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Surgical management of cholecystoduodenal fistula complicated by ileus (Mirizzi syndrome type Vb). A case report

Mirizzi Syndrome (MS) is a rare gallbladder disease described by Argentine surgeon, Pablo Luis Mirizzi in 1948. It concerns a wide range of clinical manifestations, including gallstone obstruction of the intestine. The modified classification by Csendes distinguishes five types of MS. The case described by the authors of this paper could be classified as type Vb, which means MS with a complication of obstruction. A 74-year-old woman with nonspecific, permanent, diffuse abdominal pain and bile vomiting was admitted to the emergency department where conservative treatment was administered. Based on the preoperative diagnosis of ileus, the patient was qualified for a laparotomy. The patient had a cholecystoduodenal fistula which was responsible for the displacement of the stone into the intestinal lumen and consequently for the mechanical obstruction of the intestine. It must be taken into account that the clinical manifestation of MS may be biliary obstruction, as occurred in the case presented below. Biliary symptoms may occur, but this is not a common situation, whereas half of patients with an obstruction have a history of biliary disease. The case presented here can be a valuable lesson in being mindful of the possibility of elderly and female patients developing biliary obstruction, even without having a history of chronic gallstone disease. Therefore, a meticulous intraoperative inspection should be performed in such cases in order to search for possible fistulas.

KEY WORDS: Cholecystoduodenal fistula, Gallstone disease, Mirizzi syndrome

Introduction

Mirizzi Syndrome (MS) is a rare gallbladder disease described by Argentine surgeon Pablo Luis Mirizzi in 1948. It consists of the light of the neck of the gallbladder or the gallbladder duct being closed by a gallstone. This results in the development of mechanical

jaundice^{1,2}. The condition of patients with this disease can vary greatly. Clinically, the patient may be both asymptomatic and have severe cholangitis. For this reason, it is often impossible to diagnose the patient preoperatively and the final diagnosis is made postoperatively³. The correct preoperative diagnosis can only be made in 43-73% of cases⁴. Among the possible surgical variants, a subtotal cholecystectomy, choledochoplasty, and bilio-enteric anastomosis (depending on the type) are the most common ones. The method used should be selected on the basis of the patient's clinical condition to minimise risk. A surgical intervention which is carried out as quickly as possible improves the prognosis⁴. Patients with MS have a higher risk of cancer of the gallbladder (than patients with uncomplicated cholelithiasis), as well as bilio-enteric and bile tract injury during surgery^{3,5}. In Western developed countries, this

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ABBREVIATIONS

BP: blood pressure
 CA19-9: carbohydrate antigen 19-9
 CRP: C-reactive protein
 CT: computed tomography
 ERCP: endoscopic retrograde cholangiopacreatography
 FFP: fresh frozen plasma
 HB: hemoglobin
 HCT: hematocrit
 HR: heart rate
 MR: (MRI) – magnetic resonance (imaging)
 MRCP: magnetic resonance cholangiopacreatography
 MS: Mirizzi Syndrome
 POD: postoperative day
 PRBC: packed red blood cell concentrate
 PTC: percutaneous transhepatic cholangiography
 RUQ: right upper quadrant
 TPN: parenteral nutrition
 US: ultrasound

The patient was defecating normally and did not complain of flatulence. A physical examination confirmed a moderately painful abdomen with the presence of peristalsis. Bowel sounds were audible on auscultation. There were no acute peritoneal symptoms. The temperature was within normal limits; HR was 78 and BP was 150/80 mmHg. Her C- reactive protein was elevated (24.2 mg%). The patient was chronically treated due to hypertension and dizziness. Based on the preoperative diagnosis of ileus, the patient was qualified for a laparotomy.

disease occurs with a frequency of no more than 1% per year. However, in underdeveloped countries such as in Latin-America, it reaches 5.7%^{2,5}. There is no difference in frequency between men and women. It most often occurs between 40 and 70 years of age². The modified classification by Csendes distinguishes five types of this disease. Type I refers to the external pressure of the bile duct through a stone located in the infundibulum of the gallbladder or the cystic duct. Types II, III, and IV mean the presence of a cholecystocholedochal or cholecystohepatic fistula. In type II it occupies no more than 1/3 of the circumference of the gallbladder, in type III up to 2/3, and in type IV it leads to the complete obstruction of the bile duct. Type V refers to cases in which any type of cholecystoenteric fistula occurs. Subtype Va is when there is a cholecystoenteric fistula without a gallstone ileus, while in subtype Vb this fistula is complicated by a gallstone ileus^{1,2}. The case presented below concerns a type Vb MS.

CASE PRESENTATION

A 74-year-old woman (S.L.) with nonspecific, permanent, diffuse abdominal pain and bile vomiting was admitted to the emergency department, where conservative treatment was administered. The next day there was an increase in the pain and vomiting and an X-ray revealed features of an obstruction within the small intestine loop (Fig. 1).



Fig. 1: Abdominal radiograph showing the multiple air-fluid levels.

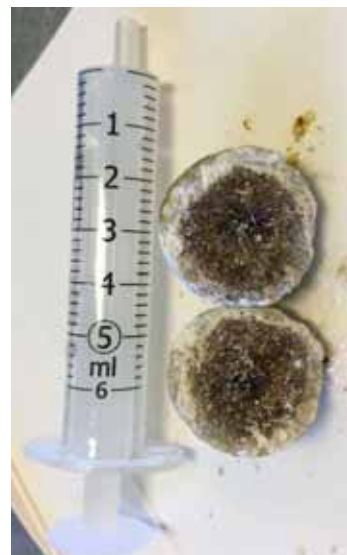


Fig. 2: Operative view of the gallstone from the ileum measuring 25mm in diameter.

my. The abdominal cavity was opened using a midline incision. Intestinal loops of jejunum were distended to the border with the ileum. The intestine was cut and a spherical stone with a diameter of 2.5cm was excavated (Fig. 2).

Then the ileum were transversely sutured and the anastomosis was covered with Tachosil[®]. Reactive fluid was evacuated from the peritoneal cavity during the procedure. No palpable abdominal disorders and no visible abnormalities were reported. However, on POD 1 (post-operative day 1) subileus symptoms appeared, which were successfully cured conservatively. A week later the patient had to undergo another operation in order to treat eventration by reconstructing the abdominal wall. During the second operation purulent contents were detected, with fibrin on the intestines and a small fistula at the anastomosis site. It was decided to perform a segmental bowel resection with side-to-side anastomosis using a mechanical suture. After reoperation, TPN (total parenteral nutrition) was administered due to the patient's low nutrition parameters. A few days later the patient had bloody diarrhea without any other symptoms, but was in good general condition. Laboratory test results revealed decreased blood parameters: HCT was 23% (N: 36.0- 47.0 %), and HB was 7.5 g/dl (N: 12.0- 16.0 g/dl). A gastroscopy was carried out, which excluded bleeding, visualised gastritis and also revealed a cholecystoduodenal fistula in the descending part of the duodenum with a diameter of 10 mm. Effective treatment including 4 units of PRBC (packed red blood cell concentrate) and 4 units of FFP (fresh frozen plasma) normalized the blood parameters and the patient was discharged from the hospital in a good general and local condition. 2.5 years later, the patient was admitted to the ward with the following symptoms: diffuse abdominal pain, nausea and vomiting. The symptoms receded after conservative treatment and the patient was discharged from the ward in a good general condition.

Discussion

From the pathophysiological point of view, MS can be understood as an infrequent and late complication of gallstone disease. The pathological process consists of external compression of a common hepatic duct caused by stones in the neck of the gallbladder or the cystic duct and concomitant inflammation within the triangle of Calot. One or more gallstones may cause pressure and ischemia and may then penetrate the gallbladder wall, leading to the formation of a fistula which, depending on the location, could be cholecystobiliary, cholecystogastric or cholecystoenteric, which include cholecystoduodenal and cholecystocolonic fistulas⁵⁻¹⁰. Displaced by such a fistula, bile deposits may cause the obstruction of the gastrointestinal tract, which occurs in 1-4% of all the cases of obstruction within the small

intestine, and in 25% of cases it affects people over 65 years of age. The size of the stones causing the biliary obstruction is over 2 cm, and on average 4 cm. Most gallstones up to 2-2.5 cm pass through the gastrointestinal tract by themselves and are expelled with a stool without health consequences^{4,11}. A gallstone enters the gastric outlet through the cholecystoduodenal fistula and causes obstruction there. Only 1 in 10 cases of gallstone ileus end with the gallstone stuck in the duodenal bulb because of its spectacular size. This is called Bouveret syndrome, first described in 1896. This syndrome is characterised by non-specific symptoms that may resemble MS¹¹.

In this case, the patient had a cholecystoduodenal fistula, which is the most common in biliary obstruction due to the anatomical proximity of the gallbladder to the duodenum. The fistula was responsible for the displacement of the stone into the intestinal lumen and consequently, for the mechanical obstruction of the intestine⁷⁻⁹. MS is more common in women aged 50 to 70 with a history of long-term gallstone disease, having suffered from the disease for an average of 29.6 years^{5,12}. There are no pathognomonic symptoms specific to Mirizzi syndrome. The most common symptoms include obstructive jaundice, RUQ (right upper quadrant) pain and fever. MS may coexist with cholecystitis, acute cholangitis, or pancreatitis, and leukocytosis is then observed. Laboratory tests show hyperbilirubinemia as well as elevated levels of aminotransferases and alkaline phosphatases^{3,5,9,13,14}. Some studies point to a high level of CA19-9 (carbohydrate antigen 19-9)^{5,8,15}. This is especially significant in patients with cancer of the gallbladder. MS needs to be differentiated from gallbladder cancer because cancer is present in 6% to 27% of patients with a preoperative diagnosis of MS. It is worth noting that xanthogranulomatous cholecystitis may be confused with malignant lesions^{3,13}. It must be taken into account that the clinical manifestation of MS may be biliary obstruction, as it occurred in the case presented here. Typically, the patient reports nausea, vomiting, diffuse pain, flatulence or constipation. Biliary symptoms may occur, but this is not a common situation, whereas half of patients with an obstruction have a history of biliary disease^{7,8,16}. Many sources emphasise that, due to biliary tract injuries, a preoperative diagnosis of Mirizzi syndrome is very important. The absence of abdominal surgery or an inguinal hernia in the past should raise suspicion of a gallstone ileus¹⁶. This seems to be possible in 8% to 63% of cases, while an intraoperative diagnosis concerns over 50% of patients with Mirizzi syndrome^{5,8,12,13,17}. Ultrasound is a commonly used diagnostics tool which is recommended as the best screening method, with the sensitivity of the test varying from 8 to 48%. Typical pathologies seen in an ultrasound are a contracted gallbladder with thin, atrophic walls, the presence of bile or concrements located in the cystic duct, and hepatic ductal dilatation above the level of the

obstruction^{5,8,12,13,15,17-19}. A CT examination, similarly to an US examination, enables the visualisation and measurement of wall thickness and biliary dilatation. It also visualises bile deposits, but it is mainly beneficial in distinguishing Mirizzi's syndrome from the malignant process within the extrahepatic biliary system^{5,8,13,15,17,19}. MRCP, which is considered the best non-invasive imaging method, can visualise the features typical of MS, such as: external compression of the bile duct, gallstones, dilatation of the intrahepatic ducts, as well as enabling the assessment of inflammation around the follicle. It can also differentiate it from other pathologies in this area, such as the neoplastic process and is useful in imaging fistulas. Similar results can be obtained with a percutaneous transhepatic cholangiography (PTC)^{5,8,13,14,17-19}. Although the endoscopic retrograde cholangiopancreatography procedure (ERCP) is invasive, it is the gold standard of diagnosis that depicts an obstruction of the biliary tract with a widening of the segment above. ERCP also shows the cause, place and size of a biliary obstruction and locates fistulas. This method makes it possible to perform treatment procedures such as stone removal or palliative stent insertion when a patient has jaundice^{1,5,8,13,14,17,19}. In the case presented above, the diagnosis of biliary obstruction was difficult due to the accompanying non-specific symptoms. A plain abdominal x-ray is a useful and cheap test used in the diagnosis of small bowel obstruction. Formerly, the basic diagnostics tool was a radiograph and in 1941 a Rigler's triad was described showing typical radiological features such as: partial or complete intestinal obstruction, pneumobilia, and a displaced gallstone in the intestinal lumen, but their presence varies between 7% and 35% in the literature. Pneumobilia may also appear as a result of surgical or endoscopic biliary interventions or as a result of an incompetent sphincter of Oddi. However, only 10% of gallstones are calcified enough to be radiographically visible¹⁶. Balthazar described a symptom consisting in the presence of two air fluid levels in the right upper quadrant on abdominal radiograph^{7,9,10,12,19}. Currently, the US has no diagnostic significance in biliary obstruction. A CT with high sensitivity and specificity shows obstruction, gallstones or pneumobilia. MRCP can be used as an alternative to a CT in patients contraindicated for a CT^{3,7,8}. The standard treatment of MS, associated with low operative morbidity and mortality, is open surgical treatment depending on the type of disease. Laparoscopic treatment is not recommended due to possible damage to the bile duct. Endoscopic treatment methods mean biliary drainage or gallstone removal by using a basket or a balloon. Various authors agree that the best treatment for older patients with multiple comorbidities is enterolithotomy alone⁴. Other options for removing bile deposits include an electrohydraulic or extracorporeal lithotripsy, shock wave, dissolution therapy or laser. For type 1 MS, a total or partial cholecystectomy is preferred. In the case of severe

inflammation and obturation the triangle of Calot is recommended to open the fundus of the gallbladder and remove the deposits before performing a cholecystectomy. For the following types of MS, in the presence of fistulas with little loss of the gallbladder wall, some of the wall can be used to cover the fistula. When the fistula exceeds the possibilities of the above-described treatment, bilenteric anastomosis or hepatojunostomy using the Roux-en-Y method is recommended. The T-tube, which is inserted in place or distally from the fistula, is used in choledochoplasty as well as for the closure of the primary bile duct and enables radiological intervention. Laparotomy and enterolithotomy should be the first goal in treating Vb MS as they will overcome the obstruction of the intestine. Although it is a standard procedure, it should be borne in mind that it is not devoid of important complications in the form of a risk of acute cholecystitis, cholangitis and a relapse. It is controversial whether the biliary-enteric fistula should be closed at the same time as the cholecystectomy, but in over 50% of cases it closes spontaneously. Crushing the gallstone in situ is not recommended, but extracorporeal shock wave lithotripsy may be used successfully^{1,3,5-8,12-15,17-19}. Our patient was treated in accordance with the preoperative diagnosis of obstruction. A laparotomy was performed to remove the cause of the obstruction, which turned out to be a gallstone. During the operation, the cholecystoduodenal fistula was not visualised, but this was made possible by the gastroscopy performed due to the symptoms of a hemorrhage. Only at that point could our case be classified as type Vb MS, which means MS with a complication of obstruction.

Conclusion

Mirizzi's syndrome is a disease with a wide range of clinical manifestations, including gallstone obstruction of the intestine. Our case can be a valuable lesson in being mindful of the possibility of elderly female patients developing biliary obstruction, even without a history of chronic gallstone disease. The gold standard is unblocking an obstruction by laparotomy. When considering the gallstone as the cause of the obstruction, a meticulous intraoperative inspection should be performed in search of possible fistulas.

Riassunto

La Sindrome di Mirizzi (SM) è una rara malattia della colecisti descritta dal chirurgo argentino Pablo Luis Mirizzi nel 1948. Nei paesi sviluppati occidentali questa malattia si manifesta con una frequenza non superiore all'1% all'anno. La classificazione modificata di Csendes distingue cinque tipi di questa malattia. Il caso presentato riguarda il tipo Vb di MS.

Descriviamo un caso di una donna di 74 anni con dolore addominale non specifico, permanente, diffuso e vomito biliare. L'esame obiettivo ha confermato un addome moderatamente doloroso con peristasi presente e rumori intestinali all'auscultazione. Secondo la diagnosi preoperatoria di ileo, il paziente era qualificato per una laparotomia urgente. Una pietra biliare sferica con un diametro di 2,5 cm è stata scavata dall'ileo. Il paziente è stato sottoposto a un nuovo intervento e sono stati rilevati i contenuti purulenti con fibrina sull'intestino e una piccola fistola nel sito di anastomosi. La SM è una complicanza rara e tardiva di calcoli biliari. In questo caso il paziente aveva una fistola colecistoduodenale che è la più comune nell'ostruzione biliare dovuta alla vicinanza anatomica della colecisti con il duodeno. Considerando i calcoli biliari come la causa dell'ostruzione, è opportuno eseguire una meticolosa ispezione intraoperatoria alla ricerca di eventuali fistole.

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