

MIRIZZI SYNDROME DIAGNOSED ON MRCP: A CASE REPORT

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ABSTRACT

Mirizzi syndrome is a rarely observed phenomenon that results from extrinsic compression of common hepatic duct due to gallbladder or cystic duct calculi presenting with obstructive jaundice and cholangitis. Mirizzi syndrome is difficult to differentiate from other serious conditions such as cholangiocarcinoma, biliary strictures and choledocholithiasis clinically and on imaging. Making a pre-operative diagnosis using imaging modalities is a challenge. MRCP is a non invasive accurate modality for diagnosis of Mirizzi syndrome.

KEYWORDS: Gall Stones, Mirizzi Syndrome, MRCP, Obstructive Jaundice.

INTRODUCTION

Mirizzi syndrome is an important but benign complication of chronic cholecystitis in which stones in the neck of the gallbladder or cystic duct cause mechanical obstruction, inflammatory stricture and eventually penetration of the common hepatic duct. An Argentinean surgeon, Pablo Mirizzi described Mirizzi syndrome for the first time in 1948 which established the eponym Mirizzi¹. Mirizzi Syndrome is an uncommon presentation of chronic cholelithiasis with incidence of less than 1% in a year in western population². According to another study conducted by Kamalesh et al incidence of Mirizzi Syndrome is approximately 1% in patients with cholelithiasis³. However incidence of Mirizzi Syndrome in our region is not known. Since replacement of open cholecystectomy by laparoscopic approach, Mirizzi Syndrome is gaining interest of biliary surgeons. Failing to recognize this condition pre-operatively results in inadvertent bile duct injury because Mirizzi Syndrome causes dangerous alteration of normal biliary anatomy as a result of chronic inflammation. Successful pre-operative diagnosis of Mirizzi syndrome requires clinical correlation, high index of suspicion, imaging and endoscopic methods. Risk of bile duct injuries without pre-operative diagnosis is up to 17%. Despite combining all these factors pre-operative diagnosis of Mirizzi Syndrome can only be made in 8-62.5% cases^{4,5}. That is why laparoscopic cholecystectomy is contraindicated and open cholecystectomy is advised in treating patients with Mirizzi Syndrome⁶. Literature review from several case series shows a strong correlation between Mirizzi Syndrome and gallbladder cancer, further emphasizing importance of this condition⁷. Mirizzi Syndrome must also be differentiated from choledocholithiasis, bile duct stricture and cholangiocarcinoma. We report a case of 47 years

old female patient presenting with obstructive jaundice, diagnosed as Mirizzi syndrome on MRCP.

CASE REPORT

A 47 years old female patient was referred with complaints of vomiting, intermittent epigastric and right hypochondrium pain radiating to the back and over scapular region and generalized skin itching. General physical examination revealed yellowish discoloration of skin, discoloration of urine and clay colored stool. Lab investigations showed markedly raised levels of bilirubin 28 mg / dl (normal value 0.1-1 mg / dl), alkaline phosphatase 2485 (normal value 45-130 IU / L) and serum amylase 120 IU / L (normal value 26-100 IU / L). Ultrasound abdomen demonstrated calculus cholecystitis and intrahepatic cholestasis. However the cause of obstructive jaundice was still unjustified. Though ERCP has high sensitivity to diagnose Mirizzi syndrome and also superior due to its therapeutic purpose it was not performed because of its contraindication in acute pancreatitis. We performed MRCP of this patient to rule out the cause of obstructive jaundice. MRCP demonstrated gallbladder filled with multiple small calculi with evidence of pericholecystic edema in SSFP sequence (Figure1). There was a filling defect in common hepatic duct as shown in Figure 2 (T2 FASE sequence). Figure 3 is 3 D view of MRCP demonstrating a large filling defect in common hepatic duct caused by gallbladder neck producing external compression on common hepatic duct associated with mildly dilated intrahepatic ducts. Based on these findings she was diagnosed as having Mirizzi Syndrome. Open cholecystectomy was performed by surgery department. Intra operative findings include thick walled, distended gallbladder with multiple calculi within gallbladder and a large calculus in Hartmann pouch causing external compression of common hepatic duct (Mirizzi Syndrome). However there was no evidence of cholecystobiliary fistula (Type I Mirizzi syndrome). Gallbladder was removed and sent for histopathology which proved negative for gallbladder carcinoma. Biliary system was drained temporarily by T-tube. On 8th post operative day patient was discharged with normal alkaline phosphatase and serum bilirubin level and progressively decreasing levels of serum amylase. Follow up of patient after one month showed normal lab investigations and normal hepato-biliary system on ultrasound.

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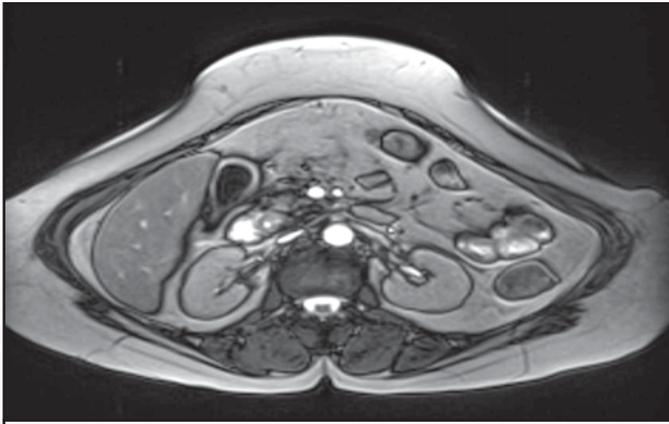


FIGURE-1: STEADY-STATE FREE PRECESSION (SSFP) SEQUENCE, SHOWING MULTIPLE CALCULI IN GALLBLADDER AND PERICHOLECYSTIC EDEMA.

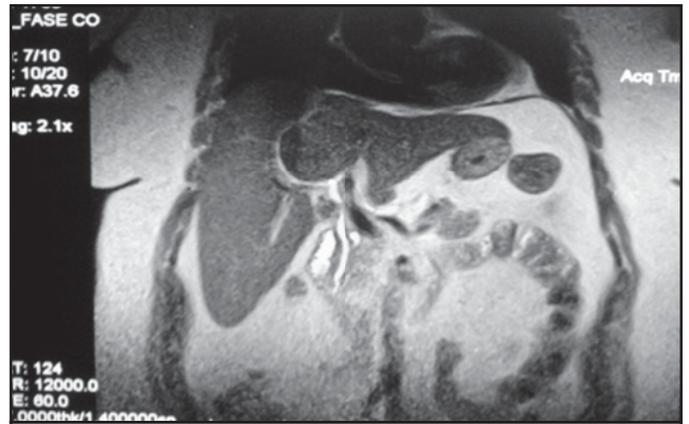


FIGURE-2: T2 FAST SPIN ECHO (FASE) SEQUENCE SHOWING A FILLING DEFECT IN COMMON HEPATIC DUCT.

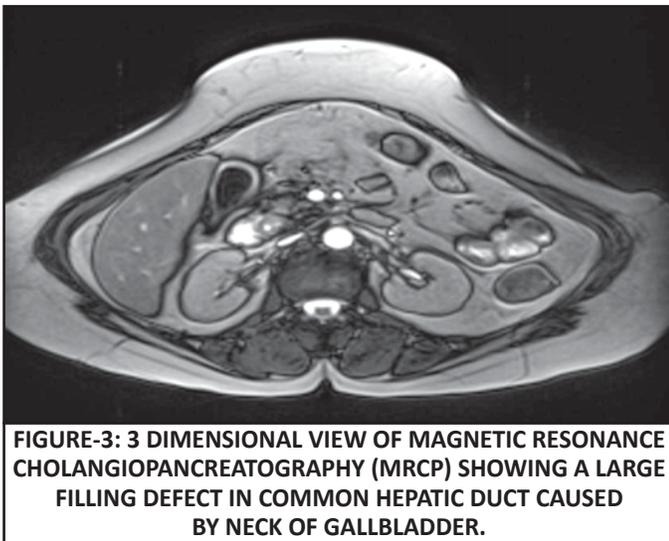


FIGURE-3: 3 DIMENSIONAL VIEW OF MAGNETIC RESONANCE CHOLANGIOPANCREATOGRAPHY (MRCP) SHOWING A LARGE FILLING DEFECT IN COMMON HEPATIC DUCT CAUSED BY NECK OF GALLBLADDER.

DISCUSSION

Mirizzi syndrome is a complication of long standing symptomatic cholelithiasis. Incidence of Mirizzi Syndrome following cholecystectomies is reported to be approximately 0.35%⁸. Compared to western world incidence of Mirizzi syndrome in underdeveloped countries is high ranging between 4.7-5.7%. Mirizzi syndrome is more commonly seen in female patients probably due to high incidence of cholelithiasis in females⁹. However acalculus cholecystitis can rarely lead to a condition mimicking Mirizzi Syndrome clinically and on imaging. The underlying mechanism is sub-hepatic inflammation and adhesions which distort normal anatomy¹⁰. Important predisposing factors in causing Mirizzi Syndrome may be Long segment of cystic duct intramurally and low lying insertion of the cystic duct into the common bile duct. Various classifications by different authors describe types of Mirizzi Syndrome. McSherry classified Mirizzi syndrome into two types. Common hepatic duct compression is present in both types. They are differentiated on the basis of presence or absence of fistula. Type I is without fistula formation, whereas in type II external compression by stone forms a cholecystocholedochal fistula¹¹. Csendes proposed another classification in 1989. It classified Mirizzi Syndrome into four types. In type I there is

mechanical obstruction of hepatic duct without fistula formation whereas in type II, type III, and type IV a cholecystobiliary fistula forms, with erosion of hepatic duct. Erosion of less than one-third of circumference of the hepatic duct is classified in type II, erosion of two-thirds of circumference of hepatic duct is classified in type III and complete destruction of the hepatic duct is classified in type IV¹². Modified Csendes classification was presented in 2007 which added type V Mirizzi Syndrome, defined as presence of a cholecystoenteric fistula along with any type of the above classification. It is further divided into two subtypes Va and Vb depending on the absence (Va) or presence (Vb) of gallstone ileus¹³. Accurate definition of the biliary anatomy and recognition of fistula formation preoperatively is crucial for optimal surgical planning due to increased morbidity and mortality associated with this condition¹⁴. MRCP is sensitive imaging modality for detection of fistulous tracts¹⁵. In our patient no cholecystocholedochal or cholecystobiliary fistula was observed, so classified into type I Mirizzi Syndrome according to McSherry and Csendes classification.

Despite improvements in radiological investigations, diagnosis of Mirizzi syndrome before surgery is not easy. The condition is difficult to differentiate from cholangiocarcinoma, bile duct stricture or choledocholithiasis because these conditions present with similar clinical features as Mirizzi Syndrome and radiological findings may also be confusing. Due to rarity of Mirizzi Syndrome the condition may also be overlooked. Imaging modalities available to diagnose Mirizzi syndrome include Ultrasound, CT scan, MRCP and ERCP (endoscopic retrograde Cholangiopancreatography). Ultrasound and CT scan have very low sensitivity to diagnose Mirizzi syndrome. MRCP has a sensitivity of 50% and ERCP has a sensitivity of 55-90%. ERCP is also superior to MRCP due to its therapeutic purpose¹⁶. However ERCP carries risk of inducing acute pancreatitis. MRCP is an optimal non invasive method to define biliary anatomy pre operatively¹⁷. It demonstrates extrinsic compression on common hepatic duct caused by stones in cystic duct or Hartmann pouch. It also demonstrates dilatation of intrahepatic biliary ducts proximal to obstruction and normal caliber of biliary channels distally¹⁸. In 50% cases Mirizzi Syndrome is diagnosed intra-operatively¹⁹.

CONCLUSION

Mirizzi Syndrome is a rare condition and high index of suspicion is required to make a pre surgical diagnosis. MRCP is a sensitive, non invasive imaging modality to diagnose Mirizzi Syndrome.

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