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RESEARCH ARTICLE

AN UNDIAGNOSED MIRIZZI SYNDROME: A NOTE OF CAUTION

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Abstract

Mirizzi syndrome, one of the chronic complications of symptomatic gall stone disease is rare in Western developed countries with an incidence of less than one percent (1%) per year.

The importance and implications of this condition are related to their associated and potentially serious surgical complications such as bile duct injury, and how it should be managed when encountered especially during laparoscopic cholecystectomy.

The patho-physiological sequence of Mirizzi syndrome has been explained by means of a pressure effect caused by an impacted gallstone at the gallbladder infundibulum, leading to an inflammatory response causing first external obstruction of the bile duct, and eventually eroding into the bile duct and evolving to a cholecystocholedochal or cholecystohepatic fistula.

Its pre-operative diagnosis is usually difficult because the clinical signs, the laboratory data and the instrumental findings are not pathognomonic. A high incidence of suspicion is necessary before proceeding to definitive diagnostic imaging techniques.

It used to be diagnosed intra-operatively; but recently in centers where endoscopic retrograde cholangio-pancreatography (ERCP), magnetic resonance cholangiopancreatography (MRCP) facilities are available; it could be diagnosed confidently pre-operatively.

The objective of presenting this paper is to consider this syndrome in the differential diagnosis during surgical treatment either by open or laparoscopic method in patients with long standing symptomatic gall bladder disease with or without features of obstructive jaundice especially at centers where ERCP and or MRCP are not readily available.

Introduction

The Mirizzi syndrome first described by Pablo Luis Mirizzi in 1948 [1] is an uncommon cause of obstructive jaundice secondary to a common hepatic duct obstruction or narrowing caused by a gallstone impacted in the gallbladder's infundibulum.

The condition was classified by McSherry et al into types 1 and 2 in 1982, and later re-classified by Csendes and colleagues in 1989 into classes 1 through 4 [2 3].

This rare complication of gallstones occurs in about 0.1% to 0.7% of patients who have gallstones [4].

Additionally a high coincidence of Mirizzi syndrome and gallbladder cancer has been reported in several studies [5 .6 7].

Depending upon the degree of impingement and the chronicity of the condition, there may be a cholecysto–choledochal fistula [8 9].

Mirizzi syndrome represents a dangerous alteration of the anatomy of the biliary tree and bears the potential to lead to significant morbidity and biliary duct injury, particularly in this laparoscopic era [10 11] especially at centers where biliary abdominal ultrasound are not at expert levels and non-availability of ERCP [12] and MRCP facilities.

This is a fifty-six (56) year-old female with long-standing history of cholelithiasis, diabetes, jaundice, recurrent chills and right upper quadrant abdominal pain.

This case is to sound a note of caution to practicing surgeons who only rely on abdominal ultrasound for the diagnosis of gall bladder stones; to have better awareness [13 14] and to consider this syndrome in the differential diagnosis during surgical treatment either by open or laparoscopic [15 16] method for patients with long standing symptomatic gall bladder disease with or without features of obstructive jaundice.

Case Report:

A fifty six (56) year-old female presented with long-standing history of cholelithiasis, diabetes, recurrent chills and right upper quadrant abdominal pain.
Clinical examination revealed a moderately ill-looking slightly dehydrated, icteric lady; Temperature 39.8^{o}C; BP 100/70mmHg; Pulse 80/min; respiratory rate (RR) 28/min. Abdomen was tender at the upper abdomen maximally at the right hypochondrium.
Her past medical history revealed type II diabetes mellitus controlled on oral medication for the past five (5) years.
Routine complete blood count revealed leucocytosis.
Liver function tests showed SGOT 89 U/L (normal 5–40 U/L), SGPT 115 U/L (normal 5–40 U/L), Alkaline Phosphatase 153 U/L (normal < 106 U/L), γGT 107 U/L (normal < 50 U/L) with bilirubin level 5.7 mg/dl (normal values for total bilirubin 0.2-1 mg/dl).
A complete coagulation profile was done and found to be within normal limits.
Abdominal U/S examination revealed a large stone impacted in a shrunken gallbladder and mildly dilated common hepatic duct (see figs 1 and 2 below).

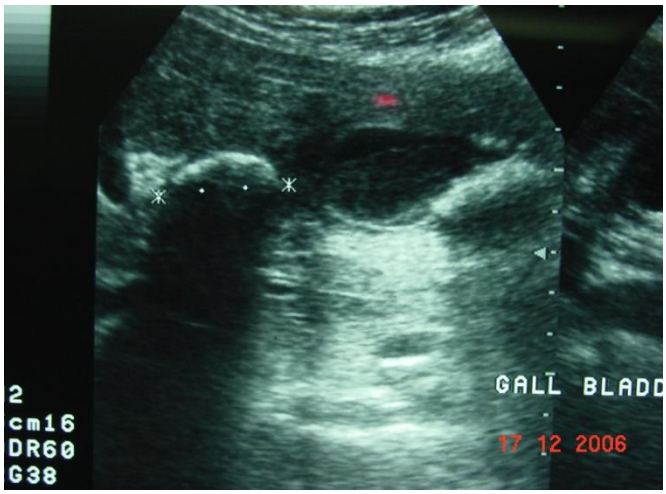

Figure 1

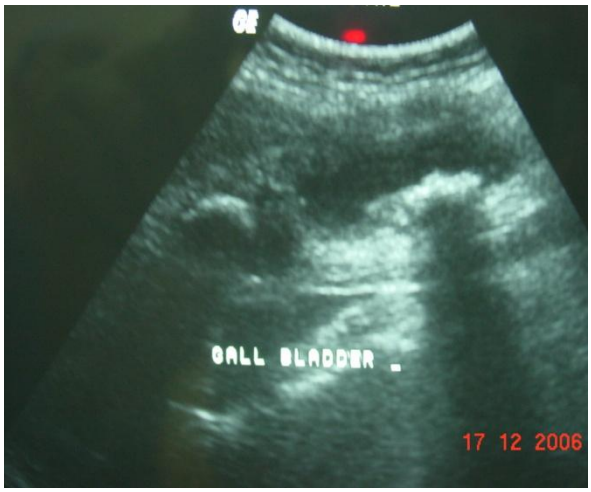

Figure 2

A provisional diagnosis of chronic calcular cholecystitis, impending cholangitis and diabetes mellitus was made.
Patient was admitted to the ward, placed on intravenous fluids, parenteral combination antibiotics, and metronidazole.
She was then scheduled for surgery on the next operating list which was within forty eight (48) hours of admission.
She was offered open cholecystectomy which was carried out through a right Kocher's or subcostal incision.
At operation a shrunken gallbladder was found completely collapsed on an impacted stone at its infundibulum grossly adherent to the lower common hepatic duct and upper part of the common bile duct and causing partial obstruction to their lumen but no fistulous connection depicting type I Mirizzi syndrome.

There was also empyema of the gallbladder (see figures 3 and 4 below).

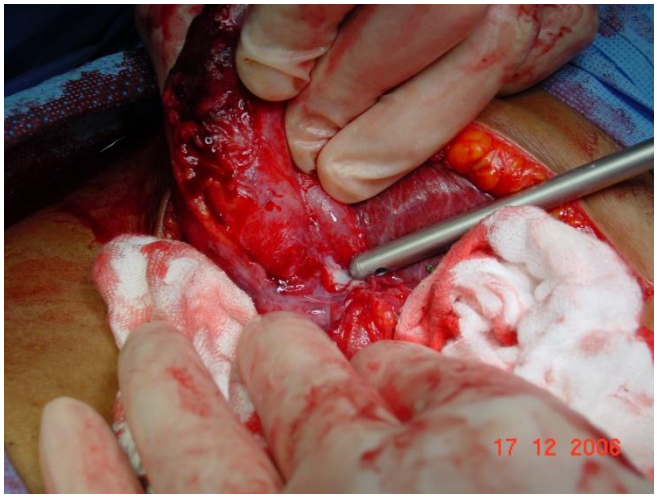

Figure 3

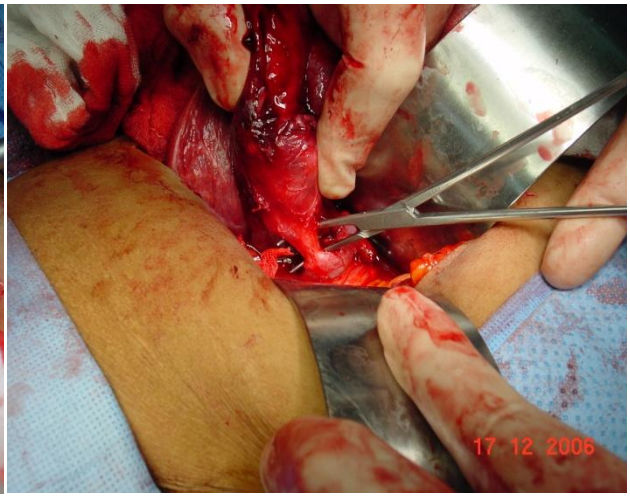

Figure 4

Intra-operative swab culture of the bile was taken but did not yield any growth.
Gallbladder was opened, the impacted stone at its infundibulum extracted; a finger was inserted into the gall bladder thereby aiding its dissection, from the common hepatic duct, and intra-operative cholangiogram was done before the final removal of the gallbladder (see figs 5 and 6 below).

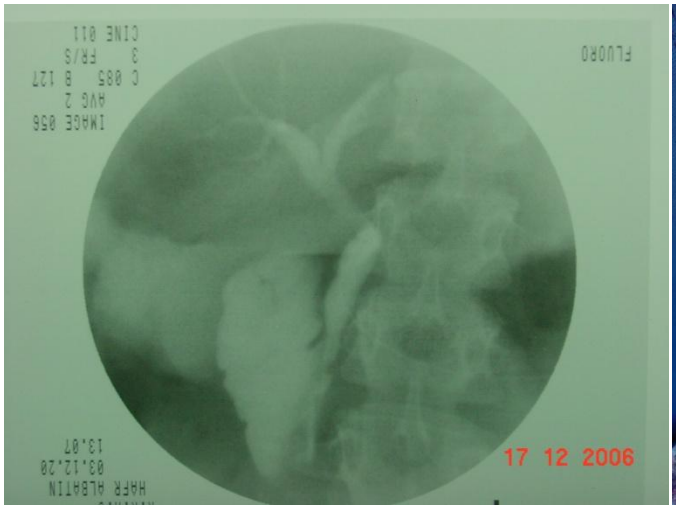

Figure 5

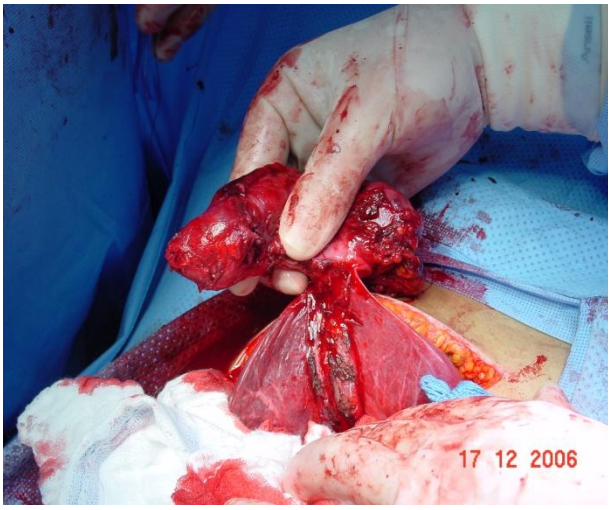

Figure 6

Abdomen was then closed after adequate haemostasis and a haemovac drain was left in-situ.
Post-operative period was uneventful and patient was discharged home on the fifth post-operative day.

Discussion and Conclusion:

Mirizzi syndrome (MS) is a rare complication (frequency about 1%) [4] of chronic cholecystitis and prolonged cholelithiasis and is one of the uncommon causes of obstructive jaundice secondary to a biliary duct obstruction.
Its differential diagnosis includes gallbladder carcinoma, [5 6] sclerosing cholangitis and metastatic lymph nodes.

When one considers that this syndrome occurs in 0.1% to 0.7% of patients who have symptomatic gallstones, the average general surgeon will probably encounter MS at some time during his or her career [17 18 19].

MS consists of inflammatory process of gallbladder wall and direct compression (Mirizzi syndrome type I) or erosion (Mirizzi syndrome types II, III, IV) of the common bile duct and subsequent fistulous formation (Csendes et al1989) [3 8 20 21].
Its pre-operative diagnosis is usually difficult because the clinical signs, laboratory data are not pathognomonic of the syndrome and the usual clinical presentations are essentially those of cholecystitis or choledocholithiasis with epigastric or right upper quadrant abdominal pain, jaundice [22 23 24], or manifest systemic symptoms of fever, chills, tachycardia, and anorexia as in this case presentation and elevated liver function tests.

The condition may be intermittent and relapsing, or fulminant, presenting as cholangitis.

Imaging is essential [25 26] in the pre-operative diagnosis of MS.
Ultrasonography (US) especially in expert hands can diagnose MS pre-operatively.
US typical findings of Mirizzi syndrome are a shrunken gallbladder, with impacted stone(s) in the cystic duct, a dilated intra-hepatic tree, and common hepatic duct with a normal-sized common bile duct.
Computerized abdominal scan (CT) could also be helpful but its main role is to differentiate MS from a malignancy in the area of porta hepatis or in the liver.

Endoscopic retrograde cholangiopancreatography (ERCP) has been used regularly and has allowed almost one hundred (100%) correct pre-operative diagnosis as revealed by Yeh's group [12].

It delineates the cause, level, and extent of biliary obstruction, as well as ductal abnormalities, including fistula. ERCP also offers a variety of therapeutic options, such as stone extraction and biliary stent placement.
Magnetic resonance cholangio-pancreatography (MRCP) is also used in the pre-operative diagnosis of suspected Mirizzi syndrome.

ERCP and MRCP are now considered the gold standards in the diagnosis of MS.

The only problem is the availability of these facilities in some centers especially in developing countries.

In addition to making the diagnosis, the endoscopist [27 28] can stent the jaundiced patient to allow a planned exploration when the patient becomes stable.

The belief is that any patient presenting with gallstones and altered liver function should have some more imaging of the biliary tree after the usual abdominal US such as ERCP and or MRCP before exploration.

Percutaneous cholangiogram (PTC) can also provide information similar to ERCP; however, ERCP has an additional advantage of identifying a low-lying cystic duct that may be missed on PTC.

When the diagnosis is made, there should be a planned open procedure [11 19 21 29] or by laparoscopic method in selected cases.

Type 1 is treated by cholecystectomy [30 31], partial cholecystectomy with stone extraction; however laparoscopic cholecystectomy may be applicable in selected patients.
In fistulous types II-IV MS, cholecystectomy, fistula excision and biliary-enteric anastomosis with Roux-en-Y loop connection appears to be the most appropriate surgical intervention [32 3334 35].
In all cases, a frozen section of the gallbladder wall should be done to rule out co-existent cancer [6 7].

In conclusion, one recommends taking into consideration MS even though rare as a possible differential diagnosis, as one of the long standing cholelithiasis complications; and to approach this syndrome with extreme caution [13 14 21 36 37].
One dissuades from the laparoscopic approach if the diagnosis is made pre-operatively unless the surgeon has the laparoscopic skill; otherwise the patient could be subjected to greater risk of common bile duct injury and its later consequences.

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