

Original Article

Incidence, Diagnosis and Management of Mirizzi Syndrome in Kuwait

Abdul-rahman Al-Mutairi¹, Deena Al-Refai², Obaid Al-Harbi¹, Mahmoud Marzouk¹

¹Department of Surgery, Farwaniya Hospital, Kuwait

²Department of Radiology, Farwaniya Hospital, Kuwait

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ABSTRACT

Aims/Background: Mirizzi syndrome is an obstructive jaundice associated with pressure on the common hepatic duct from gallstones in Hartmann's pouch or the cystic duct. The stones sometimes erode through the main duct, leaving a fistula. We reviewed cases encountered between January 2001 and November 2002.

Design: Retrospective review of seventeen patients with diagnosis of Mirizzi syndrome managed in the surgical wards of Farwaniya Hospital. Patients were fully investigated including liver function tests, abdominal ultrasonography, ERCP and/or intra-operative cholangiography.

Results: During the study period 625 cholecystectomies

were performed. Out of these, 17 were found to have Mirizzi syndrome which accounts for an incidence of 2.72%. In 11 patients, jaundice resulted from gallstone pressure (type I) and in the remaining six patients, the stones had eroded into the common hepatic duct producing a fistula (type II).

Conclusion: The diagnosis can usually be made preoperatively, especially if a large single stone is seen in conjunction with a dilated common hepatic duct and normal caliber common bile duct. We favor partial cholecystectomy, adding choledochoplasty using the gallbladder remnant, to close the fistula in type II cases.

KEYWORDS: diagnosis, management, Mirizzi syndrome

INTRODUCTION

Pablo Mirizzi reported the syndrome in 1948 for the first time, thus the syndrome was named after him. He described the syndrome as a partial or spastic obstruction of the common hepatic duct secondary to an impacted gallstone in the cystic duct or infundibulum of the gall bladder^[1].

In 1982, McSherry *et al*^[2] suggested a sub-classification of the syndrome into two types: type I which involves external compression of the common hepatic duct by a stone impacted in the cystic duct, and type II which occurs when a cholecystocholedochal fistula is caused by stone migration into the common hepatic duct. Further modification was suggested by Csendes *et al*^[3]. In this modification, type II is a cholecystocholedochal fistula that involves less than one third of the circumference of the bile duct, type III is a fistula involving up to two thirds of the duct circumference, and type IV is complete obstruction of the bile duct.

In the literature, Mirizzi syndrome is encountered in 0.1% - 1% of all cholecystectomies performed^[4-7].

This study aims to recognize and evaluate the frequency of this problem, with special emphasis on the preoperative and intra-operative diagnosis, the approach to its management and follow-up.

PATIENTS AND METHODS

Seventeen patients were diagnosed with Mirizzi syndrome at our hospital, between January 2001 and November 2002. The age, gender, incidence, admission complaint including the duration of symptoms and previous attacks were evaluated. During this period a total of 625 cholecystectomies were performed in the unit.

All patients underwent a complete assessment which included hematology, coagulation, biochemical profile including complete liver function tests, and abdominal ultrasonography. They also underwent pre-operative ERCP and/or intra-operative cholangiography.

The operative strategy was determined following a complete assessment of the stage of disease at laparotomy. Accordingly, patients were classified into two types as suggested by Mc Sherry^[2]. All patients were followed up with serial liver function tests for six months post operatively.

RESULTS

There were ten male and seven female patients with a mean age of 49 years (range 32 - 77 years). They accounted for 2.72% of a total of 625 cholecystectomies done during the same time period.

Address correspondence to:

Dr Abdul-Rahman Al-Mutairi, P.O. Box 18115, Zip code 81002, Farwaniya, Kuwait. E-mail :arahman3@hotmail.com



Fig. 1: ERCPtype I MS

All patients in this study presented late with long standing biliary symptoms. In ten patients with no previous attack the duration of symptoms ranged between 5-60 days. The remaining cases presented with symptoms of less than five days duration and had more than one previous attack of biliary symptoms in the past. The clinical presentation included right upper quadrant abdominal pain, jaundice, and fever in 14, 14, and 10 cases respectively.

An admission diagnosis of obstructive jaundice was made clinically, in ten patients, acute cholecystitis in six patients and biliary pancreatitis in one patient. Serum bilirubin and liver enzymes were found to be elevated in all 17 patients.

Ultrasound allowed the correct diagnosis of cholelithiasis in all patients. A large stone, more than 1.5 cm was seen in 12 out of 17 patients. The sonographic findings of acute cholecystitis were reported in 13 cases and common hepatic duct (CHD) dilatation in 11 cases; two cases were diagnosed by ultrasound as type II Mirizzi syndrome, a stone impaction in Hartman's pouch or in the cystic duct was detected in seven cases.

CT allowed the diagnosis to be made in one case with liver cirrhosis and associated hepatocellular carcinoma suspected to have lymph node compression of the CHD by ERCP. In fact, a large stone impacted in the cystic duct with type I Mirizzi syndrome was detected.



Fig. 2A: ERCPtype II MS

ERCP demonstrated extrinsic compression of the CBD (type I Mirizzi syndrome) in seven cases and cholecystobiliary fistula with a stone eroding into the CBD in four cases. Operative diagnosis of type I was clearly evident in two cases with stone impaction in the cystic duct and compression of the CBD with CHD dilatation.

During surgery, Calot's triangle was found to be obliterated by dense fibrosis and/or the CBD was eroded by a stone in almost all the patients who underwent surgical intervention.

The gall bladder was approached from the fundus, stones were evacuated by transverse cholecystotomy and the interior of the gallbladder was inspected for the presence and extent of erosion involving the CBD. An intra-operative cholangiogram, when indicated, was then obtained at this stage (Fig.3) and was performed in nine cases out of 17 patients.

Type I and II Mirizzi syndrome were managed by partial cholecystectomy, while type II also underwent CBD exploration through a fresh choledochotomy, gallbladder flap choledochoplasty with a drainage T-Tube.

In our study, laparoscopic cholecystectomy was initially attempted in nine patients. Six of them had Mirizzi syndrome diagnosed intraoperatively and were converted to an open procedure because of the unexpected difficult anatomy. Three of these cases were diagnosed as Mirizzi syndrome preoperatively. Cholecystectomy was completed laparoscopically in only one of these patients, due to the presence of soft adhesions between the gallbladder neck and the CHD



Fig. 2B: ERCP type II MS

Open operative approach was undertaken for the remaining cases correctly diagnosed preoperatively as Mirizzi syndrome. Three of these patients were type II and two were type I.

Intraoperative cholangiography helps to avert injury to important structures and was helpful in this study for those patients who were not diagnosed preoperatively (two cases each with type I and type II Mirizzi).

The operative diagnosis correlated well with the preoperative diagnosis in all patients who were suspected preoperatively as type I (5 cases) and type II (3 cases). ERCP and biliary stenting alone was the only therapeutic method available for three cases determined to be poor surgical candidates.

All patients remained well six months postoperatively with normal liver functions.

DISCUSSION

The higher incidence of Mirizzi syndrome in this study is attributed to a delayed presentation of patients to hospital and long standing biliary symptoms in most of the patients, as noted by other authors^[9]. Although no specific clinical course or symptomatology is pathognomonic for Mirizzi syndrome, certain clinical and biochemical parameters may be indicative of or provide a high degree of suspicion for this syndrome^[10].

In the authors' experience, delayed presentation with a previous history of repeated similar attacks, an admission diagnosis of obstructive jaundice with or without acute cholecystitis, high bilirubin level and elevated alkaline phosphatase are clinical findings which are highly suspicious for this syndrome.

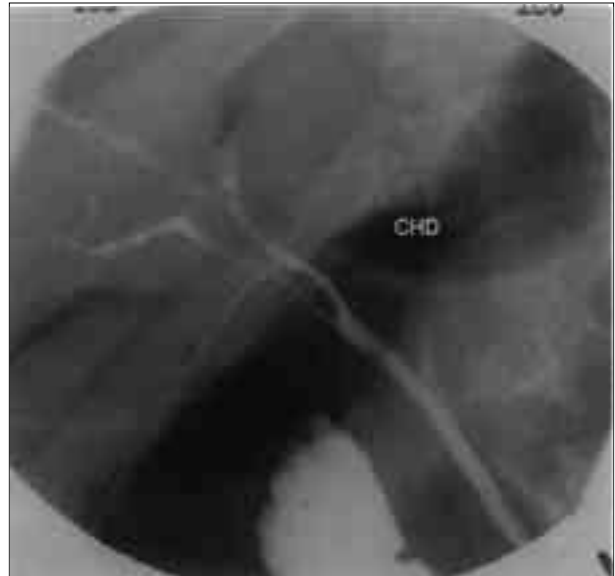


Fig. 3: intraoperative cholangiogram type I MS

Ultrasound is the commonest screening investigation in patients who present with biliary symptoms. Although there are lists of ultrasound features for Mirizzi syndrome, they have a low predictive value^[2,8,11,12]. Most commonly, a single large stone in the neck of the gallbladder raises the suspicion, but patients with multiple small stones may also develop the syndrome if the stones become impacted in the cystic duct or neck of the gallbladder. In our experience and that of others, a large stone more than 1.5 cm, impacted in the neck of the gallbladder or cystic duct and a dilated common hepatic duct are findings strongly suggestive of Mirizzi syndrome^[13,14]. Two cases of type II Mirizzi syndrome were clearly identified by ultrasound.

A normal-sized CBD distal to the calculus was seen in a majority of our cases similar to that reported in the literature^[15,16].

ERCP is the investigation currently recommended for further evaluation of the biliary tree^[2,3,17]. In our study, ERCP demonstrated extrinsic compression of the CBD (type I) in seven cases (Fig. 1) and cholecystobiliary fistula with stone eroding into the CBD (type II) in four cases (Fig. 2). The operative diagnosis correlated with the preoperative findings detected at ERCP in all cases.

ERCP and biliary stenting may be the only therapeutic method available for cases determined to be poor surgical candidates^[18] (as seen in three of our patients).

The role of CT with intravenous contrast remains to be evaluated. In our study it was used to exclude the diagnosis of obstructive jaundice secondary to portahepatic lymph node compression^[16].

Intraoperative cholangiography helps to avert injury to important structures and should be performed, if possible, in cases suspected to have Mirizzi syndrome during surgery. It will demonstrate anatomic variations, stones and fistulas of the biliary system^[14].

To date, a variety of techniques have been described in the surgical management of this syndrome. Our management in majority of patients with type I disease consisted of partial cholecystectomy (leaving behind the posterior layer of Hartmann's pouch), as recommended by others^[9], because the cystic duct is frequently occluded and obscured by inflammatory changes in the region of Calot's triangle. Type II disease was managed by subtotal cholecystectomy (leaving a small cuff of the gallbladder), gallbladder flap choledochoplasty and T-Tube choledochostomy through a fresh choledochotomy site as described by Sandblom *et al* (1975)^[20].

Bilio-enteric anastomosis was not performed in our study, due to the absence of a biliary stricture in this patient population.

Although successful laparoscopic cholecystectomy in the treatment of Mirizzi syndrome has been reported^[21,22], it has been suggested by others that Mirizzi syndrome is a contraindication to laparoscopic management^[12].

Accordingly, many authors still prefer the conventional celiotomy approach for the management of these patients to avoid any laparoscopy related complications. This has been our preference in the management of such patients^[23, 24].

The complex anatomy of an unexpected Mirizzi syndrome can create a definite hazard to the CBD at the time of laparoscopic surgery^[25]. In our study, only one patient's cholecystectomy was completed laparoscopically.

CONCLUSION

The incidence of Mirizzi syndrome in Kuwait seems to be higher than that reported in the literature. The diagnosis can usually be made preoperatively, especially if a large single stone is seen in conjunction with a dilated common hepatic duct and normal caliber common bile duct on ultrasound and/or confirmed by ERCP. When this syndrome is diagnosed or is strongly suspected preoperatively, an open biliary operation is the procedure of choice. Intraoperative cholangiography should be performed, if possible, in cases suspected during surgery. We favor partial cholecystectomy in type I cases, supplemented by choledochoplasty using the gallbladder remnant, to close the fistula in type II cases.

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