



# Diagnosis and Treatment of Mirizzi Syndrome: Case Report and Literature Review

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## Abstract

The Mirizzi syndrome is a set of symptoms related to the obstruction of the common hepatic duct by an impacted stone in the cystic duct or gallbladder infundibulum. The objective of article is to describe a case of a woman with obstructive jaundice and cholangitis diagnosed by Endoscopic Retrograde Cholangiopancreatography (ERCP), and the subsequent surgical management of the patient.

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## Introduction

The Mirizzi Syndrome (MS) was first reported by Pablo Luis Mirizzi (1893-1964), one of the major biliary surgeons in the past century. Mirizzi is mainly known for conceiving and performing the first intraoperative cholangiography in 1931, a procedure that had a strong impact on biliary surgery of the XX century. Mirizzi first described the eponymous syndrome in 1948, presenting the case of a patient with a big stone impacted in the gallbladder infundibulum. The impact of the stone caused jaundice by extrinsic compression of the Common Bile Duct (CBD) and a productive inflammation extending from the gallbladder to the CBD. The Mirizzi syndrome is an important complication of gallbladder stones and requires a differential diagnosis from gallbladder cancer involving the CBD. In some cases, the stone

erodes from the gallbladder into the CBD producing a fistula [1]. In rare instances, the Mirizzi syndrome causes benign obstructive jaundice triggered by impacted calculus in Hartmann bag or cystic duct. This condition triggers the extrinsic obstruction of the common hepatic duct, which is difficult to treat when intense inflammation occurs [2].

A classification of the severity of the Mirizzi syndrome has been proposed based on ERCP findings. Type I involves only external obstruction of the common duct, whereas type II involves fistula formation between the two ducts. This classification scheme further grades the severity of the syndrome based on the degree of CBD destruction (Table 1) [3]. As it might be expected, the more severe forms of the Mirizzi syndrome are less common [4].

**Table 1:** Csendes classification of the severity of Mirizzi syndrome.

Type of MS	Description	Incidence
1	External compression of the common hepatic duct by an impacted stone	11-78%
2	Presence of a cholecysto-biliary fistula involving <1/3 duct wall	15-41%
3	Presence of a cholecysto-biliary fistula involving up to 2/3 duct wall	3-44%
4	Complete destruction of the wall of the bile duct	1-4%

## Case presentation

Here we report the case of a 56-year-old female patient who presented abdominal pain in the upper right quadrant associated in the last month with jaundice, and in the last week with intermittent high fever. In the physical examination, jaundice scleras and abdominal pain were observed at the level of the upper right quadrant. For this reason, we decided to admit the patient for the study and treatment of her symptoms.

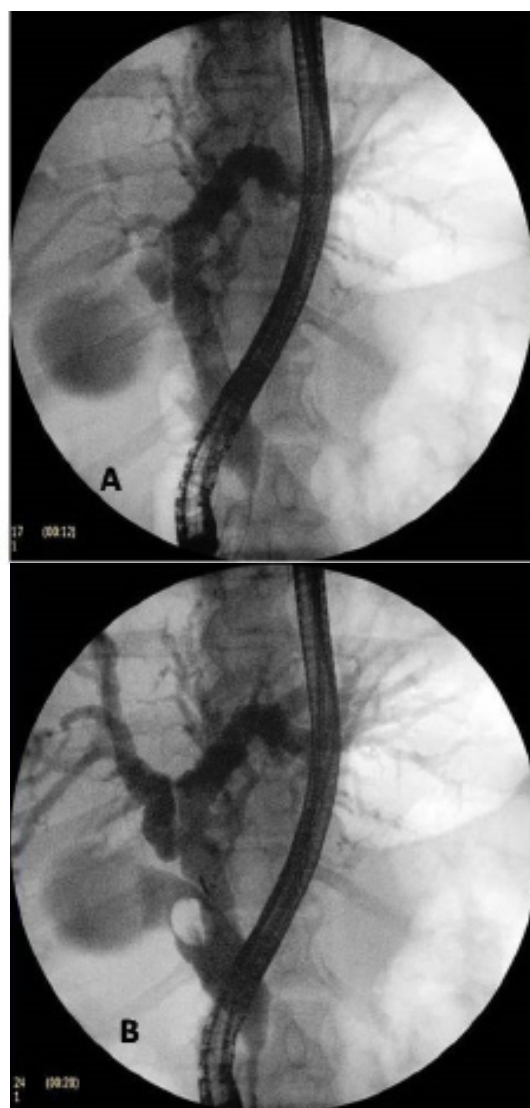
Next, we summarize the results of the blood tests: Hemoglobin: 9.9 g/L, WBC:  $13.9 \times 10^9/L$ , ALT: 249 U/L (normal: 0–40 U/L), AST: 131 U/L (normal: 0–40 U/L), ALP: 303 U/L (normal: 60-290 U/L),  $\gamma$ -GGT: 635 U/L (normal: 5- 45 U/L), total bilirubin levels: 168  $\mu\text{mol/L}$  (normal values for total bilirubin: 0–25  $\mu\text{mol/L}$ ), direct bilirubin levels: 162  $\mu\text{mol/L}$

The abdominal ultrasound showed slight dilatation of the intrahepatic bile ducts and a gallbladder with a non-thickened wall. The CBD was dilated (measuring 14 mm) with lithiasis in its interior.

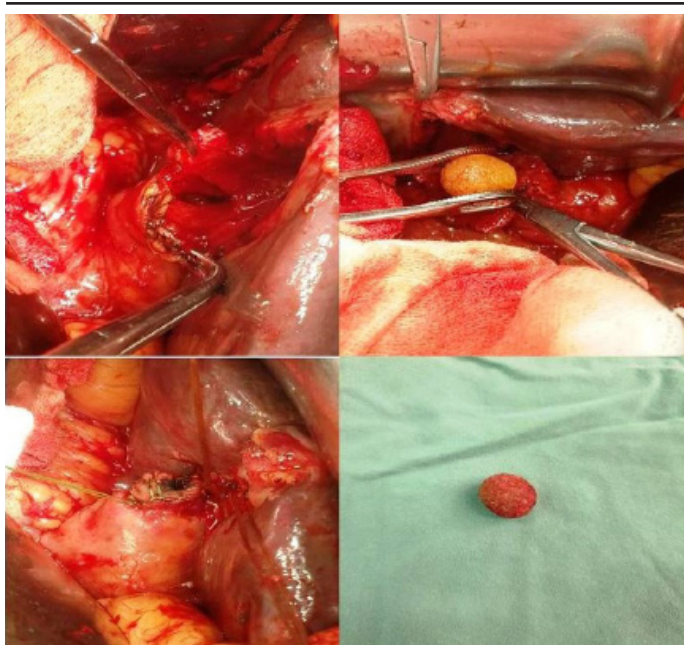
We decided to start treatment with intravenous Ceftriaxone and indicated ERCP with a presumptive diagnosis of Choledochal Lithiasis.

The ERCP showed a slight dilatation of the intrahepatic bile ducts. The common bile duct was dilated to its distal portion where extrinsic compression by lithiasis of approximately 2 cm was observed located in the cystic duct, which was also dilated. The gallbladder was normal. In addition, a biliary sphincterotomy procedure was performed and a 5 cm, 10 Fr plastic biliary stent was placed without complications. Our conclusion was the patient suffered from Mirizzi syndrome type I (Figure 1).

The patient had a good evolution after the ERCP and we discussed with the surgery service options for a definitive treatment. Accordingly, the patient was transferred to surgery where a subtotal open cholecystectomy was performed without complications and with satisfactory evolution (Figure 2).



**Figure 1:** ERCP pictures. (A). Common bile duct dilated. (B). Lithiasis located in the cystic duct. Normal gallbladder.



**Figure 2:** The intraoperative pictures were consistent with Type I Mirizzi Syndrome. A calculus at the cystic duct was removed by subtotal cholecystectomy.

### Discussion

The Mirizzi syndrome is an uncommon but clinically important complication of gallbladder disease. Extrinsic compression of the common hepatic duct from gallstones or debris within the cystic duct or from within the gallbladder itself causes intrahepatic ductal dilatation. As a consequence, obstructive jaundice and cholangitis may ensue. Significant morbidity and mortality can be associated with these disease states [5].

Reverdito and collaborators [2] reported that, out of 3691 patients admitted for treatment of gallstones at from December 2001 to September 2013, 23 (0,6%) had MS. The grades III and IV of the disease occurred in 13 patients (0,35%), ten being women (76,9%) and three men (23,1%). The mean age was 55,6 years, ranging from 31 to 89. Two patients had hypertension and diabetes mellitus type II as pre-existing conditions. The average time of the disease was 375 days, ranging from one to 60 months. Seven patients were admitted to the emergency room and six, on an outpatient basis. The most prevalent symptom was abdominal pain (92%), followed by jaundice in 84,6% of the patients; fever was present in two of the seven patients with colangitis. Abdominal pain was the most common symptom, followed by jaundice and, less frequently, by the classical triad of cholangitis [6].

Another study by Tataria and coworkers [7] also reported abdominal pain as the most common symptom in 54–100% of the patients, followed by jaundice in 24–100% and cholangitis in 6–35% of the patients. Biochemical parameters of liver function showed a cholestatic pattern. Serum bilirubin measurements delivered values as high as 30 mg/dl with an average value of 7–10 mg/dl. The serum alkaline phosphatase levels ranged from normal to about 3–10-fold rise [7].

In all cases, abdominal ultrasound was utilized as a primary investigation technique for patients with abdominal pain and jaundice. MS was confirmed on the evidence of dilated intrahepatic ducts and common hepatic duct by observing the narrowness of the compressed external bile duct that caused by the calculus [8,9].

Percutaneous trans-hepatic cholangiography provided an alternative option in cases where ERCP failed. Secondly, Magnetic Resonance Cholangio Pancreatography (MRCP) was used in 7,1% to 80% of cases. Interestingly, Computed Tomography (CT) was often performed to exclude tumors. However, most authors preferred the combination of  $\geq 2$  modalities for diagnosis of MS. 18% to 62% of the patients were diagnosed with the Mirizzi syndrome prior surgery. This number increased to 85,9% when MRCP and ERCP techniques were used in combination [9]. ERCP is considered the gold standard for the diagnosis of the Mirizzi syndrome given the sensitivity (55% to 90%) of this technique. In addition to being a diagnostic procedure, ERCP is also a therapeutic technique that enables the extraction of gallstones from the CBD or biliary drainage using stents, allowing for elective surgery to be performed [10].

Endoscopic Ultrasonography (EUS) is another important diagnostic modality in cases where the abdominal ultrasound delivered negative results of suspected biliary pancreatitis for microlithiasis. However, while endoscopic ultrasonography is used in some units for confirmation of choledocholithiasis prior to therapeutic ERCPs, EUS does not seem to play any major role in the diagnostic or management planning protocols of Mirizzi's syndrome. On the other hand, intra-operative laparoscopic ultrasound, if available, could be used to diagnose MS and in aid management planning [6].

Surgery remains the preferred approach for the treatment of the Mirizzi Syndrome. Laparoscopic Cholecystectomy (LC), which has been used since 1987, was first reported to successfully treat Type I MS. However, most surgeons do not recommend LC as a viable standard treatment due to the increased risk of bile duct injury and a high conversion rate with this condition. In the reports discussed here open surgery was still the favorite treatment modality, accounting for 40% to 100% of cases [9]. One systematic review, by Antoniou et al, [11] associated laparoscopic treatment for the Mirizzi Syndrome with low success rates; consequently, the authors do not recommend this technique.

The choice of appropriate surgical technique depends on the type of Mirizzi syndrome. It is well established that the treatment for type I MS is cholecystectomy or sub-total cholecystectomy. Cholecystectomy in the context of MS was initially considered an absolute contraindication to laparoscopic surgery, but it can be attempted laparoscopically in experienced hands. All other types of Mirizzi syndrome should be managed with open surgery [12].

The surgical management of Csendes type I and IV MS is well established, with subtotal cholecystectomy as the procedure of choice in Type I MS and cholecystectomy with Roux-en-Y hepatico-jejunostomy for Type IV. However, for small to moderate size cholecysto-choledochal fistula, Csendes type II and type III MS, the procedure (choledochoplasty and bilio-enteric anastomosis) of choice for type II and type III MS is unclear. Some authors have raised concerns related to choledochoplasty procedures, as direct repair using gallbladder flap is prone to failure due to associated inflammatory process; rather, they recommend bilio-enteric anastomosis [13].

### Conclusion

Mirizzi syndrome is a rare cause of abdominal pain and obstructive jaundice that requires a high diagnostic suspicion and constitutes a therapeutic challenge for gastroenterologists and surgeons.

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