Research Article
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Mirizzi syndrome: case series and literature review

Abstract

Introduction: Mirizzi syndrome (MS) is defined as extrinsic obstruction of the main bile duct by a stone impacted in the cystic duct or in the infundibulum of the gallbladder.

Objective: To determine the incidence and describe the management of 15 cases diagnosed in the General Hospital of Zacatecas "Luz González Cosió".

Material and methods: Retrospective, observational, cross-sectional and comparative study, age, sex, diagnostic method, surgical management are described.

Results: 719 cholecystectomies were performed in a period of 3 years, 15 cases of MS were found, with an incidence of 2.07%, the diagnosis was made intraoperatively in 11 cases, 10 cases (66%) were type I, 3 cases (19.8%) type V, 1 case (6.6%) type III and 1 case (6.6%) type IV, for type I 10 cholecystectomies were performed, the other types had a specific surgical management.

Conclusion: It is a complex pathology of difficult pre-surgical diagnosis. It must be treated by experienced surgeons to reduce the risk of the dreaded bile duct injury.

Keywords: Mirizzi syndrome; Cholelithiasis; Cholecystitis; Surgical management

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Abbreviations: MS, Mirizzi syndrome; MRCP, magnetic resonance cholangiopancreatography; ERCP, endoscopic retrograde cholangiopancreatography; US, ultrasound.

Introduction

In 1948, Pablo Luis Mirizzi, an Argentine surgeon, described the "hepatic duct syndrome". Mirizzi suggested that the obstruction of the common hepatic duct was functional and the mechanical obstruction of the gallbladder with the consequent inflammatory process predisposed the contraction of a "muscular sphincter" located in the common hepatic duct. It is now well known that there is no sphincter in hepatic duct.[1]

However, a partial obstruction was described for the first time in 1905 by Kehr[2] and in 1908 by Ruge,[3] while in 1982 McSherry et al classified MS into two types, based on the findings of endoscopic retrograde cholangiography, the type Type 1 involves partial or complete obstruction of the common hepatic duct by an impacted stone in the cystic or Hartmann's pouch, resulting from inflammation of Calot's triangle, while type 2 referred to the formation of a fistula between the cystic duct and common hepatic duct.[4]

Subsequently, in 1989, Csendes et al.[5] classified MS into four types, which categorized cholecystocholedochocal fistulas according to the degree of destruction, which in 2007 was validated by Beltrán et al., who added type five to the classification, which involves a cholecystenteric fistula and subdivides it according to whether the latter is causing ileus.[6] The objective is to communicate the experience in the treatment of MS in the General Hospital of Zacatecas, compare the results with those published in the literature and discuss the possible handling options.

Material and methods

We carried out a retrospective, observational, cross-sectional and comparative study on patients admitted to the General Hospital of Zacatecas with a diagnosis of MS over a period of 3 years (January 2018 to January 2020). The data was analyzed using descriptive statistical tools. Age, sex, diagnostic method, surgical management are described. Analytical laboratory data, hepatobiliary-pancreatic ultrasound were studied, in selected cases magnetic resonance cholangiopancreatography (MRCP), endoscopic cholangiopancreatography or intraoperative cholangiography were performed. To evaluate the results, measures of central tendency and dispersion were applied by descriptive statistics using the SPSS 25 program.

Ethical considerations. The research was carried out in accordance with the principles of the Declaration of Helsinki and the protocol was approved by the Bioethics and Research Committee of the General Hospital of Zacatecas, informed consent was not requested for the publication of this article because in this article no personal data that allows the patient to be identified is published.

Results

In the period studied, 719 cholecystectomies were performed, 472 (65.3%) uncomplicated cholelithiasis were found, while 158 (21.8%) were acute cholecystitis, 45 cases (6.2%) of pyocholecyst, 29 (4.01%) hydrocholecyst, of the total 15 cases (2.07%) of MS were found, 11 cases (73%) were in the female gender and 4 cases (27%) in the male gender, with a mean age of 48 years, in terms of symptoms, the most common it was pain in the right hypochondrium in 93% of the patients, jaundice in 53%, nausea and vomiting in 46%, in laboratory studies, hyperbilirubinemia was found in 50% of the patients, all at the expense of direct bilirubin. The diagnosis of MS was intraoperative in 11 of the 15 patients (73%), in 3 cases it was performed by endoscopic retrograde cholangiopancreatography (ERCP), in which a stent was placed, and only 1 case by ultrasound (US), the 4 cases were complemented with MRCP with which diagnostic confirmation was possible.

The therapeutic approach of 93% of the patients was open, while only 1 patient was approached by laparoscopy, in all patients the intervention was scheduled, according to the current classification of MS (Figure 1), 10 cases (66%) they were type I, 3 cases (19.8%) type V, 1 case (6.6%) type III and 1 case (6.6%) type IV.

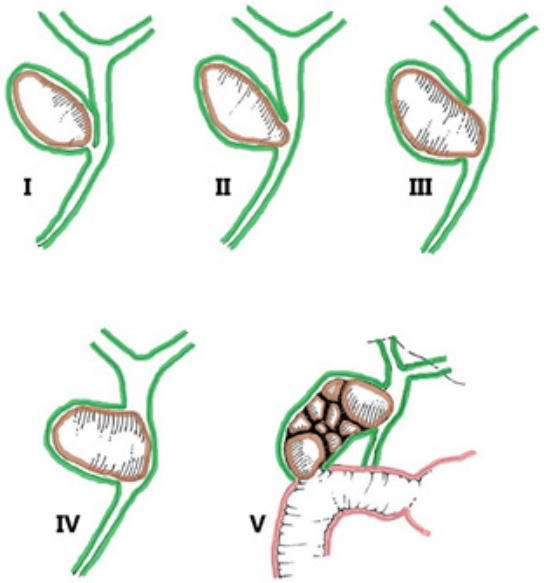

Figure 1 Representation of the Csendes[6] classification of Mirizzi Syndrome.

Regarding the management given for each type, in type I MS, 80% of the cases (8 patients) were treated with cholecystectomy alone, 20% of the cases (2 patients) were treated with cholecystectomy, cholangiography and exploration of bile ducts, for MS type III, which in our sample was only 1 patient, the procedure that was performed was cholecystectomy, cholangiography and exploration of the bile ducts, for the case of MS type IV subtotal cholecystectomy, cholangiography and exploration of the bile ducts were performed bile ducts, for the 3 cases of MS type V (with fistula to the transverse colon), subtotal cholecystectomy was performed in 2 of them, cholecystectomy, cholangiography and exploration of the bile ducts in 1 case, in the 3 cases extraction of the embedded stones and primary closure of the transverse colon in 2 planes (Table 1).

Discussion

MS is an extrinsic compression of the bile duct, it is a rare complication of chronic cholecystitis, accompanied by obliteration of the infundibulum (Hartmann's pouch) or the cystic duct, resulting in partial or total obstruction, accompanied by inflammation of the gallbladder and in some cases a cholecystocholedochal fistula.[7] With an annual incidence of less than 1% in developed Western countries and 4.7 to 5.7% in developing countries.[8] The incidence reported in Mexico is 4.7% without existing established difference between men and women, prevails between the fourth and seventh decade of life.[9] In our setting, the incidence found was 2.07%. Women are especially prone to developing gallstones, since a 4.5:1 female/male ratio has been shown, probably secondary to estrogenic effects on the enterohepatic circulation, thus causing biliary obstruction secondary to cholecystitis or choledocholithiasis.[10] In our series we found 11 cases (73%) in women.

Table 1 Demographic characteristics of the population

Case	Tipe MS	Diagnosis	Age (Years)	Sex	LEUC	TB	DB	IB	AP	Procedure
1	IV	Intraoperative	54	F	10.7	10.4	9.31	1.12	486	STCL + Intraoperative Cholangiography + BTE
2	V	Intraoperative	45	F	15.5	0.27	0.15	0.12	82	CLT + Intraoperative Cholangiography + Colon closure
3	I	Intraoperative	37	F	11.2	0.60	0.25	0.35	74	CLT
4	I	Intraoperative	38	F	18.5	0.72	0.52	0.20	89	CLT
5	I	US	72	M	7.4	1.52	1.31	0.31	158	CLT
6	I	ERCP	18	F	17.7	3.21	2.86	0.20	258	CLT
7	I	Intraoperative	36	M	9.6	0.71	0.26	0.45	80	CLT
8	V	Intraoperative	92	M	7.7	3.35	2.84	0.51	200	STCL + Intraoperative Cholangiography + Colon closure
9	III	Intraoperative	67	M	6.9	6.90	6.20	0.60	267	CLT + Intraoperative Cholangiography + BTE
10	I	Intraoperative	54	F	14.6	7.57	7.04	0.53	211	CLT + Intraoperative Cholangiography + BTE
11	I	ERCP	45	F	12.0	4.50	3.80	0.70	201	CLT + Intraoperative Cholangiography + BTE
12	I	Intraoperative	43	F	11.4	0.31	0.21	0.10	82	CLT
13	V	Intraoperative	28	F	5.5	1.46	1.30	0.16	79	STCL + Intraoperative Cholangiography + Colon closure
14	I	Intraoperative	40	F	8.6	0.52	0.24	0.28	88	CLT
15	I	ERCP	51	F	10.2	3.46	1.98	1.43	78	CLT

Epidemiology, diagnosis, laboratory, surgical variables. SM, Mirizzi syndrome; US, Ultrasound; ERCP, endoscopic retrograde cholangiopancreatography; F, Female; M, Male; LEUC, Total white blood cell count in cells/mm3; TB, Total bilirubin in mg/dl; DB, Direct bilirubin in mg/dl; IB: indirect bilirubin in mg/dl; AP, alkaline phosphatase expressed in IU/l; STCL, subtotal cholecystectomy; BTE, biliary tract exploration; CLT, cholecystectomy.

The pathophysiology of the disease is based on the obstruction of the infundibulum or cystic duct of the gallbladder, causing compression of the adjacent duct. In addition, the inflammatory process secondary to erosion favors the formation of adhesions, by eventually fusing its walls with inflammatory tissue. it will become fibrotic to neighboring structures, most often the common bile duct, duodenum, and colon,[10,11] adherence to the bile duct contributes to external compression of the bile duct, leading to obstructive jaundice and ultimately liver dysfunction.[12] We found jaundice in 53% and hyperbilirubinemia in 50% of the patients studied. It is accompanied by pain in the right hypochondrium or in the epigastrium (50 to 100%), fever, nausea, vomiting (60%) and can also occur in the context of acute cholecystitis, pancreatitis and choledocholithiasis.[13] Nausea and vomiting occurred on 46 % of patients, no pancreatitis data found. The mucosa eventually erodes, progressing towards a cholecystocholedochal or cholecystoenteric fistula that presents different degrees of communication between the adjacent structures.[14] These adhesions, distortion of the anatomy and the presence of the fistula increase the risk of injury to the bile duct during cholecystectomy.[15]

There are some anatomical predispositions that include the location of the cystic and close relationship with the hepatic ducts, or a long cystic inserted low in the biliary tree.[14,15]

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The diagnosis of MS before surgery occurs in 18 to 62% of cases, it is crucial for the patient's prognosis, it can avoid exposing the surgeon to the risk of facing unexpected conditions or difficult surgery, and allows for a more adequate therapeutic planning.[15,16] In only 4 patients (27%) was the preoperative diagnosis made, in the rest 11 patients (73%) it was intraoperative. It has been observed that diagnostic sensitivity can increase to 85% when using the combination of 2 or more diagnostic modalities, however, this practice is not supported by solid evidence and there is currently no consensus among experts as to the additional benefit of this technique. In practice, an early and accurate diagnosis has a great impact on management, morbidity and mortality, as well as the prevention of future complications, reducing them to 54%.[17-18] US of the liver and bile ducts is a technique with low diagnostic accuracy, it can reveal atrophic gallstones and an ecstatic common hepatic duct, it has a diagnostic accuracy of between 8.3-29%.[18,19] Only 1 case was diagnosed by US in our series, which corresponds to the low diagnostic precision described in the literature.

Computed axial tomography is effective in detecting the cause and location of biliary obstruction, although it has a sensitivity of 22.5%.[19] It was not used in our series due to its low diagnostic sensitivity. One of the most favorable diagnostic tools is ERCP, it has a sensitivity of 31 to 100%, it can provide information on the obstruction of the bile duct and the location of the fistula, it allows therapeutic decompression through sphincterotomy or stent placement.[20] In 3 cases preoperatively was performed in our series.

MRCP can delineate the typical characteristics of MS, such as a stone in the common hepatic duct or its extrinsic compression, it has a sensitivity of 62.5%, according to several reviews it is considered to be performed routinely before surgery when the patient suspects SM.[18] In 4 cases, Type IV and V allowed diagnostic confirmation, which prevented feared surgical complications such as bile duct injury.

A variety of signs associated with MS are revealed during surgery, such as an edematous or atrophic gallbladder with distortion of Calot's triangle, an impacted gallstone in the infundibulum or neck of the cystic duct, and hepatic adhesions, if removal of an impacted stone is followed by a bile leak from the bile duct, a cholecystobiliary fistula should be strongly suspected. Subsequently, an additional intraoperative cholangiography can be performed to determine the position and dimension of the fistula and to verify the integrity of the wall.[20] How it was performed intraoperatively in cases where there were anatomical alterations that made us suspect MS in our series. The differential diagnosis must include other causes of obstructive jaundice such as gallbladder carcinoma and cholangiocarcinoma, in addition to the fact that it is considered a risk factor for these oncological pathologies, this occurs mainly in MS type II onwards, the underlying risk factor seems to be a permanent inflammation due to gallstones and stasis.[19,20] In this study, no patient was suspected of gallbladder carcinoma.

Cholecystectomy is considered the treatment of choice in cases with moderate inflammation, when severe inflammation prevents safe dissection of Calot's triangle, retrograde cholecystectomy or subtotal cholecystectomy can be applied, an incision can be made at the bottom of the gallbladder and removal of the impacted stone, when the intraoperative diagnosis is made, cholangiography should be performed through a Kher tube after removing any stone from the decompressed gallbladder, while in cases where there is a fistula, closure of the itself is the main problem, Roux-Y hepaticojejunostomy is recommended instead of anastomosis of the bile ducts to prevent early biliary stricture, in some cases it will be necessary to insert a Kher tube into the bile duct, to decompress and to shape the duct with in order to minimize the risk of leaks, although drainage and stenting may be applied preoperative by ERCP to avoid the insertion of a Kher tube, for type IV, there is consensus that the best surgical technique is cholecystectomy and Roux-Y hepaticojejunostomy.[20] In 12 cases cholecystectomy was resolutive and 3 cases cholecystectomy was performed subtotal, it was not required to perform biliodigestive anastomoses because adequate passage of bile was found. A morbidity rate of 13% has been reported for patients with MS types I and II, and significantly higher with patients with MS types III and IV.[19,20]

Laparotomy has been considered the technique of choice for the treatment of MS, this is largely due to its relative safety compared to the laparoscopic technique, which was described for the first time to successfully treat type I MS, it is convenient only use the laparoscopic approach to manage type I, since the minimally invasive technique is associated with high conversion rates (31-100%),[7-10] in addition to a higher incidence of bile duct injury, the most common reasons for conversion are severe and dense adhesions, acute inflammation, and unclear anatomy.[20] Bile leak, surgical site infection, and retained common bile duct stone, transitory hyperamylasemia, and common bile duct stenosis have been documented as the most frequent complications of the laparoscopic approach. In our series, only 1 case was managed by laparoscopy, 14 were resolved openly, which explains why there were no complications in our patients. In patients who are not candidates for surgery, endoscopic or percutaneous biliary decompression may be the initial treatment.[18-20]

Conclusion

This work provides important data on MS. It is important to establish a timely diagnosis and a well-established suspicion in a case of MS to be clear about the difficulties associated with cholecystectomy, as well as to be clear about the therapeutic management for each case.

Through preoperative studies, the main characteristics can be found, such as adhesions between the gallbladder and common bile duct, extrinsic compression of the common hepatic duct, fistula, dilation of intrahepatic ducts and common hepatic duct. With all this, emphasis is placed on the development of the surgeon's skills, since it is usually an intraoperative finding. This pathology must be treated by experienced surgeons to reduce the risk of the dreaded bile duct injury.

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Conflicts of interest

The authors have no conflicts of interest to declare.

References

1. Mirizzi PL. Syndrome del conducto hepático. Journal International du Chirurgie. 1948;8:731–777.
2. Rohatgi A, Singh KK. Mirizzi syndrome: laparoscopic management by subtotal cholecystectomy. *Surgical endoscopy.* 2006;20(9):1481–1477.
3. Sharma AK, Rangan HK, Choubey RP, et al. Pitfalls in the management of Mirizzi's syndrome. *Trop Gastroenterol.* 1998;19:72–74.
4. Kehr H, Die in meiner Klinik geübte Technik der Gallen– steinoperationen mit einem Hinweis auf die Indikationen und die Dauererfolge In: Wentworth Press. Auf Grund eigener, bei 1000 Laparotomien gesam–melter Erfahrungen. Lehmann; München. 1905; pp. 301–112.

5. Posta CG. Unexpected Mirizzi's anatomy: a major hazard to the common bile duct during laparoscopic cholecystectomy. *Surg Laparosc Endosc.* 1995;5(5):412–414.

6. Beltran MA, Csendes A, Cruces KS. The relationship of Mirizzi syndrome and cholecystoenteric fistula: validation of a modified classification. *World J Surg.* 2008;32(10):2237–2243.

7. Safioleas M, Stamatakos M, Safioleas P, et al. Mirizzi Syndrome: an unexpected problem of cholelithiasis. Our experience with 27 cases. *International Seminars in Surgical Oncology.* 2008;5:12.

8. Ibrarullah MD, Mishra T, Das AP. Mirizzi síndrome. *Indian J Surg.* 2008;70(6):287–281.

9. Waisberg J, Corona A, de Abreu IW, et al. Benign Obstruction of the common hepatic duct (Mirizzi Syndrome): diagnosis and operative management . *Arq Gastroenterol.* 2005;42(1):13–18.

10. Zhong H, Gong JP. Mirizzi syndrome: experience in diagnosis and treatment of 25 cases. *Am Surg.* 2012;78(1):61–65.

11. Cui Y, Liu Y, Li Z, et al. Appraisal of diagnosis and surgical approach for Mirizzi syndrome. *ANZ J Surg.* 2012; 82(10):708–713.

12. Erben Y, Benavente–Chenhalls LA, Donohue JM, et al. Diagnosis and treatment of Mirizzi syndrome: 23–year Mayo Clinic experience. *J Am Coll Surg.* 2011;213(1):114–119.

13. Karademir S, Astarcioglu H, Sokmen S, et al. Mirizzi's syndrome: Diagnostic and surgical considerations in 25 patients. *J Hepatobiliary Pancreat Surg. 2000*;7:72–77.

14. Chen H, Siwo EA, Khu M, et al. Current trends in the management of Mirizzi Syndrome. *Medicine.* 2018;97(4):e9691.

15. Valderrama Treviño AI, Granados Romero JJ, Espejel Deloiza M, et al. Updates in Mirizzi syndrome. *Hepatobiliary Surg Nutr.* 2017;6(3):170–178.

16. Oladini O, Zangan SM, Navuluri R. Delayed Diagnosis of Mirizzi Syndrome. *Semin Intervent Radiol.* 2016;33(4):332–336.

17. Zhao J, Fan Y, Wu S. Safety and feasibility of laparoscopic approaches for the management of Mirizzi syndrome: a systematic review. *Surgical endoscopy.* 2020;34(11):4717–4726.

18. Ramia JM, Villar J, Muffak K, et al. Síndrome de Mirizzi y cáncer de vesícula [Mirizzi syndrome and gallbladder cancer*]. Cirugia espanola.* 2007;81(2):105–106.

19. Yasojima EY, Lopes Filho GJ. Systematic Cholangiography during laparoscopic cholecystectomy. *Rev Col Bras Cir.* 2002;29(2).

20. Shah OJ, Dar MA, Wani MA, et al. Management of Mirizzi syndrome: a new surgical approach. *ANZ J Surg.* 2001;71:423–427.

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