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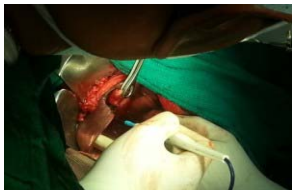
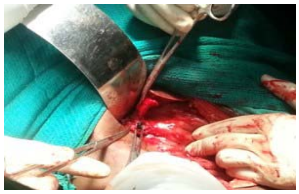
MIRIZZI SYNDROME - A CASE REPORT

BIRAVINTH SOLOMON F
Department of General Surgery, MADURAI MEDICAL COLLEGE AND HOSPITAL

Abstract : The Mirizzi syndrome is an unusual presentation of gallstones which occurs when a gallstone becomes impacted in either Hartmann's pouch of the gallbladder or the cystic duct, causing obstruction of the common hepatic duct by extrinsic compression. The diagnosis of this syndrome is of importance because surgery in its presence is associated with an increased incidence of bile duct injury.
Keyword :MIRIZZI SYNDROME

CASE REPORT :

A 40 year old male patient admitted with complaints of passing yellow coloured urine, clay coloured stools, and itching for past two months. No history of fever, vomiting, melena, altered bowel habit, urinary tract infection or abdominal pain. On examination patient is icteric with no palpable mass and digital rectal examination shows roomy rectum. Ultrasound of abdomen showed gall bladder wall thickening with stone in the biliary tree and fistulous tract lying between gallbladder and biliary tree. liver function test alered with elevated total bilirubin of 8.5 mg/dl and there is prolonged prothrombin time. Complete hemogram done and since Jaundice persists we have done ERCP guided stenting of CBD . After that patient is observed for a while. But jaundice still persists and we decided of doing Laparotomy and CBD exploration. After adequate preoperative preparations laparotomy was done which reveals gall bladder found adherent to the liver bed. Adhesiolysis done. Moreover kocherisation of duodenum done and fistulous tract found between fundus of gallbladder and biliary tract. Stone of 1 x 1 cm found at the confluence of right and left hepatic duct along with debris. Retroduodenal portion of CBD found to be normal. Gall bladder opened in the fundus and bisected upto the fistulous junction. CBD exploration done. Stone and debris retrieved through the fistulous opening. Infant feeding tube introduced through the fistulous opening and flushed with normal saline. Patency of CBD confirmed. Subtotal cholecystectomy done. ERCP stent retained in situ. Choledochoplasty done with the remnant gall bladder with interrupted 3-0-vicryl sutures and drain kept over the hepatorenal pouch His postoperative period found to be uneventful. liver function test returns to normal level within the same week. sips of fluid started on sixth day and sutures and drain are removed on tenth postoperative day. The patient is in regular follow-up for six months and is perfectly alright now.

DISCUSSION:

It is a benign condition resulting from a chronically impacted stone or stones in the neck of gall bladder or cystic duct which over time induces sufficient pericholecystic inflammation to narrow and obstruct the adjacent common hepatic duct. Mirrizi was the first to describe this phenomenon as functional hepatic syndrome in 1948.

PATHOPHYSIOLOGY

The current concept of Mirizzi syndrome, includes the external compression of the bile duct and the later development of cholecystobiliary and cholecystoenteric fistulas as different stages of the same disease process[4,7,8]. Mirizzi syndrome can be caused by an acute or chronic inflammatory condition secondary to a single large gallstone or multiple small gallstones impacted in the Hartmann's pouch or in the gallbladder infundibulum and cystic duct[1,4,7,8,14,25-37].].

SYMPTOMATOLOGY AND OTHER CLINICAL MANIFESTATIONS

Patients with Mirizzi syndrome present with a mean age varying from 53 to 70 years, and are female with a frequency of around 70% of all cases[2,4,5,7,25,26,28,32,36]. The most common form of clinical presentation of Mirizzi syndrome is obstructive jaundice (60%- 100%), accompanied by abdominal pain over the right upper abdominal quadrant (50%-100%), and fever in the context of a patient with known or suspected gallstone disease [1,14,26-33,35-37]. A previous recent history of jaundice can sometimes be elicited. Frequently, patients with Mirizzi syndrome

present in the setting of acute cholecystitis, acute cholangitis or acute pancreatitis[1,2,27, 28,33,35]. Recently, Mirizzi syndrome in the setting of gallstone ileus has been described and validated as another clinical presentation that surgeons must bear in mind[4,8]. The most common laboratory finding present in these patients is hyperbilirubinemia. ,are elevated aminotransaminase levels and leukocytosis, frequently seen when acute cholecystitis, cholangitis or pancreatitis is present[1,25,26,28,29,33,35]. Recently, extremely high levels of the malignancy marker, CA19-9, have been consistently found in patients with Mirizzi syndrome type II or higher [38- 40]. The differential diagnosis of Mirizzi syndrome includes any other benign or malignant cause of obstructive jaundice, such as gallbladder cancer, cholangiocarcinoma, pancreatic cancer, sclerosing cholangitis, metastatic disease, and others [1,27,31,37]. Csendes classification includes:

- Mirrizi type 1 is the extrinsic compression of CBD by an impacted gall stone.
- Mirrizi type 2 consists of a cholecysto-biliary fistula involving one third of the circumference of bile duct.
- Mirrizi type 3 where cholecysto-biliary fistula comprises upto two-thirds of circumference of bile duct.
- Mirrizi type 4 where cholecysto-biliary fistula has destroyed the bile duct wall and compresses the whole circumference of bile duct.
- Mirrizi type 5 corresponds to any type of Mirrizi associated with a bilio-enteric fistula with or without gall stone ileus.

DIAGNOSIS

Preoperative diagnosis of Mirizzi syndrome followed by thoughtful surgical planning is of utmost importance[6]. The incidence of bile duct injuries in patients operate d on with Mirizzi syndrome without preoperative diagnosis could be as high as 17%[14]. Preoperative diagnosis of Mirizzi syndrome is difficult and can be made in only 8% to 62.5% of patients[26,29,37,41].

Ultrasonography

A contracted gallbladder with thick or extremely thin walls with one large gallstone or multiple smaller gallstones impacted in the infundibulum may be appreciated[32]. The hepatic duct would be dilated in its extra and intrahepatic portions above the level of the obstruction site, and the common bile duct would be within normal size under the level of obstruction[1,14,28,32]. The reported diagnostic accuracy for ultrasonography in Mirizzi syndrome is 29% [29], with a reported sensitivity varying from 8.3% to 27%[33,41].

Computed tomography

The main utility of CT would be the exclusion of malignancy in the porta hepatis area or in the liver[1,14,32].

Magnetic resonance

Magnetic resonance imaging can also show the extent of the inflammatory process surrounding the gallbladder and has the advantage of avoiding the complications associated with endoscopic cholangiography[14]. However, the diagnostic accuracy for MRCP is 50%[29].

Endoscopic retrograde cholangiopancreatography

Endoscopic retrograde cholangiopancreatography (ERCP) is an invasive procedure not only useful to confirm the presence of Mirizzi syndrome with or without cholecystobiliary or cholecystoenteric fistulae, but also for therapeutic means allowing stone retrieval, stent placement and other procedures[1,14,28,41

Intraoperative diagnosis

Over 50% of patients with Mirizzi syndrome are diagnosed during surgery[29]. Surgical characteristics include the presence of a shrunken gallbladder with distorted anatomy or a dilated gallbladder with thick walls and a large stone, or multiple gallstones, impacted at the gallbladder neck or infundibulum, an obliterated Calot's triangle, a dense fibrotic mass at the Calot's triangle, and dense adhesions at the subhepatic space. Intraoperative cholangiography could be useful and help to confirm the diagnosis, determine the location and size of the fistula, detect bile duct stones, and detect whether there is any loss of integrity of the bile duct wall [14

TREATMENT

The treatment of Mirizzi syndrome is surgical. Mirizzi syndrome is important to surgeons because preoperative diagnosis is not always possible and because the surgical treatment of this condition is associated with a significantly increased risk of bile duct injury[48-56]. Besides, the severe inflammatory process with thick dense hard adhesions and associated edematous tissues distort the anatomy[37,48,51,53,55]. Additionally, the presence of a cholecystobiliary fistula further increases the risk of biliary duct injury[48,52,53]. During the operation, dissection of the Calot's triangle may lead to bile duct injury or excessive bleeding and other morbidity such as sepsis, delayed bile duct stricture, and secondary biliary cirrhosis [14,26,28,48,53]. Subtotal cholecystectomy may be the best treatment for Mirizzi typeIand most cases of Mirizzi type II and III[48-57]. Subtotal cholecystectomy was described in 1985 by Recently, subtotal cholecystectomy has been described in laparoscopic cholecystectomy, following the same technical details described for the open technique[59].

The reflux of bile indicates the presence of a fistula between the gallbladder and the bile duct, because the cystic duct is usually occluded[48-55]. If no fistula is macroscopically evident or diagnosed by intraoperative cholangiography, a partial cholecystectomy leaving the gallbladder neck or infundibulum is performed and the gallbladder stump is closed with absorbable monofilament sutures over the bile duct. If a fistula is present (Mirizzi III and IV), besides partial cholecystectomy, a biliary-enteric anastomosis could sometimes be performed between the duodenum and bile duct or between the bile duct and a loop of jejunum *en-Y-de-Roux*[7,14,29,33,37,48,51,55]. The bile duct should be explored for the presence of stones through a different incision over the common hepatic duct or the choledochus, because common bile duct stones are a common occurrence in Mirizzi syndrome and have been found in 25% to 40% of cases[26,53].

The common bile duct suture must be protected with a Kehr tube placed through a different hepaticotomy or choledochotomy proximal or preferable distal to the fistula[7,28,30,33,37,41,52]. Operative cholangiography through the Kehr tube should be performed in all patients with Mirizzi syndrome before concluding the surgical procedure[26-33,51,53].

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