

## Mirizzi syndrome: a challenging diagnosis. Case report

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**SUMMARY: Mirizzi syndrome: a challenging diagnosis. Case report.**

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*Mirizzi Syndrome (MS) is an uncommon complication of chronic gallstone disease defined as a common bile duct (CBD) obstruction secondary to gallstone impaction in the cystic duct or gallbladder neck. MS is still a challenging clinical situation: pre-*

*operative diagnosis of MS is complex and can be made in 18-62.5% of patients. Over 50% of patients with MS is diagnosed during surgery. In most of cases, laparotomy is the preferred surgical approach.*

*We report the case of a 70-year-old woman with a history of asthenia, jaundice, abdominal pain and preoperative imaging that suggest the presence of biliary stones with a choledocal stenosis. Intraoperatively, a MS with cholecysto-biliary fistula involving less than two-thirds of the circumference of the bile duct was diagnosed and successfully treated.*

KEY WORDS: Cholelithiasis - Chronic gallbladder disease - Mirizzi syndrome.

## Introduction

Mirizzi Syndrome (MS) is an uncommon complication of chronic gallstone disease. Kehr first described partial duct obstruction secondary to an impacted gallstone and its associated inflammatory process in 1905 (1), followed by Ruge in 1908 (2). However, the famous article that established this eponym for this condition was published in 1948 by Professor Pablo Luis Mirizzi (3). MS is defined as a common bile duct (CBD) obstruction secondary to gallstone impaction in the cystic duct or gallbladder neck. The pathophysiological process leading to the different MS stages has been explained as an inflammatory phenomenon secondary to a pressure ulcer

caused by an impacted gallstone at the level of the gallbladder infundibulum (4). The impacted gallstone together with the inflammatory response first causes an external obstruction of the main duct, eventually eroding into this latter. Such a condition should then evolve into cholecysto-choledochal or cholecysto-hepatic fistula, with different degrees of communication between gallbladder and bile duct (5, 6).

Anatomical predispositions include a side-by-side location of the cystic and CBD, coupled with a long, low inserting cystic duct to the biliary tree.

MS is detected in 0.06-5.7% of patients during cholecystectomy and in 1.07% of patients undergoing endoscopic retrograde cholangio-pancreatography (ERCP) (7).

## Case report

We report the case of a 70-year-old woman with a recent recovery in the Gastroenterology Department of our hospital for asthenia, jaundice, and ab-

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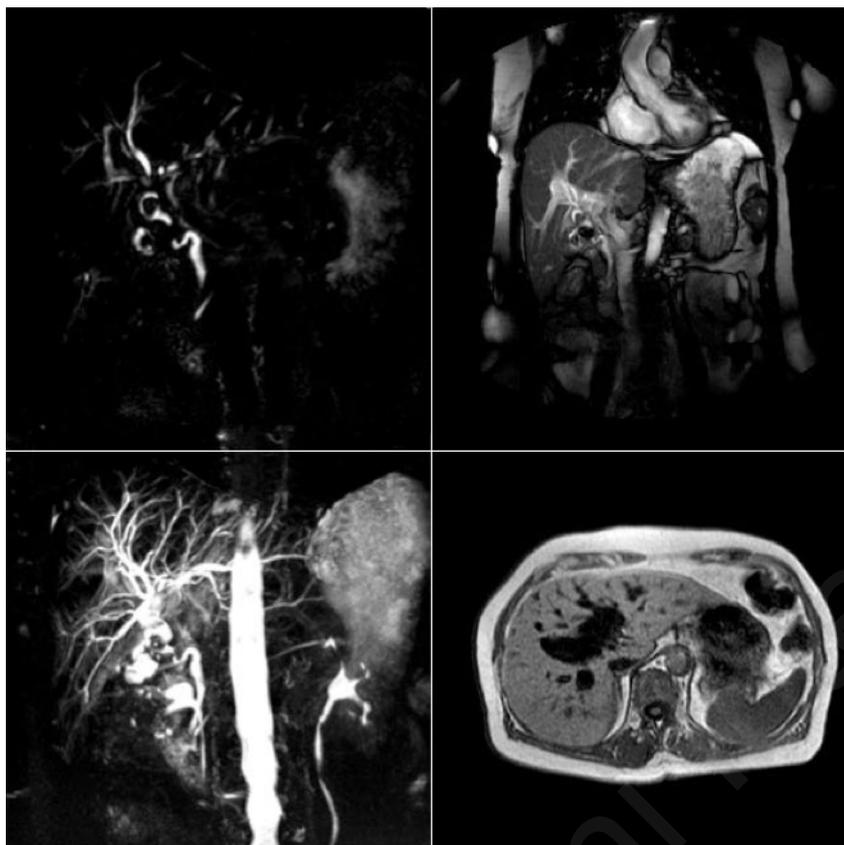


Figure 1 - Preoperative MRI showing severe intrahepatic biliary tree dilatation, impacted gallstones at the biliary confluence and cystic duct with concomitant choledochal stenosis.

dominal pain. Blood exams showed increased transaminases and gamma-GT. Abdominal ultrasound demonstrated cholecysto-choledochal lithiasis. The patient performed a Cholangio-MRI that evidenced severe intrahepatic biliary tract dilatation, impacted gallstones at the biliary confluence and choledochal stenosis (Figure 1). ERCP confirmed the MRI report, but it was unable to remove the biliary stones (Figure 2). A brushing in the stenotic tract was performed, resulting negative for malignancy. A trans-papillary biliary stent was placed. The patient was admitted to our surgical division and scheduled for a complete removal of the biliary stones and, possibly, to a bilio-enteric diversion.

A rooftop (Chevron) laparotomy was performed, with a contextual lysis of hepato-duodenal adhesions. After an antegrade gallbladder mobilization, we noticed a complete fusion between the gallbladder and CBD wall. After the cholecystotomy, a big impacted stone was identified and removed. A sizeable cholecysto-choledochal fistula was then evidenced, consenting to diagnose a MS (Figures 3, 4).



Figure 2 - Preoperative ERCP confirming multiple cholecysto-choledochal lithiasis.

Frozen section of the gallbladder wall was achieved with no evidence of malignancy. The biliary stent was left in place, the CBD was explored and a second big stone was removed from the hepatic duct confluence. A third stone was then removed from the left hepatic duct. Intraoperative cholangiography with a 12 Fr Foley's Catheter through the fistula demonstrated no residual biliary lithiasis (Figure 5). Cholecystostomy was sutured with absorbable 3/0 detached stitches.

Postoperative course was uneventful leading to a progressive normalization of biohumoral exams. Subhepatic drainage was left in place until 4<sup>th</sup> POD with no evidence of biliary leakage. Patient was discharged on 8<sup>th</sup> day after surgery.

## Discussion

Beltrán (4) observed some anatomical elements associated with MS: an atrophic gallbladder with thick or thin walls and impacted gallstones at the infundibulum or the Hartmann's pouch; an obliterated

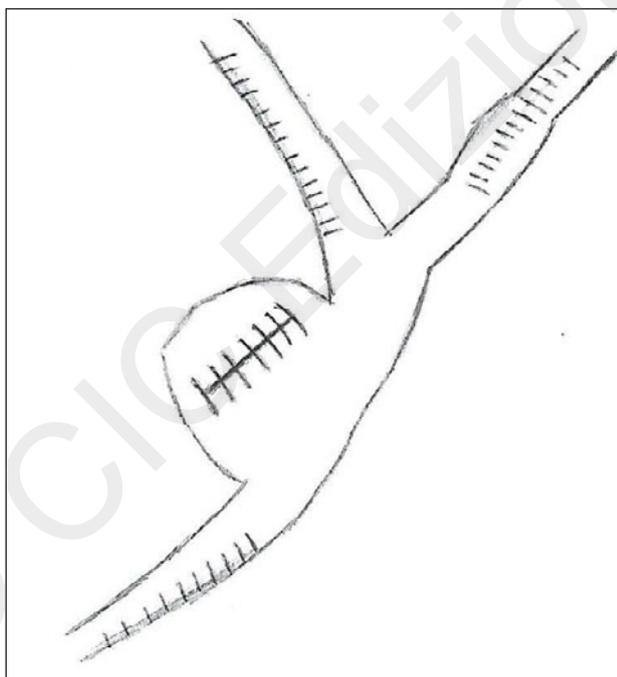


Figure 3 - Preoperative anatomical condition: complete fusion between gallbladder and CBD walls with a cholecysto-biliary fistula involving less than two-thirds of the circumference of the bile duct with a big impacted gallstone into it. Gallstones are also reported at the biliary confluence and in the left hepatic bile duct.

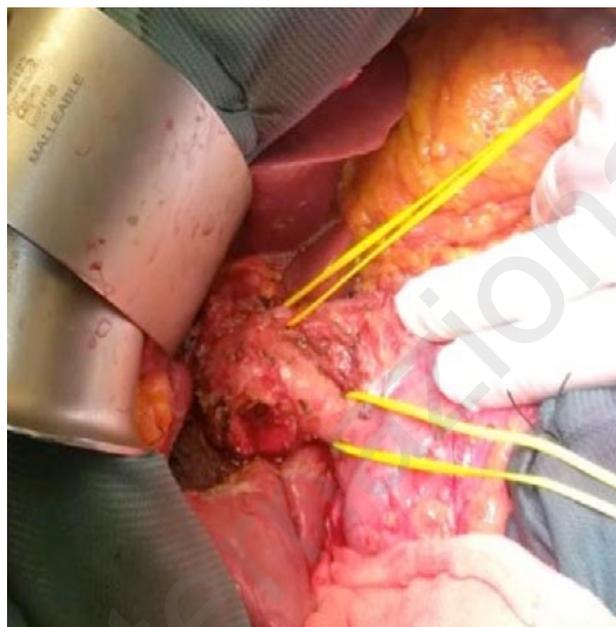


Figure 4 - Intraoperative picture. Partial cholecystectomy with evidence of the cholecystobiliary fistula after cholecystostomy. Vessel loops are passed behind the hepatic artery and the common bile duct.



Figure 5 - Intraoperative cholangiography through the fistula showing no residual biliary stones.

ed cystic duct; a long cystic duct running parallel to the CBD with low insertion; a partial obstruction of the bile duct by external compression or erosion caused by a gallstone; a normal-caliber distal bile duct with walls of average thickness; a dilated proximal bile duct with thick inflamed walls; an abnormal communication between the gallbladder and the

bile duct; an anomalous communication between the gallbladder and the stomach, duodenum, colon, or other abdominal viscera (8-14). Post-cholecystectomy residual cystic duct stones have also been implicated in MS.

McSherry et al. in 1982 (15), classified the MS into two types based on ERCP findings: 1) Type I is characterized by external compression of the CBD by large stones placed in the cystic duct or the Hartmann's pouch; 2) type II consists of a proper cholecysto-biliary fistula, caused by gallstones eroding into the CBD.

Afterwards, Csendes et al. (5) modified the McSherry classification dividing MS into four types. Csendes MS type I corresponds to McSherry MS type I, with external compression of the CBD by an impacted gallstone in the infundibulum or cystic duct. MS type II consists of a cholecysto-biliary fistula resulting from erosion of the bile duct wall

by a gallstone; the fistula must involve less than one-third of the circumference of the bile duct. In type III, it involves less than two-thirds of the circumference of the bile duct. In type IV, the cholecysto-biliary fistula completely destroys the bile duct wall, with the gallbladder fused to the bile duct forming a single structure with no recognizable dissection planes between both biliary tree structures (5). In 2008, Beltrán et al. (6) classified the MS type V, which includes the presence of a cholecysto-enteric fistula together with any other type of MS with (type b) or without (type a) gallstone ileus (Figure 6).

Preoperative diagnosis of MS is complex and can be made in 18-62.5% of patients: over 50% of patients with MS is diagnosed during surgery (16).

In our case, a MS type III was evidenced after laparotomy. Cholangio-MRI and ERCP were unable to clarify the anatomy of the gallbladder and CBD

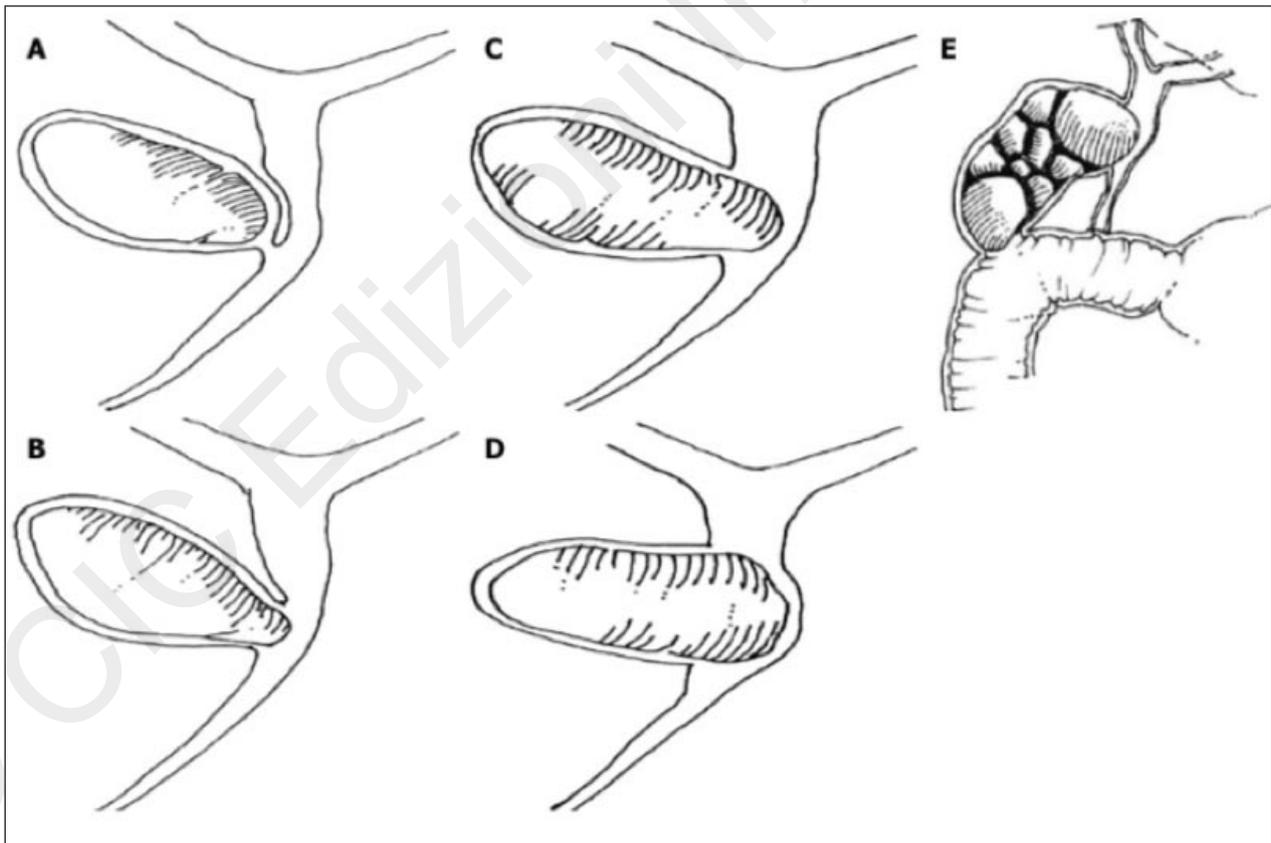


Figure 6 - A) Mirizzi type I is the extrinsic compression of the common bile duct by an impacted gallstone; B) Mirizzi type II consists of a cholecystobiliary fistula involving one third of the circumference of the bile duct; C) In Mirizzi type III, the cholecystobiliary fistula compromises up to two-thirds of the circumference of the bile duct; D) In Mirizzi type IV, the cholecystobiliary fistula has destroyed the bile duct wall, and comprises the whole circumference of the bile duct; E) Mirizzi type V corresponds to any type of Mirizzi associated with a bilioenteric fistula with or without gallstone ileus (original drawings from World Journal of Surgery 2008 (6).

lithiasis. As a consequence, a correct knowledge of the treatment of a MS is necessary for the surgeon, since preoperative diagnosis is not always possible. Moreover, the surgical treatment of MS should be associated with a significantly increased risk of bile duct injury due to the severe inflammatory process associated with the edematous tissues that distort the anatomy.

Surgical treatment of MS is not standardized and must be individualized based on the stage and the anatomy of each case. However, a few Authors have drawn some guidelines based on their experiences. Standard cholecystectomy can resolve type I MS with laparoscopic or open approach (4-14, 16-22) but severe inflammation in the area of Calot's triangle could make hard to perform the dissection of the cystic duct and cystic artery (23-25). A laparoscopic approach could be reserved to few selected cases of MS type I. In type II and III, the initial approach should involve a partial cholecystectomy with gallstones extraction leaving a remnant of gallbladder around the cholecysto-biliary fistula to close it, as in our case. The exploration of the bile duct should be performed from another incision and always protected by a Kehr tube. In some cases, when a severe grade of inflammation is present, such as in the presence of a MS type IV, partial cholecystectomy is not enough, with a bilio-enteric anastomosis often required to avoid biliary stric-

tures. MS type V needs treatment of either the bilio-enteric fistula (division and suture with the absorbable material) and the cholecysto-biliary fistula depending on the stage. When gallstone ileus occurred, its treatment is needed, and the cholecysto-biliary fistula repair can be secondarily performed few weeks after the index operation.

## Conclusions

MS is often an intraoperative diagnosis. Radiological and endoscopic preoperative exams may not recognize the real anatomical features of the extrahepatic biliary tract: in our case ERCP and Cholangio-MRI where not able to describe the relationship between gallbladder and CBD. Surgery was performed to remove biliary stones and permitted type III MS diagnosis clarifying the extrahepatic biliary anatomy. In our opinion, MS is a dangerous condition because can easily lead to iatrogenic lesions of biliary tract due to an impaired anatomy and severe chronic tissutal inflammation.

In conclusion, we suggest to consider Mirizzi Syndrome in all cases of cholecysto-choledochal lithiasis with unclear preoperative imaging. When the intraoperative report confirms MS, a laparotomic approach can be safer and more feasible specially if performed by Hepato-Biliary skilled surgeons.

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