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Case Report

Mirizzi syndrome - differential diagnosis with cholangiocarcinoma: case report

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ABSTRACT

Mirizzi syndrome is a rare complication of gallstone disease and poses a diagnostic challenge for general surgeons due to its clinical presentation, which resembles other pathological entities such as cholangiocarcinoma. We present a clinical case of a 68-year-old female patient with symptoms of jaundice and right upper quadrant pain. During the patient's diagnostic workup, altered liver function tests were documented, highlighting obstructive pattern hyperbilirubinemia, elevated tumor marker CA 19.9, and suspicion of biliary tract tumor based on imaging studies. However, surgical and pathological confirmation revealed Mirizzi syndrome. This case emphasizes the importance of considering differential diagnoses in patients presenting with obstructive jaundice to provide timely and appropriate treatment.

Keywords: Jaundice syndrome, Mirizzi syndrome, Cholangiocarcinoma, Tumor markers, Differential diagnosis

INTRODUCTION

Mirizzi syndrome is a rare complication of gallbladder stones, with an incidence ranging from 0.7 to 1.4% in the general population. It was described by Kehr in 1905, later described by Ruge in 1908 and finally in 1948 by Pablo Luis Mirizzi as an inflammatory process secondary to a pressure ulcer caused by an impacted lithium at the level of the infundibulum, generating obstruction of the main bile duct, erosion and a fistula between both structures or the digestive tract.[1,2]

Clinical picture is nonspecific, with the appearance of jaundice of obstructive pattern (60-100%), accompanied by abdominal pain in the right upper quadrant (50-100%) and fever in patients with presence of cholelithiasis. Laboratory studies present with hyperbilirubinemia of direct predominance and in some patients, there may be elevation of the tumor marker CA 19-9. This syndrome can resemble a gallbladder cancer or a cholangiocarcinoma, while it is considered a precancerous lesion.[3]

The non-specific clinical picture, the difficulty of diagnosis by imaging studies and the elevation of tumor markers makes Mirizzi syndrome a diagnostic and therapeutic challenge for the general surgeon. The available diagnostic methods such as tomography and ultrasound are neither sensitive nor specific, but they are useful to detect data of suspicion such as intrahepatic and extrahepatic biliary tract dilatation. Endoscopic retrograde cholangiopancreatography has a diagnostic certainty of 55 to 90%.[4,5]

Treatment depends on the type of Mirizzi syndrome, ranging from cholecystectomy in type I to reconstruction of the biliary tract as in type II to V, according to the new classification of Csendes et al in 2007 (Table 1).[6]

Table 1: Mirizzi syndrome classification.[6]

Type	Description
I	Extrinsic compression of the common bile duct by and impacted gallstone
II	Cholecystobiliary fistula secondary to an eroded gallstone involving one third of the circumference of the common bile duct
III	Cholecystobiliary fistula involving two thirds of the circumference of the common bile duct
IV	Cholecystobiliary fistula comprising the whole circumference of the common bile duct
V	Any type plus a cholecystoenteric fistula
Va	Without gallstone ileus
Vb	With gallstone ileus

CASE REPORT

We present the clinical case of a 68-year-old female patient, clinical picture characterized by jaundice, weight loss, mild pain in the right hypochondrium and general discomfort.

She was admitted to the general surgery department for icteric syndrome. Admission labs showed total bilirubin (BT) 12.8 mg/dl, direct bilirubin (BD) 9 mg/dl, indirect bilirubin (BI) 3.7 mg/dl, alkaline phosphatase (ALP) 360 U/l, gamma glutamyltransferase (GGT) 272 U/l, leukocytes 9.39×103 ul, and CA 19.9 178 mg/dl.

In imaging studies with biliary tract dilatation by ultrasonography (USG) (common bile duct 10 mm, gallbladder not assessable by WES phenomenon) (Figure 1). Simple and contrasted abdominal computed tomography (CT) scan documented intra and extrahepatic biliary tract dilatation, choledochus 13 mm without visible lithium, non-assessable gallbladder content, imaging diagnosis probable cholangiocarcinoma (Figure 2). Three-phase CT was performed, without other relevant findings, without documenting obstruction, or other data of tumor activity.

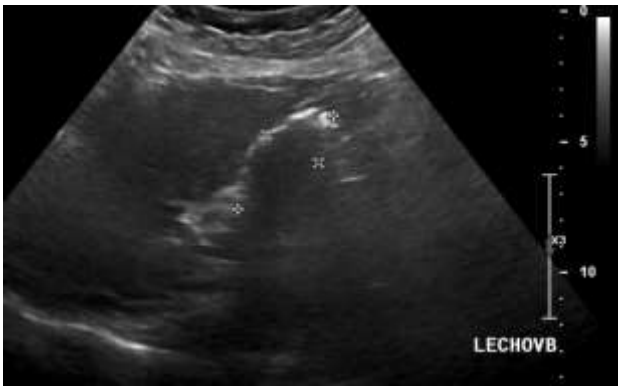

Figure 1: Ultrasound of liver and biliary tract showing gallbladder with WES (wall, echo, shadow) phenomenon, dimensions of 49×21 mm wall and non-assessable content.

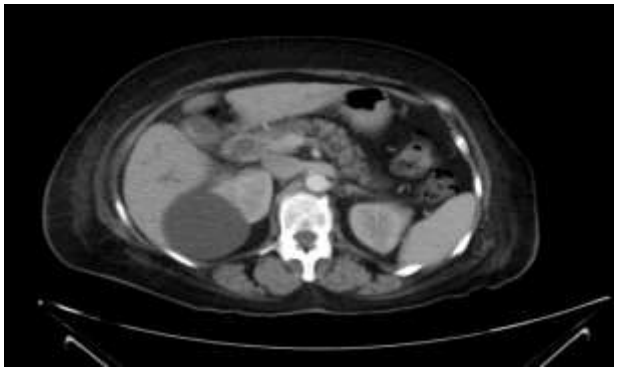

Figure 2: Simple and contrasted abdominal CT showing intra and extrahepatic biliary tract dilatation, 10 mm choledochus and decrease of caliber in intrapancreatic segment with enhancement upon contrast medium application, considering a probable infiltrative process of cholangiocarcinoma.

Patient candidate to bile duct bypass due to risk of cholangitis, reason for which ERCP was performed with placement of stent type Amsterdam 10 Fr×10 cm, documenting intra and extrahepatic bile duct dilatation, without appreciating filling defects or areas of stenosis, balloon sweep was performed without obtaining lithos, endoscopic diagnosis probable sphincter of Oddi dysfunction and with endoscopic brushing of the bile duct (Figure 3).

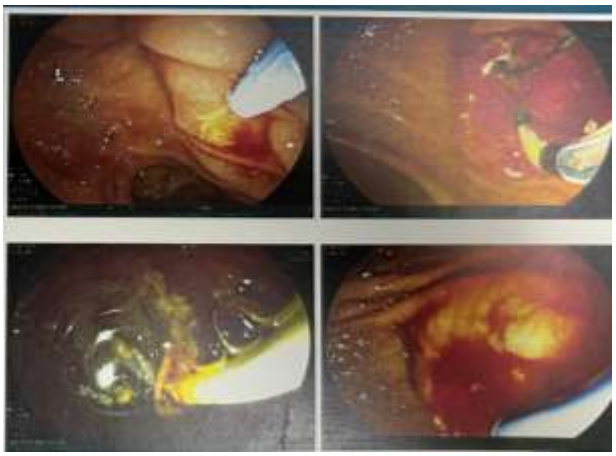

Figure 3: ERCP showing intra and extrahepatic biliary tract dilatation, without filling defects or stenosis, common bile duct brushing, 10 Fr×10 cm Amsterdam stent placed.

Posterior signs of moderate cholangitis despite stent, with tumor markers remaining elevated, without identifying by imaging or endoscopic studies conclusive data of malignancy, it was decided to perform diagnostic laparoscopy but due to anatomical difficulties open biliary tract exploration was performed, findings of ascites, cholecystitis, pitcher mouth communication with main biliary tract were reported, friable tissue, probable Mirizzi syndrome type IV versus cholangiocarcinoma, performing

partial reconstituted cholecystectomy with 2-0 polyglycolic acid suture to close the fistula with gallbladder remnant, choledochotomy with exposure and removal of biliary stent, followed by negative biliary tract exploration, so it was decided to place a T-tube and choledochorraphy with 3-0 polyglycolic acid suture simple stitches, transoperative cholangiography without alterations, placing Penrose type open drainage into Winslow's hiatus. Adequate post-surgical evolution without complications, functional T-tube, no signs of biliary leakage and improvement in laboratory parameters. Pathology report of common bile duct brushing with definitive diagnosis of chronic inflammatory process without evidence of neoplasia, definitive histopathological report of the surgical specimen with a diagnosis of acute chronic cholecystitis, without data of tumor activity. Follow-up by the outpatient clinic, with cholangiography by T-tube without evidence of leakage, stenosis or filling defects, liver function tests within normal, T-tube was removed after 3 weeks, without evidence of subsequent complications.

DISCUSSION

Mirizzi syndrome is a rare, complex entity with a variable clinical presentation that can simulate different pathologies, including cholangiocarcinoma. Before offering surgical treatment, it is diagnosed in only 12.5 to 22% of the cases that present. In our environment it is reported that the incidence of Mirizzi's syndrome presents in 4.7%.[5,6]

The diagnostic approach is currently performed with clinical presentation, laboratory studies including liver function tests, imaging studies such as computed tomography, magnetic resonance or even the surgical approach, the latter also considered as the gold standard of treatment.[7]

Endoscopic retrograde cholangiography is considered an adequate diagnostic method for those patients who are not considered candidates for the surgical approach. Ultrasound is reported to have a diagnostic accuracy of 29% with a sensitivity ranging from 8.3-27%. By computed tomography, in the presence of periductal inflammation it can be confused with a cholangiocarcinoma, however, its usefulness is to exclude tumor activity in the porta hepatis or in the liver. The diagnostic accuracy of magnetic cholangioresonography (MRCP) is 50%, besides being useful in identifying the presence or not of a fistula. ERCP has a diagnostic accuracy ranging from 55-90% with a failure rate of 5-10%, with the possibility of being diagnostic-therapeutic.[8,9]

Surgical treatment is the gold standard for the management of Mirizzi's syndrome, however, it is challenging due to low suspicion, lack of preoperative diagnosis and anatomical distortion due to chronic inflammation. Open surgery used to be considered the approach of choice because of its relative safety, however laparoscopy has increasingly gained ground with the disadvantage of conversion rates ranging from 11-80%. Patients with Mirizzi syndrome type I, II and III undergo sphincterotomy, stent placement and subsequent partial cholecystectomy. Peroral cholangioscopy with electrohydraulic lithotripsy has a success rate for lithotripsy removal of 100%.10 The minimally invasive era continues to develop new techniques for the treatment of these types of conditions.[10,11]

This clinical case illustrates the difficulty of preoperative diagnosis and the importance of surgical exploration in defining the diagnosis. The clinical history of this patient initially suggested cholangiocarcinoma, with jaundice, weight loss and right hypochondrium pain. Laboratory findings and imaging studies confirmed intrahepatic and extrahepatic bile duct dilatation, dilated common bile duct and non-assessable gallbladder. However, the lack of evidence of frank obstruction or tumor activity by three-phase CT and ERCP led to a surgical exploration, which allowed the diagnosis of Mirizzi syndrome type III with a jug mouth communication with the main bile duct. This case highlights the importance of the surgical approach in the differential diagnosis and the need to carefully explore the bile ducts to avoid complications, including inadvertent bile duct injury and biliary leakage.

CONCLUSION

Mirizzi syndrome is a rare, complex entity with a clinical presentation that can resemble different pathologies, including malignant pathology such as cholangiocarcinoma, leading to therapeutic delay and erroneous treatment with serious complications. Diagnosis can be challenging due to the lack of sensitivity and specificity of extension studies, as well as the elevation of the tumor marker CA19-9 which can be elevated in non-oncologic pathology due to the presence of chronic inflammation. Differentiating Mirizzi's syndrome from cholangiocarcinoma requires exhausting all available resources in care centers, as well as a multidisciplinary approach.

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