www.jmscr.igmpublication.org
Impact Factor (SJIF): 6.379
Index Copernicus Value: 79.54
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossrefDOI: https://dx.doi.org/10.18535/jmscr/v6i8.83

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Yellow Eyed Man with a Stone in the Pouch

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Abstract
Mirizzi Syndrome is a rare complication of gallstone disease which is always caused by a calculus in the cystic duct or neck of the gallbladder, resulting in mechanical compression of common bile duct and the gallbladder with an incidence of less than 1% in general population and is a challenging clinical entity to manage.
Herein we report a rare case of a 62-year-old male who was treated for hepatic abscess now came with stone in the neck of gallbladder obstructing common bile duct. This case highlights the fact that there might be difficulty in diagnosing Mirizzi Syndrome.
The importance and implications of this condition are related to their associated and potentially serious surgical complications such as bile duct injury, and to its modern management when encountered during laparoscopic cholecystectomy.

Introduction

Mirizzi Syndrome refers to partial common hepatic duct obstruction resulting from compression by a gallstone impacted in the cystic duct or Hartmann's Pouch.It is rare and occurs in only in less than 1% of patients with cholelithiasis. Clinically, it is characterised by abdominal pain, fever and jaundice. Argentinean surgeon Pablo Mirizzi was the first one to explore this condition and to publish an authentic article about it based on cholangiography. The famous article that established the eponym of Mirizzi for this condition was published in 1948.

USG generally reveals gallstones with a contracted gallbladder and moderate intrahepatic ductal dilatation with normal extrahepatic biliary anatomy. ERCP and MRCP are useful in making out the hepatic duct anatomy. The typical findings are a dilated intrahepatic biliary tract, with a normal-sized bile duct, secondary to obstruction at the level of the cystic duct insertion into the common hepatic duct. The appearance of the obstruction and surrounding inflammation may be confused with a Klatskin tumor.

During cholecystectomy, it is seen as a dangerous adherent and inflammatory tissue in the area of Calot's triangle. In the general population, aberrant right posterior hepatic duct is present in 4%–8% of people and is one of the causes of bile duct injury during duct surgery.

Case Presentation

A 62-year-old gentleman was admitted with history of severe generalised itching and yellowish discoloration of eyes and urine. Patient is non-alcoholic and non-smoker.

Past History

He is known to have type II diabetes mellitus.

Before six months he had presented with fever, chills, tender abdomen, and mild jaundice. He was admitted and investigated. HRCT thorax showed a well-defined non enhancing area with peripheral enhancement in liver suggestive of liver abscess. A diagnosis of liver abscess was made and he was treated for liver abscess. His symptoms subsided and he was discharged.

Examination

The patient is well built and moderately nourished. Examination showed severe jaundice with greenish tinge and no external manifestations of chronic liver disease, tuberculosis or malignancy. No ascites. Vitals were within normal limits. Abdominal examination showed tender hepatomegaly with liver span of 16 cm, no splenomegaly and no para-aortic lymph node involvement. Other system examination revealed no abnormality.

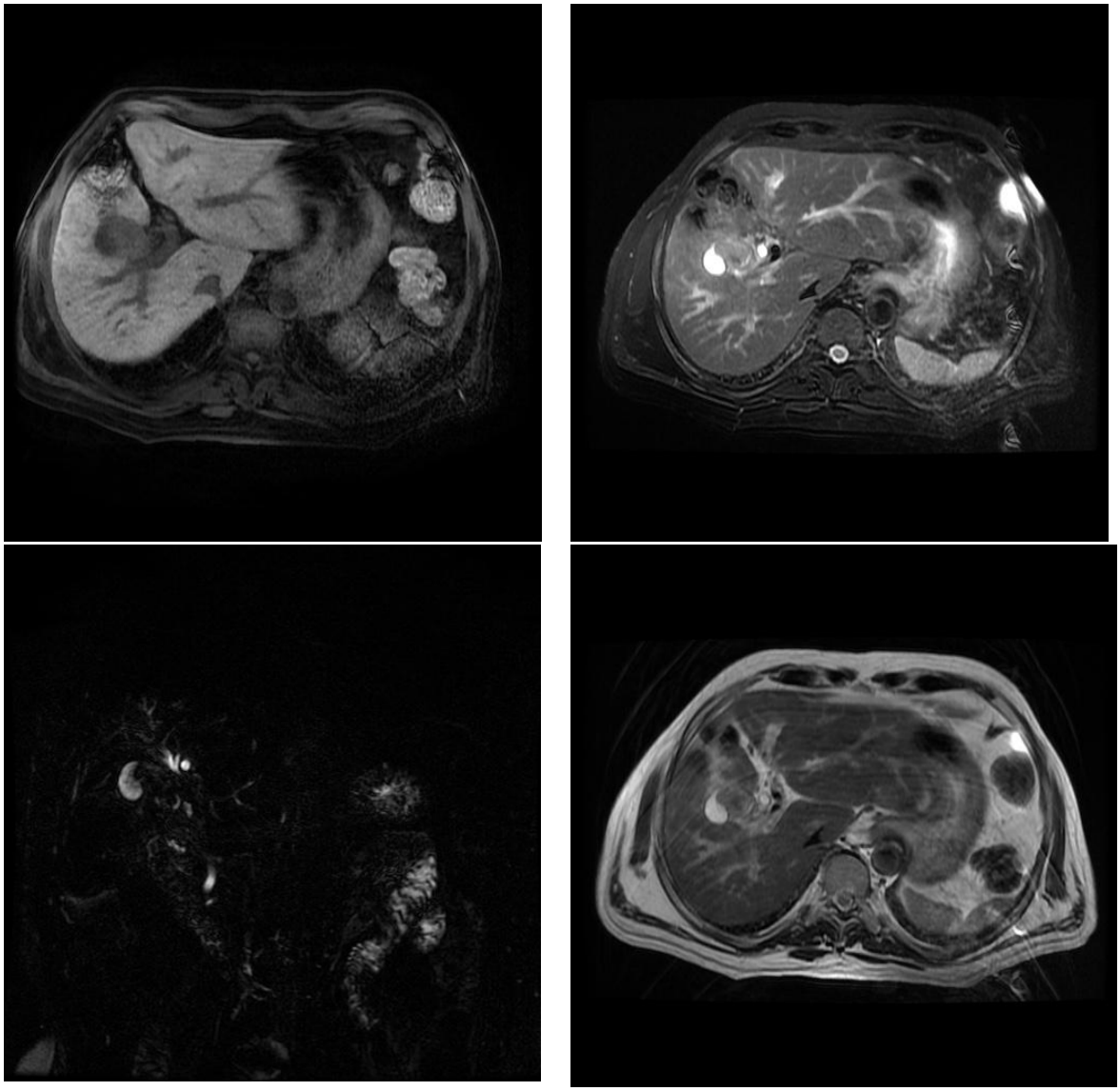
In view of extensive pruritis with jaundice, in a known patient of diabetes mellitus, we clinically suspected a possibility of pancreatic carcinoma, which was investigated for. Other possibilities like cholestatic jaundice and viral hepatitis were also considered.

Blood investigations showed:

- HB: 13.2
- Total WBC: 9200
- Platelets: 2.87 Lakhs
- ESR: 15
- MCV: 90
- MCH: 31.7
- MCHC: 35
- Random Blood Sugar: 224
- Blood Urea: 26
- Serum Creatinine: 1.0
- Uric Acid: 6.0 mg/dL
- PT: 14.3
- INR: 0.98
- aPTT: 33.2
- **ALT**: 43 U/L (normal: 0–40 U/L)
- **AST**: 37 U/L (normal: 0–40 U/L)
- **ALP:** 95 U/L (normal: <106 U/L)
- **γ-GGT**: 63 U/L (normal: <50 U/L)
- **Total bilirubin**: 3.8 mg/dL (normal values for total bilirubin:0.1 to 1.2 mg/dL).
- Serologic tests for hepatitis B and C were all negative.
- Tumor markers including α-fetoprotein, carcinoembryonic antigen, and CA 19-9 were within normal limits.
- Serology for hydatid cysts : Negative
- Total Cholesterol: 198
- ANA, RA Factor: Negative
- HIV: Negative
- VDRL: Negative
- Mantoux Test: Negative
- CRP: Positive
- Chest X-ray, ECG, ECHO were all normal

USG abdomen: showed cholelithiasis with a distended gallbladder, multiple calculi within its lumen, and moderate intrahepatic biliary dilatation.

MRI: Calculus in the neck of the gall bladder causing smooth stricture of proximal common bile duct resulting in the upstream dilatation of the biliary radicles. Smooth thickening of the gall bladder wall possibly secondary to inflammatory changes.



Finally, after doing MRCP, a possibility of Mirizzi Syndrome was diagnosed.

Discussion

The pathophysiology leading to Mirizzi syndrome is attributed to inflammatory process which occurs secondary to a pressure ulcer caused by an impacted gallstone at the gallbladder infundibulum first causing an external obstruction of the bile duct, and eventually eroding into the bile duct and evolving to a cholecystocholedochal or cholecysto-hepatic fistula. And accordingly some authors have classified it into different types. McSherry et al classified into two types. Type I, the hepatic duct is compressed by a large stone impacted in the cystic duct or Hartman's pouch. Associated inflammation may contribute to the obstruction and formation of a stricture in the central section of the extra- hepatic bile duct. In type II, the calculus has eroded into the common hepatic duct to produce a cholecystocholedochal fistula.

Significance of Mirizzi Syndrome is that the diagnosis of this condition is often missed preoperatively. Clinically Mirizzi Syndrome is characterised by bouts of abdominal pain, fever and jaundice and many times is asymptomatic. A large number of patients are only diagnosed with Mirizzi Syndrome during surgery. An impacted gallstone in the infundibulum or the neck of the cystic duct, edematous or atrophic gallbladder, fibrosis and distortion of Calot's triangle are the signs of Mirizzi Syndrome that can be observed during surgery.

Ultra Sound shows dilated intrahepatic and common hepatic ducts with a contracted gallbladder. But the cause of dilatation might not often be seen in USG.

At present, MRCP is the preferred diagnostic tool. Typical characteristics of Mirizzi Syndrome, such as, extrinsic compression of the common hepatic duct, and dilatation of the common hepatic duct with normal-sized common bile duct can be made out clearly with MRCP. Moreover, it is non-invasive too. MRCP confirmation is warranted when USG examination detects a dilated bile duct with evidence of obstructive jaundice or stone in the bile duct. Biliary ducts and pancreatic ducts can also be assessed by MRCP, which can create superior images of inflammation around the gallbladder. Such inflammation is characteristic of Mirizzi Syndrome, and can therefore be used to distinguish biliary conditions including cancer. ERCP is considered gold standard diagnostic tool for Mirizzi Syndrome.

Surgical management is the mainstay treatment for Mirizzi Syndrome, although this is challenging for several reasons. First, there is a low index of suspicion for this condition among surgeons, largely owing to its rarity. Usually gallbladder surgery is often performed in patients with relatively shorter histories of illness, long before the onset of Mirizzi Syndrome. Secondly, preoperative diagnosis is often missed pre operatively.

Differential Diagnosis are Caroli's Disease, Klatskins Tumor, Biliary Cyst Tumor, Cholangio carcinoma.

Conclusion

This patient had presented with mild jaundice and fever which was diagnosed as hepatic abscess from which patient has completely recovered. Significance of this is that, it might have been a dilatation of biliary radicles which has been mistaken as hepatic abscess. So in an elderly person with jaundice and extensive pruritis, a possibility of other underlying causes must also be considered and must be followed up. Here we present this case due to the rarity of Mirizzi syndrome and due to the rarity of presentation.