonstrate with precision the presence of biliary dilation, the degree of obstruction, the intraluminal or external location of the biliary stones, and also identifies and evaluates the degree of inflammation around the gallbladder. It can also can reveal extrinsic narrowing of the common bile duct, complications such as fistula and helps to show anatomical variants and malformations (Becker et al,1997; and Fulcher et al,1999).

Some authors consider preoperative diagnosis of MS is essential in avoiding bile ducts injuries (Baer et al,1990 and Dewar et al, 1990). Others concluded that with a cautious intra-operative approach to periductal inflammation and judicious dissection, preoperative diagnosis of MS is not necessary for successful management (Curet and Rosendale, 1994 and Vezakis et al, 2000). Additional imaging is often needed to obtain details of the biliary pathology because of lack of sensitivity of US and CT scans in discerning the underlying pathology (Johnson et al, 2001). The most frequently used modality was ERCP and this was successfully diagnostic in 4 of our patients,(30.8)%. All cases were reported as MS type 1. In the present study US was helpful in diagnosis of 15.4% of cases of MS, CT in 25% of cases and MRCP in 25% of cases. In the study of Shah et al 2001, US was helpful in diagnosis of MS in 14.7% and
ERCP was helpful in diagnosis of 50% of cases. Janes et al, 2005 stated that ERCP is the primary method of diagnosing fistulæ, and also has an important therapeutic role including stone retrieval and stent placement. ERCP diagnosed 50% of cases of MS in the study of Tan et al, 2004 which included 24 cases of MS. He reported that US and CT failed to reach the diagnosis of any case of MS during the whole study.

In the present study, the surgical strategy aimed at tackling the two difficult problems when faced with MS - firstly, the safe completion of the cholecystectomy without inflicting injury to the bile duct; secondly, the appropriate management of the choledocho-choledochal fistula. During cholecystectomy, the fundus first approach is favoured over the conventional Calot’s first dissection. In acute cholecystitis or when the gallbladder is distended and tense, decompressing it can facilitate dissection. A cholangiogram should be done to confirm the diagnosis, to assess the location and size of the fistula, as well as to exclude the presence of stones or strictures in the bile duct. In Type 1 MS, the minimum necessary surgery (a cholecystectomy) is adequate. In the absence of CBD stones on pre or per-operative cholangiogram, stones impacted in the cystic duct or the neck of the gallbladder are milked back into the gallbladder which is then removed and the cystic duct oversewn (Pemberton and Wells, 1997 and Johnson et al, 2001). However, the cystic duct is frequently occluded and obscured by inflammatory changes in the region of Calot’s triangle. In these cases, a partial cholecystectomy, fundus first dissection and leaving the neck of the gallbladder behind, is a more prudent approach as was done for one of the 4 cases of MS type 1 in the present study. Routine CBD exploration is not necessary unless stones are noted on IOC (Csendes et al, 1989; and Venketesh et al, 1998). Bile duct stenosis generally resolves as inflammation subsides following cholecystectomy (Baer et al, 1990 and Pemberton and Wells, 1997). A small fistula may be closed primarily by interrupted stitches and a larger defect can be closed using a cuff of gallbladder.
remnant following subtotal cholecystectomy (Csendes et al, 1989). Sandblom et al, 1975, described a technique using a well-vascularised flap from the gallbladder or cystic duct for the closure of a large defect. Putting of the T-tube following closure of the fistula remains contentious. While some advocate placement through the fistula opening, others suggest that the T-tube to be placed through a separate choledochotomy distal to the fistula (Csendes et al, 1989; Baer et al, 1990 and Dewar et al, 1990). Baer et al, 1990, advocated routine biliary bypass of the choledochal fistula to the duodenum or jejunal loop. In Csendes Type IV fistula, where there is a wide fistula of the CHD and questionable vascularity of the CHD, Roux-en-Y hepaticojejunostomy is the procedure of choice (Johnson et al, 2001). In the present study, primary closure of the fistula with insertion of T-tube in CHD above the fistula was the procedure done for MS type 11, choledochoduodenostomy for MS type 111 and Roux-en-Y hepaticojejunostomy for MS type IV. The technique of closure of the fistula with a flap from the gall-bladder or cystic duct was not used in the present study.

Among the nine patients with Type 11, 111 and 1V MS in the present study, the overall outcomes were good, there was no incidence of bile leak or stricture formation, liver function tests returned to normal for all patients and no patient required re-operation.

Some literatures support an increased coincidence of gallbladder carcinoma and MS (Redaelli et al, 1997 and Nishimura et al, 1999), however, no single case of carcinoma of the gallbladder was recorded in the present study.

Surgery for MS has a high morbidity and mortality rate, which in large series was reported as 23% and 4%, respectively for MS types 11, 111 and 1V (Johnson et al, 2001). In the present study, there was no post-operative complications or mortality.

It could be concluded that MS is an uncommon form of benign obstructive jaundice identified with a frequency of 0.7% of pa-
patients with cholelithiasis. The preoperative diagnosis of Mirizzi syndrome is difficult and an awarded suspicion is necessary to avoid injury of the biliary tree. The problem may only become evident during the operation due to firm adhesions around Calot’s triangle. The success of the treatment is related to a precocious recognition of the condition, during surgery, and adapting the management according to the individual characteristics of each case.

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