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CASE STUDY

MIRIZZI SYNDROME: A RARE ENTITY

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ABSTRACT

The Mirizzi syndrome (MS), a rare condition, is a late complication of chronic cholecystitis. It is a spectrum of disease ranges from simple compression of common hepatic duct to cholecystobiliary fistula (CBF). The clinical presentation is similar to symptomatic cholelithiasis like abdominal pain, vomiting, jaundice etc, hence it is difficult to diagnose preoperatively. Even peroperatively it is diagnosed by expert surgeon who has experience in hepatobiliary surgery, otherwise chances of biliary duct injuries are high.

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INTRODUCTION

Mirizzi syndrome, being rare entity, is also a one of the cause of obstructive jaundice. It is first described by Kehr and Ruge in 1905 and 1908 respectively but got recognition in 1948 when Argentinansurgeon Pablo LuisMirizzi described it as a functional hepatic syndrome (Simoglou *et al*., 2013; Solis-Caxaj, 2009). One of the component of functional hepatic syndrome is the presence of functional sphincter in common hepatic duct that causes stasis and subsequently obstructive jaundice. As such there is no physiologic or anatomical sphincter present in common hepatic duct. Later on McSheery classified this Mirizzi syndrome into two types in 1982 (Targarona *et al*., 1997). Type 1 is external compression of common hepatic duct and type II being the fistula i-e cholecystocholedochal fistula. Csendes *et al* in 1989 have proposed a classification consisting of four different types: type I, in which an impacted gallstone leads to an extrinsic compression of the common bile duct (CBD); type II corresponds to a cholecysto-biliary fistula (CBF) secondary to an eroded gallstone involving one-third of the circumference of the common bile duct; type III the fistula involves two-thirds of the circumference; and type IV the fistula involves the whole circumference (Ibrarullah *et al*., 2008) (Fig. 1).

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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 neck of the gallbladder (GB),
(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 (Chatzoulis *et al*., 2007).

Case Report

65 years old female known case of Diabetes Mellitus, Hypertension and Ischemic Heart Disease admitted through emergency department with complaints of high grade fever and abdominal pain for last 10 days. Pain was in right upper quadrant radiating to back, associated with vomiting, not relieved with over the counter medications. Fever was high grade associated with rigors and chills. On examination, pulse was 110 beats per minute, blood pressure 100/60mmhg and temperature 102^0F. She was dehydrated, jaundiced and tender in right hypochondrium. The patient was diagnosed as a case of cholangitis with classic Charcot's triad. She was resuscitated and investigations were done. Ultrasound demonstrated prominent proximal CBD of size 1.5cm with stone and mild intrahepatic duct dilatation.

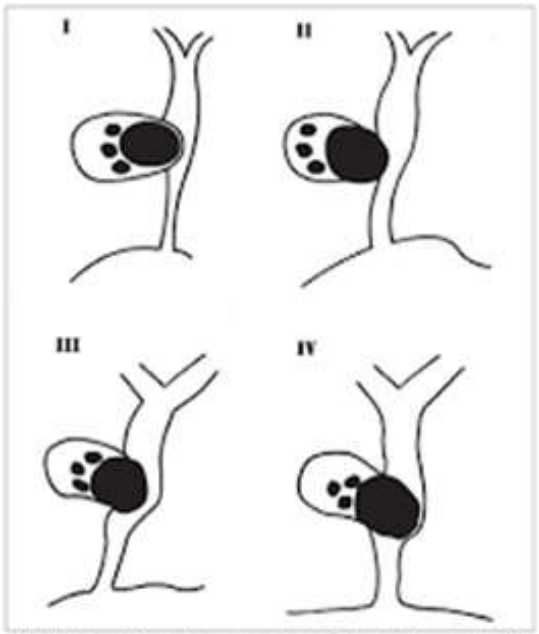

FIGURE 1 – Schematic representation of Csendes classification for Mirizzi syndrome

Figure 1. Schematic representation of Csendes classification of Mirizzi syndrome

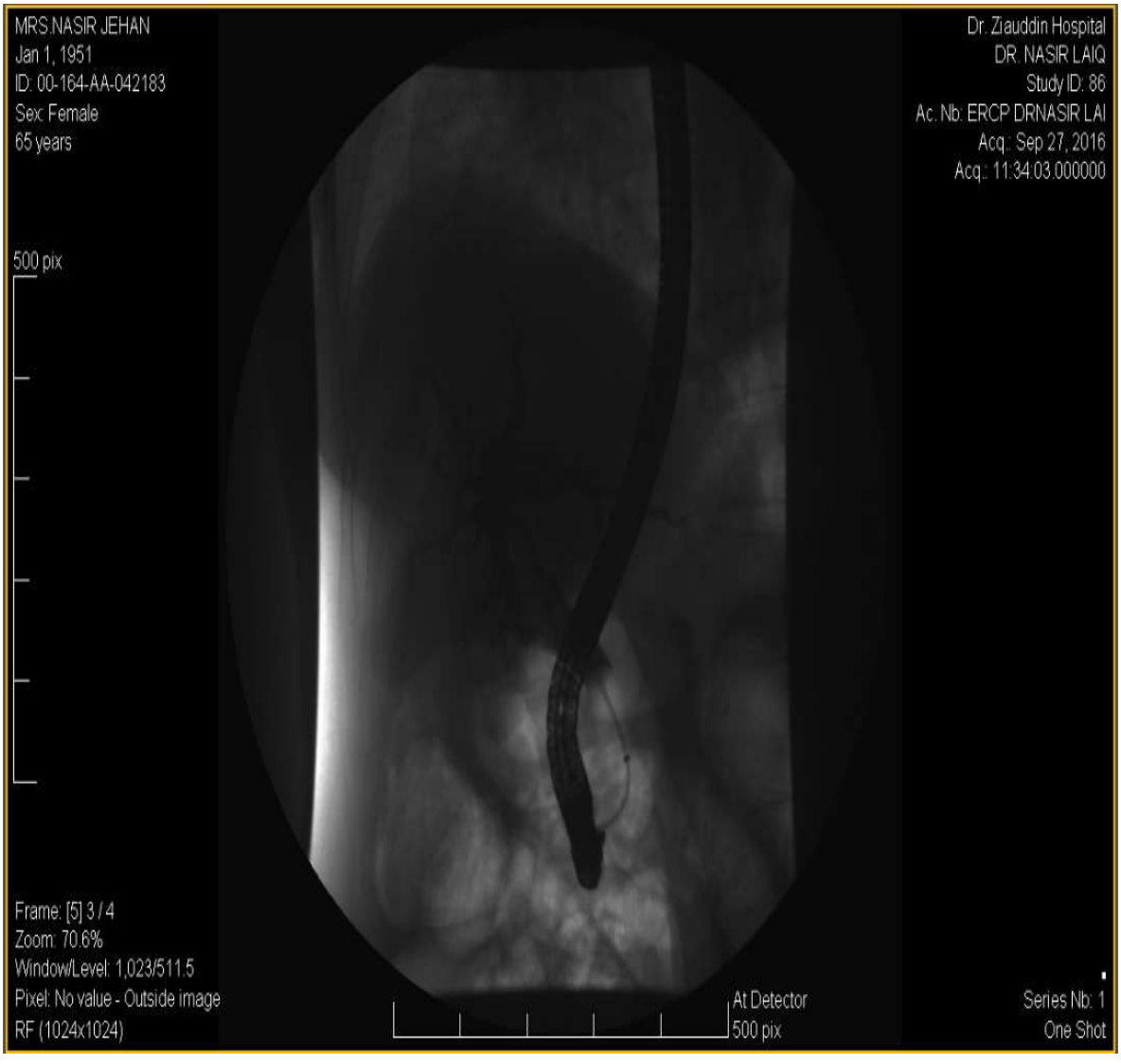

Figure 2. ERCP cholangiogram

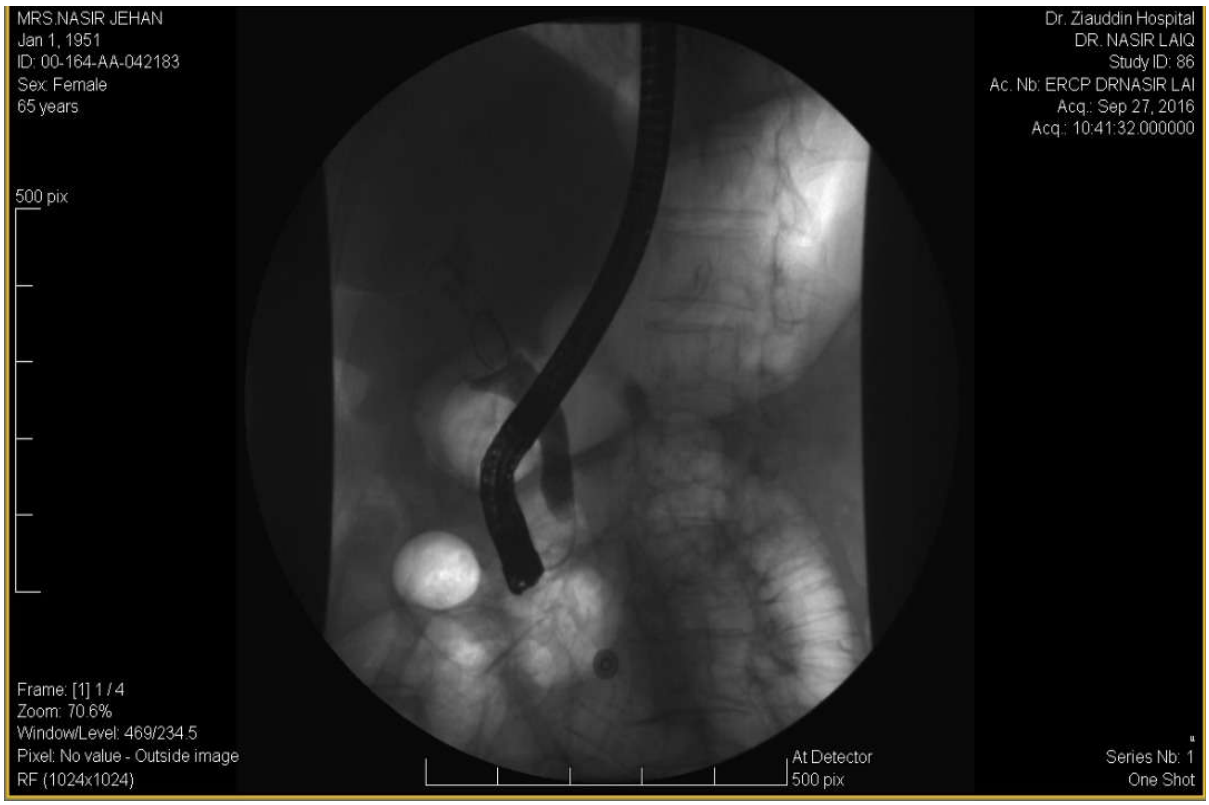

Figure 3. ERCP cholangiogram

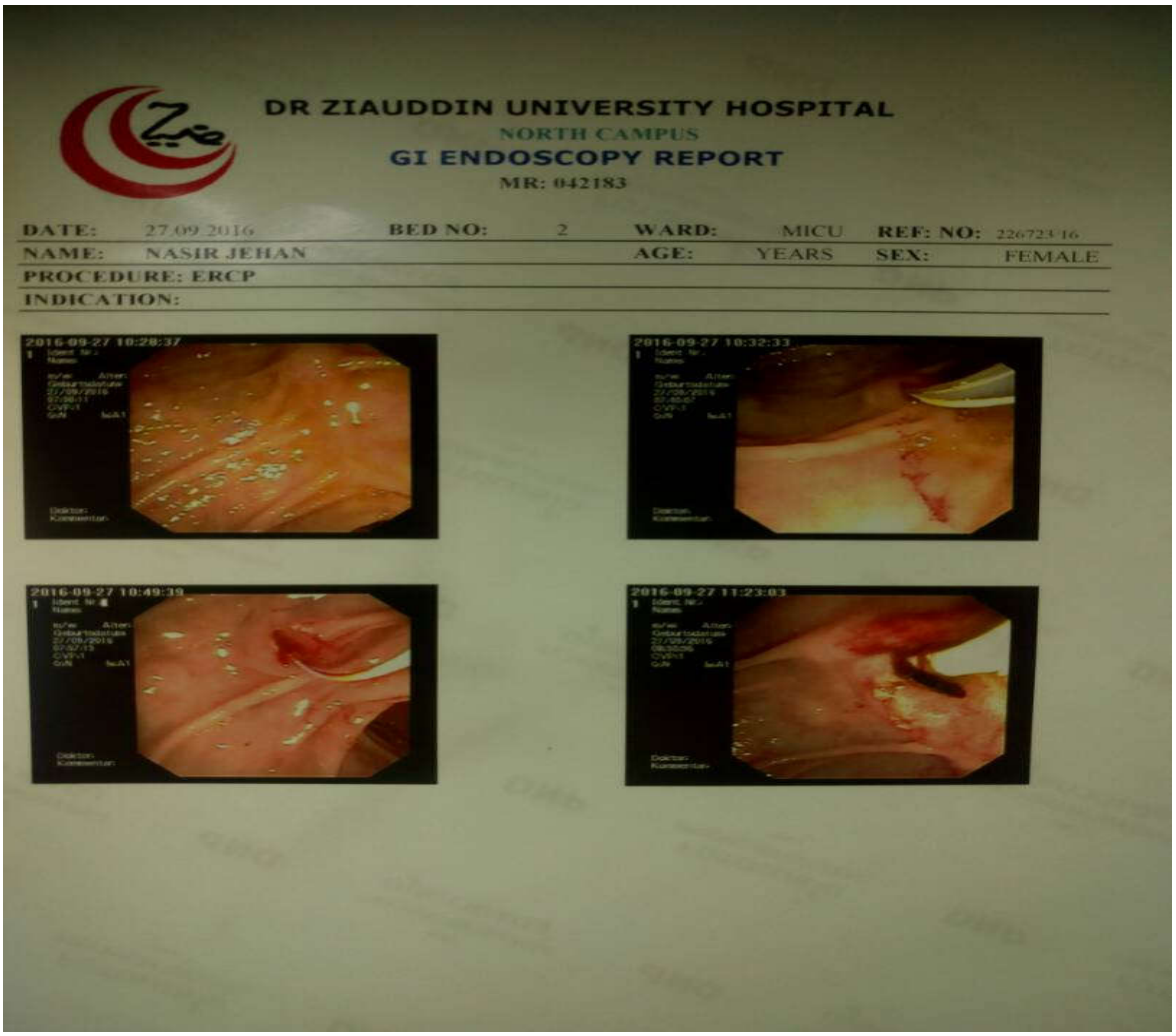

Figure 4. ERCP

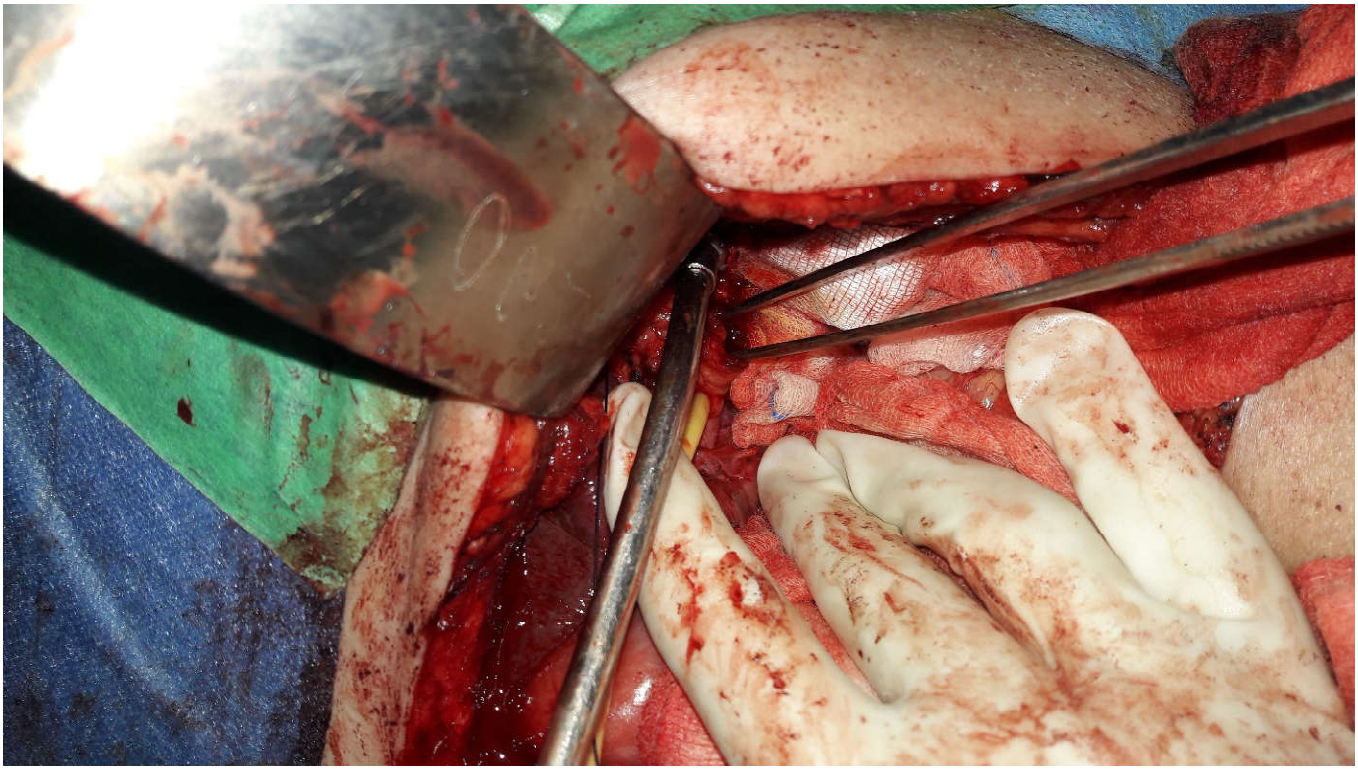

Figure 5. The fistula is above shown by forceps, T-tube is placed distally in healthy CBD

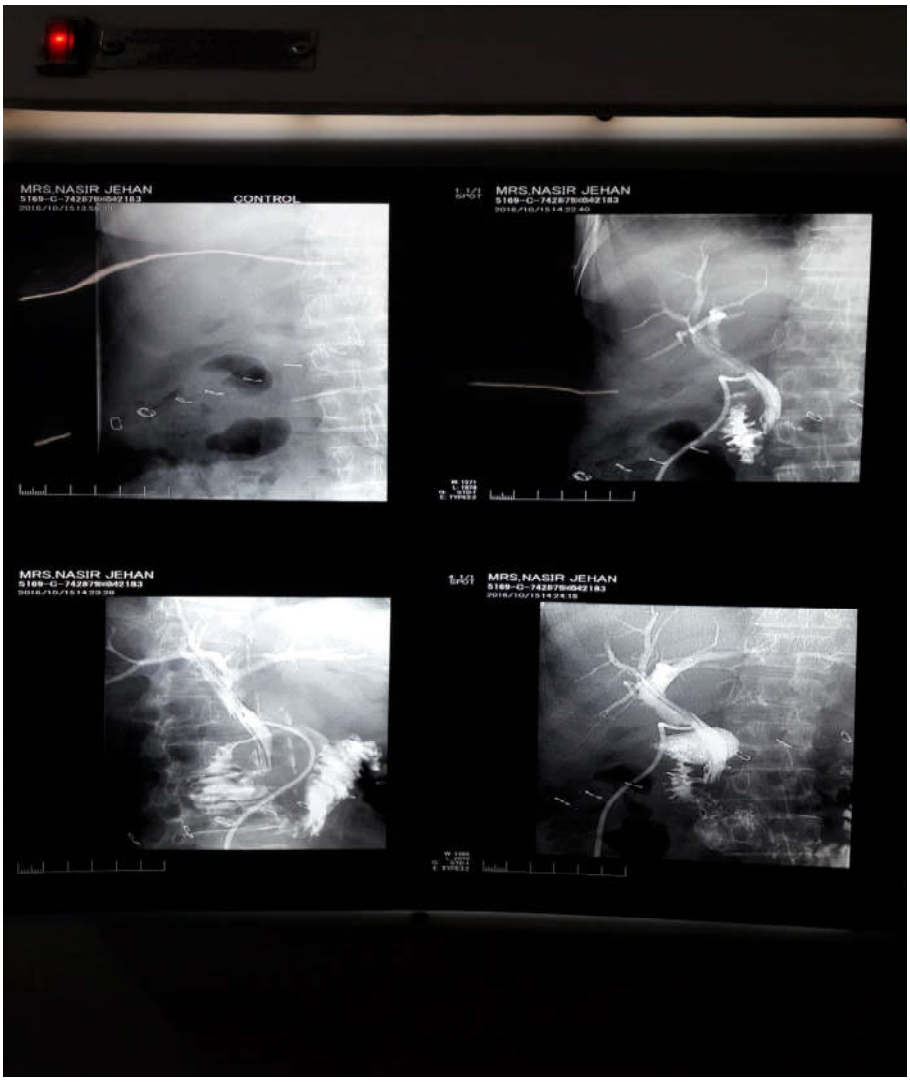

Figure 6. Post operative T-Tube cholangiogram

Total bilirubin was 4.08, Direct bilirubin was 3.57, alkaline phosphatase 748, gamma GT 458. Her total leucocyte count was 18000/cm^3. Her ERCP was planned that shows large stone in common bile duct which could not be retrieved (Figure 2,3,4). Patient was proceeded for open cholecystectomy and CBD exploration. Peroperatively we found very small fibrosed, shrunken gall bladder with no prominent cystic duct and stone in common hepatic duct. Fundus first dissection was carried out and gall bladder was amputed at about 1.5cm in fish mouth fashion, which was incorporated in reconstruction of anterior wall of common hepatic duct after removing the stone and T-tube placement distally through healthy common bile duct (Figure 5). Patient did well post operatively and was discharged. Two weeks later T-tube cholangiogram done (Figure 6) that was normal and T-tube was removed.

DISCUSSION

MS is a rare entity, but it demands importance because of high chances of biliary injuries that needs complex surgical procedures. Although very good radiologic imaging is available but most of the time it is diagnosed preoperatively (Kulkarni *et al*., 2016). Its presentation is similar to that of symptomatic gallstones like pain, vomiting, jaundice, charcots triad etc (Kok *et al*., 1998). Magnetic resonance cholangiopancreatography (MRCP), CT scan abdomen and Endoscopic retrograde cholangiopancreatography (ERCP) along with abnormal Liver function tests (LFT) might suggest preoperatively MS[6]. Even the challenge remains in diagnosis peroperatively because of pericholecystic adhesions, shrunken and sessile gall bladder with or without cholecystocholedochalfistula, obliterated calot's triangle etc. The impacted stone may mimic carcinoma of gall bladder neck complicating the situation even more. The peroperativecholangiogram in early part of surgery confirms the diagnosis and helps in delineating the anatomy. Dealing with this type of situation there are certain guidelines that should be followed. An antegrade approach to gall bladder is recommended (Beltran *et al*., 2008). The fundus of GB is opened and the stone extracted. The cystic duct stone should be milked back in to the GB. In rare instances when the Hartman's pouch or cystic duct lies behind the CBD, retrieval of the offending stone may be difficult. A transcholedochal approach has been recommended in such situation. It is often surprising to find the so called growth at the neck of GB disappear with evacuation of stone/s. A frozen section biopsy may be done if concomitant malignancy is strongly suspected. In the absence of any bile duct erosion i.e. type I MS, partial cholecystectomy alone is adequate. After removal of the offending stone the Hartman's pouch or the parallel cystic duct is left behind.

The inflammatory adhesion between these structures and CBD precludes any dissection in this area and is bound to result in bile duct injuries if insisted upon. When bile duct erosion is significant late strictures has been reported with simple closure of fistula or end to end repair. In most of the cases the defect in the CBD can be managed by retaining a cuff of GB around the fistula which is approximated, the procedure thus known as choledochoplasty. The CBD in such cases should be drained by a T-tube placed through a fresh choledochotomy or the fistula itself. Csendes reported an increased incidence of bile leak when the tube was bought out through the fistula rather than a fresh choledochotomy. Though choledochoplasty alone may suffice in nearly all cases of CBF, there are patients who present with complete or near complete obstruction of the bile duct at the initial exploration itself. In these cases for better long-term results it is safer to perform bilioenteric anastomosis. A well defined management guideline was provided by Csendes *et al* who classified MS on the basis of extent of erosion of CBD circumference. The recommended procedures for different types are: type I – partial cholecystectomy, type II – suture closure of fistula or choledochoplasty, type III – choledochoplasty, type IV – bilioenteric anastomosis[8]. In our case ERCP done that demonstrated large stone in CBD, stone could not be retrieved. So we planned open cholecystectomy with CBD exploration. Peroperatively we found a lot of adhesions, very small fibrosed gall bladder with stones in CBD. We immediately recognize it as a case of MS and we did choledochoplasty.

Conclusion

Mirizzi syndrome is a rare entity. Presence of it makes cholecystectomy an extremely hazardous procedure. It should be in mind while dealing with small, thick walled and fibrosed gall bladder. Most of the time it is diagnosed peroperatively. Csendes described its classification and management very well.

REFERENCES

Beltran, M.A., Csendes, A. and Cruces, K.S. 2008. The relationship of Mirizzi syndrome and cholecystoenteric fistula: validation of a modified classification. *World journal of surgery,* Oct 1;32(10):2237-43.

Chatzoulis, G., Kaltsas, A., Danilidis, L., Dimitriou, J. and Pachiadakis, I. 2007. Mirizzi syndrome type IV associated with cholecystocolic fistula: a very rare condition-report of a case. *BMC surgery,* May 27;7(1):6.

Ibrarullah, M., Mishra, T. and Das, A.P. 2008. Mirizzi syndrome. *Indian Journal of Surgery.* Dec 1;70(6):281-7.

Kok, K.Y., Goh, P.Y. and Ngoi, S.S. 1998. Management of Mirizzi's syndrome in the laparoscopic era. *Surgical endoscopy,* Oct 21;12(10):1242-4.

Kulkarni, S.S., Hotta, M., Sher, L., Selby, R.R., Parekh, D., Buxbaum, J. and Stapfer, M. 2016. Complicated gallstone disease: diagnosis and management of Mirizzi syndrome. *Surgical Endoscopy,* Sep 1:1-8.

Simoglou, C., Simoglou, L. and Babalis, D. 2013. Mirizzi syndrome. *Hellenic Journal of Surgery.* Mar 1;85(2):109-12.

Solis-Caxaj, C.A. 2009. Mirizzi syndrome: diagnosis, treatment and a plea for a simplified classification. *World journal of surgery*. 2009 Aug 1;33(8):1783-4.

Targarona, E.M., Andrade, E., Balague, C., Ardid, J. and Trias, M. 1997. Mirizzi's syndrome. *Surgical endoscopy,* Aug 1;11(8):842-5.

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