Mirizzi’s syndrome in a cystic duct variation

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ABSTRACT

Mirizzi’s syndrome is a rare complication of gallstone disease, that gallbladder lumen can lie alongside the common hepatic duct, resulting in inflammation and resultant bile duct stricture. Most cases are not identified preoperatively. Variations of the cystic duct and its point of union with the common hepatic duct are surgically important. Here, we present an unusual case of type I Mirizzi’s syndrome with an uncommon anomalous cystic duct that long cystic duct joins common hepatic duct behind the duodenum. Also, there is a high bilirubin level due to gallstone and bile duct stone.

Keywords: Mirizzi’s syndrome; common bile duct; cholecystobiliary fistula; cystic duct.

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Introduction

Mirizzi’s syndrome (MS) was described as a functional hepatic syndrome in 1948, which consists of the common hepatic duct secondary to compression, by an impacted gallstone in the cystic duct or in the infundibula of gallbladder (1). This is often referred to as Mirizzi’s syndrome. This process is typically subdivided into two categories.

Type I Mirizzi’s syndrome occurs when the process result in either mechanical compression of the duct or an inflammatory stricture of the common hepatic duct.

In contrast, type II consists of erosion of the stone in the duct, resulting in cholecystocholedochal fistula (2). Many cases have so far been reported, and various operations have been suggested depending on the type of MS.

Many authors have adopted laparoscopic cholecystectomy for type I MS.

However, a high rate of bile duct injury indicates that it is not a safe treatment for MS (1, 3-5). In this report, we present a case of type I Mirizzi’s syndrome with anomalous cystic duct due to biliary duct stone, with a bilirubin level highly raised.

Case Report

A 19-year female attended our hospital complaining of abdominal pain for 7 days and prolonged jaundice. At physical examination, she had icteric sclera and mild tenderness at right upper quadrant.

In laboratory data: Total bilirubin/Direct bilirubin (19.7/16.3), Alkaline Phosphatas (1900), aspartate aminotransferase (50), alanine aminotransferase (57).

Ultrasonography (US) defined one 9mm gallstone in the neck of gallbladder, dilated CBD
with diameter of 11mm, and increased diameter of intrahepatic duct.

But GB wall was normal. Endoscopic ultrasonography (EUS) revealed CBD with 11mm in diameter and much sludge and stone was seen. Thick wall GB (6mm) with sludge and sludge ball was seen.

Endoscopic retrograde cholangiopancreatography (ERCP) was conducted for an examination and removal of the CBD stone. In ERCP, CBD was dilated and had no filling defect and sphincterotomy was done and with balloon, gravel <5mm was extern, bile flow was commission.

However, bilirubin level and patient’s jaundice was not reducing in 15-20 days. In (MPCP) MR cholangiopancreatography, intrahepatic bile ducts to CHD were dilated but CBD showed severe stenosis. Gall bladder was not dilated and multiple filling defects (maybe stones) up to 8mm were seen in the cystic duct.

Also MRCP showed Mirizzi’s syndrome with low insertion of cystic duct, which was preoperatively diagnosed (Figure 1).

![Figure1. MRCP showing multiple stones in cystic duct. Biliary ducts dilatation is seen due to severe stenosis of CBD.](image)

Therefore, surgery treatment was planned. During laparotomy via a right sub costal incision, GB was thickened and collapsed and cystic duct was explored. The long and dilated cystic duct with multiple stones seen on MRCP was explored and GB did not attached to the CBD, but cystic duct was inserted to low portion of CHD behind the duodenum. After total cholecystectomy, the remnant of cystic duct was thickened and dilated and multiple stones with 5-7mm in diameter located in the cystic duct was removed by a stone forceps made. Then, intraoperative cholangiography was performed via the remnant of cystic duct with a feeding tube inserted in cystic duct. During this procedure CBD lying behind duodenum is defined and jejunum was filling with die (Figure 2). Feeding tube was placed in cystic duct and fixed, by absorbed suture.

![Figure2. Cholangiography inter operative. The Arrow is distinguished feeding tube that inserts to the remnant of cystic duct](image)

Also, a closed suction drain was placed in liver bed and both the drain and feeding tube were pulled out from right upper quadrant abdomen wall. The post-operative, minimal drainage from the closed suction drain and free drainage biliary from feeding tube was visible on the first postoperative day.

On post-operative day 7, bilirubin level was reduced to 11mg/dl, then we removed closed suction and patient was discharged.

At 2 weeks postoperative follow up visit, bilirubin level was 3mg/dl, and feeding tube was functional, but it was not discharged. After 3
months, the patient was visited, jaundice completely resolved and cholangiography via feeding tube was done. Extra and intra hepatic duct were patent and filling defect wasn’t detected and feeding tube discharged.

**Discussion**

Variation of the cystic duct and its points of union with the common hepatic duct are surgically important and 8 types variations in cystic duct described:

- **A:** Low junction between the cystic duct and common hepatic duct.
- **B:** Cystic duct adherent to the common hepatic duct.
- **C:** High junction between the cystic duct and hepatic duct.
- **D:** Cystic duct drains into the right hepatic duct.
- **E:** Long cystic duct that joins common hepatic duct behind the duodenum.
- **F:** Absence of cystic duct.
- **G:** Cystic duct crosses posterior to common hepatic duct and joins in anteriorly.
- **H:** Cystic duct courses anterior to common hepatic duct and joins it posteriorly.

In particular, low medial insertion of the cystic duct deserved special attention. This anatomical variant causes a wrong diagnosis by imaging and has a negative effect on therapeutic intervention.

Mirizzi’s syndrome is a rare disease entity that accounts for about 0.1%-0.2% of all operations performed for gallbladder stones. Eventually, the gallbladder lumen can lie alongside the common hepatic duct, resulting in inflammation and resultant bile duct stricture often referred to as Mirizzi’s syndrome. This process is typically subdivided in two categories. Type I Mirizzi’s syndrome occurs when the process results in either mechanical compression of the duct or an inflammatory stricture of the common hepatic duct. In contrast, type II consists of erosion of the stone in the duct, resulting in choledystocholedochal fistula.

Also, in 1989 the other classification of Mirizzi’s syndrome was described by Csendeset et al. (7), which includes 4 types. Type I involves external compression of the common bile duct due to stone impacted in cystic duct or Hartmann’s pouch. Lesions classified as Type II are choledystocholedochal fistula with erosion presented in less than a third of the bile duct.

Type III lesion is a fistula involving up to two-thirds of the duct circumference. Type IV lesion is a complete destruction of the bile duct. Ultrasound is the best method for diagnosis of this situation.

Also, ERCP or MRCP to confirm the diagnosis are very useful. Like ERCP, MRCP can be used in recognizing this condition but its disadvantages are inability to confirm the presence of a fistula and insertion a therapeutic stent.

The presence of Mirizzi’s syndrome obliterates the triangle of calot and makes laparoscopic cholecystectomy particularly difficult and will often require conversion to an open procedure.

The surgical technique depends on the type of MS. Most inflammatory stricture returns to normal when the inflammatory process resolves and retrograde dissection is contraindicated. This is due to risks of injury to the color’s triangles in the presence of inflammation resulting in adhesions and distorted anatomy.

Type I MS requires a partial Cholecystectomy and leave the neck of the gallbladder. Type II, partial cholecystectomy is recommended and the cuff of remaining gallbladder is used to repair the bile duct over a T. tube.

In this present case, she is a known case for Mirizzi’s syndrome combined with cystic duct anomaly. After right sub costal incision, a safe total cholecystectomy is done. The remnant of cystic duct has multiple stone with 5-7 mm diameter. We removed the stones using a stone forceps made and a feeding tube insert to cystic duct and intra operative cholangiography was performed. Extra and intra hepatic duct and CBD was patent. The feeding tube is placed within
remnant cystic duct, the other end of this, was pulled out of the abdomen wall.
In conclusion, we recommend the operator should prefer open surgery for Mirizzi’s syndrome, and dissect carefully, while considering anatomical deformities associated with chronic inflammation. Successful treatment depends on early diagnosis of this situation when the unique features of each case are considered (8).

References

1. Jung CW, Min B, Song TJ, Son GS, Lee HS, Kim SJ, et al. Mirizzi’s syndrome in anomalous cystic duct: a case report. Word J Gastroenterol 2007; 13:5527-29.
2. Zinner MJ, Stanley S, Editors. Maingot's abdominal operations. 12th ed. New York: McGraw-Hill; 2013. P.1052-53.
3. Al-Akeely MH, Alam MK, Bismar HA, Khalid K, Al–Teimi I, Al –Dossary NF. Mirrizzi syndrome: ten years experience from a teaching hospital in Riyadh Word J Surg 2005; 29:1687-92.
4. Brunicardi F, Andersen DK, Billiar TR, Editors. Schwartz’s principles of surgery. 9th ed. New York: McGraw-Hill; 2010. P.1138-39.
5. Tan KY, Chng HC, Chen CY, Tan SM, Poh BK, Hoe MN. Mirizzi syndrome: noteworthy aspects of a retrospective study in one centre. ANZ J Surg 2004; 74:833-37.
6. Shah OJ, Dar MA, Wani MA, Wani NA. Management of Mirizzi syndrome: a new surgical approach. ANZ J Surg 2001;71:423-27.
7. Csendes A, Diaz JC, Burdiles P, Malvenda F, Nava O. Mirrizzi syndrome and cholecystobiliary fistula: a unifying classification. Br J surg 1989; 76: 1139-43.
8. Safioleas M, Stamatakos M, Revenas C, Chatziconstantinou C, Safioleas C, Kostakis A. An alternative surgical approach to a difficult case of Mirizzi syndrome: a case report and review of the literature. World J Gastroenterol 2006; 12:5579-81.
9. Corlette MB, Bismuth H. Biliobiliary fistula trap in the surgery of cholelithiasis. Arch Surg 1975; 110:377-83.
10. Schafr M, Schneider R, Krahenbuhal L. Incidence and management of Mirizzi syndrome during laparoscopic cholecystectomy. Surg Endosc 2003; 17: 1186-90.