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Case Report

Mirizzi Syndrome Secondary to Impacted Stone in A Low Inserted Cystic Duct Variant: Case Report

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Introduction: Cystic duct anatomic variations are encountered frequently during surgical procedures. In this report, a female patient underwent laparoscopic cholecystectomy for acute cholecystitis and developed symptoms of obstructive jaundice subsequently, which was diagnosed as low insertion of the cystic duct and an impacted stone that was causing common hepatic duct obstruction.

Case report: A 42-year-old woman presented to the emergency room complaining of persistent right upper quadrant pain that was radiating to the back. White blood cell count was slightly elevated with normal liver function tests. An abdominal ultrasound was performed and confirmed cholelithiasis with no signs of acute cholecystitis or biliary dilatation. The patient underwent laparoscopic cholecystectomy as an emergency procedure for persistent biliary colic. Eight days later, the patient returned to the emergency room with obstructive jaundice and continued right upper quadrant pain with elevated white blood cell count and liver function tests. Further tests were conducted, and the patient was found to have a long cystic duct with a low insertion variant to the common hepatic duct. An impacted stone was identified in the cystic duct, referred to as Mirizzi syndrome type I. The patient underwent endoscopic retrograde cholangiopancreatography (ERCP) several times, but the common bile duct could not be cannulated. In the last ERCP session, SpyGlass and electrohydraulic lithotripsy were performed with a balloon sweep.

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Conclusion: This case demonstrates the importance of understanding Mirizzi syndrome and cystic duct variation to achieve optimal treatment, and careful assessment and investigation are essential for proper diagnosis. In experienced hands, Mirizzi syndrome secondary to an impacted cystic duct stone can be managed successfully with ERCP, electrohydraulic lithotripsy, and SpyGlass.

Key words: Mirizzi syndrome – Cystic duct variant – ERCP – Post cholecystectomy

Cystic duct anatomic variations are encountered frequently during surgical procedures, and sufficient understanding of the extrahepatic biliary system is essential for an optimal surgical intervention.[1] The level and site of the cystic duct's insertion into the extrahepatic biliary tree vary; the duct is inserted commonly in the lateral aspect at the middle of the extrahepatic biliary tree.[2,3] Mirizzi syndrome is a rare condition characterized by a compression of the common hepatic duct by stones in the cystic duct or the gallbladder's neck.[4] The prevalence of Mirizzi syndrome is rare in Western countries, with an annual incidence of less than 1%.[5] In Riyadh, Saudi Arabia, the incidence of the syndrome was reported to be 0.7% in 2415 cholecystectomies.[6] Although surgical intervention is the common option to manage Mirizzi syndrome, new techniques of endoscopic cholangioscopy with laser lithotripsy have proven to be effective.[7,8]

A low insertion cystic duct variant has been reported in 9% to 11% of cystic duct variations.[2,3] Preoperative diagnosis is essential to avoid iatrogenic injury and other complications,[9] including misdiagnosis, bile duct formation, and stones retained after cholecystectomy.[2,10] Jung *et al*[11] reported a young patient with a low insertion cystic duct that was obstructed by a stone and caused Mirizzi syndrome. The case was diagnosed properly and managed with open cholecystectomy and surgical removal of the impacted cystic duct stone.[11]

In this report, a female patient underwent laparoscopic cholecystectomy for persistent biliary colic. A week later, the patient presented to the hospital again with obstructive jaundice, which was diagnosed as a low insertion of the cystic duct with an impacted stone that was causing common hepatic duct obstruction. The patient underwent successful endoscopic diagnosis and management of Mirizzi syndrome using cholangioscopy, SpyGlass, and electrohydraulic lithotripsy.

Case Report

A 42-year-old woman presented to the emergency room complaining of a 7-day history of persistent right upper quadrant pain that radiated to the back. The pain is exacerbated by fatty meals, relieved moderately by painkillers, and associated with bilious vomiting. The patient was vitally stable with a temperature of 36.8°C, blood pressure of 138/87, respiratory rate of 19, and SpO_2 of 98%. On examination, there was right upper quadrant tenderness with a positive Murphy sign. White blood cell count was slightly elevated (12.1×10^9/L). Liver function tests were normal (alanine transaminase, 40 units/L; total bilirubin, 6.6 μmol/L). An abdominal ultrasound was performed and confirmed multiple gallbladder stones, pericholecystic fluid, and no wall thickness, as cholelithiasis with no signs of acute cholecystitis or biliary tree dilatation. The patient had a similar history 9 years previously receiving a diagnosis of calculous cholecystitis and treated conservatively. The patient was admitted for acute care surgery service and underwent laparoscopic cholecystectomy as an emergency procedure for persistent biliary colic. The *intraoperative* finding was a chronically inflamed gallbladder with a sporadic calcified wall and wide cystic duct. The *postoperative* course progressed smoothly, and the patient was discharged after 2 days.

Eight days later, the patient returned to the emergency room with obstructive jaundice, persistent right upper quadrant pain, frequent vomiting, and dark tea-colored urine. The patient was vitally stable with no fever. White blood cell count was elevated, with 12.9×10^9/L. Alanine transaminase and total bilirubin were high (1311 U/L and 94.4 μmol/L, respectively) with mainly direct bilirubin (78.4 μmol/L). Targeted ultrasound was performed for the biliary system, which showed mild edematous changes at the surgical bed following the recent cholecystectomy, but no localized collections were found. The extrahepatic biliary tree was dilated up to 1.1 cm with echo-free lumen of its proximal part, whereas the distal part was concealed by bowel gasses. Moderate intrahepatic biliary radicles' dilation was noticed as well (Fig. 1). The patient was admitted for further investigation as a case of obstructive

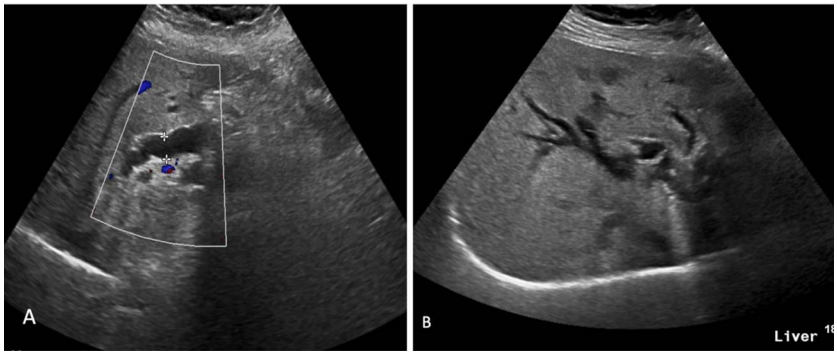

Fig. 1 Postcholecystectomy, a targeted ultrasound of the biliary system showed (A) mild edematous changes seen at the surgical bed with (B) evidence of moderate intrahepatic and extrahepatic biliary tree dilation.

jaundice. She was started on intravenous hydration and given nothing by mouth for endoscopic retrograde cholangiopancreatography (ERCP).

The patient underwent ERCP, but the common bile duct (CBD) could not be cannulated even when the needle knife-assisted technique was used. A pancreatic duct stent was placed (single tail stent 7 cm*5f) because it had been cannulated multiple times during the procedure. The second part of the duodenum and the ampulla of Vater were normal.

Magnetic resonance cholangiopancreatography (MRCP) was performed the next day to delineate the biliary tree anatomy, and the patient was found to have a long cystic duct with a low insertion variant to the common hepatic duct. The cystic duct was dilated up to 1.4 cm with multiple stones. The lowermost stone was compressing the distal extrahepatic bile duct and causing upstream dilation of both the intrahepatic and extrahepatic biliary tree, which was identified as Mirizzi syndrome type I. No evidence of biliary wall thickening or abnormal enhancement was found at the time of the exam (Figs. 2 and 3).

A second attempt at ERCP failed to cannulate the biliary tree, and the wire continued to go into the cystic duct. However, a cholangiogram showed a prominent biliary tree with multiple filling defects in the cystic duct. Limited sphincterotomy was performed (6 mm), and a double pigtail stent was inserted in the cystic duct to facilitate CBD cannulation for the next attempt.

Interventional radiology was used to perform percutaneous transhepatic cholangiogram for biliary drainage. A cholangiogram showed biliary tree dilation with severe narrowing of the CBD. Subsequently, an 8.5-Fr internal/external biliary drainage catheter was placed and connected to a draining bag (Fig. 4). The patient tolerated the procedure well and no immediate complications were encountered.

In the third attempt at ERCP, electrohydraulic lithotripsy was used with SpyGlass and balloon sweep. Multiple stones in the cystic duct were fragmented

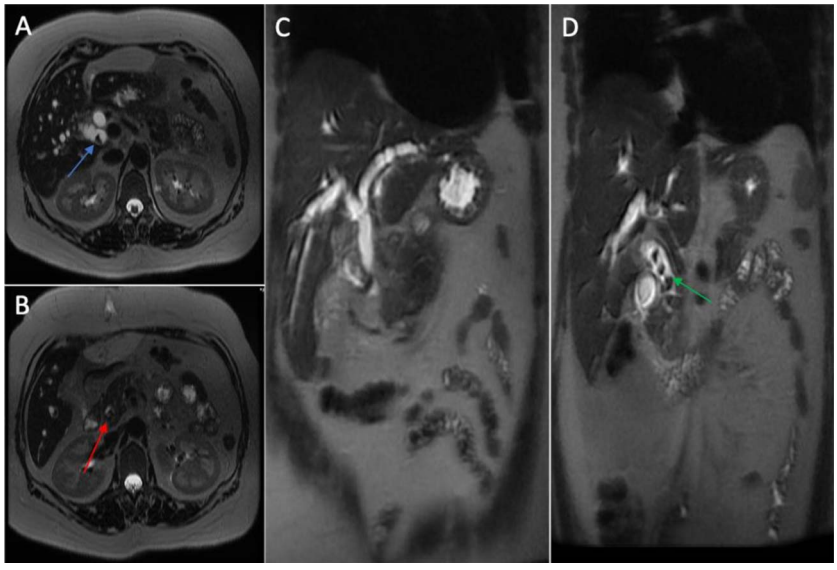

Fig. 2 MRCP images: axial (A and B) and coronal (C and D) views show a long cystic duct that has a parallel course and inserts into the intrapancreatic portion of the bile duct (distal third of extrahepatic duct) posterior-laterally (blue arrow). The cystic duct is dilated and contains multiple gallstones (green arrow), causing a compression of the extrahepatic bile duct (red arrow) and subsequent immoderate intrahepatic and extrahepatic bile duct dilation.

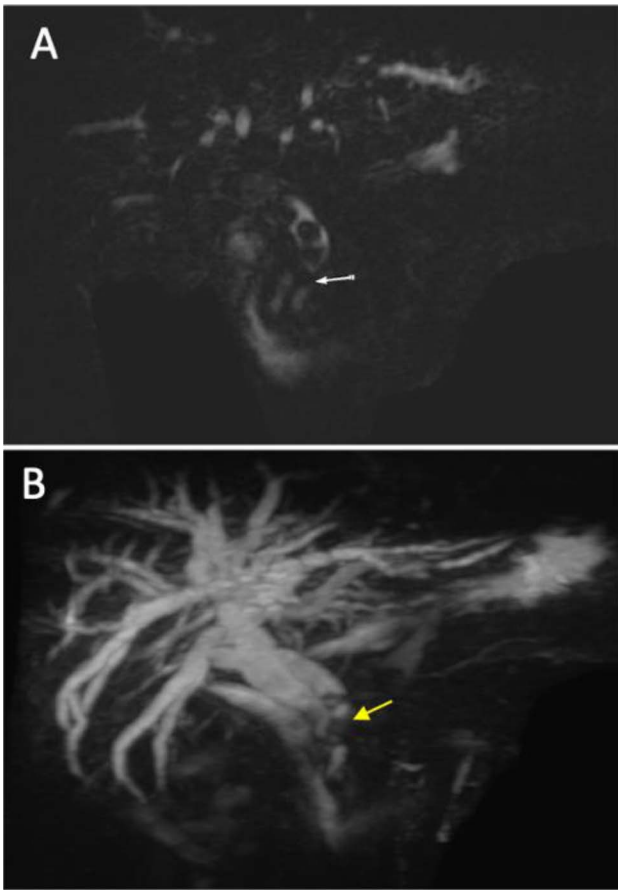

Fig. 3 (A) Coronal oblique magnetic resonance cholangiopancreatogram demonstrating dilated long parallel cystic duct measuring 1.7 cm which contains multiple intraluminal rounded filling defects that denote gallstones. The most distal stone (white arrow) exerts a mass effect and obstructs the extrahepatic bile duct for 1.3 cm with subsequent diffuse biliary tree dilation (Mirizzi syndrome). (B) Coronal MRCP 3-dimensional maximum intensity projection reconstruction image shows that the cystic duct has a parallel posterior course (yellow arrow) to the extrahepatic duct and is inserted distally 2 cm above the ampulla of Vater.

and swept out. The pancreatic and cystic ducts' stents were removed, and a double pigtail (size 8 Fr × 3 cm) plastic biliary stent (size 10 Fr × 9 cm) was placed (Fig. 5).

The percutaneous transhepatic cholangiogram catheter was removed under fluoroscopy 6 days later, and there was no immediate complication.

The patient was discharged 14 days after the second admission, coming regularly to the clinic for follow-ups. She is healthy, with no identified complications. She is also following up with the gastroenterology clinic with a plan of biliary stent removal after 6 weeks.

Discussion

Our patient, a 42-year-old woman, was found to have a long cystic duct with a low insertion variant to the CBD after an uneventful laparoscopic cholecystectomy. The lowermost stone in the cystic duct was compressing the distal extrahepatic bile duct, causing upstream dilation of both the intrahepatic and extrahepatic biliary tree, which is defined as Mirizzi syndrome.

The preoperative diagnosis of Mirizzi syndrome is very important to reduce the risk of injury to the bile duct and adjacent organs. A single-operator endoscopic cholangioscopy, combined with laser lithotripsy, is a safe alternative for the treatment of this condition, with a success rate of 90% to 100% and lower complication rates.[7]

Mirizzi syndrome is a rare complication of long-standing cholelithiasis.[4] It has 5 types according to the severity and presence of fistula. As in our case, type I Mirizzi is an external compression of the common hepatic duct without evidence of fistula.[7,12] The initial investigation for biliary disease is ultrasound, but its sensitivity to Mirizzi syndrome is very low. Another noninvasive imaging modality of choice is an MRCP, which reaches a 50% diagnostic accuracy rate.[13] However, the gold standard method to diagnose Mirizzi syndrome is ERCP, with an accuracy of 55% to 90%. Further, ERCP provides therapeutic functions, such as stone extraction and stent placement.[8,14] Cholecystectomy or subtotal cholecystectomy is sufficient to manage type I Mirizzi syndrome in most cases, although more complex operations with biliary reconstruction are needed in the other types.[15] There were no indications for MRCP or ERCP on the initial presentation of our patient. Uneventful laparoscopic cholecystectomy was performed on our patient initially for persistent biliary colic. However, 1 week after the surgery, she presented with Mirizzi syndrome secondary to an impacted cystic duct stone.

Low insertion of the cystic duct has been reported in 8% to 11% of cases in previous studies.[2,3] This cystic duct variant is reported to have a high recurrence rate of CBD stone formation.[10] Moreover, the inability to identify a low insertion of the cystic duct may result in technical difficulties during ERCP procedures and lead to devastating complications.[2] Preoperative diagnosis of a low insertion cystic duct is crucial to plan the operation and avoid iatrogenic injury to the biliary tree.[9,11]

Laparoscopic cholecystectomy was performed successfully in a 70-year-old patient who received a

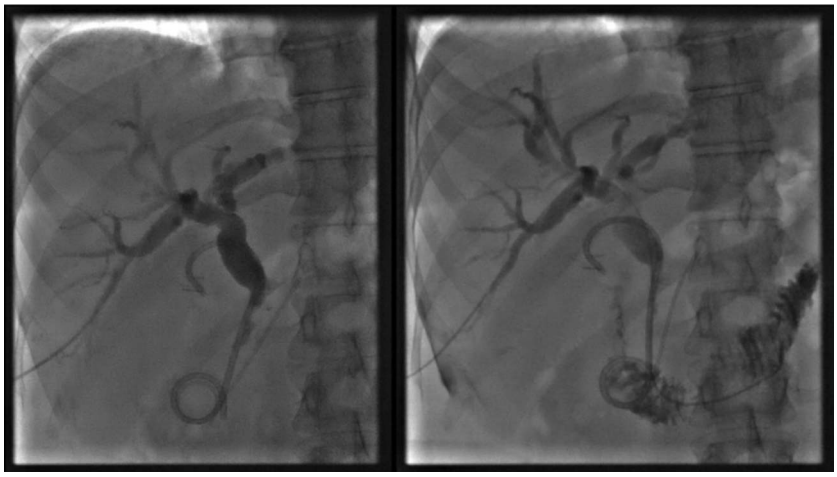

Fig. 4 Percutaneous transhepatic cholangiogram shows biliary tree dilation with severe narrowing of CBD. Subsequently, an 8.5-Fr internal/external biliary drainage catheter was placed.

preoperative diagnosis of low medial insertion of the cystic duct. An endoscopic biliary stent was placed in preparation for the surgery.[9] Another case reported a patient with a preoperative diagnosis of Mirizzi syndrome and low medial insertion of the cystic duct with an impacted cystic duct stone. Open cholecystectomy was performed and the cystic duct stone was extracted surgically after multiple failed trials.[11]

Difficulties in managing Mirizzi syndrome have prompted research into new treatment options to reduce the risk of complications.[7] SpyGlass was found to be clinically feasible because it provided adequate visualization and guided laser lithotripsy for difficult cystic duct stones successfully in type I Mirizzi syndrome.[8] In 1993, Binmoeller *et al*[16] demonstrated a 100% success rate when 14 patients with a diagnosis of Mirizzi syndrome were treated using electrohydraulic lithotripsy. In a following study, Tsuyuguchi *et al*[17] treated 23 of 25 patients with Mirizzi syndrome successfully with the endoscopic approach, achieving a 92% success rate. In a recent study, Bhandari *et al*[14] treated 34 patients with either Mirizzi syndrome or cystic duct stones successfully using SpyGlass and laser-guided lithotripsy with a 100 % success rate.

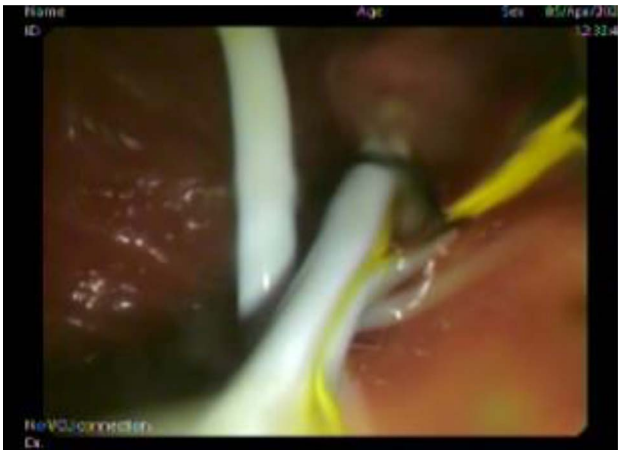

Fig. 5 Endoscopic picture of the third ERCP session: Electrohydraulic lithotripsy was used with SpyGlass and balloon sweep, and multiple stones in the cystic duct were fragmented and swept out. The pancreatic and cystic ducts stents were removed, and a double pigtail (size 8 Fr × 3 cm) plastic biliary stent (size 10 Fr × 9 cm) was placed.

Unusual complications with a rare anatomic variation may occur following uneventful laparoscopic cholecystectomy. It is crucial to evaluate the patient comprehensively and to individualize the management accordingly. In our case, the patient presented with Mirizzi syndrome secondary to an impacted stone in an unusual cystic duct variant. The patient underwent multiple trials of ERCP. In the third attempt, the impacted cystic duct stones were cleared and extracted successfully using SpyGlass and electrohydraulic lithotripsy.

Conclusion

The preoperative diagnosis of cystic duct variation is crucial in planning surgery to avoid complications. Further, the management of Mirizzi syndrome secondary to impacted cystic duct stones in a low insertion variant is challenging. Endoscopic cholangioscopy combined with SpyGlass and laser lithotripsy is an adequate and safe alternative to surgery in selected cases.

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