CASE REPORT

MIRIZZI'S SYNDROME: AN INTERESTING ON TABLE FINDING

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The Mirizzi's syndrome is a rarely observed disorder that refers to common hepatic duct obstruction caused by an extrinsic compression from an impacted stone in the cystic duct or Hartmann's pouch of the gallbladder. It has been estimated to occur in 0.7–1.8 percent of all cholecystectomies. The preoperative diagnosis of Mirrizi's syndrome is a not always easy despite the availability of latest advances in the radiological tests which can lead to significant morbidity and bile duct injury. We present the case of a 50-year-old Asian female with Mirizzi's syndrome who was initially having an impression of cholangiocarcinoma with cholecystitis and cholelithiasis based on radiological findings. Our patient was diagnosed as having Mirizzi's syndrome on an IV contrast CT scan. Cholecystectomy was performed with a restoration of biliary drainage by placing a t-tube. The on-table findings of the Mirizzi's syndrome can vary considerably as compared to the preliminary diagnosis based upon the biochemical tests and the radiological studies. The grade of the fistula, the extent of involvement of the biliary channels can only be found out on proper on-table surgical assessment.

Keywords: Mirizzi's syndrome; Cholangiocarcinoma; Obstructive jaundice

J Ayub Med Coll Abbottabad 2014;26(4):621–4

INTRODUCTION

Mirizzi's syndrome (MS) is a rarely observed condition characterized by impaction of single large stones or multiple small gallstones impacted in the Hartmann's pouch or in the gallbladder infundibulum and cystic duct, causing mechanical obstruction of the common hepatic duct and presents clinically as intermittent or persistent jaundice.[1–3] Predisposing factors include a long intramural cystic duct or a low insertion of cystic duct into the common hepatic duct.[4–6]

This syndrome was first described in 1948 by an Argentinean surgeon, Pablo Mirizzi's.[7–10] The accurate diagnosis of MS is of particular importance to surgeons as the condition may be confused with choledocholithiasis, bile duct stricture or cholangiocarcinoma on initial presentation and hence the surgical treatment is associated with a significantly increased risk of inadvertent bile duct injury.[11,12]

We report the case of a 50-year-old female with MS, who was initially diagnosed as having cholangiocarcinoma based on her ultrasonography but her CT abdomen findings were highly suggestive of MS.

CASE REPORT

We present a case of 50 year old Noor Asia from the periphery who presented to us through out-patient department complaining of: severe right hypochondrium pain on and off, nausea, indigestion to the fatty meals, and progressively increasing jaundice with pruritus for the last two months. On examination she was deeply jaundiced, icteric, dehydrated with marked tenderness in the right hypochondriac fossa. On palpation there was a tender right hypochondriac mass. We therefore proceeded with ultrasonography on which there was marked dilatation of intrahepatic biliary channels: a 2.3 cm stone in the gall bladder with a wall thickness of 3.7 mm. The overall findings were most likely suggestive of cholangiocarcinoma with cholecystitis and cholelithiasis. A CT abdomen with IV contrast was advised.

On CT abdomen there was a moderate dilatation of intra-hepatic biliary channels. The gall bladder was massively distended containing a large calculus. Findings were suggestive of MS. Magnetic resonance cholangiopancreatography (MRCP) was advised for further evaluation which could not be carried out as the patient was non-affording.

According to her latest laboratory findings 2 days before the planned cholecystectomy her serum bilirubin raised up to 10.8 mg/dl (bilirubin was 15.8 mg/dl 03 weeks ago), alkaline phosphatase up to 1300 U/L, Haemoglobin level was 11.3 and ALT was 480/L. Patient was therefore placed on the elective list for the surgical intervention three weeks after her initial presentation of the symptoms.

A subcostal Kockers incision was made; the gall bladder was markedly distended and badly stuck to duodenum, omentum and colon. A purse string with 2.0 Vicryl was applied at the fundus of the gall bladder and opened, yellowish secretion in the gall bladder aspirated. The index finger was then passed in the cavity of gall bladder. A large stone measuring up to 2×2 cm got impacted in the neck that was retrieved and extracted. Purse string sutures were closed. Cholecystectomy was done by first fundus approach as the Callot's triangle was not discernable. Two fingers were passed in the foramen of Winslow, and the

contents were palpated within the wall. The entire contents were matted, fibrosed and inflamed with stony hard consistency and a distorted anatomy.

Right from the Porta Hepatisto the duodenum common bile duct (CBD) was identified, dissected from the surrounding contents, lifted with Lahey feeding tube, and passed around it to sling it up. CBD was opened through 0.5 cm incision and white bile was drained. Feeding tube was passed into the lumen and was tried to left and right hepatic ducts but it failed. Then Bake's dilator passed to negotiate the confluence which succeeded to right and left ducts, patency was ensured, and a few cc yellowish bile came out after the white bile. CBD, and confluence of the right and the left hepatic duct were flushed with the normal saline. All the gravel and debris were washed. The same feeding tube was passed distally to check the distal CBD patency. Feeding tube could be palpated in the duodenum.

Her perioperative findings were: markedly distended thick walled gallbladder, and the surrounding omentum, duodenum and colon was badly stuck with the inferior wall of the gallbladder. CBD and common hepatic duct (CHD) were badly obscured and not clearly visible. There was a large stone measuring 2×2.5 cm in the gall bladder which was taken out after draining out the collection causing the markedly distended gall bladder which was presenting as a tender right hypochondrial mass on initial examination. The CHD was completely obscured; it was very difficult to identify the Callots triangle as it was completely obliterated and there were dense adhesions at the sub-hepatic space and difficult to identify other landmarks as the surrounding duodenum was badly stuck with the gall bladder wall as was presenting a hard fibrotic mass. Intra-operative cholangiography could not be done due to the non-functional C-arm.

Eventually a T-tube was placed in the bile duct for temporary external drainage of bile. Post-operative T-tube drainage was 100ml/day without any features of cholangitis. The position of the T-tube was confirmed with the T-tube cholangiogram that showed free flow of the contrast into the duodenum with no evidence of the leak. Gradually T-tube drainage reduced and was removed on the 8th post-operative day. Tissue from the gall bladder were sent for histopathology that tested negative for malignancy. The post-operative course of our patient was uneventful, and our patient was discharged on the ninth post-operative day with progressive normalization of the hepatic stasis parameters and the transaminases with normal bilirubin level of 0.8 and normal hepatic enzyme levels. The post-operative cholangiogram was performed. Our patient remains well at 3 months follow-up period

DISCUSSION

Mirizzi's syndrome is a rarely observed disorder which is often confused with cholangiocarcinoma preoperatively. Despite the availability of the latest biochemical and the radiological tests the preoperative diagnosis of the MS presents as a challenging task as the on-table findings of the MS vary considerably.

Mirizzi's syndrome is caused by acute or chronic condition secondary to a single large gallstone or multiple small calculi impacted in the Hartman's pouch or in the gallbladder infundibulum and the cystic ductus often demonstrates a gall stone impacted at the neck of the gall bladder or cystic duct and associated dilatation of the biliary tract. It presents in approximately 0.35% of cholecystectomies performed.[13] The clinical presentation of the MS is very unspecific. The commonest form of the clinical presentation is obstructive jaundice in 60–100% of the cases accompanied by the pain right hypochondrium.[14–16] There are two types of classifications of MS. The pathophysiological process leading to the subtypes of the MS has been explained by means of pressure ulcer caused by the impacted gallstones at the gall bladder infundibulum leading to the inflammatory response causing first the external obstruction of the bile duct and then eventually leading to cholecystodochal fistula or cholecysto-hepatic fistula.[16,18]

Mirizzi's syndrome was first described in 1948 as obstructive jaundice due to a gallstone impacted in the cystic duct or Hartmann pouch compressing the common hepatic duct. McSherry *et al* in 1982 suggested a sub classification of MS into two types.[19]

Mcsherry's Classification:

- Type-I=External Compression on hepatic duct without fistula
- Type-II=External Compression on hepatic duct with fistula

Furthermore, in 1989 a new classification of patients with MS and cholecystobiliary fistulae was presented by Csendes *et al*.[2]

Csendes Classification:

- Type-I=External Compression and obstruction of hepatic duct
- Type-II=Cholecysto biliary fistula with the erosion of the 1/3rd of the circumference
- Type-III=Cholecystobiliary fistula with the erosion of 2/3rd of the circumference
- Type-1V=Total destruction of the hepatic duct
- Type-V=This includes any of the above classification with or without gallstone ileus

Surgical treatment for type-I MS is partial cholecystectomy leaving the neck of the gallbladder in place. In some cases, open or laparoscopic total cholecystectomy may be performed. However some authors consider this a contraindication for laparoscopic cholecystectomy. Surgical treatment of type-I MS is less clearly defined. Corlette and Bismuth have recommended partial cholecystectomy, over suturing of the gallbladder cuff and insertion of a T-tube through the fistula as an adequate treatment for type-2 MS. Choledochoplasty is an acceptable therapeutic approach but the amount of the gallbladder tissue required for this has not been standardized. Furthermore cholecystoduodenostomy has been described and hepaticojejunostomy can also be used if complete destruction of the common hepatic duct occurs. Since the accurate diagnosis of MS preoperatively is not possible due to its nonspecific symptoms. The most common form of presentation is hyper-bilirubinemia and the elevated aminotransferases and leukocytosis.

A very careful surgical planning is required as the incidence of the bile duct injuries in patients operated with MS without preoperative diagnosis could be as high as 17%.[20] Preoperative diagnosis of MS is difficult and can be made in only 8–62.5% of patients.[21–23] The diagnosis of MS is based on the clinical characteristics previously described and a high index of suspicion or surgical intuition, which might be complemented by radiological images and endoscopic procedures.

The diagnostic accuracy of ultrasonography for MS is 29% with a reported sensitivity varying from 8.3–27%.[14,24,25] CT abdomen can be able to identify gallbladder and measure its wall thickness and bile duct dilatation. However the peri-ductal inflammation can be misinterpreted as gallbladder cancer.[24]

Pre-operative diagnosis of MS is important in order to avoid complications during the surgical exploration due to cholecystobiliary fistulas and damage to the common hepatic duct during the surgery. Even ultrasonography and CT abdomen findings are not that accurate and definitive, but findings such as impaction of stone in gall bladder neck and dilatation of biliary tree above the level of gallbladder neck, and abnormal caliber of common bile duct distal to the level of impaction are highly demonstrative of MS.

Endoscopic retrograde cholangiopancreatogram (ERCP) is considered an effective pre-operative method for diagnosing the condition in these patients and can provide a relatively accurate localization and characterization of the cause of the biliary obstruction.[25] Typical findings of MS at ERCP include: (1) mid-bile duct obstruction with dilated proximal common hepatic duct and intra-hepatic ducts combined with normal duct caliber distal to the obstruction, (2) insertion of the cystic duct at the point of obstruction and/or complete obliteration of the cystic duct, and (3) a stone visualized at the point of obstruction either within the cystic duct or the common duct. If a stone is not seen or suspected, however, the findings may be misleading towards a stricture or malignancy. In addition, an interesting finding that suggests MS indirectly during ERCP is the fact that biliary tree dilatation may subside when a patient is placed in an anti-Trendelenburg position.

The on-table findings during the surgical exploration markedly vary as compared to the preliminary diagnosis as this condition is often misdiagnosed as cholangiocarcinoma because of the close resemblance with the preliminary radiological findings. Over 50% of patients with MS are diagnosed during surgery.[24] Despite the availability of the advanced radiological diagnostic tests the peri-operative assessment of the MS presents as a challenging task. Careful and vigilant peri-operative assessment and exploration remains the definite treatment for the majority of the patients and should satisfy three goals, extraction of the stone, removal of the gall bladder and the restoration of the biliary drainage

CONCLUSION

Due to the rarity of the condition and non-specific symptoms, the diagnosis of MS presents as a difficult task because this condition can easily be confused with cholangiocarcinoma, biliary duct stricture or acute cholecystitis. In our patient the ultrasonographic appearance of the gall bladder was misinterpreted as cholangiocarcinoma but CT abdomen was highly suggestive of MS. The correct diagnosis was made on surgical exploration. Tissue samples were sent for histopathology which were tested negative for malignancy. Our patient remained well on a 6thweek's follow-up period with no complications postoperatively.

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