Mirizzi syndrome, an uncommon complication of cholelithiasis

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Abstract
We hereby describe a case of 38 years old young man, known diabetic and alcoholic, presenting with history of pain in right hypochondriam, fever and progressive jaundice. His investigations revealed marked derangement of liver functions with predominant conjugated hyperbilirubinemia and raised alkaline phosphatase suggestive of obstructive jaundice. Subsequent investigation by ultrasound and magnetic resonance cholangiopancreatography (MRCP), revealed a large stone in the cystic duct compressing the common bile duct with intrahepatic biliary radical dilatation, suggesting the rare diagnosis of Mirizzi Syndrome. In the present report the authors will discuss the importance of recognizing this relatively rare entity, its classification and management related issues.

Keywords: Obstructive Jaundice, Hyperbilirubinemia, Mirizzi syndrome, Gallstone.

Introduction
Mirizzi syndrome is a rare complication of gall stone disease in which there is compression of common hepatic duct by the stone present in the neck of gall bladder or cystic duct, with an estimated incidence of 0.05- 2.7%.[(1)] The presentation of this syndrome can be diverse. It can manifest as obstructive jaundice, bile duct obstruction or biliary fistula. Diagnosis is difficult initially and one should have a very high index of suspicion for the disease but advances in imaging in the form of endoscopic retrograde cholangiopancreatography (ERCP) and MRCP results in precise diagnosis of this condition. We are reporting a rare case of Mirizzi syndrome presenting as obstructive jaundice.

Case Report
A 38 years old male presented with history of pain in right hypochondriam of eight days duration, which was mild and dull aching in nature initially, but later increased in intensity along with intermittent radiation to back. Abdominal pain was associated with nausea, bloating sensation in abdomen and vomiting. After two days patient developed intermittent high grade fever (103°-104° F) associated with chills and rigors which was relieved by antipyretics. Later patient developed yellow discoloration of urine and sclera which was gradually increased. There was no history of hematemesis, malena, bleeding from any site, loose stools, constipation and distention of abdomen. There was no history of genitourinary symptoms, cough and chest pain. Patient was a known case of diabetes type 2 for ten years on irregular treatment and also had daily alcohol consumption of 200 ml of whisky for 8 years. There was no significant past history of any chronic illness.

On clinical examination, icterus was present but there was no pallor, cyanosis, clubbing and pedal edema. His pulse was 100 per minute, blood pressure, 130/80 mm Hg and temperature 103°F. On systemic examination abdomen was soft with marked tenderness of right hypochondriam, and a tender palpable lump below coastal margin. Murphy's sign was positive. There was no free fluid in the abdomen. Rest of the systemic examination was unremarkable.

Laboratory examination demonstrated a high leucocyte counts (12000 cell per cumm) with predominance of neutrophils (82 %), liver function test showed raised total bilirubin (7.05 mg/dL), mildly increased transaminases (serum glutamic oxaloacetic transaminase 146 U/L, serum glutamic pyruvate transaminase 294 U/L) and marked increase in alkaline phosphatase (849 U/L) and γ-glutamyltransferase (748 U/L) with a normal serum protein levels. (Table 1)

Table 1: Showing the laboratory variables of patient during hospital stay

Variables	Day 1	Day 3	Day 6
Hemoglobin (gm %)	14	13.6	13.8
Total Leucocyte Count (cell/ cumm)	12000	11600	7600
Differential Leucocyte Count (%)	$N_{82}\,L_{14}M_2\,E_2$	$N_{78}\,L_{20}\,M_0\,E_2$	$N_{65}\,L_{27}\,M_5\,E_3$
Platelet count cell (cell/ cumm)	2.3 Lac		
ESR (mm FHR)	35		
Blood sugar Random (mg %)	145	150	148

Blood Urea (mg %)	28		24
Serum creatinine (mg %)	0.8		0.6
Na+ (Meq/L) K+ (Meq/L)	140.4 3.81	139.9 3.71	
Serum Bilirubin (mg %) (Total) (Direct)	3.36 3.33	4.18 2.66	7.04 3.95
Serum Proteins (gm %) (Total) Albumin Globulin	5.91 2.49 3.42	6.7 3.6 3.1	
SGOT (AST) (U/L) SGPT (ALT) (U/L)	146 294	120 229	149 184
GGTP	748	932	
Alkaline Phosphatase (IU/L)	129	849	
HBsAg/ Anti HCV Antibody	Negative		

Ultrasound imaging demonstrated distended gall bladder along with neck and cystic duct dilatation (Fig. 1). A 1.07 cm calculus was seen in the cystic duct which is compressing the, common bile duct suggesting the diagnosis of Mirizzi syndrome.

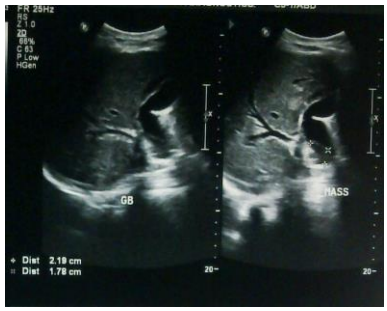

Fig. 1: Ultrasound showing calculus in cystic duct compressing the common bile duct and distention of gall bladder along with neck & cystic duct.

Magnetic resonance cholangiopancreatography (MRCP) confirmed the presence of calculus in cystic duct, causing compression over the proximal common bile duct (CBD) leading to intra hepatic biliary dilatation (IHBD) in both lobes of liver (Fig. 2 and 3).

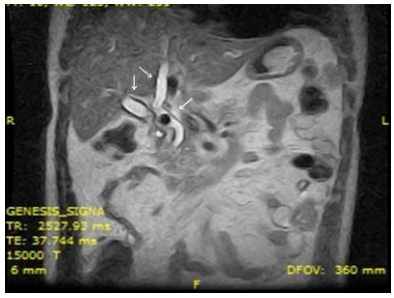

Fig. 2: In saggital view MRCP image showing calculus in cystic duct causing compression over proximal CBD leading to IHBD in both lobes of liver.

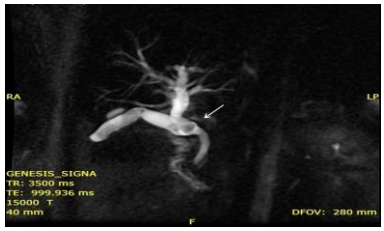

Fig. 3: MRCP image showing calculus in cystic duct causing compression over proximal CBD leading to dilatation of intra hepatic biliary radicals.

The patient was managed with antibiotics, intravenous fluids, and regular insulin. He improved symptomatically and was later planned for endoscopic retrograde cholangiopancreatography (ERCP) with stenting followed by elective cholecystectomy for which patient was guided to super-specialty centre.

Discussion

Mirizzi syndrome is a rare complication of gallstone disease, first described in 1948 by Argentinean Surgeon Pablo Luis Mirizzi, as a syndrome of common hepatic duct obstruction in the setting of longstanding cholelithiasis and cholecystitis, causing inflammation and extrinsic compression of the main bile duct leading to obstructive jaundice and cholangitis.[2] Usually the patients of Mirizzi syndrome present with recurrent bouts of cholangitis presenting as pain, fever and jaundice.

Classification of Mirizzi syndrome has evolved over many years, based initially on diagnostic methods and surgical approach and later-on complications. McSherry et al classified Mirizzi syndrome in two types based on ERCP findings.[3] Type 1 was characterized by extrinsic compression of the common hepatic duct and type 2 by an erosion of common hepatic duct wall secondary to the gall stone leading to the formation of a cholecystobiliary fistula. To clarify the definition of cholecystobiliary fistulae and to guide an appropriate surgical management, Csendes et al classified Mirizzi syndrome in four types.[4] According to this classification, in type 1 there is an external compression

of the CBD due to a stone impacted at the neck of the gallbladder or at the cystic duct (commonest). In type 2 there is the presence of a cholecystobiliary fistula (cholecysto-hepatic or cholecysto-choledochal) due to erosion of the anterior or lateral wall of the common hepatic duct (CHD) or common bile duct (CBD), respectively, with the fistula involving less than one-third of the circumference of the CBD. In type 3 there is presence of a cholecystobiliary fistula with erosion of the wall of the CBD involving up to two-thirds of its circumference and in type 4, there is a presence of a cholecystobiliary fistula with destruction of the entire CBD wall. Later Beltran et al suggested that Mirizzi syndrome, apart from producing cholecystobiliary fistula, may further complicate into a superimposed cholecysto-enteric fistula.[5] Therefore, a modified classification was proposed i.e. aforementioned types plus a cholecysto-enteric fistula without gallstone ileus (type 5a), or with gallstone ileus (type 5b). Mirizzi's syndrome is rare, occurring in about 1% of all patients who undergo cholecystectomy.[6]

Transabdominal ultrasound and CT are often the initial imaging studies performed in suspected cases of Mirizzi syndrome but these studies are limited by regional inflammation and bowel gas.[7,8] Direct cholangiography during ERCP or via the percutaneous transhepatic route is the diagnostic method of choice. A distended gallbladder, extrinsic compression of the main bile duct, and a dilated intrahepatic biliary tree are seen in type I Mirizzi syndrome. A fistulous tract between the gallbladder and bile duct may be seen in type II Mirizzi syndrome.[9] A stone that is fixed in the region of the cystic duct or gallbladder should also raise the possibility of this diagnosis. MRCP is an alternative noninvasive imaging modality that can be utilized to diagnose Mirizzi syndrome.[10] Treatment of Mirizzi syndrome has traditionally been surgical. Endoscopic management consists of drainage and decompression of the bile duct and/or gallbladder.[11,12] This is essential in relieving jaundice and treating cholangitis before surgical intervention. Management of type I Mirizzi's syndrome includes cholecystectomy with or without bile duct exploration. In the presence of severe inflammation, in which identification of the anatomy is difficult, partial cholecystectomy with postoperative endoscopic sphincterotomy to ensure clearance of bile duct stones is preferable. Management of type II Mirizzi's syndrome is best accomplished by partial cholecystectomy and cholecystocholedocho-duodenostomy.[6] A systemic review of Mirizzi syndrome by Uppara M. et al has discussed the various aspects of surgical management of Mirizzi syndrome and found that laparoscopic procedures had more morbidity (40%) that open approach (16%) and combined approach had even less morbidity (9.26%).[13]

The present case fits into Mirizzi syndrome type one in which there was stone in the cystic duct and distended gall bladder with extrinsic compression on CBD with IHBR dilatation with cholangitis and associated diabetes mellitus. The patient referred to higher center for operative intervention.

Conclusion

Mirizzi syndrome is a rare complication of gall stone. The diagnosis is suspected on ultrasound and confirmed on ERCP/MRCP imaging. The sub categorization is very important for management protocols. Although there is no association of Mirizzi syndrome with diabetes mellitus we postulate that the presence of diabetes may add to complication of Mirizzi syndrome.

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