Mirizzi Syndrome: A Case Report

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Mirizzi syndrome is a seldom-observed complication of gallstone disease, which if not diagnosed hitherto, may cause major biliary problems. It has a prevalence ranging from 0.05% to 2.7% among patients with cholelithiasis and is characterized by mechanical obstruction of the common hepatic duct caused by impaction of stones in the neck of the gallbladder (most frequently in the Hartmann pouch) or cystic duct and presents clinically as intermittent or persistent jaundice. This syndrome was described, in 1948, by an Argentinean surgeon Mirizzi. Here, we report the case of a 50-year-old female patient, who presented with acute right upper abdominal pain, predominantly in the epigastria and right subcostal region, which was colicky in nature, nausea, and high-grade fever since 1 day with clinically unremarkable abdominal findings. Abdominal ultrasonography revealed a partially distended gallbladder showing the presence of an intraluminal calculus measuring 1.6 cm x 1.3 cm, impacted at the neck region. Further investigations resulted in the diagnosis of Mirizzi syndrome.

Keywords: Cholelithiasis, Common hepatic duct, Gallstone, Jaundice, Mirizzi syndrome

INTRODUCTION

In 1948, the Argentinean surgeon Mirizzi described a case with partial obstruction of the common hepatic duct secondary to an impacted biliary calculus in the cystic duct, associated with an inflammatory response involving the cystic duct and the common hepatic duct. This presentation became known as Mirizzi syndrome.¹ Mirizzi syndrome is a seldom-observed complication of gallstone disease, which if not diagnosed hitherto, may cause major biliary problems. It is characterized by mechanical obstruction of the common hepatic duct caused by impaction of stones in the neck of the gallbladder (most frequently in the Hartmann pouch) or cystic duct and presents clinically as intermittent or persistent jaundice.¹

Mirizzi syndrome has a prevalence ranging from 0.05% to 2.7% among patients with cholelithiasis.²³ Mirizzi syndrome and other chronic complications of gallstone disease are rare in Western developed countries with an incidence of <1% a year.⁴ Mirizzi syndrome is, however, a more common condition in underdeveloped countries, with an incidence reportedly ranging from 4.7% to 5.7%.⁵⁶

CASE REPORT

A 50-year-old female patient presented to the Surgical OPD with upper right abdominal pain, predominantly in epigastria and right subcostal region, which was colicky in nature, nausea, and high-grade fever (38.5°C) since the previous night. The patient also gave a history of burning chest pain and bloating sensation in abdomen since the past 8-10 days. The patient gave a history of having experienced similar symptoms in the past.

There was no history of vomiting, excessively yellow-colored urine or pale stools.

The patient had no history of diabetes mellitus, hypertension, bronchial asthma, or tuberculosis.

The patient gave a history of normal sleep, appetite, bowel, and bladder habits and gave no history of any vices.

On examination, the patient was febrile, with a pulse rate of 110/min, blood pressure of 140/90 mm of Hg, mild icterus. There was no pallor icterus, cyanosis, clubbing, lymphadenopathy, or edema.

On per abdominal examination, there were no dilated veins/sinuses, no scars, the abdominal girth was 71 cm, the abdomen was soft, and there was no guarding, rigidity, or tenderness.
Abdominal ultrasonography was carried out which revealed a partially distended gallbladder. A follow-up scan was advised after 6 h NBM for gallbladder evaluation. The follow-up scan revealed a partially distended gallbladder showing the presence of an intraluminal calculus measuring 1.6 cm × 1.3 cm, impacted at the neck region. The walls of the gallbladder appeared edematous, and there was minimal pericholecystic collection noted. The proximal common bile duct (CBD) was dilated, measuring 1.2 cm in diameter. A solitary intraluminal calculus measuring 1.1 cm × 0.6 cm was noted in the proximal CBD causing dilatation of the proximal biliary tree. In addition, a solitary, simple cyst measuring 1.2 cm × 1.2 cm was noted in segment V of the right lobe of the liver, abutting the fundus of the gallbladder. A computed tomography scan of the abdomen and pelvis was advised for further evaluation.

A blood workup was carried out which included a hemogram and erythrocyte sedimentation rate (ESR), liver function tests, prothrombin time (PT), and activated partial thromboplastin time (aPTT). The hemogram revealed a hemoglobin level of 12.2 g/dL, total leukocyte count of 21,100 cells/mm³ (neutrophils - 91%), platelet count of 2.7 lacs/mm³, and ESR of 18 mm/1 h. The liver function tests revealed serum bilirubin (total) - 1.3 mg/dL, serum bilirubin (conjugated) - 0.8 mg/dL, serum glutamate oxaloacetate transaminase - 720 IU/L, serum glutamic pyruvic transaminase - 746 IU/L, serum alkaline phosphatase - 400 U/L, serum proteins (total) - 7.4 gm/dL, and serum albumin - 3.8 gm/dL. PT and aPTT were within normal limits.

Endoscopic retrograde cholangiopancreatography (ERCP) was conducted under total intravenous anesthesia, which revealed normal papilla. The CBD was selectively cannulated, and cholangiogram revealed dilated CBD with large filling defects at the cystic duct junction suggestive of stones; highly suggestive of Mirizzi syndrome. In view of cholangitis and purulent bile, stone retrieval was deferred. A 10F biliary stent was inserted, and free bile and pus drainage were noted (impression - choledocholithiasis, cholangitis, biliary sphincterotomy, and biliary stenting done) (Figure 1). Since our patient presented with Type II Mirizzi syndrome, a partial cholecystectomy was carried out. Intraoperatively, a fibrotic gallbladder with frozen Calot’s triangle with a short cystic duct with thickening of the gallbladder wall and thick adhesions surrounding the gallbladder were revealed. The patient was kept under observation postoperatively for 4 days and was kept on intravenous antibiotics. The patients’ condition on discharge was satisfactory, and the patient came for follow-up 2 weekly as advised, and a repeat ERCP was advised after 1 month.

DISCUSSION

Mirizzi syndrome is a rare complication of prolonged cholelithiasis, characterized by narrowing of the common hepatic duct due to mechanical compression and/or inflammation due to biliary calculus impacted in the infundibula of the gallbladder or the cystic duct. Presentation of this condition may vary from extrinsic compression of the common hepatic duct to the presence of cholecystobiliary fistula and thus represents a precarious alteration in the anatomy during the performance of cholecystectomy, by predisposing to an inadvertent lesion of the common hepatic duct resulting in iatrogenic CBD damage. Further, the presence of an intense fibrotic process and an eventual communication between gallbladder and common hepatic duct is what poses a difficulty in the surgical management of the disease.7-9

In 1989, a new classification of patients with Mirizzi syndrome and cholecystobiliary fistula was presented. It includes four types:

(1) Type I - No fistula found - Type IA - Presence of the cystic duct - Type IB - Obliteration of the cystic duct and
(2) Type II-IV - Fistula present - Type II - Narrowing smaller than 33% of coronary heart disease (CHD) diameter - Type III - Narrowing between 33% and 66% of CHD diameter - Type IV - Narrowing larger than 66% of the CHD diameter.10

Clinical presentation includes symptoms of obstructive jaundice: Recurrent cholangitis, jaundice, fever, right upper quadrant pain, and elevated liver enzymes.

Diagnosis is made by physical examination, imaging studies such as abdominal ultrasonography, ERCP, Magnetic resonance cholangiopancreatography (MRCP), and liver functional tests.
Differential diagnosis includes choledocholithiasis, Caroli’s disease, hilar cholangiocarcinoma, pancreatic head cancer.

Treatment of Mirizzi syndrome is challenging for a surgeon, and operative methods depend on the type of pathology. Management is by determining the type and best surgical procedure at the time of laparotomy. In Type I, simple cholecystectomy is the method of choice. If CHD wall inflammatory changes are found, T-tube placement is recommended to avoid disruption, leaks, and stricture formation. Type II, III, and IV patients require complex management. Total isolation of inflamed segment with Roux-en-Y hepaticojejunostomy may have the best long-term outcome.10

CONCLUSION

In conclusion, Mirizzi syndrome is a rare pathological condition that cannot be diagnosed during a physical examination. It requires imaging study for diagnosis. An awarded suspicion is necessary to avoid lesions of the biliary tree as the problem may only become evident during surgery due to firm adhesions around Calot’s triangle. The success of the treatment is related to a precocious recognition of the condition, cautiousness at the time of surgery and adapting the management based on the individual characteristics of each case.

REFERENCES


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