Surgical management of Mirizzi syndrome

Mirizzi sendromunun cerrahi tedavisi

Ünal AYDIN[1], Pınar YAZICI[1], İsmail ÖZSAN[1], Galip ERSÖZ[2], Ömer ÖZÜTEMİZ[2], Murat ZEYTUNLU[1], Ahmet ÇOKER[1]

Departments of [1]General Surgery and [2]Gastroenterology, Ege University, School of Medicine, İzmir

Background/aims: *Mirizzi syndrome is an unusual presentation of prolonged cholelithiasis. This study aimed to analyze the diagnostic methods, operative strategies, and outcome of the surgical treatment of patients with Mirizzi syndrome.* ***Methods:*** *We retrospectively evaluated the patients with Mirizzi syndrome treated in our General Surgery Clinic. The data collected included demographic variables, clinical presentation, diagnostic methods, surgical procedures, and postoperative complications.* ***Results:*** *The study included 13 male and 21 female patients, with a mean age of 67.2 years. The incidence of Mirizzi syndrome was determined as 0.6% (34/5632), and type II was more frequently observed (52.9%); no patient was determined as type IV. The incidences of types I and III were 35.2% and 11.7%, respectively. Among the preoperative diagnostic evaluations, ultrasonography was the initial imaging study that was performed in all patients. Computerized tomography, magnetic resonance cholangiopancreatography, and endoscopic retrograde cholangiopancreatography were the other radiological studies. Surgical procedures included cholecystectomy for 83% of the patients with type I. The remaining cases and 14 of the type II patients (77.7%) underwent choledochotomy and T-tube insertion following cholecystectomy. Four of the patients with type II variety and all of the type III patients underwent cholecystectomy and roux-en-Y hepaticojejunostomy. All of the patients had complete recovery, with a morbidity rate of 5.8%, and there was no hospital mortality.* ***Conclusions:*** *The essential part of the management of patients with Mirizzi syndrome is to determine the best surgical procedure in the preoperative period. In type I patients, simple cholecystectomy is generally enough, but sometimes T-tube insertion may be required, while the cases with types II-IV require more complex surgical approach, such as cholecystectomy and bilioenteric anastomosis. Roux-en-Y hepaticojejunostomy is an appropriate procedure with good outcome.*

Key words: Mirizzi syndrome, cholecystectomy, roux-en-Y hepaticojejunostomy

Amaç: *Mirizzi Sendromu uzamış taşlı kolesistitin nadir bir formudur. Bu çalışmada tarafımızdan takip edilen Mirizzi Sendrom'lu hastalarda tanı metodları, operatif teknikler ve cerrahi tedavi sonuçları değerlendirildi.* ***Yöntem:*** *On yılı aşkın süredir genel cerrahi servimizde tedavi edilen Mirizzi Sendrom tanılı hastalar demografik verileri, klinik bulguları, tanı metodları, cerrahi prosedürler ve postoperatif komplikasyonlar açısından araştırıldı. Sınıflama Csendes'in klasifikasyonuna göre yapıldı.* ***Bulgular:*** *Yaş ortalaması 67.2 yıl olan 21 bayan 13 erkek tespit edildi. Mirizzi Sendrom insidansı %0.6 (34/5632) olarak bulundu. Tip II (%52.9) hastalarda artmış bir insidans mevcutken Tip IV hastaya rastlanmadı. Tip I ve Tip III görülme sıklığı ise sırası ile %35.2 ve %11.7 olarak bulundu. Preoperatif tanı yöntemleri arasında tüm hastalarda ultrasonografi ilk tanı yöntemi olarak tespit edildi. Bilgisayarlı tomografi, manyetik rezonans kolanjiografi, ve endoskopik retrograd kolanjiopankreatografi diğer radyolojik tetkikler arasında izlendi. Uygulanan cerrahi prosedürler arasında Tip I hastalarda kolesistektomi %83 ile birinci sırada iken geriye kalan Tip I hastalara ve Tip II Mirizzi Sendrom tanılı 14 hastaya (%77.7) kolesistektomi sonrası koledokotomi ve T-tüp drenaj uygulandı. Tip II tanılı geriye kalan 4 hastada ve tüm Tip III tanılı olgulara biliyoenterik anastomozla birlikte kolesistektomi uygulandı. Tüm hastalar genelinde %5.8 morbidite oranı hariç problemsiz iyileşme sağlandı ve mortalite gözlenmedi.* ***Sonuç:*** *Mirizzi Sendrom tanılı hastalarda tedavide en önemli noktalardan biri de preoperatif dönemde cerrahi tedavinin belirlenmesidir. Tip I olgularda basit kolesistektomi yeterli olurken bazen t- tüp yerleştirilmeai gerekebilir. Öte yandan Tip II-IV MS olguları kolesistektomi ve biliyoenterik anastomoz gibi kompleks prosedürler gerektirebilir. Bu durumda Roux-en-Y hepatikojejunostomi iyi sonuçlarla uygulanabilir bir yöntemdir.*

Anahtar kelimeler: Mirizzi sendromu, kolesistektomi, roux-en-Y hepatikojejunostomi

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. Its

Address for correspondence: Pınar YAZICI
Ege Üniversitesi Tıp Fakültesi Gastroenteroloji Bilim Dalı
35100 Bornova-İzmir, Turkey
Phone: + 90 232 390 40 20 • Fax: + 90 232 339 88 38
E-mail: drpinaryazici@gmail.com

Manuscript received: 09.11.2007 **Accepted:** 03.07.2008

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 MS (1). The prevalence of this rare complication varies from 0.3 to 3% (2-6). 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.

MATERIALS AND METHODS

From February 1996 to February 2007, 5690 patients with a diagnosis of cholelithiasis were included in a retrospective review of case notes. According to the observations in the operation, 34 patients with the final diagnosis of MS, who were managed in our surgical clinic, 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, postoperative complications, and follow-up period were evaluated. To determine the surgical procedure, Csendes classification (4) was applied prospectively to the patients, generally considering preoperative and/or intraoperative observations (Table 1).

RESULTS

During the study period, 5632 cases underwent cholecystectomy. Incidence of MS diagnosis among the patients was 0.6%. There were 13 male (38.3%) and 21 female patients (61.7%), with a mean age of 67.2 years (range: 45-82 years). Jaundice and itching were the major symptoms, followed by abdominal pain. The mean duration of symptoms was 26.19 weeks (range: 1-168 weeks). Twenty-six patients (76.4%) presented with jaundice and 13 of these patients (50%) had associated cholangitis. The most frequent signs and symptoms are shown in Figure 1. Only 12 patients (35.2%) experienced recurrent symptoms of epigastric or right hypochondrial pain during the last year prior to presentation. The results of the hepatic function tests are shown in Table 2. Mean total bilirubin in the patients with jaundice was 3.8 mg/dl (range: 1.7-7.7 mg/dl). All the patients underwent ultrasonography initially, which revealed the gallstones in both the common bile duct (CBD) and gallbladder-associated dilatation of the intrahepatic bile ducts in 22 patients (64.7%), whereas presence of gallstones only in the gallbladder was reported for 8 patients (23.5%). The remaining 4 patients were aggressively investigated with advanced screening methods including computerized tomography (CT) and magnetic resonance cholangiopancreatography (MRCP) to determine whether there was a gallbladder cancer formation. The

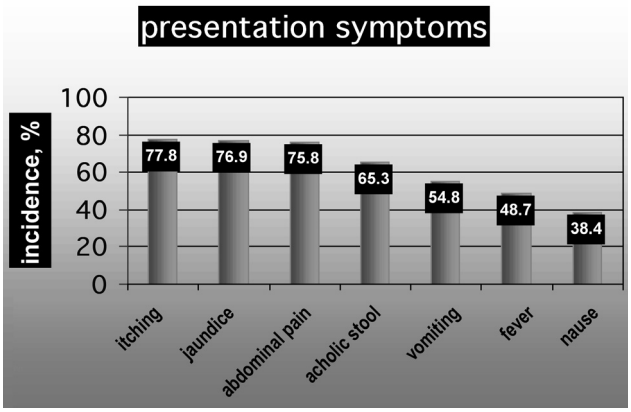

Figure 1. Signs and symptoms presented by patients with Mirizzi syndrome.

Table 1. Classification of Mirizzi syndrome

Classification type	Csendes (1989)	Mc Sherry (1982)
Type I	External compression of the common duct because of a stone impacted at the neck of the gall bladder or at the cystic duct.	
Type II	CCBF involving less than one-third of the circumference of the common duct	
Type III	CCBF that involves up to two-thirds of its circumference	cholecystobiliary fistula
Type IV	CCBF with complete destruction of the entire wall of the common duct	

CCBF: Cholecystobiliary fistula

Table 2. Hepatic function tests in patients with Mirizzi

Laboratory test	patient' results (minimum and maximum)	normal range	n*
Total bilirubin (mg/dl)	0.17-7.78	0.1-1	26(76%)
AST (UI/L)	11-347	<35	13(38%)
ALT (UI/L)	7-325	<45	13(38%)
GGT (UI/L)	16-896	<55	27(78%)
ALP (UI/L)	144-2248	35-104	18(52%)

AST: Aspartate aminotransferase. ALT: Alanine aminotransferase. GGT: gamma-glutamyl transpeptidase. ALP: Alkaline phosphatase. *n: The number of patients whose laboratory parameters were out of the normal ranges.

other diagnostic modality, endoscopic retrograde cholangiopancreatography (ERCP), was performed in 27 patients (79.4%), and MS was suspected in 16 patients, preoperatively (Figures 2a, b). The remaining 11 patients who underwent ERCP could be diagnosed intraoperatively. Figure 3 presents the categorization of the patients with MS based on Csendes classification. Twelve patients (35.2%) were classified as type I; 18 (52.9%) as type II, and 4 (11.7%) as type III. No patient was determined as type IV. During ERCP, endoscopic nasobiliary drainage (NBD) was performed with sphincterotomy in 7 patients (20.5%) who had an obstruction of the CBD and dilatation of the intrahepatic bile ducts and/or CHD.

Cholecystectomy without additive surgical procedure was performed in 10 (20.5%) patients with type I. Laparoscopic cholecystectomy was attempted in 7 of these patients but could be successfully performed in only 4 patients (57%). Surgery was converted to the open technique in 3 patients because of dense adhesions and distorted anatomy due to edematous tissue and inflammatory process. The open technique, retrograde fundus-first cholecystectomy (7), was applied as the initial procedure in the remaining 3 patients, and partial cholecystectomy was performed in 2 of them. The other patients with type I (n=2) and additionally, 14 patients with type II, underwent cholecystectomy, choledochotomy and insertion of T-tube for biliary drainage through a separate choledochotomy (77.7%). All of the patients with type III variety and the remaining 4 patients (22%) with type II underwent cholecystectomy with excision of the external bile ducts and reconstruction with Roux-en-Y hepaticojejunostomy, except for 2 who underwent choledochoduodenostomy. One of these patients was diagnosed as periampullary diverticulum. The results of all the histological examinations were reported as chronic inflammatory reaction, but 2 revealed porcelain gallbladder. The mean hospital stay was 8.3 days (range: 4-18 days). There were no complications including biliary problems in the early postoperative period except pneumonia. However, in the late postoperative period, 1 patient (2.9%) was diagnosed with benign biliary stricture, in the ninth postoperative month, and was managed with balloon dilatation, but it recurred and hepaticojejunostomy had to be performed six months after the first dilatation procedure.

DISCUSSION

Different stages of MS were defined in the 1980s. In 1982, McSherry et al. (8) classified MS into two types based on ERCP findings. However, in 1989, Csendes et al. subclassified MS into four types. This classification further categorized the cholecystocholedochal fistula according to its extent of destruction to enable identification of the appropriate management of MS (Table 1) (4). 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

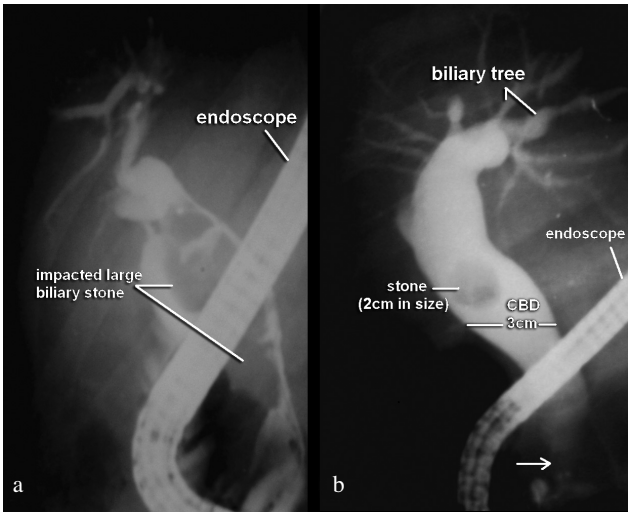

Figure 2. a) A dilated CBD due to impaction of a huge biliary stone; cholangiography was performed through a fistula tract just above the papilla, and **b)** obstruction of the distal CBD (white arrow) and proximal bile duct dilatation, which also included a biliary stone.
CBD: Common bile duct.

formation associated with necrosis of the adjacent ductal walls (6, 9). 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. Dietrich (10) reported a low CBD insertion in as many as 18% of his patients who underwent cholangiography (10, 11).

In a large study (219 patients), Csendes et al. (4) 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. In this study, the categorization of the patients is shown in Figure 3. 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 (4,6,9,12,13). The incidence rate in our study was 0.6%, which correlates well with the incidence rate reported in the literature.

Although MS is an unusual condition, suspected cases should be examined with ultrasonography or CT for anatomical evaluation in order to avoid any serious surgical consequences, including biliary injury or postoperative biliary leakage related to undetected fistula formation. ERCP has been recommended as the best screening method (14,15). In addition, it has been proposed as the best modality in the preoperative diagnosis and especially for initial management of MS with therapeutic decompression. It also helps determine the ideal surgical intervention method (16-18). If there is a suspicion of malignancy, advanced screening methods need to be performed. It has been reported that the incidence of malignancy in patients with MS (27%) is significantly higher than in patients with long-standing cholelithiasis (2%) (19).

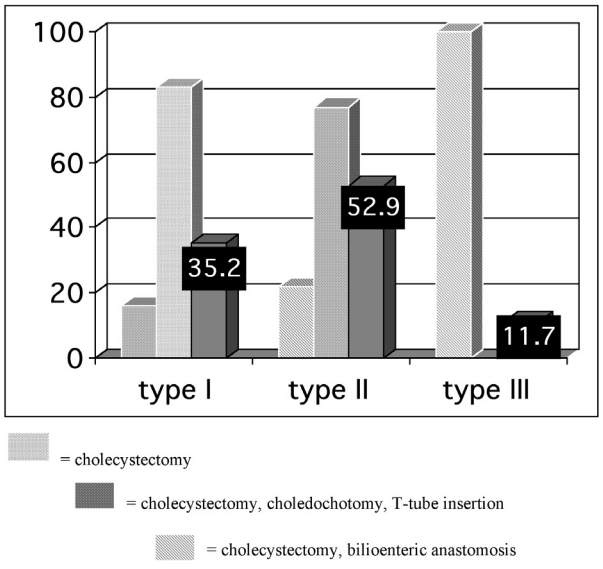

Figure 3. The categorization of cases with Csendes classification (type; n, %) in all patients (plain grey colons) and type of the surgical procedure performed in the patients with Mirizzi syndrome.

The reports on the incidence of the MS classification vary. Whereas Csendes et al. (4) reported a dominance of incidence of type III, Chan et al. (16) in a study on 18 cases and Waisberg et al. (20) also reported the dominance of type I. In the present study, higher incidence of type II (53.8%) lesions was observed. This difference could be due to both increased knowledge and early diagnosis.

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 (13-15), 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 (14). In this study, intraoperative cholangiography was performed on 7 of 12 patients due to advanced fibrosis and subsequent increased risk of injury to the bile duct. Tan et al. (13) reported bile duct injury in 4 cases (16.7%); two of them occurred during open surgery while the others occurred during laparoscopic dissection.

The surgical treatment of type I MS generally involves minimal interventions such as partial (4, 14, 21) or total (open or laparoscopic) (3) cholecystectomy. The reported incidence rate of conversion to open cholecystectomy was remarkably high, with a range of 37-78% (22), and the incidence of 57% in our study was well correlated with the literature. However, some authors consider this a contraindication for laparoscopic cholecystectomy (15, 23, 24). In our series, most of the type I cases underwent only cholecystectomy, while only two of them needed T-tube insertion with choledochotomy after cholecystectomy for temporary decompression. However, endoscopic therapy has recently been used in the evaluation and treatment of patients with MS, mainly in types I and II (21,

22, 25). Nevertheless, because they involve cholecystocholedochal fistula with respect to both Csendes (4) and McSherry (8) classification, MS types II-IV varieties generally require more complex interventions. Incision of the CBD via longitudinal choledochostomy directly over the gallstone, followed by cholecystectomy and subsequent suturing of the remaining gallbladder flaps around a T-tube is one option (10, 11). 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 (6). However, isolated cholecystectomy in the patients with type II should be avoided to prevent likely postoperative biliary complications. Baer and colleagues (14) 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. These authors also suggested biliary-enteric bypass via Roux-en-Y choledochojejunostomy or a choledochoduodenostomy to reduce the mortality and morbidity risk of CBD stricture (6, 14). In the present study, most of the patients (77%) with type II were submitted to cholecystectomy with choledochotomy and insertion of T-tube for drainage. The remaining cases with type II and all of the type III MS cases underwent cholecystectomy with excision of the external bile ducts and reconstruction with Roux-en-Y hepaticojejunostomy except for two of the cases who underwent choledochoduodenostomy - one due to difficulty in repairing the fistula (type III MS) and the other due to advanced dilatation of the extrahepatic biliary tract (type I MS), in whom a periampullary diverticulum was also detected. Because choledochoduodenostomy in the absence of an adequately dilated CBD may not yield satisfactory results in the long term, it is usually not preferred. Roux-en-Y hepaticojejunostomy procedure is safer and easier to perform. Postoperatively, no significant bile leakage was observed. Benign biliary stricture occurred in one patient with type II who underwent only cholecystectomy without any additive surgical procedure. Fifteen months after the operation, Roux-en-Y hepaticojejunostomy was performed in this patient. Because excessive dissection may do further harm by enlarging the fistulous opening, ERCP control study was performed to confirm no fistula formation persisted. The clamping of the T-tube was routinely performed to test patient tolerance just two days before discharge. T-tube drain was removed 4-6 weeks postoperatively.

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 proceduse 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. Alternative surgical strategy may be used if advanced inflammatory process is observed. Thus, T-tube insertion was performed for two of those who were classified as type I. 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. If this is not possible, individual surgical management with respect to the intraoperative observation should be sought.

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