Abstract
Mirizzi syndrome is a rare complication of gallstone disease, causing major biliary problems, if the diagnosis is not made early. This condition was also reported to occur in the setting of acute acalculous cholecystitis. It was first described in 1948 by Pablo Mirizzi and presented itself in the form of unusually lodged gallstone in either the cystic duct or most frequently in Hartmann pouch of the gallbladder. We report a 50-year-old female with exquisite pain in right hypochondrium and epigastrium with nausea and vomiting for the last 20 days. She also had yellowish discoloration of urine, clay-colored stools, and tender right hypochondrium. Laboratory investigations revealed raised liver enzymes and ultrasonography (USG) was inconclusive in describing an impacted stone in the gallbladder neck. Computerized tomography (CT) scan and magnetic resonance cholangiopancreatography (MRCP) revealed it as suspected case of a mirizzi syndrome which was later confirmed by surgery. The patient was recovered after surgery without complications.

Keywords: Mirizzi syndrome; Cholelithiasis; MRCP; gallstones.

Introduction
Mirizzi syndrome is a form of obstructive jaundice occurring as an infrequent complication of gallstones [1]. This condition can be developed by impaction of gallstone in the neck of gallbladder, cystic duct and/or in the setting of acute acalculous cholecystitis [2]. This syndrome clinically presents as a lodged calculus in either the Hartmann pouch, infundibulum of the gallbladder or in its cystic duct [1]. The impaction of the gallstone and the acute obstruction with wall ischemia results in the inflammation and abscess formation [3]. The compression of the external common hepatic bile duct and its obstruction is responsible for the signs and symptoms of constant or intermittent jaundice [2]. The symptomatic gallstones are having laparoscopic cholecystectomy as their standard treatment nowadays [4]. The first-line treatment for common bile duct (CBD) stones is the endoscopic stone extraction during pre-, intra- and/or post-operative endoscopic retrograde cholangiography (ERC) or laparoscopic CBD exploration.
However, the recent data have indicated that the laparoscopic approach might be more preferable on the ground of the number of interventions, complications and cost-effectiveness. Most surgeons still consider it difficult to take out the stones in the mirizzi syndrome due to the intense fibrotic process, communication between the common hepatic duct and the gallbladder. Although the outcomes of laparoscopic and open surgery for mirizzi syndrome are not superior or inferior as compared to one another, but the former is associated with a higher conversion rate. Thus, currently the laparoscopic cholecystectomy for mirizzi syndrome is not recommended as a standard of care procedure. Non-surgical methods such as endoscopic sphincterotomy, stone extraction, and endobiliary drainage are useful methods to deliver definitive treatment of mirizzi syndrome in patients who is unfit for surgery. Furthermore, endoscopy can serve as a bridge therapy to postpone definitive treatment option as surgery to be performed in the possible best conditions to lower the postoperative complications.

**Case Report**

A 50 year- old female presented in emergency department of Capital Development Authority (CDA) Hospital, Islamabad, Pakistan, with a history of diffuse pain in right hypochondrium and epigastrium, yellow discoloration of urine, clay-colored stools for last 20 days. The pain was colicky in nature, aggravated by consuming fatty food and associated with nausea/vomiting. Examination revealed pulse 96/min, blood pressure 100/70 mmHg, temperature 98.6°F, scleral icterus, tender epigastrium and right hypochondrium. The other systems, as well as her past medical and surgical history, were unremarkable. Investigations revealed hemoglobin (Hb) 11.4gm/dl; total leukocyte count (TLC) 12700; platelet count (PLC) 380,000; total bilirubin 11.4mg/dl; direct bilirubin 8mg/dl; bilirubin ++ in urine; alanine aminotransferase (ALT) 52 IU/L; aspartate aminotransferase (AST) 54 IU/L; alkaline phosphatase (ALP) 802mg/dl; and gamma-glutamyl transferase (GGT) 53 IU/L. Ultrasound abdomen showed fatty liver, intrahepatic cholestasis, multiple tiny echogenic foci with posterior acoustic shadowing collectively measuring 43x24 mm in the gallbladder. CT scan of the abdomen with contrast showed multiple stones in the gallbladder (Figure 1).

MRCP showed gallbladder is filled with multiple small calculi, the average size of each calculus is approx 3mm. The gallbladder neck is producing external compression on the common hepatic duct. Intrahepatic biliary ducts are mildly prominent. Right, and left hepatic ducts are mildly prominent. There is evidence of choledocholithiasis, the CBD is of normal caliber measures 5 mm in diameter and normal pancreatic ducts. The diagnosis on the basis of MRCP report is cholelithiasis associated with mirrizi syndrome (Figure 2).
The patient underwent surgery, in accordance with the guidelines, which led to the confirmation of the diagnosis: a stone eroding from the gallbladder neck into the common hepatic duct (type II). The pus had collected in the gallbladder and the surrounding tissues were acutely inflamed. Distal CBD was found to be patent. Subtotal cholecystectomy was performed on the patient. Histopathological report of removed gallbladder showed no evidence of malignancy. Within one month, the patient had a complete recovery without any complication.

**Discussion and Literature Review**

Mirizzi syndrome is a chronic complication of symptomatic gallstone disease. This complication of chronic gallstone disease is so rare in western developed countries with an incidence of less than 1 percent a year that this syndrome would be encountered by an average general physician only a few times during his career. Since there are no pathognomonic features in either the history or the physical examination of the patient with mirizzi syndrome, it's clinical diagnosis is difficult. No diagnostic method or clinical feature has 100 percent sensitivity and specificity. The preoperative diagnosis of mirizzi syndrome can be made in only 8% to 62.5% of patients. If the preoperative diagnosis is not made, it is essential to go for intraoperative recognition and proper management.

The reported diagnostic accuracy for ultrasonography in the mirizzi syndrome is 29% [8], with a reported sensitivity varying from 8.3% to 27% [7, 9]. On ultrasonography, it presents as a contracted gallbladder with thick or extremely thin walls. The radiologist may appreciate one large gallstone or multiple smaller gallstones impacted in the infundibulum [10]. There would be dilatation of the hepatic duct in its extra and intrahepatic portions above the obstruction level, while the common bile duct would be within normal size under the level of obstruction [1].

Abdominal CT scan can identify, measure the wall thickness of gallbladder and the bile duct dilatation in the case of mirizzi syndrome. However, the most useful tool to demonstrate extrinsic compression of the bile duct in case of this unusual complication of gallstones is magnetic resonance cholangiopancreatography (MRCP) [1]. The diagnostic accuracy for MRCP is 50% [8]. It also helps to determine whether a fistula is present or not. Furthermore, MRCP helps to rule out choledocholithiasis and other causes of bile tract obstruction. MRCP helps to reveal some typical features of Mirizzi syndrome such as the extrinsic narrowing of the common hepatic duct, a gallstone in the cystic duct, dilatation of the intrahepatic and common hepatic ducts, and a normal choledochus [1]. Diagnosis using MRCP avoids the complications associated with endoscopic retrograde cholangiopancreatography (ERCP), in addition, it can also show the extent of the inflammatory process surrounding the gallbladder clearly without any invasive procedure [11].

The ERCP is an invasive procedure which helps to diagnose miririzi syndrome, allows stone retrieval and stent placement but multiple complications are associated with it [1]. Diagnostic accuracy of this procedure is around 55 to 90% [8], with a failure rate ranging from 5% to 10%. The ERCP findings of mirizzi syndrome include a narrowing or curvilinear extrinsic compression involving the lateral portion of the distal common hepatic duct with proximal ductal dilatation and normal distal caliber [1].

Over 50% of patients with mirizzi syndrome are diagnosed during surgery [8]. The surgeon appreciates the presence of a shrunken gallbladder with distorted anatomy. The gallbladder may get dilated with thick walls in case of a large stone, or multiple gallstones, impacted at the neck of the gallbladder or its infundibulum. An obliterated calot’s triangle or a dense fibrotic mass at the calot’s triangle may be observed. Dense adhesions at the subhepatic space may also be observed [1]. Intraoperative cholangiography could be useful [11] but may be difficult to perform. Intraoperative ultrasonography is a useful procedure to assess the
anatomy of the biliary tree and guide the accurate dissection of bile duct in the inflamed area.\textsuperscript{11,12}

In our case at initial presentation, despite the absence of clinical and laboratory features of acute inflammation, the gallbladder was grossly inflamed and contained pus. The pericholeductal tissues were also acutely inflamed. The presence of this inflammation made it difficult to explore CBD. So, surgery got confined to partial cholecystectomy.

**Conclusion**

In our patient, the diagnosis was made on USG, CT scan, MRCP and was confirmed during surgery. Mirizzi syndrome is a rare disease and clinicians should be aware of the presentation of this syndrome to prevent life-threatening complications. The patient recovered completely without any complications after surgery.

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**References**


