ECRONICON OPEN ACCESS

EC GASTROENTEROLOGY AND DIGESTIVE SYSTEM

Case Report

Mirizzi Syndrome. Case Report

Fernanda Gutiérrez[1]*, Florencia Molfeso[1], Tomás Flores[2], Guillermo Di Palma[1], Marcelo D´Alessandro[1], Luis Sarotto[2], Alexander Napolitano[2] and Juan Sordá[1]

[1]Gastroenterology Division, Hospital of Clinics José de San Martín, Autonomous City of Buenos Aires, Argentina

[2]Surgery Division, Hospital of Clinics José de San Martín, Autonomous City of Buenos Aires, Argentina

***Corresponding Author:** Fernanda Gutiérrez, Gastroenterology Division, Hospital of Clinics José de San Martín, Autonomous City of Buenos Aires, Argentina.

Received: November 10, 2020; **Published:** December 10, 2020

Abstract

Mirizzi Syndrome is an uncommon complication secondary to chronic gallbladder stones and is defined by external compression of the common bile duct, with its consequent obstruction. It is a pathology that, by itself, increases the risk of biliary injury during cholecystectomy, so preoperative identification is key for careful planning of the surgical procedure and reducing of the associated morbidity and mortality. We present the case of a 74-year-old patient who presented jaundice and abdominal pain. Endoscopic Retrograde Cholangiopancreatography (ERCP) was performed showing an immobile 15 mm lithiasic image with a dilated bile duct that obstructed the common hepatic duct leading to the diagnosis of type II Mirizzi Syndrome.

Keywords: *Biliary Tract Diseases; Bile Duct Diseases; Cholestasis; Cholelithiasis; Chronic Gallbladder Disease; Gallstone Disease*

Introduction

Mirizzi Syndrome was described by the Argentinean Cordoba surgeon Pablo Mirizzi. It is defined as a rare entity and difficult to diagnose, causing cholestasis due to extrinsic compression of the common bile duct secondary to the impaction of gallstones in the Hartmann's bag. This chronic impaction leads to inflammation and ulceration with formation of cholecystobiliary and/or cholecystoenteric fistulas. This pathology, by itself, increases the risk of biliary injury associated with cholecystectomy, being a challenge, even for expert surgeons.

Clinical Case

We present a 74-year-old male patient, with no history of hierarchy, who was admitted to the ward of our hospital due to a 2-week history characterized by jaundice and colicky abdominal pain in the right upper quadrant. In the laboratory analysis, it is observed: Leukocytosis (13,400/mm^3), total bilirubin 9.4 mg/dl (directly predominant); aspartate aminotransferase 82 IU/l; alanine aminotransferase 58 IU/l; alkaline phosphatase 227 IU/l. An abdominal ultrasound was performed, which reported a 3.2-mm intrahepatic bile duct with a 9-mm common bile duct and a multilithiasic collapsed gallbladder. Abdominal tomography shows dilatation of the intrahepatic bile duct and both hepatic ducts with significant dilation of the common bile duct immediately below the Carrefour secondary to a voluminous 12-mm calcium-density lithiasic image (See figure 1).

It was decided to perform ERCP: where, after selectively cannulating and opacifying the bile duct, a 15 mm lithiasic image was observed at the level of the upper third of the common bile duct, eccentric with regard to the axis of the bile duct, which was not mobilized with the instrumental maneuvers. The adjacent and intrahepatic bile ducts show significant dilation (See figure 2). Papillotomy is per-

formed, a 7 French plastic stent is placed, crossing the stone for drainage, which reveals a Mirizzi Syndrome with the obstructing stone in the cystic duct, which causes compression of the common hepatic duct.

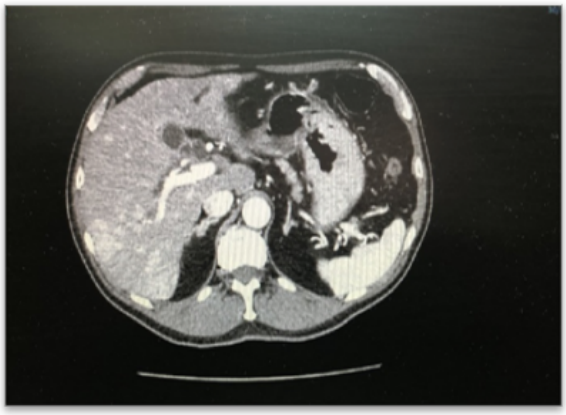

Figure 1: *TAC shows via figure 1: CT scan showing dilated bile duct and choledochal lithiasis.*

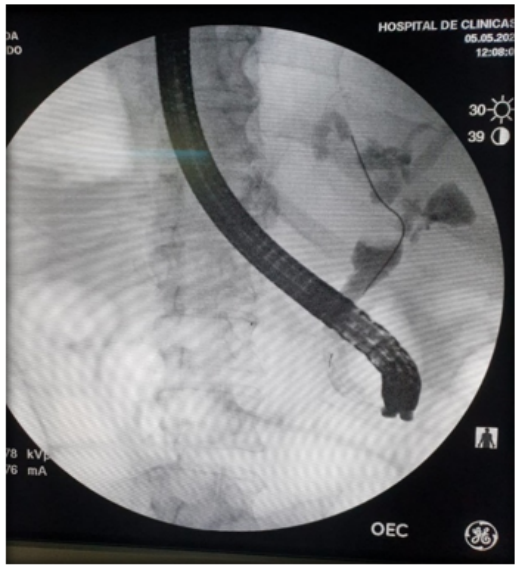

Figure 2: *Fluoroscopic image of ERCP showing a large, fixed stone in the upper third of the common bile duct.*

The patient evolves stable with improvement of his clinical and laboratory parameters with surgical treatment 72 hours later. In laparoscopic (converted) cholecystectomy, firm cholecystocolonic adhesions without fistulization are identified to the colon (See figure 3). The gallbladder is scleroatrophic with an intense inflammatory process. Tutored by the biliary stent placed in the ERCP, a cholecystocholedocian fistula is identified that covers one third of the circumference (See figure 4 and 5). The known stone is removed and the common bile duct is closed primarily. Intraoperative diagnosis: Mirizzi syndrome type II. Currently, two weeks after surgery, the patient is progressing favorably.

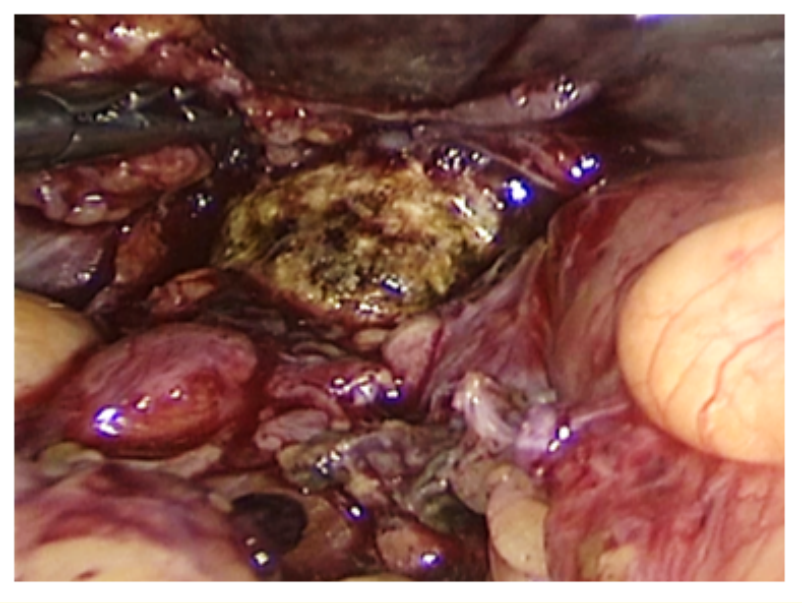

Figure 3: *After the release of adhesions, a calculus located in the vesicular fundus is observed in intimate contact with the colonic wall (arrow).*

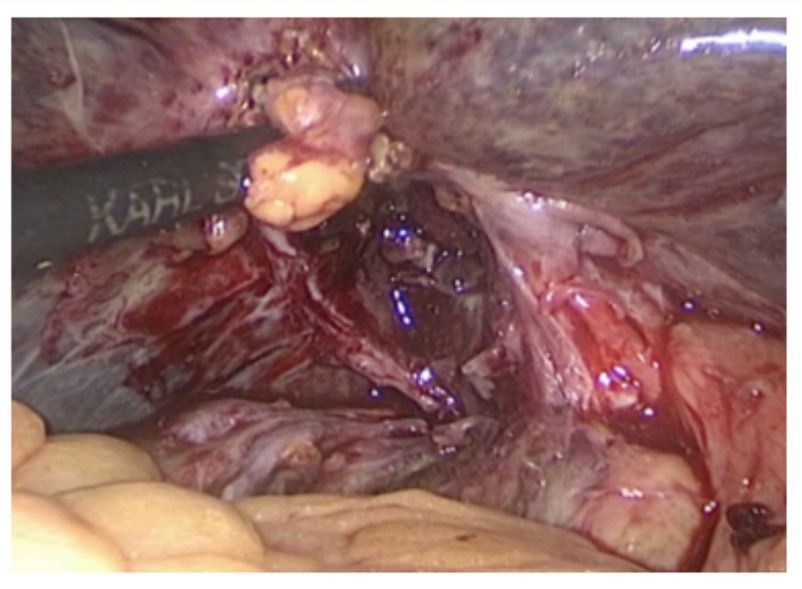

Figure 4: *The calculus is removed from the gallbladder fundus and a scleroatrophic gallbladder [1] is identified, generating a fistula with the bile duct [2].*

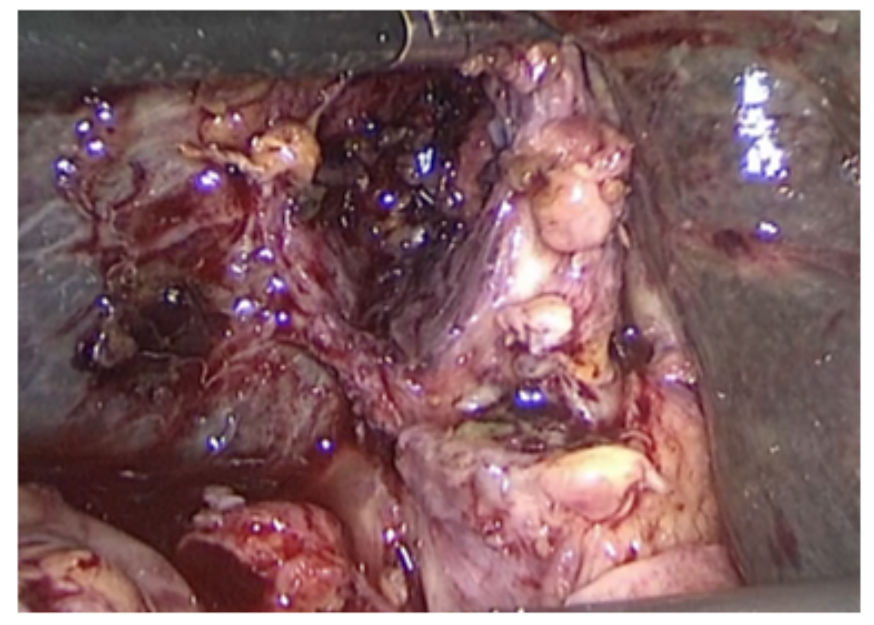

Figure 5: *Dissection of the gallbladder [1] and identification of cholecystocholedocian fistula [2].*

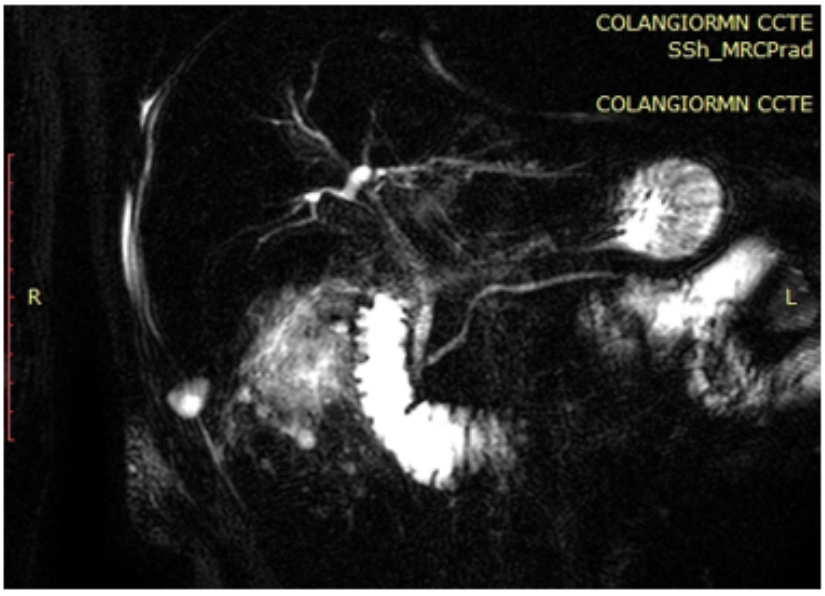

Figure 6: *Control with cholangioresonance at two weeks postoperative. Expedited bile duct without residual lithiasis.*

Discussion

Mirizzi syndrome, also known as extrinsic biliary compression syndrome, is a rare complication of symptomatic cholecystitis and chronic cholelithiasis secondary to obliteration of the gallbladder infundibulum (Hartmann's bag) or the cystic duct, caused by the impact of one or more calculations. This stone impaction generates compression of the adjacent bile duct, resulting in partial or complete obstruction of the common hepatic duct with the consequent cholestasis. It is accompanied by chronic cholecystitis and cholecystocholedocal fistula (Mirizzi syndrome type II, III, IV) and/or cholecystoenteric (Mirizzi syndrome type V) [1].

It has an incidence of less than 1% per year in developed Western countries and 4.7% to 5.7% in developing countries. It prevails between the fourth and seventh decade of life [1], with a higher prevalence in women (3: 1 ratio) [2].

It is detected in 0.06% to 5.7% of patients during cholecystectomy and in 1.07% of patients undergoing ERCP [3].

Several types were described:

- Type I: External compression of the bile duct by a gallstone impacted in the infundibulum of the gallbladder or the cystic duct (10.5% - 78%).
- Type II: Consists of a cholecystobiliary fistula resulted from the erosion of the bile duct wall by a stone. The fistula must involve less than one third of the circumference of the bile duct (15% - 41%).

  (Figure 7)
- Type III: Consists of a cholecystobiliary fistula that involves up to two thirds of the circumference of the bile duct (3% - 44%).
- Type IV: Cholecystobiliary fistula with complete destruction of the bile duct wall with the gallbladder completely fused with the bile duct forming a single structure without recognizable dissection planes between both structures of the biliary trees (1% - 4%).
- Type V: Includes the presence of a cholecystoenteric fistula together with any other type of Mirizzi, it is further subdivided into type Va, it includes a cholecystoenteric fistula without gallstone ileus and type Vb refers to a cholecystoenteric fistula complicated by gallstone ileus (29%) [2].

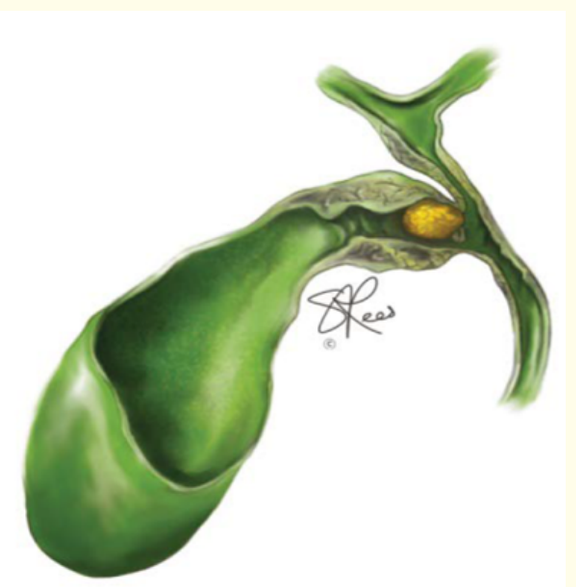

Figure 7: *Mirizzi syndrome type II.*

It is common for patients to report a long clinical course, presenting recurrent pain in the right upper quadrant (60% -100%) and jaundice (50% -100%) at the time of diagnosis. Fever (20%) has also been reported in patients with acute cholecystitis, acute cholangitis, or acute pancreatitis [4].

The most common laboratory finding is directly predominant hyperbilirubinemia. Other abnormalities include elevated levels of aminotransaminases, alkaline phosphatase, and leukocytosis. Recently, extremely high levels of the malignancy marker CA19-9 have been found in patients with Mirizzi syndrome type II, which should be interpreted with caution [1,2].

Preoperative diagnosis remains difficult despite advances in imaging, and in half of cases it is performed during surgery.

The differential diagnosis should include benign and malignant causes of obstructive jaundice such as: gallbladder carcinoma, cholangiocarcinoma, pancreatic cancer, metastatic lymph node disease with compression of the bile duct, and sclerosing cholangitis [1,2,5].

Depending on the situation, and with timely pre-surgical diagnosis, it may be necessary, in patients with Mirizzi Syndrome type I, the placement of one or more plastic stents for ERCP, followed by an almost total cholecystectomy (since a small portion remains adhered to the main bile duct that works as a patch to resolve the defect). It is advisable to remove the stent before 3 months after surgery [5]. The stent also facilitates the identification of the main bile duct during surgery, as in the case of our patient [1,3].

In Mirizzi syndrome type I, treatment with ERCP can be considered definitive; however, the impacted stone can create real difficulties. Frequently, ERCP is used to make the diagnosis and insert a stent to relieve jaundice and allow planning of an elective operation. If used as a definitive treatment, sophisticated techniques may be necessary for these cases, such as electrohydraulic or laser lithotripsy. Any of these sophisticated ERCP techniques would have required an endoscopic sphincterotomy [6].

This pathology constitutes a surgical challenge due to the high rate of postoperative biliary lesions, which can reach 17% [2]. It is for this reason that laparotomy has been considered the technique of choice. The laparoscopic technique is associated with high conversion rates (31 - 100%) and more complications than the conventional technique. Approximately 8 - 25% of patients require biliary tract repair due to fistulization of the main bile duct [1,3].

When making the intraoperative diagnosis of Mirizzi Syndrome, cholangiography should be performed through a Kehr tube after removing the gallstones from the decompressed gallbladder.

Depending on the type of Mirizzi we will have to take into account some questions. Type I could be resolved by classical cholecystectomy. Type II with a subtotal cholecystectomy that begins with dissection of the fundus of the gallbladder into Hartmann's pouch. Type III can be treated by subtotal cholecystectomy leaving a gallbladder wall flap to repair the bile duct, however, some cases will require a bilioenteric anastomosis to the duodenum or a Roux-en-Y hepaticojejunostomy anastomosis. In Type IV, due to extensive destruction of the bile duct wall, treatment consists of a bilioenteric anastomosis, preferring a Roux-en-Y hepaticojejunostomy. Type V differs depending on the subtype, with chronic active or inactive bilioenteric fistulas, but the safest approach is always laparotomy [1-3,5].

Conclusion

In conclusion, Mirizzi syndrome continues to be a fascinating subject of study, due to the challenge it represents and the unexpectedness of its presentation. Despite the fact that more than 60 years have passed since the description of Dr. Pablo Luis Mirizzi, it seems that the last word on this issue has not yet been pronounced. The approach to suspect Mirizzi syndrome should be prudent and robust. Every effort should be made to establish a correct preoperative diagnosis, and if this is not possible, an attempt should be made, through precise and cautious surgery, to try to identify the type of Mirizzi and perform the most appropriate treatment for each case in particular.

The case of our patient is a type II Mirizzi Syndrome, which is the most frequent. It was studied through images (CT and MRCP) without being able to reach an accurate diagnosis until ERCP was performed and a stent placed for decompression of the bile duct with subsequent cholecystectomy and repair of the biliary tract, the patient presented a good evolution continuing with outpatient control.

Conflict of Interest

The authors declare no conflict of interest.

Bibliography

1. Valderrama-Treviño AI., *et al.* "Updates in Mirizzi syndrome". *Hepatobiliary Surgery and Nutrition* 6.3 (2017): 170-178.
2. Beltrán MA. "Mirizzi syndrome: History, current knowledge and proposal of a simplified classification". *World Journal of Gastroenterology* 18.34 (2012): 4639-4650.
3. Chen Hang., *et al.* "Current trends in the management of Mirizzi Syndrome: A review of literature". *Medicine* 97.4 (2018): e9691.
4. Uppara M and Rasheed A. "Systematic Review of Mirizzi's Syndrome's Management". *JOP. Journal of the Pancreas* 18.1 (2020).
5. Clemente Gennaro., *et al.* "Mirizzi Syndrome: Diagnosis and Management of a Challenging Biliary Disease". *Canadian Journal of Gastroenterology and Hepatology* (2018).
6. Kelly Michael D. "Acute Mirizzi Syndrome". *Journal of the Society of Laparoendoscopic Surgeons* 13.1 (2009): 104-109.
7. Kim Hong Jun., *et al.* "The Clinical Usefulness of Simultaneous Placement of Double Endoscopic Nasobiliary Biliary Drainage". *Clinical Endoscopy* 48.6 (2015): 542-548.

Volume 8 Issue 1 January 2021