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Mirizzi's syndrome: diagnostic and surgical considerations in 25 patients

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Abstract: Mirizzi's syndrome is a rare complication of longstanding cholelithiasis. Many surgical approaches of varying complexity have been advocated for treatment. However, the distorted extrahepatic biliary anatomy continues to be threatening, with a high risk of biliary complications. Presented here is a series of 25 patients with Mirizzi's syndrome who were treated at the Dokuz Eylul University Hospital since 1985. Type I lesion (without cholecystocholedochal fistula) was encountered in 11 patients, while the remaining 14 had type II lesions (with cholecystocholedochal fistula). Preoperative diagnoses were made in 14 of the 25 patients (56%). Follow-up in 17 patients ranged from 1 to 96 months (mean, 40 months). Unfortunately, the remaining 8 patients were lost to follow-up after discharge. The morbidity rate in our series was 32%, while no mortality was encountered. During long-term follow-up, no biliary stricture was diagnosed. Following an uneventful postoperative course, all of our patients are symptom-free and doing well, with normal liver function. We conclude that partial cholecystectomy alone is a safe and sound surgical approach for the treatment of type I lesions. For type II lesions, depending on the size of the fistula, either primary closure over a T-tube, or bilio-digestive anastomosis, preferably Roux-en-Y, can be an appropriate treatment modality, with a low morbidity rate.

Key words: Mirizzi's syndrome, cholelithiasis, biliary fistula

Introduction

Mirizzi's syndrome is an uncommon complication of longstanding gallstone disease. It was first described in 1948 by a surgeon of the same name as obstructive jaundice due to extrinsic compression of the extrahepatic bile duct by an impacted stone in the cystic duct or gallbladder neck.[1] The roentgenological features of the syndrome were described in 1965.[2] In 1982, McSherry et al.[3] classified Mirizzi's syndrome into two types according to the progression of the inflammatory process. They described a type I lesion as an extrinsic compression of the adjacent common hepatic duct and a type II lesion as a pressure necrosis of the septum between the cystic and common bile duct (CBD) leading to a cholecystocholedochal fistula. Total cholecystectomy in the patients with Mirizzi's syndrome may be challenging because of marked inflammation and adhesions around Calot's triangle.[4] To avoid a major CBD injury, partial rather than total cholecystectomy is a safer alternative in type I lesions.[5,6] Despite various modes of operative treatment, however, the problem of fistula repair in type II lesions remains unsettled. Here, we report the diagnostic and surgical techniques and clinical outcomes in 25 patients with Mirizzi's syndrome.

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Patients and methods

Between the years 1985 and 1999, 25 patients with Mirizzi's syndrome were diagnosed at our hospital, representing approximately 1.13% of all cholecystectomies. The clinical charts and X-rays of these patients were retrospectively reviewed for presentation, preoperative evaluation, intraoperative findings, management, and complications. In 17 patients, follow-up was carried out for a mean of 40 months (range, 1 to 96 months) and follow-up data were obtained by outpatient visits or telephone inquiries. For 8 patients who were lost to follow-up, no data were available beyond the third month after the operation. The study population consisted of 9 men and 16 women with a mean age of 53.2 years (range, 39 to 74 years). Diagnosis at presentation was obstructive jaundice in 18 patients, acute cholecystitis in 5 and acute biliary pancreatitis in 2. Preoperative maximum levels of serum bilirubin, alanine aminotransferase (ALT), and alkaline phosphatase

were 6.3 ± 3.8 mg/dl, 123 ± 65 IU/l, and 512 ± 165 IU/l, respectively. In 2 patients, the clinical symptoms of acute pancreatitis (Ranson score of 3 at admission) were resolved with medical treatment. The 5 patients who suffered acute cholecystitis were taken to the operating room within the first 72 h after admission. All our patients received intravenous broad-spectrum antibiotics for at least 5 days, started either pre- or postoperatively.

Preoperative examinations

All 25 patients underwent preoperative ultrasonographic (USG) examination of their gallbladders. Cholelithiasis was demonstrated in all patients. Endoscopic retrograde cholangiopancreatography (ERCP) was performed in 18 patients, but was unsuccessful in 3. In 8 patients, both USG and ERCP revealed CBD stones; these were removed by endoscopic sphincterotomy in 2 patients (type I) and by CBD exploration in 6 patients (type II). In 1 patient, ERCP did not confirm the diagnosis of choledocholithiasis which was indicative on USG. In 7 patients, negative ERCP findings for CBD stones were also negative at laparotomy. Computed tomography (CT) was required to exclude malignancy in those patients who had a mass lesion in the gallbladder bed shown by USG ($n = 3$) and to obtain images of the pancreas in patients with suspected acute pancreatitis ($n = 2$). The interpretation of CT was indicative of Mirizzi's syndrome in 1 patient. Preoperative diagnoses were made in 14 of the 25 patients (56%) by the diagnostic studies (Table 1).

Surgical procedures

Cholecystectomy was performed in all 25 patients. However, the cholecystectomies were partial except in 4 patients with type I lesions (Table 2). Mucosa of the remnant gallbladder was electrocauterized to avoid mucoclasis. One patient with type I lesion underwent pancreaticoduodenectomy (Whipple procedure) due to a large duodenal defect that occurred during the dissection of cholecystoduodenal fistula. In 1 patient, an attempted laparoscopic approach was converted to laparotomy due to difficulties in dissection of fibrotic gallbladder and lack of appropriate exposure.

In patients with type II lesions ($n = 14$), the cholecystocholedochal fistulas were repaired in various ways. Suture closure over a T-tube was performed in 7 patients. In these patients, T-tubes were placed into the CBD via fistula ($n = 4$) or a separate incision ($n = 3$). All T-tubes were removed at a mean of 21.8 days (range, 14 to 36 days) when cholangiograms were normal. The remaining 7 patients underwent bilio-

Table 1. Preoperative radiologic studies in diagnosis of Mirizzi's syndrome

	No of patients	Preop diagnosis	Perioperative diagnosis
USG	7	2	5
USG + CT	2	0	2
USG + ERCP	12	10	2
USG + CT + ERCP	3	2	1
Total	25	14 (56%)	10

Preop, Preoperative; USG, ultrasonographic; CT, computed tomography; ERCP, endoscopic retrograde cholangiopancreatography

Table 2. Surgical procedures

No of patients		Surgical procedures
Type I	5	Partial cholecystectomy
	4	Total cholecystectomy
	1	Partial cholecystectomy + T-tube drainage via cystic duct
	1	Pancreaticoduodenectomy (Whipple procedure)
Type II	7	Partial cholecystectomy + primary closure + T-tube drainage
	3	Cholecystocholedochoduodenostomy
	1	Cholecystocholedochojejunostomy (Roux-en-Y)
	1	Cholecystocholedochojejunostomy + Braun anastomosis
	2	Hepaticojejunostomy (Roux-en-Y) + external bile duct excision

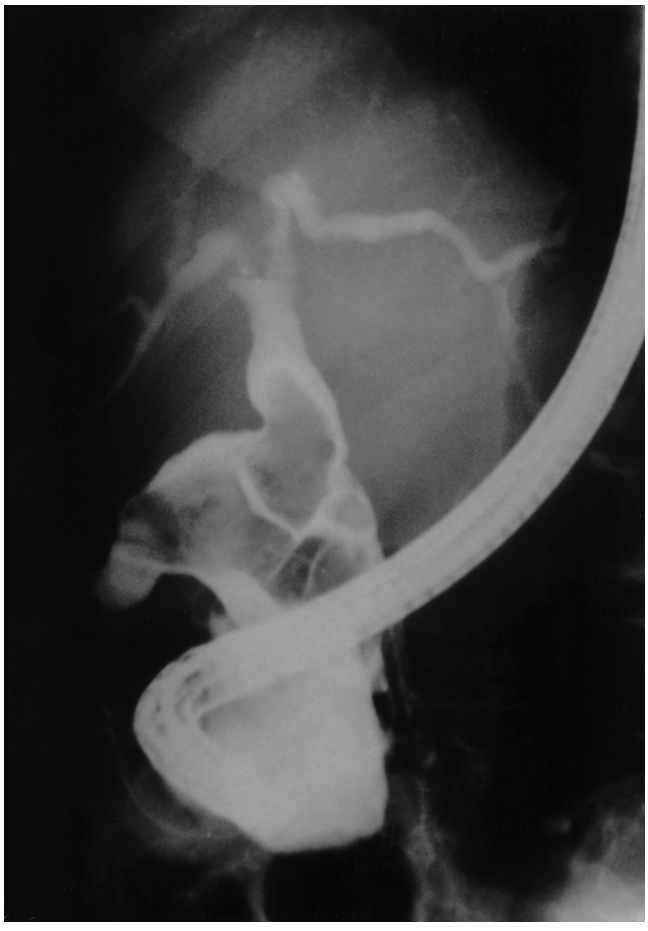


Fig. 1. Extensive deformation of the common bile duct in a patient in whom the diseased segment was excised and reconstructed by hepaticojejunostomy

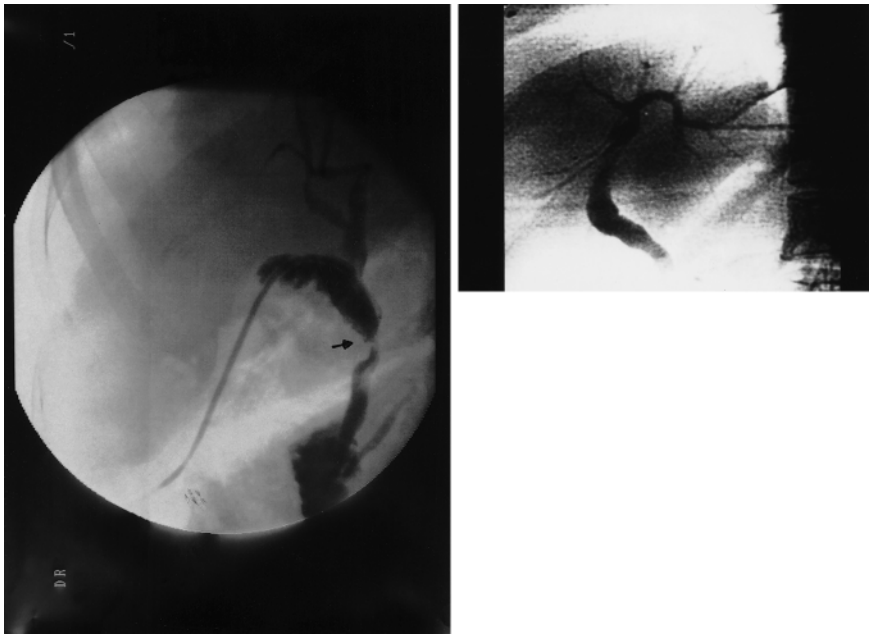
digestive bypass either with cholecystocholedocho duodenostomy ($n = 3$) or cholecystocholedocho jejunostomy (Roux-en-Y, $n = 3$; loop of jejunum, $n = 1$). In 2 of the 4 patients with jejunostomy, the CBD was extensively dilated and deformed by multiple stones in totally fused gallbladder (Fig. 1). Excision of the diseased bile duct segment in continuity with Roux-en-Y hepaticojejunostomy was performed in both of these patients. In another patient, a loop of jejunum was preferred to establish side-to-side choledochojejunostomy because of difficulties in mobilization of the duodenum or proximal jejunum.

CBD exploration with choledochotomy was not performed in any patients with type I lesions. However, CBD exploration was performed through the transected duct (during the Whipple procedure) in 1 patient, and through the patent cystic duct in another 2 patients. In contrast, all patients with type II lesions ($n = 14$) underwent CBD exploration through the fistula by flexible choledochoscope or Bakes dilators.

Intraoperative cholangiograms were taken through the fistula ($n = 4$), T-tube ($n = 3$), or the cystic duct ($n = 2$) when needed (Fig. 2a). Frozen sections were obtained in order to rule out suspected malignancy in 7 patients. No malignancy was determined in any frozen sections or cholecystectomy specimens.

Results

There were no perioperative complications in our patients except for the patient with the large duodenal



a **b**

Fig. 2a,b. Cholangiograms taken from a patient **a** intraoperatively and **b** 14 days after operation. Bile duct stenosis was significantly resolved as the inflammatory process subsided 2 weeks after simple decompression by partial cholecystectomy

Table 3. Perioperative morbidity rate

Complications	No of patients
Duodenal injury	1/25
Atelectasis	2/25
Wound infection	3/25
Subhepatic collection	1/25
Cholangitis	1/25
Total	8/25 (32%)

defect occurring from cholecystoduodenal fistulization. This patient was treated for gastric atony for 2 months after pancreaticoduodenectomy was performed. In one patient in whom the duodenum was used to repair the CBD defect, medical therapy was established for cholangitis that was diagnosed in the fourth postoperative week. In another patient, there was spontaneous incisional bilio-serous drainage (about 100 cc) that occurred once on the tenth postoperative day following side-to-side cholecystocholedochojejunostomy. Liver scintigraphy (HIDA), which was done in both these patients to elucidate the underlying pathology, did not reveal any bile leakage or stricture formation at the site of anastomosis. In a fourth patient, percutaneous USG-guided aspiration and drainage was done for a subhepatic hematoma detected during the fever work-up early after the operation. Of the 17 patients who were followed-up (14 patients for more than 1 year; of whom 5 patients were followed-up for more than 5 years), none showed any symptoms attributable to the biliary system and their liver function test results were within normal limits. The perioperative morbidity rate was 32% (Table 3) and the mortality rate was zero.

Discussion

Cystic duct obstruction from cholelithiasis occurs in as many as 40% of patients with gallstones.[7] These patients mostly present with the symptoms of acute cholecystitis. However, on rare occasions, longstanding gallstone impacted in the Hartmann pouch or the cystic duct may externally compress the CBD, which presentation is called Mirizzi's syndrome. As a consequence, obstructive jaundice becomes the prominent clinical symptom in these patients. Some offending stone(s) may also play a causative role in the pathogenesis of gallbladder cancer by inducing chronic irritation and marked biliary inflammation in the wall of the gallbladder.[8] The incidence of unsuspected malignancy in patients with Mirizzi's syndrome is significantly higher than the incidence in longstanding gallstone disease (27% vs 2%, respectively).[8] In contrast to findings reported in the literature, gallbladder cancer has not been shown in any of our patients with Mirizzi's syndrome. This may be attributed to an error occurring due to the relatively small patient population or to an error in sampling. However, when we consider the mean follow-up of 40 months in 17 of our patients, the risk of unrecognized malignancy due to sampling error is unlikely.

The literature classifies Mirizzi's syndrome in various ways, but the most widely used classification is the division of the syndrome into two types by McSherry et al.;[3] type I (without cholecystocholedochal fistula) and type II (with cholecystocholedochal fistula). Despite the advances in imaging techniques the diagnosis of Mirizzi's syndrome is mostly made during the surgical procedure. Although cholecystectomy and the removal of the offending stone(s) are the mainstays of treatment in these patients, dissection around the hepatoduodenal ligament is usually difficult, due to dense fibrosis and edema.[4] Under these circumstances, insisting on the removal of the entire gallbladder may entail a high risk of inadvertent bile duct injury.[4,9] Thus, partial cholecystectomy alone is considered to be a safe and definitive surgical treatment in type I lesions in which the cystic duct is mostly obliterated secondary to longstanding inflammation.[5,6] In these patients, bile duct stenosis due to external compression of the offending stone(s) and inflammatory tissue generally resolves as the inflammatory process subsides following simple decompression by partial cholecystectomy[6,10] (Fig. 2a,b). Therefore, in the absence of any stone inside the gallbladder, CBD exploration just for stenosis is mostly unnecessary and dangerous.[6] If a retained stone is demonstrated, postoperative ERCP and stone extraction may provide a safer alternative.[5,6,9]

Bile spilling from the site where the impacted stone is removed could indicate the presence of a bilio-biliary fistula (type II lesion), and cholangiogram with or without CBD exploration can be carried out through the same access.[6,11,12] Although various surgical techniques have been advocated, the best method of bilio-biliary fistula repair varies according to the size of the fistula, the quality of surrounding tissues, and the experience of the surgeon. If primary suture closure with or without a flap from the remnant gallbladder wall is used to repair the fistula, a T-tube can be inserted into the CBD either through the fistula or from a separate incision to provide maximum safety of the suture line, which may consist of fibrotic and edematous tissue.[9–11,13] This procedure may both decompress the CBD and reduce the risk of stenosis.[14] In our seven patients in whom T-tubes were used, the T-tubes were removed at a mean of 21.8 days (range, 14 to 36 days) without any complication. If the fistula cannot be closed primarily, then biliodigestive bypass may be the choice of treatment, and this

is widely used.[6,9] A wide enough bilio-digestive anastomosis is essential to avoid early and late complications, such as bile leakage, stricture formation, or cholangitis. Although cholecystocholedocho-duodenostomy or Roux-en-Y hepaticojejunostomy can be used for anastomosis, the latter may be preferred to minimize the risk of reflux cholangitis. This has been a problem in one of our three patients with cholecystocholedochoduodenostomy who needed hospitalization for an episode of cholangitis 1 month after the operation.

In two of our patients, the gallbladder was almost entirely fused with the CBD, and the CBD was extensively dilated and contained many stones. In both patients, the diseased segment of the CBD was resected to avoid the risk of anastomotic leakage, cholangitis, or stricture formation. Reconstruction was established with Roux-en-Y hepaticojejunostomy. Segmental CBD resection followed by end-to-end bile duct anastomosis as an alternative surgical approach has a high risk of biliary stricture in the early postoperative period.[10] In our series, no biliary stricture was demonstrated. Although most patients with benign bile duct strictures present soon after their initial operation, this may be delayed for years in some patients.[15] Hence, in order to reach a firm conclusion, at least 5 years of follow-up duration need to be completed.

The role of minimally invasive surgery in the treatment of Mirizzi's syndrome remains controversial. Some authors regard this condition as inappropriate for laparoscopic surgery due to the dense adhesions around Calot's triangle, whereas others have reported that the laparoscopic approach or mini-laparotomy is feasible, particularly for type I lesions, but technically demanding.[16–20] In contrast, other than some sporadic cases which were reported as successful, conventional laparotomy is usually needed for effective repair of the cholecystocholedochal fistula.[19] In our series, laparoscopic cholecystectomy was attempted in only one patient, with a preoperative diagnosis of acute cholecystitis. However, that approach was converted to open surgery because of the unclear anatomy in Calot's triangle, due to dense adhesions.

Although the primary treatment is surgical, endoscopic techniques can be helpful at different stages of treatment in Mirizzi's syndrome. A nasobiliary stent placed into the CBD before the operation may both serve as a guide to avoid the risk of bile duct injury and as a good access for obtaining multiple cholangiograms.[13] In patients with co-morbidity, endoscopic stent placement facilitates bile drainage into the intestine, which, in turn, helps recovery from cholangitis or local inflammatory processes and renders the patient suitable for subsequent surgical intervention.[18,20]

In 14 of our patients (56%), the diagnosis of Mirizzi's syndrome was made preoperatively. Retrospective evaluation of our experience indicated that these patients may be successfully managed without preoperative diagnosis of Mirizzi's syndrome. In our series, there seemed to be no difference between patients with or without preoperative diagnosis regarding peri- and/or postoperative complications (early and late). We conclude that a safe surgical approach in Mirizzi's syndrome can be accomplished by: (1) avoiding extensive dissection, or possibly uncontrolled cholecystectomy, in the presence of contracted and fibrotic gallbladder associated with dense inflammatory adhesions around the hepatoduodenal ligament and (2) preferably performing the Roux-en-Y bilio-digestive bypass in type II lesions, using the remnant gallbladder wall following the partial cholecystectomy.

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