World Journal of Surgery

Published Online: 16 November 2005

World J Surg (2005) 29: 1687-
DOI: 10.1007/s00268-005-0

Mirizzi Syndrome: Ten Years Experience from a Teaching Hospital in Riyadh

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Abstract

Objective: Mirizzi syndrome (MS) is an uncommon presentation of cholelithiasis. This study aims to find the incidence and analyze the outcome of management of this condition at Riyadh Medical Complex (RMC) with particular reference to diagnostic methods and outcome of surgical treatment.

Methods: Retrospective study on 17 consecutive patients of MS diagnosed and managed at RMC over ten year period. The records were reviewed for demography, clinical presentation, diagnostic methods, operative procedures, postoperative complication and follow up.

Results: The incidence of MS syndrome was 0.7% of 2415 cholecystectomies. There was preponderance of Type I variety (58.8%). Ultrasonography was able to diagnose 82% cases. ERCP suggested the diagnosis in all cases and helped further in classifying and management of these patients. All Type I cases were managed with partial cholecystectomy, two underwent laparoscopic surgery. Three Type II patients were managed by partial cholecystectomy alone. Three patients with Type III variety had choledochoplasty whereas one remaining patient with Type IV variety underwent hepatico-jejunostomy. All patients had complete recovery with 17.6% procedure-related morbidity and no hospital mortality. All patients are doing well over a mean follow up 6.5 years.

Conclusion: Preoperative diagnosis of Mirizzi syndrome by ultrasound and ERCP is essential to prevent serious complications during surgery. Partial cholecystectomy is an adequate procedure for Types I & II MS. Choledochoplasty provides an effective surgical repair in Type III cases. Although laparoscopic cholecystectomy in MS may be hazardous, it may still be tried in preoperatively diagnosed type I cases, provided the surgeon is experienced and keeps a low threshold for conversion open surgery.

Obstructive jaundice in patients with gallstones commonly occurs when the stones migrate through the cystic duct to bile ducts. On rare occasions stones may become impacted in the cystic duct or the neck of the gallbladder, leading to various grades of inflammation of the common hepatic or common bile duct and associated jaundice and cholangitis. In 1950, Behrend and Cullen noted that the latter condition had originally been reported by Kehr in 1905 and Ruge in 1908.[1] However Mirizzi was the first to describe this phenomenon a

Table 1.
Clinical characteristics of 17 patients according to types of Mirizzi syndrome

Features	Mirizzi type I	Mirizzi type II	Mirizzi type III	Mirizzi type IV
Mean age	58 (38–70 years)	60.3 (57–65 years)	59.3 (58–61 years)	59 years
Male:female	7:3	2:1	1:2	0:1
Upper abdominal pain	10/10 (100%)	3/3 (100%)	3/3 (100%)	1/1 (100%)
Jaundice	7/10 (70%)	2/3 (66.6%)	3/3 (100%)	1/1 (100%)
Cholangitis	2/10 (20%)	1/3 (33.3%)	3/3 (100%)	0
Mean total bilirubin (icteric)	118 μmol/l	127 μmol/l	148 μmol/l	264 μmol/l
Mean alkaline phosphatase (icteric)	368 U/l	571 U/l	723 U/l	960 U/l
Preoperative USG diagnosis	7/10 (70%)	3/3 (100%)	3/3 (100%)	1/1 (100%)

USG: ultrasonography.

stula, various stages of which have gradually become an ntegral part of the classification of MS.[3–5] The overall ncidence of MS is low, reported in 0.7%–2.53% of all patients undergoing cholecystectomy.[6–9] Preoperative diagnc is important to avoid injury to bile ducts and to exclude more common causes of bile duct obstruction of a benign or malignant nature. At present ultrasonography and cholangiography are the two main imaging techniques employed in the diagnosis of MS.

This report on 17 consecutive cases of Mirizzi syndrome managed in the Riyadh Medical Complex over a 0-year period analyzes the incidence, clinical presentation, diagnosis, and surgical procedures for different types of MS, as well as associated morbidity and mortality, as well as follow up.

PATIENTS AND METHODS

This is a retrospective study on records of patients with ne final diagnosis of Mirizzi syndrome managed at iyadh Medical Complex over the 10 years between ary 1994 and December 2003. Various parameters valuated included demographic details, clinical presentation, diagnostic modalities, therapeutic procedures, omplications, and follow up. The Csendes classification[5] as followed to categorize patients intraoperatively when holecystobiliary fistula was present. All patients were een in the surgical clinic within 3 months of initial surgery nd every 6 to 12 months thereafter. Patients were xamined clinically, and liver function tests were evaluated for each follow-up visit. An ultrasound study was uggested in those with persistently elevated alkaline hosphatase during the first year of follow-up.

RESULTS

diagnosed with Mirizzi syndrome. There were 10 mens (58.8%) and 7 women (41.2%). Mean age of the 17 patients was 58 years (range: 38–70 years). All patients presented with upper abdominal pain. Thirteen (76%) patients were jaundiced at presentation, and 6 (35.3%) of these had associated cholangitis. Two patients (11.8%) presented with recurrent attacks of cholangitis. The mean duration of symptoms in icteric patients was 3.8 weeks (range: 2–7 weeks) and 6.25 months (range: 3–9 months) in non-icteric cases. Liver function tests were altered in all patients. Mean total bilirubin in jaundiced patients was 139.7 μmol/l (range: 38–264 μmol/l) and 19.3 μmol/l (range: 17.8–21 μmol/l) in non-jaundiced patients. Mean alkaline phosphatase in icteric patients was 467 U/l (range, 243–1091 U/l) and 287 U/l (range, 230–315 U/l) in non-icteric patients. Ultrasound was the initial imaging investigation performed in all patients. It revealed gallstones in every case (100%) and features of acute cholecystitis with distended edematous gallbladder in 4 (23.5%) cases. A thick-walled contracted gallbladder and intrahepatic bile duct dilatation with a normal common duct were reported in the remaining 14 cases (82%). These findings led the radiologist to suggest the diagnosis of MS in these patients. Because all of the patients had altered liver function tests and 13 were clinically jaundiced at presentation, endoscopic retrograde cholangiopancreatography (ERCP) was performed in all cases, and it led to preoperative suspicion of MS in every case (100%). Ten (58.8%) patients were reported as type I, whereas the remaining 7 (41.2%) patients were found to have various grades of cholecystobiliary fistula (types II–IV). Table 1 summarizes the clinical characteristics of the 17 patients according to the Csendes classification.

Endoscopic sphincterotomy with nasobiliary drainage (NBD) was performed in 10 patients who had multiple

phylaxis included intravenous ceftriaxone and metronidazole in all patients prior to ERCP, and three further doses were administered during surgery, commencing on induction of anesthesia. Antibiotics were continued in patients with cholangitis. Endoscopic retrograde cholangiopancreatography had to be repeated preoperatively in four patients (two for reinsertion of NBD and two for retraction of CBD stones). Partial cholecystectomy was performed in the 10 patients with type I disease. Laparoscopic cholecystectomy was attempted in six of these patients but could be successfully completed in only two patients. Surgery was converted to the open technique in four patients because of dense fibrosis and distorted anatomy in Calot's triangle. The open technique was employed as the initial procedure in the remaining four cases. An on-table operative cholangiogram (OTC) was attempted in all cases, but it was conclusive in only six patients with no evidence of CBD stones or fistula.

All seven patients with cholecystobiliary fistula (types II–IV) underwent open cholecystectomy by the "fundus first" technique. The gallbladder was opened at the fundus and impacted stones were extracted. The fistula was visualized to assess its size, and an OTC was performed through the fistula to determine the presence of common duct stones or stricture. Type II and III varieties were observed in three patients each (17.6%), whereas one patient (5.9%) had type IV disease. The stone was impacted in the cystic duct in one patient and in the Hartmann's pouch in two others with type II disease. The fistula was classified as small (less than one third the circumference of common duct), so partial cholecystectomy alone was undertaken, with oversewing of the gallbladder remnant. Although OTC had revealed a CBD stone in one patient and mild CHD narrowing in two others, no attempt was made to explore the common duct, nor was a T-tube placed in any of these type II patients. All Type III patients were treated by choledochoplasty employing a gallbladder flap. In addition, a T-tube was placed distal to the repair and was retained for 6 weeks. In one type IV patient the stone was large (4 cm) and so was the fistula. This patient underwent Roux-en-**Y** hepaticojejunostomy. All patients had a tube drain left in the subhepatic space that was removed within 3 to 12 days.

Overall procedure-related morbidity was 17.6% (3 cases). Two patients developed wound infection necessitating open drainage. One of these patients developed chest infection necessitating antibiotics appropriate to culture reports (*Escherichia coli*). One pa[illegible] pneumonia [illegible] 4,5,11 Tan and [illegible]

Three type II patients remained persistently jaun[illegible] postoperatively. All these patients underwent ERCP during the early postoperative period (5–11 days follo[illegible] surgery). A common duct stone was extracted in these patients, followed by smooth recovery. The remaining patients were successfully managed by e[illegible] scopic placement of a biliary stent for suspected nar[illegible] ing of the distal common hepatic duct. The stents [illegible] removed after 6–8 weeks. The mean hospital stay wa[illegible] days (range: 8–24 days). There was no incidence of b[illegible] injury and no hospital mortality. Final histopathology revealed cholecystitis with cholelithiasis in all patients [illegible] associated malignancy was reported in any case.

All patients were followed up in the surgery and [illegible] troenterology clinics for a period of 1–10 years (mean [illegible] years). Three non-Saudi patients (2 with type I dise[illegible] and 1 with type III) who left the Kingdom were los[illegible] follow-up 1–3 years after surgery. All were symptom[illegible] with normal liver function tests through the last follow[illegible] visit. The remaining 14 patients have been asymptom[illegible] except mildly elevated alkaline phosphatase persis[illegible] among those patients treated for cholecystobiliary fist[illegible] Follow-up ultrasound studies in these patients did [illegible] reveal any abnormality of the biliary tree or any cirrh[illegible] changes in the liver.

DISCUSSION

Mirizzi syndrome is an unusual complication of g[illegible] stone disease. The importance of MS has been hi[illegible] lighted as a clinical entity associated with a high incider[illegible] of biliary injuries and demanding complex surgical p[illegible] cedures. There are several classifications and subcl[illegible] sifications of MS. McSherry and colleagues in 19[illegible] described a classification that includes type 1, compre[illegible] sion of the bile duct by a stone impacted in the cystic du[illegible] or Hartmann's pouch, and type 2, characterized [illegible] cholecystobiliary fistula.[4] Csendes and associates [illegible] 1989 proposed a subclassification of the cholecystobilia[illegible] fistula (McSherry type 2), according to size, into thr[illegible] types (type II–IV).[5]

The MS incidence of 0.7% in the present series corr[illegible] lates well with the 0.7%–2.53% reported in the liter[illegible] ture.[5–10] The mean age of 58 years is well within the a[illegible] range of 44–62 years quoted in other studies.[5,10,11] Ma[illegible] preponderance (58.8%) in the present study differs fro[illegible] various other reports describing the syndrome as mo[illegible]

sentation in both icteric and non-icteric patients is ger than reported in earlier studies.[5,10] Of the patients the present study, 76% were jaundiced at the time of spital admission, and 35% had associated cholangitis. ese figures are slightly higher than earlier re- rts.[5,10,11] This may be explained in part by a delay in eking the medical advice, particularly among male tients.

Ultrasonography (USG) is usually the initial radiological vestigation in obstructive jaundice. The reported sensi- vity of ultrasound in the diagnosis of MS is 8.3%–7%.[5,10–12] However, it has been recommended as the est screening method.[13,14] Ultrasonography was able to aise the suspicion of MS in 82% of cases in this series. his higher figure may reflect a more cautious examination, in keeping with the difficult imaging findings in this subgroup of patients. Ultrasound and ERCP successfully sugge d the diagnosis of MS in all cases in this study. Endoscopic retrograde cholangiopancratography has been proposed as the best modality in the preoperative diagnosis and initial management of Mirizzi syndrome by permitting therapeutic decompression and providing a road map for surgery.[11,12,15] However, in addition to its inherent complications, ERCP may be limited by technical failure in 5%–10% of cases.[11] In such cases, percutaneous transhepatic cholangiography (PTC) remains a viable alternative.[16] MRCP has also been recommended as a diagnostic modality of choice. However, its disadvantage is its inability to confirm the presence of fistulas and to afford therapeutic stenting.[10] Studies differ on the incidence of different types of MS. Whereas Csendes and colleagues[5] reported an increased incidence of type II (41%) and type III (44%) lesions in their study of 219 cases in 1989, the increased incidence of type I MS (58.8%) ob ved in the present study is in accordance with various recently published reports.[10–12] This difference could have been due to small sample size in these studies, but it may reflect a changing spectrum of the disease on account of increased awareness and early diagnosis.

Preoperative stenting or nasobiliary drainage was employed in 10 patients with cholangitis or deep jaundice in this study. This selective policy of preoperative biliary drainage has also been adopted by other authors.[15,17] When the subtype of Mirizzi has not been determined preoperatively, the best initial operative strategy recommended is to go for the "fundus first" technique.[11,13] The same technique was adopted in this study. The reflux of bile into the gallbladder is an indicator of the presence of fistula, because the cystic duct is usually occluded by

ence of the fistula, to determine its size, and to exclude CBD stones.[13] The surgical treatment of type I Mirizzi syndrome is generally accepted to be removal of stones partial cholecystectomy, and closure of the remaining gallbladder cuff or enlarged cystic duct stump.[5,10,11,18] The presence of CBD stones, fistula, or stenosis at the site of impaction of the stone may necessitate CBD exploration and/or the insertion of a T-tube. Most ducts affected by impaction subsequently resume normal caliber once the inflammation subsides.[5]

In the present study, intraoperative cholangiography was performed in 6 of the 10 patients with type I MS, and it revealed mild narrowing at the site of stone impaction, but no further action was taken. Postoperatively all of those patients had uneventful recoveries.

The management of MS type II and III is more controversial. Corlette and Bismuth recommended partial cholecystectomy, oversewing of the gallbladder cuff, and insertion of a T-tube through the fistula as adequate treatment for type II disease.[19] Baer and colleagues suggested placement of a T-tube through a separate choledochotomy in the distal CBD in order to prevent excessive leakage and stricture at the fistula site.[13] All three type II patients in this study underwent partial cholecystectomy without CBD exploration or T-tube insertion, despite an on-table cholangiogram that revealed a CBD stone in one patient and mild narrowing of the common hepatic duct at the fistula site in the other two patients. No significant bile leakage was observed, although all these patients had persistent jaundice and one later underwent ERCP for CBD stone extraction and the other two had stents placed. In both cases, the stent was removed after 6–8 weeks, and the patients recovered completely. This is in accordance with the recommendation by various other studies[15,20] whose authors believe that excessive dissection in Calot's triangle in the presence of adhesions and distorted anatomy may do further harm by enlarging the fistulous opening and jeopardizing the blood supply to the common bile duct. Retrospectively, the latter two patients in our study could have been managed expectantly, awaiting resolution of the inflammation, as suggested by Csendes and colleagues.[5]

Mirrizzi syndrome type III is generally best treated with choledochoplasty using a well-vascularized gallbladder or cystic duct flap.[5,7,8,21] According to Baer and colleagues, using such a flap from a diseased gallbladder is prone to failure.[13] They therefore recommend choledochoduodenostomy, and they report satisfactory results. In the present study, all three MS type III patients were sue

authors agree with Johnson and colleagues that T-tube decompression remains integral to the management of type III MS.[18] Moreover, choledochoduodenostomy in the absence of an adequately dilated common bile duct may not give satisfactory results in the long term.

The management of MS type IV is almost established, with bilio-enteric anastomosis as the standard treatment.[5,10,11,18] One type IV MS patient in this study was successfully treated by hepaticojejunostomy.

Laparoscopic cholecystectomy has now become the gold standard in the treatment of gallstones. Its success usually depends (among other factors) on the ability to safely grasp and laterally retract the gallbladder for easy dissection around the cystic duct in the region of Calot's triangle. These maneuvers are not only difficult but may be impossible to perform in MS. For these reasons, MS has been considered by various authors to be a contraindi on for laparoscopic cholecystectomy.[22,23] However, some authors have reported success with the laparoscopic technique.[24–26] Review of these reports, however, revealed increased complication rates compared to open cholecystectomy.[18] In the present study, two of six type I patients successfully underwent laparoscopic partial cholecystectomy with an endo-GIA stapler. In the remaining four patients the procedure was converted early an open procedure. No patient with cholecystobiliary fistula had laparoscopic surgery in this study. The 17.6% overall morbidity rate correlates well with the 6%–23% figures quoted in the literature.[5,10,11] There was no incidence of biliary injury, late stricture, or cirrhotic changes over a mean follow-up of 6.5 years.

CONCLUSIONS

Preoperative diagnosis of Mirizzi syndrome by ultrasound and ERCP is essential to prevent serious complications during surgery. Partial cholecystectomy is an adequate procedure for types I and II MS, and choledochoplasty provides an effective surgical repair in type III cases. Although laparoscopic cholecystectomy in MS may be hazardous, it may still be tried in preoperatively diagnosed type I cases, provided the surgeon is experienced and maintains a low threshold for conversion to open surgery.

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