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Endoscopic Retrograde Cholangiography and Mirizzi Syndrome: A Single-Center Experience

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ABSTRACT

Objective: This study was an investigation of the experience of a single institution with cases of definitive or suspected Mirizzi syndrome (MS) based on endoscopic retrograde cholangiography (ERCP) findings and the correlation of the preliminary diagnosis to the final diagnosis.

Materials and Methods: This retrospective study was performed with the data of 58 patients treated between January 2010 and December 2019. Cases where the gallbladder or cystic duct compression of the main hepatic duct was clearly visible as the cause of biliary obstruction were classified in the ERCP report as definitive MS, and those with only a suspicion of compression were reported as suspected MS. In all, 22 patients had a definitive diagnosis of MS and 36 patients had a report of suspected MS. The ERCP reports were compared with surgical reports and the results were analyzed.

Results: Examination of the surgical reports of the 22 patients with a preliminary diagnosis of definitive MS based on the ERCP findings revealed that MS was confirmed in 15 patients (68%) and malignancy in 3 patients (14%). No pathology other than gallstones was detected in 4 patients (18%). Review of the surgical reports of the 36 patients with a preliminary diagnosis of suspected MS indicated that MS was detected in 5 patients (14%), malignant stenosis in 20 patients (55%), chronic pancreatitis in 1 patient (3%), and a choledochal cyst in 1 patient (3%). Of the 23 patients with malignancy, cholangiocarcinoma was detected in 16 patients, pancreatic cancer in 3 patients, and gallbladder cancer in 3 patients.

Conclusion: The results of this study illustrate the difficulty in accurately diagnosing MS and the frequent confusion with malignancy.

Keywords: Cholangiocarcinoma, cholangiopancreatography, endoscopic retrograde, gallbladder neoplasms, Mirizzi syndrome, pancreatic neoplasms

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INTRODUCTION

Mirizzi syndrome (MS) is a rare complication of gallstone disease, with a reported incidence of 0.06% to 5.7% in patients who underwent a cholecystectomy (1). In MS, a gallstone becomes impacted in the cystic duct or the neck of the gallbladder, causing compression of the common hepatic duct and resulting in obstruction and jaundice (2). It is clinically characterized by intermittent or persistent duct obstruction and jaundice (3). MS has been observed more frequently in women who had gallstones for a long period of time (4). The presence of MS increases the risk of bile duct injury during surgery; therefore, it is very important to diagnose it preoperatively and reveal the biliary anatomy (5).

However, since there is no pathognomonic finding for MS, it is difficult to make a clinical diagnosis before surgery (6). The first imaging method used in diagnosis is ultrasonography, but MS can be overlooked (7). The same is true for computed tomography. In the presence of periductal inflammation, MS can be mistakenly evaluated as gallbladder cancer, cholangiocarcinoma, or metastasis by ultrasonography or computed tomography (8). Furthermore, magnetic resonance cholangiopancreatography (MRCP) and endoscopic retrograde cholangiopancreatography (ERCP) can confirm the diagnosis in only half of the cases (9, 10).

MS mimics malignancy, especially gallbladder cancer, causing significant diagnostic difficulties. ERCP is accepted as the gold standard in the diagnosis of MS, with a sensitivity varying between 55% and 90% (7). This study was an examination of the diagnostic efficiency and reliability of ERCP for MS.

MATERIALS and METHODS

Ethics approval for this study was obtained from the Ethics Committee of Ankara City Hospital on November 25, 2020 (no: E1-20-1301). The study was conducted according to the ethical standards specified in the Declaration of Helsinki.

Study Location and Population

Patients with a diagnosis of biliary cholestasis treated at a Turkish tertiary referral center hospital between January 2010 and December 2019 were included in the study. The inclusion criteria were age >18 years and a diagnosis or suspicion of MS in an ERCP report. The exclusion criteria were a previous cholecystectomy or a prediagnosis of malignancy.

Data Collection

The clinical data were collected retrospectively from the medical records of the gastroenterology clinic and ERCP unit of a single center. The demographic data noted at baseline were gender, age, laboratory findings (liver function test results), and radiological/ERCP results. Subsequently, postoperative diagnosis data were also collected and reviewed.

Definitions

The classification of MS is made as follows (11):

Type I: No fistula

Type IA: Presence of the cystic duct

Type IB: Obliteration of the cystic duct

Type II-IV: Fistula present

Type II: Defect smaller than 33% of common hepatic duct diameter

Type III: Defect between 33% and 66% of common hepatic duct diameter

Type IV: Defect greater than 66% of common hepatic duct diameter

Statistical Analysis

IBM SPSS Statistics for Windows, Version 22.0 software (IBM Corp., Armonk, NY, USA) was used to perform the statistical analysis. Descriptive statistics (frequency, mean and SD, median and minimum-maximum) were calculated. Categorical variables were summarized as percentages. Comparisons of continuous variables were made by using Student's t-test or the Mann-Whitney U test, according to the normality of distribution. A chi-squared test or Fisher's exact test (when chi-squared test assumptions do not hold due to low expected cell count) was used to compare categorical variables in different groups.

RESULTS

This study was performed retrospectively using records from January 2010 through December 2019. A total of 9334 ERCP records were examined and 134 patients with a report indicating a finding of Mirizzi syndrome or possible Mirizzi syndrome were identified. A total of 60 patients were excluded from the study after the ERCP evaluation because other imaging test results could not be accessed or they were followed up at another medical center.

The remaining 74 patients were referred to surgery with a prediagnosis of MS. The pathology reports were reviewed and another 16 patients were excluded from the study because the surgical and pathological reports were insufficient to diagnose MS. The final analysis was conducted with 58 patients: 22 patients with definitive MS and 36 patients with suspected MS.

Table 1. Demographic data, ERCP findings, and postoperative diagnoses

	n	%
Age (years, mean±SD)	56±9.2	
Gender		
Female	22	38
Male	36	62
Total ERCP count	9334	
"Mirizzi"	22	38
"Mirizzi?" in ERCP report	36	62
Postoperative diagnosis		
Malignancy	23	40
Mirizzi syndrome	20	34.5
Choledochal cyst	1	1.7
Chronic pancreatitis	1	1.7
Normal except for cholelithiasis	13	22.4
Type of Mirizzi syndrome		
Type 1	14	70
Type 2	4	20
Type 3	1	5
Type 4	1	5

ERCP: Endoscopic retrograde cholangiopancreatography; SD: Standard deviation

Table 2. Other diagnoses in patients operated on with a preliminary diagnosis of Mirizzi syndrome

Diagnoses	n	%
Cholangiocarcinoma	16	64
Pancreas carcinoma	3	12
Gall-bladder carcinoma	3	12
Papillary carcinoma	1	4
Choledochal cyst	1	4
Chronic pancreatitis	1	4

SD: Standard deviation

The demographic data, ERCP findings, and postoperative diagnoses are provided in Table 1. The mean age of the patients was 56 years; 22 patients were women and 36 were men. Of the 22 patients with the preliminary diagnosis of definitive MS based on ERCP, the surgical reports indicated that MS was detected in 15 patients (68%) and malignancy in 3 patients (14%). No pathology other than gallstones was detected in 4 patients (18%). An ERCP image of MS is shown in Figure 1. Examination of the surgical reports of the 36 patients with a preliminary diagnosis of suspected MS based on ERCP who could not be diagnosed by other methods revealed that MS was detected in 5 patients (14%), malignant stenosis in 20 patients (55%), chronic pancreatitis in 1 patient (3%),

Table 3. Comparison of definitive MS and suspected MS groups by age, gender, diagnosis, and MS type

	Definitive MS (n=22)	Suspected MS (n=36)	p
Age (years, mean±SD)	53.6±10.2	57.4±9.2	0.57
Gender (female/male)	9/13	13/23	0.71
Postoperative diagnosis			<0.05
Normal except			
for cholelithiasis	4	9	
MS	15	5	
Malignancy	3	20	
Chronic pancreatitis		1	
Choledochal cyst		1	
MS type			0.74
Type 1	11	3	
Type 2	2	2	
Type 3	1	–	
Type 4	1	–	

MS: Mirizzi syndrome; SD: Standard deviation

Figure 1. **Endoscopic retrograde cholangiopancreatography image of Mirizzi syndrome**

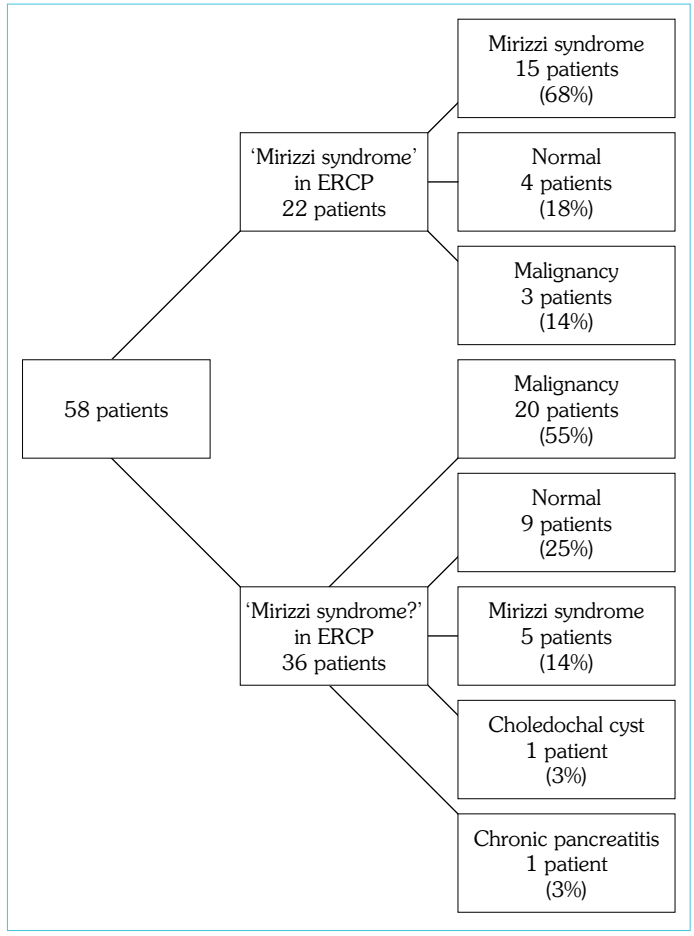

Figure 2. **Preliminary diagnoses based on endoscopic retrograde cholangiopancreatography results and final diagnoses after surgery**

and a choledochal cyst in 1 patient (3%). No pathology was found in the surgical reports of 9 patients other than gallstones (25%). The preliminary diagnoses according to the ERCP findings and the final diagnoses after surgery are provided in Figure 2.

Analysis of the results revealed that 14 patients had type I MS (70%), 4 patients had type II (20%), 1 patient had type III (5%), and 1 patient had type IV (5%) MS. Other diagnoses determined for patients operated on with a preliminary diagnosis of MS are given in Table 2.

Comparison of the definitive MS and suspected MS groups in terms of age, gender, and MS type yielded no statistically significant results. There was a statistically significant difference between the groups in the postoperative diagnosis ($p<0.05$) (Table 3).

Among the 23 patients with malignancy, cholangiocarcinoma was detected in 16 patients, pancreatic cancer in 3 patients, gallbladder cancer in 3 patients, and papillary cancer in 1 patient.

DISCUSSION

The results of this study indicated that most patients with a definitive diagnosis of MS based on the ERCP results were most often ultimately diagnosed with MS after surgery. However, in cases where the diagnosis of MS is only suspected, MS was frequently confused with malignancy.

MS is a rare condition. In previous studies, the rate of MS in cholecystectomy cases has been found to be around 1% (12). Few studies in the literature have applied a methodology similar to ours. We examined 9334 ERCP reports. "Mirizzi" was found in the report of 134 patients, and 20 patients were finally diagnosed with MS (0.2%). However, we cannot make a conclusive statement due to the patients excluded from the study.

In a study of 36 patients with MS, 20 were diagnosed with MS by ERCP and the sensitivity of ERCP in MS diagnosis was found to be 63% (13). In our study, 15 of 22 patients who were diagnosed with definitive MS by ERCP were subsequently diagnosed with MS after surgery, and the sensitivity of ERCP was found to be 68%. However, as stated above, we cannot precisely state the accuracy of this rate due to the excluded patients.

In this study, MS often presented with malignancies. It has been established that a bilirubin value of >10 mg/dL suggests malignancy. Notably, however, patients who may be diagnosed with MS and only normal choledochal stones may have a high bilirubin level upon hospital presentation due to the external compression of the biliary tract. The fluoroscopic images obtained with ERCP for MS patients are often very similar to images in cases of malignancy. In 23 of the 58 patients in our study, the final diagnosis was malignancy. Our results indicated that MS was most frequently confused with cholangiocarcinoma. Previous studies have also shown that MS can be confused with periductal infiltrative-type cholangiocarcinoma (14, 15). We found no studies in the literature related to a confusion between MS and pancreatic cancer. Pancreatic cancer was detected in 3 of our cases, and gallbladder cancer was detected in an additional 3 patients. Previous research has shown that MS can mimic gallbladder cancer (12, 16, 17). It has also been proposed that MS may increase the risk of gallbladder cancer. It may be that a similar relationship exists between MS and cholangiocarcinoma. MS can also be confused with benign diseases. In our study, the final diagnosis was a choledochal cyst in 1 case. It was noted in a case report that choledochal cysts can mimic MS (18).

The most important limitation of our study is the patients who were excluded. Although the word "Mirizzi" was mentioned in the ERCP reports of 134 patients, the study was conducted with the data of 58 patients. The most important reason for this was that most of the patients were followed up elsewhere after the ERCP and we could not obtain the final diagnosis. In addition, 16 patients were excluded from the study because their surgical reports were insufficient to permit a diagnosis of MS or other diseases. Nonetheless, according to our review of the literature, this study is one of the most comprehensive studies of MS to date. Another important limitation of our study is that detailed information about patient follow-up was not available as a result of its retrospective design.

CONCLUSION

The results of this study illustrate that an accurate preoperative diagnosis of MS is still difficult and that MS is frequently confused with malignancy. We think that malignancy should be considered in the diagnostic process, especially in cases when ERCP findings only yield a suspicion of MS. Prospective studies with larger patient groups and longer patient follow-up are needed to supplement our knowledge and inform our practice.

Ethics Committee Approval: The Ankara City Hospital Clinical Research Ethics Committee granted approval for this study (date: 25.11.2020, number: E1/1301/2020).

Informed Consent: Since the study is retrospective, written informed consent could not be obtained from the patients.

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