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Original Research Paper

MIRIZZI SYNDROME : A CASE REPORT AND REVIEW OF LITERATURE

Medicine

KEYWORDS: - obstructive jaundice, ERCP, open cholecystectomy

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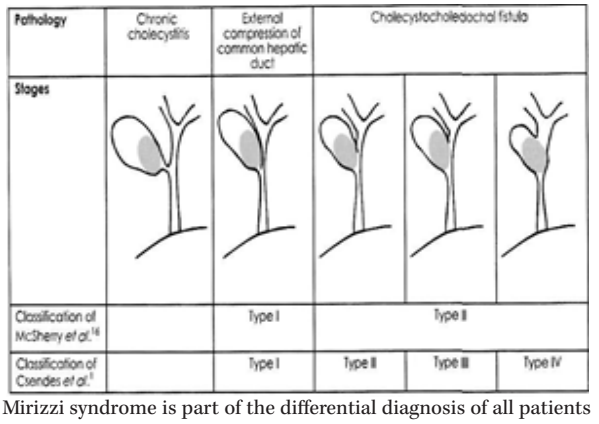
ABSTRACT

Mirizzi syndrome is a rare cause of obstructive jaundice. This entity should be considered in the differential diagnosis of all patients with obstructive jaundice. Failure to recognize the condition preoperatively can result in a major bile duct injury, particularly during laparoscopic surgery. Here we are presenting a case of 70y male patient came with complaining of severe epigastric pain with yellowish discoloration of whole body since 1 month. We performed open cholecystectomy, 1 month after ERCP, in this patient uneventfully. Pt was discharged on post-operative day 5. In conclusion, clinicians need to diagnose and evaluate carefully in cases of obstructive jaundice and adopt an optimal strategy to avoid the complication associated with this disease.

INTRODUCTION

Mirizzi syndrome is a rare cause of obstructive jaundice. This entity should be considered in the differential diagnosis of all patients with obstructive jaundice. Failure to recognize the condition preoperatively can result in a major bile duct injury, particularly during laparoscopic surgery. The syndrome refers to obstruction of the common hepatic duct by extrinsic compression usually from a gallstone impacted in Hartmann's pouch or the cystic duct. Large gallstones that become impacted in this area produce common hepatic duct obstruction by two mechanisms: mechanical obstruction by direct compression of the common hepatic duct, or they can cause obstruction secondary to repeated bouts of local inflammation. In 1948, Argentinean surgeon Pablo Luis Mirizzi, first described a syndrome of common hepatic duct obstruction in the setting of longstanding cholelithiasis and cholecystitis. The classic description of the disease includes four components: (a) a close parallel course of the cystic duct and the common hepatic duct, (b) an impacted stone in the cystic duct or the neck of the gallbladder, (c) common hepatic duct obstruction secondary to external compression by the cystic duct stone (and the surrounding inflammation), and (d) jaundice, with or without cholangitis. Mirizzi's syndrome is a rare complication of cholelithiasis, with an estimated incidence of 0.05 - 2.7%. It presents as a spectrum of disease that varies from extrinsic compression of the common hepatic duct to the presence of a cholecystobiliary fistula. Often, this dangerous alteration to anatomy is not recognized pre-operatively, and has the potential to lead to significant morbidity and biliary injury, particularly in the laparoscopic era.

Classification : There are three classifications which have been proposed to describe variants of Mirizzi syndrome, and to aid in selecting the appropriate therapeutic procedure. The original classification, by McSherry et al, described two types. Type I referred to compression of the common hepatic duct by a stone impacted in the cystic duct or Hartmann's pouch. Type II referred to erosion of the calculus from the cystic duct into the common hepatic duct, producing a cholecystocholedochal fistula. Csendes et al created a second classification taking into account the extent of fistula. Type I remained the same, external compression of the common hepatic duct due to a stone impacted at the neck of the gallbladder or at the cystic duct. Types II to IV lesion referred to the presence and extent of a cholecystobiliary (cholecystohepatic or cholecystocholedochal) fistula, due to erosion of the anterior or lateral wall of the common hepatic duct by impacted stones. The fistula involved less than one-third of the circumference of the common hepatic duct in type II. Involvement of between one-third and two-thirds of the circumference of the common hepatic duct was called a type III lesion, while destruction of the entire wall of the common hepatic duct was called a type IV lesion. In their original paper, a total of 219 patients were identified with Mirizzi's syndrome. The incidence of type I lesions was 11 per cent, type II, 41 per cent, type III, 44 per cent and type IV, four per cent. The majority had obstructive jaundice. The third classification, proposed by Nagakawa and colleagues, expanded upon the definition of the Mirizzi syndrome. Type I referred to a stone impacted in the cystic duct or gallbladder neck. Type II was characterized by a fistula of the common duct. Type III was defined by hepatic duct stenosis due to a stone at the confluence of the hepatic and cystic ducts. Type IV was characterized by hepatic duct stenosis as a complication of cholecystitis in the absence of calculi impacted in the cystic duct or gallbladder neck. In one series of 30 patients, the frequency of these four types as described by Nagakawa et al was 14, 2, 6, and 8%, respectively



Mirizzi syndrome is part of the differential diagnosis of all patients with obstructive jaundice, and requires a high index of suspicion. Most patients present with jaundice, and right upper quadrant pain. Elevations in the serum concentrations of alkaline phosphatase and bilirubin are present in over 90 per cent of patients. The clinical and laboratory findings are similar to patients who present with obstructive jaundice secondary to choledocholithiasis. Once a diagnosis of obstructive jaundice has been made an abdominal ultrasound is often the first imaging test preformed. Imaging generally reveals gallstones, dilated intrahepatic ducts, with a long parallel cystic duct and a contracted gallbladder. The presence of a stone impacted in the gallbladder neck and an abrupt change to a normal width of the common duct below the level of the stone are also very suggestive of Mirrizi's syndrome. The sensitivity of ultrasound in detecting Mirizzi's syndrome is 23-46%. In Csendes' series, ultrasound revealed dilated ducts in 81% of patients and raised suspicion of Mirizzi's syndrome in only 27% of cases. CT scanning has a similar sensitivity to ultrasound, but can be helpful in diagnosing other causes of obstructive jaundice such as gallbladder cancer, cholangiocarcinoma, or metastatic tumor.

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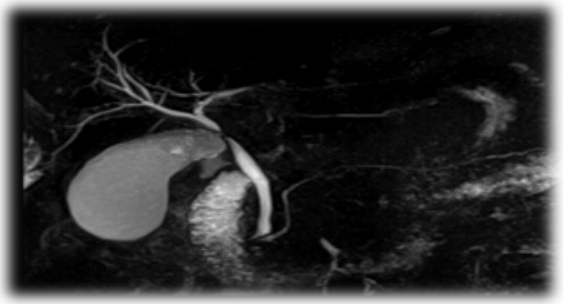
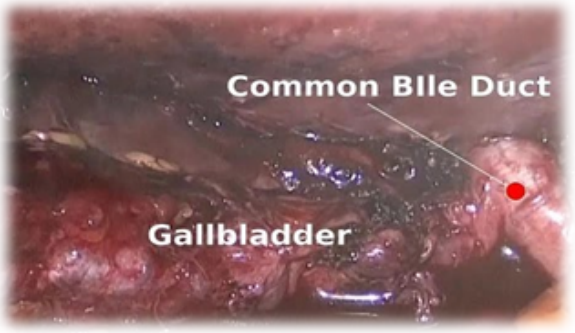
CASE STUDY

a 70 year male patient admitted in our emergency department with the chief complains of severe epigastric pain with yellowish discoloration of urine and the whole body. On admission patient temperature was normal, pulse was 92/min, BP was 110/80 mmHg, RR was 14/min with jaundice present over sclera and whole body including palms and soles. Liver was enlarged 3 finger breath below Rt subcostal margin and gallbladder was palpable. Laboratory data showed Hb 9.97 gm/dl, WBC count 13,800/cmm, total bilirubin 39.31 mg/dl with direct 29.20 mg/dl, SGPT 45.20 U/L, SGOT 73 U/L, ALP 149.3 U/L, PT (T) 14 sec (C) 13.5 sec, INR 1.05. MRCP s/o approx. 10x10 cm sized filling defect in proximal CBD just after opening of cystic duct s/o calculus with dilated CBD 11mm and abrupt cut off in its distal most part p/o stricture. RHD

- measures 5 mm & LHD 3mm. few tiny areas of filling defect noted in GB p/o ? sludge ? stone. Patient underwent for ERCP. Sphincterotomy was done with CBD stenting. Patient's jaundice were immediately improved. Later patient underwent for interval open cholecystectomy after 1 month. Operative findings revealed distended GB with indentation over CBD due to stone in cystic duct. No peri GB adhesions or fluid collection. Drain was kept in subhepatic region and removed on POD – 2. Patient was discharged on POD – 4. No post-operative complicatios were noted on patient's follow-up.





DISCUSSION

Surgery is the mainstay of therapy of Mirizzi syndrome, the dense inflammatory reaction in Calot's triangle, as well as the frequent aberrant biliary anatomy, pose a difficult challenge to the unsuspecting surgeon when dealing with a Mirizzi syndrome. The two principal aims are (a) the safe completion of cholecystectomy without injuring the biliary system and (b) the appropriate management of the cholecystocholedochal fistula. Meticulous dissection and vigilance are essential in order to avoid inadvertent bile duct injury. If the diagnosis of Mirizzi syndrome is made preoperatively , an operative strategy that minimizes the risk of injury to the biliary tract can be carried out. However, a preoperative diagnosis of Mirizzi syndrome is seldom made because ERCP and direct cholangiography are not widely used. ERCP, direct cholangiography, or magnetic resonance cholangiography should be performed in patients with clinical jaundice and signs and symptoms suggestive of biliary obstruction. A standardized surgical approach has been recommended based on the Csendes classification of the variants of Mirizzi syndrome: Type I - Cholecystectomy plus common bile duct exploration with T-tube placement. Exploration should be performed only if the CBD is easily exposed. Type II - Suture of the fistula with absorbable material or choledochoplasty with the remnant gallbladder. Type III - Choledochoplasty; suture of the fistula is not indicated. Type IV - Bilio-enteric anastomosis is preferred since the entire wall of the common bile duct has been destroyed. The approach may vary with the type of fistula present; both the operative mortality and postoperative morbidity increase according to the severity of the lesion.

LAPAROSCOPIC SURGERY : The Mirizzi syndrome presents a difficult challenge for laparoscopic surgery because the dense adhesions and edematous inflammatory tissue cause distortion of the normal anatomy and increase the risk for biliary injury. While it appears to be feasible, especially for type I anatomy, the routine use of laparoscopic surgery as the primary treatment of Mirizzi syndrome is controversial. It has been suggested, that a prudent approach for type 1 Mirizzi syndrome is to perform a trial laparoscopic dissection, but to have a low threshold to convert to an open procedure. This approach should be undertaken only by experienced laparoscopic surgeons.

ENDOSCOPIC THERAPY : Endoscopic treatment with or without electrohydraulic lithotripsy (EHL) can be effective as a temporizing measure before surgery and can be definitive treatment for unsuitable surgical candidates. One report described the experience with 14 patients with Mirizzi syndrome treated with EHL. Twelve patients had a single stone and complete clearance was achieved with one treatment session; two had multiple stones and required an additional treatment session. Asymptomatic leakage of contrast medium from the cystic duct into the peritoneal cavity was observed in one patient after removal of a large impacted cystic duct stone. This patient recovered with conservative therapy and suffered no adverse events. In another series of 25 patients with cholangiographic evidence of Mirizzi syndrome, 12 were referred for surgery after preliminary endoscopic therapy and 13 were treated solely with endoscopy. Stones were completely removed in three and nine were treated with long-term stents; complications occurred in four patients.

Endoscopic treatment of Mirizzi syndrome should be used as a temporizing measure before surgery. It can serve as a definitive treatment for those patients who are unsuitable surgical candidates when further endoscopic attempts can be made to dis impact and remove the stones. Long term success appears to be most likely in patients with type II disease who do not have residual gallbladder stones.

CONCLUSIONS

Mirizzi syndrome is a rare complication of cholelithiasis and requires a high index of suspicion in the setting of obstructive jaundice. Diagnosis preoperatively may be elusive with bloodwork, US and CT alone. Cholangiography (intraoperative and ERCP) as well as MRCP aids in both the diagnosis and identification of anatomy and may prevent serious biliary injury. Surgery is the mainstay of therapy of Mirizzi syndrome, and requires the safe completion of cholecystectomy without injuring the biliary system and the appropriate management of the cholecystocholedochal fistula. The aberrant anatomy intrinsic to this syndrome presents a difficult challenge to surgeons and the laparoscopic approach should be undertaken with caution and probably left to specialized minimally invasive centres. Endoscopic treatment may be effective as a temporizing measure before surgery and can be definitive treatment for unsuitable surgical candidates.

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