

Mirizzi syndrome: diagnosis and management of a challenging biliary disease

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Abstract

Background: Mirizzi syndrome is a condition, difficult to diagnose pre-operatively and treat, represent a particular challenge for hepatobiliary surgeons. Furthermore, it increases the risk of intra-operative biliary injury, particularly during laparoscopic cholecystectomy. The aims of this study were to point out some particular aspect of diagnosis and treatment of this condition that will be helpful for the surgeons.

Methods: We retrospectively reviewed all records of the patients, surgically treated for Mirizzi syndrome from January 2013 to January 2018 in Dhaka Medical College Hospital, Shaheed Sohrwardy Medical College Hospital and BIRDEM General Hospital. Patients' presentation, diagnostic methods, treatment and complications were recorded.

Results: During the study period, a total of 1320 cholecystectomy were performed, out of which 50 patients were diagnosed with Mirizzi syndrome, representing an incidence rate of 3.78%. Male to female ratio was 0.72:1 and mean age was 54.14 years. When we analyzed the main symptoms in the clinical debut, we found that common features were jaundice (31, 62%), cholangitis (20, 40%) and abdominal pain (22, 44%). Roux en Y hepaticojejunostomy was the treatment of choice and subtotal cholecystectomy were done for 7 cases and laparoscopic cholecystectomy for 2 cases. Mean hospital stay was 4.8±2.9 days. There was no post-operative mortality. Two cases of biliary fistula resolved with conservative management and another case required percutaneous treatment for resolution of an intraperitoneal postoperative collection.

Conclusion: Mirizzi syndrome continues to be a disease of difficult diagnosis and treatment. General surgeons without long experience in hepatobiliary surgery should refer the patient to a specialized hepatobiliary surgical center. In most of the cases biliary reconstruction is inevitable. It is important for the surgeons to know the disease and possible intra-operative challenging situations.

Key words: common bile duct, laparoscopic cholecystectomy, Mirizzi syndrome, subtotal cholecystectomy.

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Introduction

Mirizzi syndrome was first described by one of the major biliary surgeons of past century, Pablo Luis Mirizzi (1893-1948) in 1948. This syndrome is an uncommon complication of chronic gall stone disease. Pathophysiologically, this condition involves extrinsic

compression of the bile duct by pressure applied upon indirectly by an impacted stone in the infundibulum or neck of the gall bladder. In turn, the resulting chronic inflammation and ulceration leads to varying degree of cholecysto-biliary fistula. Furthermore, cholecysto-enteric fistula may also occur.¹

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Clinical presentations of Mirizzi syndrome ranges from asymptomatic to nonspecific, with obstructive jaundice (27.8-100%) being the most common, elevated liver enzymes (aspartate aminotransferase/alanine amino transaminase), right upper quadrant pain (16.7-100%) and constitutional such as fever, nausea, vomiting, diarrhoea and constipation. Mirizzi syndrome may also present with gallstone ileus.² Very often the presentation of Mirizzi syndrome mimics with biliary malignancy³ and misdiagnosis is therefore not infrequent.

Various classifications were described in the literature with McSherry's and Csendes being the most common.

Csendes classification of Mirizzi syndrome	
Type	Description
Type I	Type I (A) Extrinsic compression in the common hepatic duct (CHD), caused by stone impacted in the cystic duct or in the infundibulum
	Type I (B) Absence of cystic duct (5)
Type II	Presence of cholecystocholedochal biliary fistula involving one third of the circumference of the CHD wall
Type III	Presence of cholecystocholedochal biliary fistula with a diameter over two thirds of the CHD wall
Type IV	Presence of cholecystocholedochal fistula involving entire circumference of the CHD wall
Type V	New addition by the Csendes group include any of the Mirizzi syndrome types with cystoenteric fistula

Csendes classification is useful for planning the management strategy; so, while simple cholecystectomy is suitable for type I, partial cholecystectomy adopting the fundus first dissection technique leaving a cuff of infundibulum is needed for type II to avoid damage to the main bile ducts. Choledochoplasty can be considered for type IA and is recommended for type II and early type III to restore adequate diameter of hepatic duct. For type III to V, Roux en Y hepatico-jejunostomy is conventional.⁴

Surgical treatment should be considered after a careful assessment of the local situations and anatomy. During the past 5 years we observed some consecutive cases of Mirizzi syndrome which led us to review this field in an attempt to describe the diagnosis and treatment of this challenging condition.

Methods

This retrospective study was based on the diagnosis and treatment results of 50 patients with Mirizzi syndrome type I to V (A. Csendes, 1989), who were treated from January 2013 to January 2018. Study was conducted in Dhaka Medical College Hospital, Shaheed Sohrwardy Medical College Hospital and BIRDEM General Hospital. Patients diagnosed with cholelithiasis by ultrasonogram (USG) with raised bilirubin and/or alkaline phosphatase or clinically icteric were suspected for Mirizzi syndrome and were evaluated by magnetic resonance cholangiopancreatography (MRCP), were included in the study. Alongside 13 cases were referred by gastroenterology department with the diagnosis of Mirizzi syndrome by endoscopic retrograde cholangiopancreatography (ERCP) with or without stenting. In

few cases Mirizzi syndrome was diagnosed preoperatively without any prior suspicion specially in type I or type II. The following data were considered: sex and age of the patients, presenting symptoms and previous treatment, preoperative radiological investigations, preoperative diagnosis, surgical procedure performed, postoperative course, morbidity and mortality. Follow up data were obtained by direct clinical observation, laboratory findings and imaging.

Results

From January 2013 to January 2018, a total of 1320 cholecystectomy were performed, out of which 50 patients were diagnosed with Mirizzi syndrome representing an incidence rate of 3.78%. Male to female ratio was 0.72:1 and mean age was 54.14 years. Characteristics are shown in Table I. Frequent symptoms were jaundice, cholangitis and abdominal pain in 31 (62%), 20 (40%) and 22 (44%) cases respectively.

Preoperative imaging included USG of hepatobiliary system. Cholecystectomy was initiated by laparoscopic approach in 7 (14%) patients out of which 2 cases were converted to open cholecystectomy. In all cases of type III, IV and V, Roux en Y hepatico-jejunostomy were done and subtotal cholecystectomy for 7 cases of type II, simple open cholecystectomy for 5 patient of type I and type II. In two patients of type II, subtotal cholecystectomy was adjuncted by fistula repair. Mean hospital stay for all patients was 4.8±2.9 days.

Thirteen patients were referred by gastroenterologists, where ERCP with stenting were performed for biliary decompression. Forty patients were diagnosed preoperatively and the rest in surgery, representing 80%

versus 20%. Cholecystectomy was performed in all cases. Six patients were operated on emergency basis because of clinical suspicion and positive USG of acute cholecystitis.

There was no postoperative mortality. Morbidity included two cases of biliary fistula that resolved with conservative management and another case that required

percutaneous treatment for resolution of an intraperitoneal postoperative collection.

The average post-operative follow up was 9±3 months but 3 patients were lost to follow up. After the histopathological study of surgical specimen, we found one case of gall bladder carcinoma in stage T2A in which oncological resection was done in second surgery.

Table I Distribution of patients by type of Mirizzi syndrome, demographic, clinical, laboratory data (preoperative diagnosis and imaging with each case) (n=50)

Characteristics	Type I	Type II	Type III	Type IV	Type V
N (%)	7 (14)	13 (26)	25(50)	4 (8)	1 (2)
Mean age (year)	42.5	53.2	49.7	59.6	65.7
Sex (M/F)	2/5	8/13	8/9	1/3	1/0
Clinical presentations					
Jaundice	5	7	15	4	0
Abdominal pain	7	5	7	3	0
Cholangitis	2	06	08	3	1
Laboratory data					
Leucocytosis (×10 ³ /uL)	9.5±1.5	10.3±3.5	11.3±4.3	12.5±22.5	14.5±2.5
Serum total bilirubin(mg/dl)	1.8	2.5	3.2	4.1	2.2
Alkaline phosphatase (u/L)	37±19	53±22	61±22	89±21	98±27

Table II Preoperative diagnosis based on imaging (n=50)

Imaging (N)	Cholelithiasis	Mirizzi syndrome (%)	Carcinoma gall bladder	Cholangiocarcinoma of proximal CBD
USG (50)	15	20 (40%)	8	7
MRCP (37)	1	29 (78.3%)	5	2
ERCP (13)	0	11 (84%)	0	2

Table III Procedures performed in different types of Mirizzi syndrome and morbidity with hospital stay (n=50)

	Type I	Type II	Type III	Type IV	Type V
Procedures (%)					
Laparoscopic cholecystectomy	5 (71.4)	0 (0)	0 (0)	0	0
Open cholecystectomy	2(28.5)	3(23)	0 (0)	0	0
Subtotal cholecystectomy	0 (0)	7 (53.8)			
Subtotal cholecystectomy with fistula repair	0 (0)	2 (15.3)			
Roux en Y hepaticojejunostomy	0 (0)	1 (7.6)	25 (100)	4 (100)	1 (100)
Morbidity					
Wound infection	0	2	3	2	10
Bile leak	0	0	1	1	
Respiratory tract infection	1	1	4	3	0
Urinary tract infection	2	0	2	2	0

Discussion

The frequency of Mirizzi syndrome presented in different series of cholelithiasis is highly variable: less than 1% in western countries and around 4.5% in developing countries.⁵ In our study, this percentage is high, because of the bias being a surgical unit in tertiary hospitals, where a large number of patients with complex biliary disease are being admitted.

Mirizzi syndrome is a difficult disease to diagnose because of the absence of pathognomonic sign and symptoms. Characteristically the is often disease scarce and occur often as an intermittent jaundice, an unexpected findings in the course of laparoscopic cholecystectomy or starts in Boulder et syndrome.⁶ In our series preoperative diagnosis was 80% and jaundice was the main sign. Therefore, it is important to have Mirizzi syndrome in differential diagnosis of obstructive jaundice.

In the literature, the idea that majority of patients present pain in episodic or right upper quadrant (60% to 100%), jaundice (50% to 100%) and elevated liver enzymes (10%) is reinforced. In our series the most common two features were jaundice and abdominal pain.

Cholangitis has also been referred as initial abut of Mirizzi syndrome. The mean age of presentation of Mirizzi syndrome was high in our series in comparison to other studies.⁷ It is noteworthy that in recent years, we are experiencing an increased number of cholecystectomy in older patients with gallstone because of increased life expectancy.

Adequate preoperative diagnosis with meticulous surgical planning is vital for the management of Mirizzi syndrome. Unfortunately, this preliminary diagnosis only can be made in 18% to 62% of patients.⁸ A fundamental aspect to note in our study is that this number could increase (80% in our study) with the support of MRCP, ERCP and computed tomography scan which allowed a more selective, programmed and laparoscopic approach. This possibly contributed to minimize postoperative complications related to the biliary tract.

ERCP is useful in assessing the existence of a cholecystobiliary fistula. However, it has sensitivity of over 55% and a significant morbidity and not all patient in our series met the criteria of ERCP. Endoscopic treatment of Mirizzi syndrome includes with stenting

or prosthesis to relieve jaundice and allow elective surgery. It is also used in removal of CBD stones with a balloon or dorm basket.⁹

Current evidence suggests that laparoscopy can not be recommended as a standard procedure in Mirizzi syndrome. In fact preoperative diagnosis of the syndrome is an important predictor the success of the approach, especially in type I and type II.² This approach remains controversial because of high rates of conversion.^{10,11} Most authors suggest that type I Mirizzi syndrome is ideal for minimal invasive approach.⁸

Faced with type I Mirizzi syndrome, treatment of choice is laparoscopic cholecystectomy with retrograde dissection of the gall bladder. As mentioned subtotal cholecystectomy is an safe and easy option and definitive strategy in more complex approach especially in case of cirrhotic liver with portal hypertension.²

Faced with type II Mirizzi syndrome, treatment depends upon the severity of fistula and involvement of the surrounding tissues. In few cases, it is required to perform a partial cholecystectomy, with using the remaining wall of cystic duct or gall bladder wall to repair the fistula with T tube placement. This option is the most useful if the surrounding tissue is healthy.¹²

The Roux en Y hepatico-jejunostomy is the safest option for in case of bile duct tissue ischemia. Some authors reported that type Mirizzi syndrome could treated with laparoscopic cholecystectomy and ERCP stenting.¹³ The stent would overcome the resistance of the sphincter and allow the repaired fistula to heal.¹⁴ The prognosis is extreme favorable in type I Mirizzi syndrome, as it is solved with simple cholecystectomy. Post-operative morbidity increase with treatment of severe types (II, III, IV).

Conclusion

In our study, management of Mirizzi syndrome ranges from laparoscopic cholecystectomy to Roux en Y hepato-jejunostomy. Pre-operative imaging specially MRCP has high sensitivity which can help in operative strategy. ERCP is helpful for certain conditions where MRCP could not delineate the biliary anatomy clearly. Laparoscopic approach should be reserved for type I and open for rest of occasions by Roux en Y hepatico-jejunostomy mostly. Few cases for type II can be subtotal cholecystectomy and fistula repair. Nevertheless, for the

diagnosis and treatment of Mirizzi syndrome whether old or new studies are limited by small sample size and therefore studies with larger sample size is required to set a clear guideline for management of Mirizzi syndrome.

Conflict of interest: Nothing to declare.

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