

Mirizzi syndrome complicated by common hepatic duct fistula and left hepatic atrophy: a case report

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Abstract

Background: Mirizzi syndrome is a rare complication of chronic cholecystitis, usually caused by gallstones impacted in the cystic duct or the neck of the gallbladder. Mirizzi syndrome results in compression of the hepatic duct or fistula formation between the gallbladder and common bile duct (or hepatic duct, right hepatic duct, or even mutative right posterior hepatic duct). Clinical features include abdominal pain, fever, and obstructive jaundice. Severe inflammation and adhesion at Calot's triangle are potentially very dangerous for patients with Mirizzi syndrome undergoing cholecystectomy. Case presentation: We report the case of a 68-year-old Asian woman who presented with abdominal pain and jaundice. She had a medical history of gallstones, but no fever. Magnetic resonance cholangiopancreatography revealed cholecystitis, cholelithiasis, common hepatic duct stones, and ascites. Findings at surgery included a porcelainized, atrophic gallbladder that was full of gallstones, fistula formation between the gallbladder and common hepatic duct, and left hepatic atrophy. The prominent feature was the left hepatic atrophy, but stones were not visible pre-operatively in the left liver by radiologic examination.

Conclusions: This patient exhibited what can be considered a special type II of Mirizzi syndrome with a fistula of the common hepatic duct as well as left hepatic atrophy.

Keywords

Mirizzi syndrome, common hepatic duct fistula, left hepatic atrophy, left hemihepatectomy, gallstone impaction, chronic cholecystitis

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Background

The incidence of Mirizzi syndrome (MS) is low and only 0.06%–5.7% of patients with gallstones have the typical symptoms of right upper abdominal pain, with or without jaundice and fever.¹ However, these symptoms are not typical for all patients, and there are no special manifestations of MS in imaging studies. This can lead to difficulty in pre-operative diagnosis and inadequate evaluation of the condition. Radiologists might not consider the diagnosis of MS or be appropriately concerned about the potentially dangerous and difficult treatment or of the operative risks. Multiple diagnostic imaging tools are available: abdominal ultrasound, computed tomography (CT), magnetic resonance cholangiopancreatography (MRCP), and endoscopic retrograde cholangiopancreatography (ERCP).^{2,3} Among them, MRCP has the advantages of being noninvasive and intuitive, and it provides multiple perspectives.⁴ Even with comprehensive imaging studies, the diagnosis can still be difficult, especially in the case of combined fistula and adhesions that may cause catastrophic injuries in surgery.² Therefore, comprehensive preparation pre-operatively has been necessary for avoiding injuries such as damage to the bile duct. We report a special type of MS with upper left hepatic atrophy. Left hemihepatectomy had been planned (because of biliary stenosis), with partial gallbladder resection, hepatic duct stone removal, and hepaticojejunal Roux-en-Y anastomosis.

Case presentation

A 68-year-old female Asian patient was admitted to our hospital with epigastric pain and jaundice for 1 week. Before admission, the patient had noted epigastric pain after eating greasy food, especially affecting the upper right abdomen with sustained

pain. Gradually jaundice accompanied by darker urine and white, clay-like feces had appeared. She was diagnosed with cholelithiasis and obstructive jaundice in the local clinic, and concurrent anti-inflammatory treatment had been undertaken and her condition improved. Since the onset, she had no nausea or vomiting, no chills or fever, no significant weight loss. She had no appreciable loss of appetite. She had a 20-year history of gallstones. Physical examination showed yellow sclerae, flat abdomen, slight tenderness in the right upper abdomen, no guarding, and no rebound tenderness. The results of laboratory studies are shown in Table 1. In addition, hepatitis B surface antigen and hepatitis C virus antibody were negative.

Ultrasonography showed an atrophic gallbladder that was full of stones. Enhanced CT scanning of the upper abdomen demonstrated a smooth liver edge and uniform liver parenchyma, as well as dilatation of the intrahepatic bile duct and the upper pancreatic duct. The gallbladder was irregularly shaped, with thickened walls and uneven internal density. Radiologic observations indicated the presence of cholecystitis and gallstones; biliary stones were

Table 1. Laboratory studies.

Test	Result	Reference Range
ALP	292 U/L	35–104 U/L
ALT	145.4 U/L	7–40 U/L
AST	101.9 U/L	13–35 U/L
GGT	309.6 U/L	7–45 U/L
GLDH	10.0 U/L	0–6 U/L
TBA	30.2 μmol/L	0–10 μmol/L
TBIL	93.6 μmol/L	3–20 μmol/L
DBIL	87.6 μmol/L	0–6.8 μmol/L
IBIL	6.0 μmol/L	0–14.5 μmol/L

ALP, alkaline phosphatase; ALT, alanine transaminase; AST, aspartate transaminase; GGT, gamma-glutamyl transpeptidase; GLDH, glutamate dehydrogenase; TBA, total bile acid; TBIL, total bilirubin; DBIL, direct bilirubin; IBIL, indirect bilirubin.

observed in the hilum, with secondary biliary obstruction (Figure 1). MRCP revealed dilatation of the intrahepatic bile ducts and the left and right hepatic ducts, as well as truncation of the common hepatic duct (internal diameter was 5.1 mm). Multiple faint signals were seen in the common hepatic duct and gallbladder (Figure 2). The MRCP results indicated the presence

of cholecystitis, cholelithiasis, common hepatic duct stones, and ascites. The radiologist did not suspect MS or left hepatic atrophy pre-operatively. However, our surgical team had considered the possibility of MS from the clinical and imaging manifestations. Surprisingly, jaundice became apparent in a short time, and the left liver atrophy was revealed.

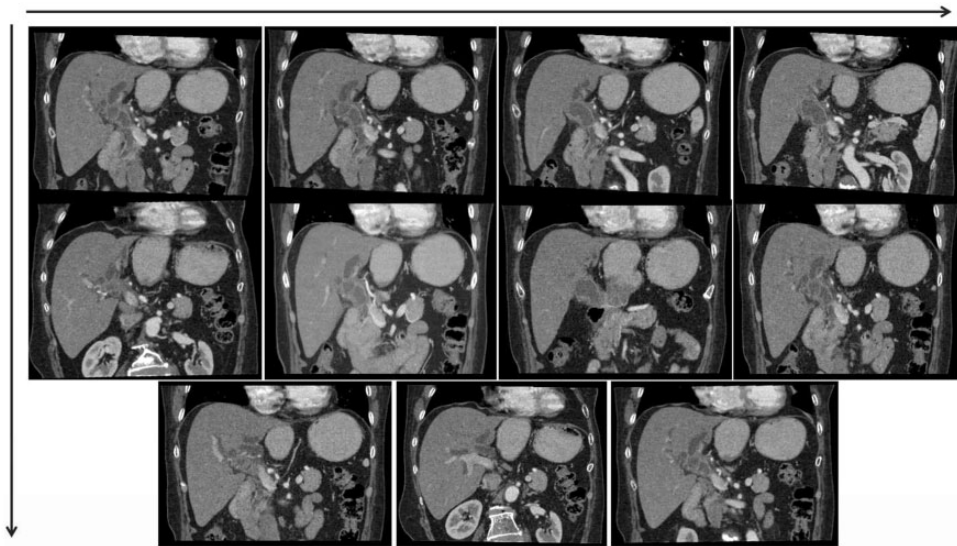


Figure 1. Enhanced CT scan showing cholecystitis as well as gallstones and biliary stones in the hilum, with secondary biliary obstruction.

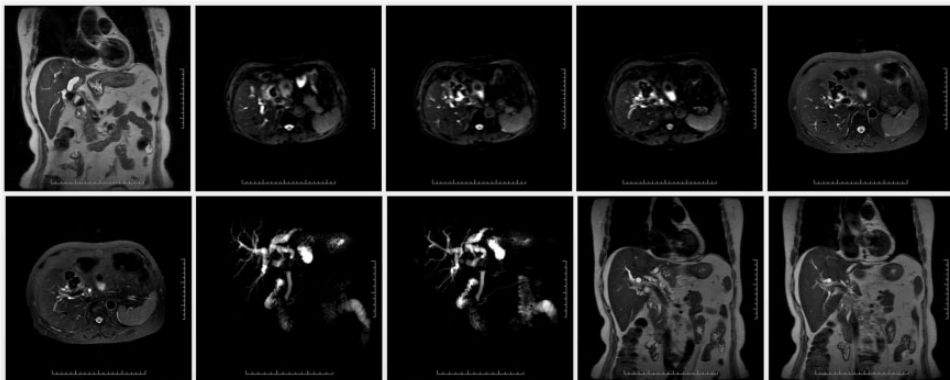


Figure 2. Magnetic resonance cholangiopancreatography showing dilatation of the intrahepatic bile ducts and left and right hepatic ducts, as well as truncation of the common hepatic duct.

Surgical findings included confirmation of the atrophic, porcelainized gallbladder. The gallbladder was incised and part of the anterior wall of the gallbladder was removed. Multiple 5 to 10 mm stones filled the gallbladder, and no bile was found in it. The stones were removed from the gallbladder to the hepatic hilum. Continuing backwards, an enlarged cystic cavity was observed and identified as the hepatic duct; bile flowed from the duct. The gallbladder and hepatic ducts were connected, and the left liver was atrophic

(Figure 3). The common hepatic duct and the left and right hepatic ducts formed a compressed and enlarged cystic cavity having a stiff, fibrotic wall. The stones in the enlarged cystic cavity were piled and layered, forming an impaction at the opening of the left and right hepatic ducts. Bile could flow through the crevices between the stacked stones; however, the opening of the left hepatic duct was completely obstructed by stones, and bile could not flow out. Thus, the chronic inflammatory stimulation had stiffened and narrowed the left hepatic

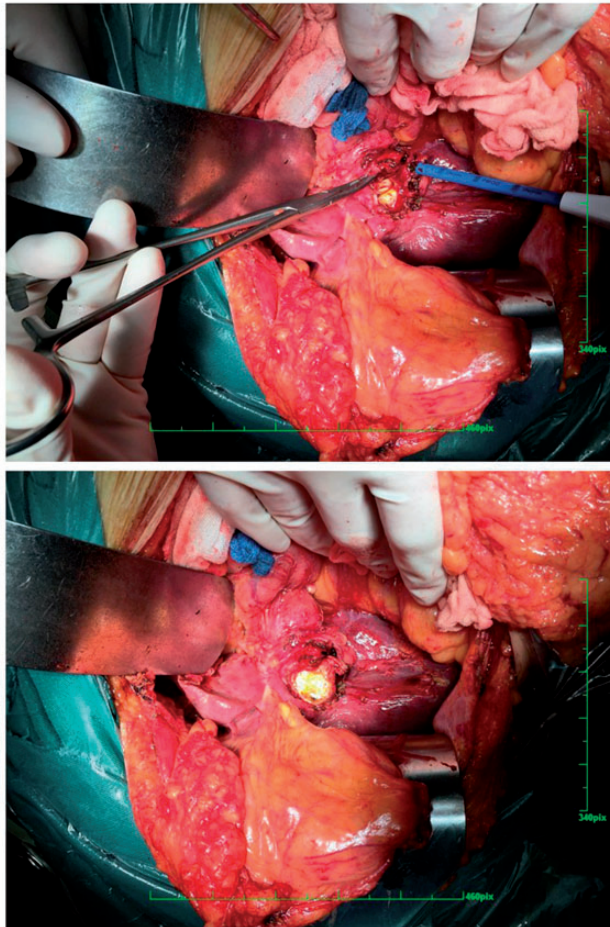


Figure 3. Intra-operative examination showing that the gallbladder and the hepatic ducts were connected; the left liver was atrophic.

duct, resulting in significant blockage of bile outflow from the left liver, ultimately causing it to atrophy. Because the opening of the right hepatic duct was not impacted completely by the stones, bile could drain into the common bile duct. After removal of the stones, the anterior walls of the common and left and right hepatic ducts were identified; the entire bile duct was stiffened and fibrotic, and reconstruction became very difficult. Therefore, left hemihepatectomy and right hepaticojejunal Roux-en-Y internal drainage operation were carried out. Postoperative pathological examination showed evidence of chronic cholecystitis with stones in the dilated intrahepatic bile duct. In addition, lymphocyte infiltration was found at the interlobular portal areas of the liver, and the gallbladder epithelium had disappeared secondary to erosion. The patient's jaundice faded after surgery and she was discharged after an uneventful postoperative course. The patient denied any discomfort at follow-up 2 years later.

All protocols were performed in accordance with the Declaration of Helsinki and approved by the ethics committee of our university (YKD2015054). Written informed consent was obtained from the patient for publication of this case report and all accompanying images.

Discussion

MS was first described by Hans Kehr in 1905⁵ and named by the Argentine surgeon Pablo Mirizzi in 1948 with cholangioangiography.⁶ MS is characterized by the mechanical compression or erosion of the common bile duct because of the impaction of gallstones in the cystic duct or in the neck of the gallbladder. In this patient, the gallbladder neck, common hepatic ducts, and left and right hepatic ducts had merged as a result of erosion and adhesions. These findings were accompanied by left hepatic atrophy. Thus, our case can be considered a special

type II MS that can result in liver atrophy from an obstructing stone at the opening of the left or right hepatic duct. Intra-operative findings confirmed an impacted stone at the opening of the left hepatic duct, causing chronic blockage of left hepatic bile outflow and left liver atrophy.

MS is generally characterized by abdominal pain, jaundice, and abnormal liver function tests. However, patients with gallstones and bile duct stones often show the same clinical characteristics. Although bile duct stones were reported by the radiological examinations, we had considered the possibility of MS in this patient. So, we fully prepared for this possibility in pre-operative planning. The cause of left hepatic atrophy was not explained before surgery. Despite the fact that there are various imaging techniques currently available, most of them have difficulties in identifying MS before surgery, especially with pre-operative fistulas and the degree of adhesion and fibrosis.

McSherry et al. has proposed classification of MS into two types based on the findings of ERCP.⁷ Csendes et al. further subclassified MS into four types according to the diameter of the choledochal obstruction; these subtypes are widely used for education.⁸ If any type is combined with a cholecystoenteric fistula, it can be classified as a Csendes type V.^{3,9} The case reported here can be considered a special type of MS not included in the above-mentioned classifications. In this patient, the common hepatic duct and left and right hepatic ducts formed an enlarged cystic cavity with a stiffened, fibrotic wall. The packed and layered stones blocked the opening of the left and right hepatic ducts. Chronic inflammation stiffened the left hepatic duct opening, and the outflow of the left hepatic bile was considerably obstructed, ultimately leading to atrophy of the left liver. The gallbladder, common hepatic duct, and left and right hepatic ducts were fused together into a cystic cavity complicated by the hepatic atrophy. On the basis of a

review of the literature, this type of MS is unique and can be considered a new subtype of type II. Several cases of MS accompanied by right hepatic atrophy have been described,¹⁰ while no MS-related left hepatic atrophy has been reported. The key point for patients with MS is to consider this special type of MS pre-operatively and formulate surgical plans that will provide the best treatment and avoid incidental injuries and complications.

In our experience, the gallbladder in MS is often atrophic and the gallbladder wall is fibrotic and porcelainized. Most of these gallbladders contain little or no bile, and they are filled with stones. In MRI, there is a low signal in the gallbladder fossa and the gallbladder border is not seen clearly. In addition, the gallbladder and bile ducts punch through and subsequently the bile duct is interrupted. The interruption of the bile ducts in MRCP is similar to a stream of water that suddenly encounters a stone and then flows around both sides of it, sometimes only slightly. The other reason for bile duct interruption is cholangiocarcinoma, which in MRCP typically is similar to the flow from a hose tied with string. As surgeons scrutinizing imaging studies, we should pay attention to the details of bile duct obstruction in patients with jaundice.

The diagnosis and treatment of MS should begin with the history of gallstones. Most of the patients present with repeated epigastric or right epigastric pain with mild or moderate jaundice, with or without fever. MRCP examination is required to make a comprehensive analysis. Careful image interpretation, study of the details, and consciously considering the possibility of MS are important for diagnosis. The location of the stones, and the degree, length, and defects of the stenosis of the bile ducts should be analyzed. We also should pay attention to the condition of the liver. Surgery can be performed based on the established plan to achieve the therapeutic

outcome to the patient's maximum benefit by avoiding intra-operative injuries.

Conclusions

This patient presented a unique type II MS with a fistula of the common hepatic duct and left hepatic atrophy, making pre-operative diagnosis difficult. MRCP can help surgeons to diagnose MS accurately and provide optimal treatment.

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Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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