

CONTRIBUTION OF ENDOSCOPIC RETROGRADE CHOLANGIOGRAPHY IN MIRIZZI SYNDROME: THE EXPERIENCE OF A MOROCCAN CENTER.

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doi:

doi url:

Published in March 2025

Abstract

Introduction: Mirizzi syndrome is a rare complication of chronic vesicular lithiasis. It has been described as an obstruction of the common bile duct by a stone embedded in the vesicular neck or cystic duct, which through an associated inflammatory reaction totally or partially compresses the common bile duct leading to the formation of a cholecysto-biliary fistula. The aim of our work is to analyze the particular role of endoscopy in Mirizzi syndrome in our clinical practice. **Patients & Methods:** This is a retrospective descriptive study carried out in the Hepato-gastroenterology department of the University Hospital of Fez; we included all patients who underwent endoscopic retrograde cholangiopancreatography (ERCP) between January 2013 and December 2022 with a confirmed diagnosis of Mirizzi syndrome (MS). Data were collected from ERCP registers. **Results:** A total of 35 MS patients were endoscopically managed between 2013 and 2022. Presenting symptoms were jaundice (60%), pain (20%) and cholangitis (62%). ERCP established diagnosis in 100 % cases. Beltran Type I was the most common type presentation (77%). All patients underwent endoscopic sphincterotomy and stone extraction was performed in 25 patients. A total of 26 patients underwent surgery with good outcome. Total cholecystectomy was performed in 18 cases, subtotal cholecystectomy in 6 cases and choledocotomy combined with biliary-digestive anastomosis was performed in 2 cases. The overall post-operative mortality rate was 2.8%. **Conclusion:** This study confirmed the place of ERCP in the diagnosis and treatment of Mirizzi syndrome.

Keywords: ERCP, Endoscopy, Mirizzi syndrome (MS), computed tomography (CT), Morocco.

INTRODUCTION

Mirizzi syndrome results from the compression of the common bile duct by one or more calculi within the cystic duct or gallbladder. The syndrome occurs approximately in 1 out of 1000 patients [1,2] with gallstones. Patients may present with recurrent episodes of jaundice and cholangitis. It can be associated with acute cholecystitis. A low insertion of the cystic duct into the common bile duct as well as a tortuous cystic duct are the main risk factors. Surgical management is the gold standard treatment for Mirizzi Syndrome, although this is challenging for several reasons.

MATERIALS & METHODS

This is a retrospective descriptive study carried out in the Hepato-gastroenterology department of the University Hospital of Fez, which included all patients who underwent ERCP between January 2013 and December 2022 with confirmed diagnosis of Mirizzi Syndrome. Data were collected from ERCP registers. All patients with radiological suspicion of Mirizzi syndrome using MRI or CT scan whatever the grade that underwent ERCP or in whom the diagnosis was made accidentally during ERCP were included in the study without age restriction.

RESULTS

During the period of the study, 2037 ERCP were performed among them 1320 for lithiasis from which 35 cases of Mirizzi syndrome were enrolled (1.7% of the total number of ERCP and 2.6% among biliary stones cases). The average age of patients was 49 years old [35- 72], with a sex ratio of 0.57. Common bile duct lithiasis was identified in 71% of patients. Of the 35 patients : 5 patients underwent cholecystectomy, while 10 patients were known to have lithiasis of the gallbladder. The clinical presentation of Mirizzi Syndrome ranged from asymptomatic to non-specific symptoms : 62% of patients had jaundice , 20% complaint from right upper quadrant abdominal pain and 34% had fever, nausea or vomiting. Patients had undergone a biological work-up, including an hepatic work-up showing cytotoxicity with biological cholestasis. All patients got a radiological work-up involving abdominal ultrasound and CT scan (**Figures 1 & 2**). Only 10 patients benefited from biliary MRI. Eight patients had a cholecysto-biliary fistula.



Figure 1 : CT scan demonstrating an impacted stone in the cystic duct [3]



Figure 2: This CT reconstruction shows the smooth narrowing of the common hepatic duct at the site of the impacted gallstone. [4]

The indications for ERCP were as following: 8 cases of acute gallstone pancreatitis (22%), 22 cases of cholangitis (62%) and 5 cases of isolated cholestatic jaundice (14%).

Regarding the diagnosis of mirizzi syndrome, all patients underwent abdominal ultrasound that found common bile duct (CBD) dilatation. The diagnosis was strongly suspected by imaging (CT scan and/or MRI) prior to ERCP in 57% of patients; it was incidental during ERCP in 42%.

All patients underwent ERCP, which enabled the diagnosis to be made and confirmed in 100% of cases. CBD Dilatation upstream of a gallstone (Mirizzi syndrome type 1 according to the Beltran classification) was found in 27 cases (**Figure3**).



Figure 3: ERCP in a case of Mirizzi type I showing the smooth and regular stricture

Six patients had a cholecysto-biliary fistula (Mirizzi syndrome type 2 according to the Beltran classification).

All patients underwent endoscopic sphincterotomy and stone extraction was performed in 25 patients (balloon extraction 70%, dormia 25%, mechanical lithotripsy 5%). The failure of stone extraction was reported in 5 cases: a plastic stent was placed in 3 cases and 2 patients were proposed for surgery.

A total of 26 patients underwent surgery (laparotomy): total cholecystectomy was performed in 18 cases with good outcome, subtotal cholecystectomy in 6 cases and choledocotomy combined with biliary-digestive anastomosis was performed in 2 cases. One patient passed away post-operatively following sepsis. For other patients, follow-up was straightforward, with disappearance of clinical signs and normalization of the biological balance within 2 months. One patient was taken back for stent debridement and replacement during follow-up. Five patients were readmitted for residual lithiasis.

Discussion:

Mirizzi syndrome has garnered increasing attention from researchers worldwide, as reflected in the development of various classifications by experts such as McSherry, Csendes, Starling, and Matallana. However, Beltrán proposed a more simplified classification system, which we have adopted for our study (**Figure 4**).

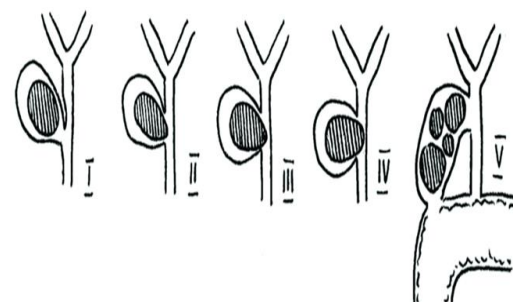


Figure 4: Beltran classification for Mirizzi syndrome [5]

A review of the literature reveals that Mirizzi type I is the most frequently reported, with incidences ranging from 10.5% to 51%, compared to 77% in our study. In contrast, Mirizzi type II, reported in 57% of cases in the literature, was found in 17% of our cases. The other types of Mirizzi syndrome are relatively rare in comparison.

The literature consistently identifies the primary symptoms of MS as upper right quadrant abdominal pain and jaundice. Several studies have also highlighted abnormal laboratory results, with MS, being a biliary disorder, typically causing elevated levels of aspartate aminotransferase (AST), alanine transaminase (ALT), alkaline phosphatase (ALP), Gamma-glutamyl transpeptidase (GGT) and bilirubin. The inflammatory nature of the condition also leads to an increase in WBC count. However, these values can fluctuate depending on the stage of the syndrome.

Ultrasonography is often the initial diagnostic tool in many studies, though its sensitivity varies, ranging from a few percent up to approximately 50%. [6] CT scans, on the other hand, are frequently noted for their ability to differentiate MS from malignant strictures. While MS can mimic cholangiocarcinoma, it is equally important to recognize the coexistence of MS with gallbladder carcinoma (GBC) [7- 10]. Research by Prasad et al. found that patients with both MS and GBC were, on average, a decade older and had a history of symptoms twice as long as those with MS alone. [7] Magnetic resonance cholangiopancreatography (MRCP) and endoscopic retrograde cholangiopancreatography (ERCP) are two competing diagnostic modalities. Both have shown high sensitivity, with MRCP ranging from 63–89% and ERCP from 63–72%, although individual studies have reported both higher and lower effectiveness. Despite this, ERCP remains the gold standard in MS diagnosis due to its superior sensitivity and its ability to offer therapeutic interventions [11, 12]. Notably, a combination of MRCP and CT has been suggested to increase diagnostic accuracy [13;14], our study has shown that CT combined with MRCP led to an MS diagnosis in 74% of cases, while ERCP confirmed the diagnosis in 100% of cases. Other diagnostic methods are also emerging in the literature. Endoscopic ultrasound (EUS) has shown comparable sensitivity to ERCP, ranging from 63-73%. [11,13] Wehrmann et al. reported promising results with intraductal ultrasound (IDUS), which demonstrated diagnostic accuracy in 97% of cases. [15] However, the adoption of IDUS has been limited due to the high costs, lengthy procedure times, and technical challenges.

Surgical intervention remains the primary treatment for Mirizzi syndrome. In our study, 74% of patients underwent surgery. However, surgical management is complicated by several factors. One significant issue is the low index of suspicion among surgeons, largely due to the rarity of the condition. Gallbladder

surgery is often performed in patients with shorter histories of illness, well before the onset of MS. Additionally, preoperative diagnosis is often missed, further complicating management.

Dense adhesions, caused by chronic inflammation, can distort the anatomy and make surgical dissection more difficult. The advancement of cholecystobiliary or cholecysto-enteric fistulas further increases the risk of bile duct injuries or severe bleeding during dissection of the Calot's triangle. Additionally, inflammation can lead to complications such as cutaneous fistulas, secondary biliary cirrhosis, delayed biliary strictures, and, in severe cases, death.

Historically, laparotomy has been the preferred approach for managing Mirizzi syndrome due to its relative safety compared to the laparoscopic technique. Laparoscopic surgery is associated with high conversion rates (ranging from 31% to 100%) and a higher incidence of bile duct injury. Laparotomy, while more invasive, offers better visualization, tactile feedback, and allows for the removal of gallbladder stones before proceeding with cholecystectomy, despite its higher complication rates, longer recovery times, and extended hospital stays. The minimally invasive laparoscopic approach offers several advantages, such as shorter hospital stay and reduced resource use. However, when used to treat Mirizzi syndrome, the conversion rates are often high, with studies reporting conversion rates between 11.1% and 80%. [14]

Treatment varies according to the type of Mirizzi syndrome. For Type I, a cholecystectomy (either laparoscopic or open) is performed. Type II typically requires subtotal cholecystectomy, leaving a 5-mm remnant of the gallbladder wall for fistula closure. Type III cases involve subtotal cholecystectomy and a 1 cm flap of the gallbladder to repair the bile duct, with more extensive inflammation possibly necessitating additional procedures such as bilioenteric anastomosis to the duodenum. For Type IV, a bilioenteric anastomosis, ideally a hepaticojejunostomy Roux-en-Y, is required due to significant damage to the bile duct wall. [16] Endoscopic retrograde cholangiopancreatography (ERCP) is widely recognized as a therapeutic option for managing Type I Mirizzi syndrome. Numerous case reports and small series have documented the successful use of endoscopic techniques, including balloon catheter passage beyond the obstruction, stone extraction with a basket, and mechanical lithotripsy. [17] Biliary decompression through nasal bile drainage or the placement of a plastic stent can not only act as a bridge to surgery but may also lead to stone fragmentation, which facilitates further endoscopic procedures. Other methods, such as extracorporeal shock wave lithotripsy combined with endoscopic nasobiliary drainage (ENBD), have also been described in case reports with favorable outcomes. In our series, all patients underwent ERCP and sphincterotomy, with 80% successfully

undergoing stone extraction, yielding excellent outcomes.

CONCLUSION

ERCP appears to be the gold standard for diagnosing Mirizzi syndrome, as it accurately identifies the cause, location, and extent of biliary obstruction, as well as associated ductal abnormalities such as fistulas. In addition to its diagnostic role, ERCP offers multiple therapeutic options, including stone extraction and biliary stent placement. While percutaneous cholangiography can provide similar information, ERCP has the added advantage of identifying a low-lying cystic duct, which might be overlooked with percutaneous methods. For even more detailed visualization, wire-guided intraductal ultrasound (US) can offer high-resolution images of the biliary tract and surrounding structures. Surgical intervention remains the primary treatment for Mirizzi syndrome, with open surgery being the standard approach. Open surgery has shown favorable short and long-term outcomes with low morbidity and mortality rates. Laparoscopic surgery, however, is generally contraindicated in many cases due to the associated higher risk of complications. Endoscopic treatment may serve as an alternative for patients who are poor surgical candidates, such as the elderly or those with multiple comorbidities. It can also act as a temporary measure for biliary drainage before elective surgery. Our study reinforces the critical role of ERCP not only in the diagnosis but also in the treatment of Type I Mirizzi syndrome.

Conflicts of Interest: The author declares no conflicts of interest.

Funding: This research received no external funding.

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