

OriginalArticle

Mirizzi syndrome: An unusual presentation of cholelithiasis

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ABSTRACT

Objective

To evaluate the diagnostic features, treatment strategies and post operative complications in Mirizzi Syndrome.

Patients and Methods

Our experience with 12 cases with Mirizzi syndrome is presented. All were diagnosed at exploration and managed surgically.

Results

Morbidity rate after surgery was 58.35% with the mortality of 8.35%. At follow-up, all patients were symptoms free after 3 months of surgery.

Conclusion

Mirizzi syndrome is rarely diagnosed preoperatively and ultra-sound even in experienced hands does not provide any clue to the diagnosis. The index of clinical suspicion should be high in long standing cholelithiasis. Surgery provides cure and definitive per operative diagnosis. (Rawal Med J 2010,35:).

Key words

Jaundice, Mirizzi syndrome, cholecystectomy.

INTRODUCTION

Mirizzi syndrome is a surprising complication of gall stone disease and occurs in about 1% of all patients with cholelithiasis.¹ It was first described in 1948 and is characterized by impaction of stones in the cystic duct or neck of the gall bladder, consequent into mechanical obstruction of the common hepatic duct (CHD) and frequent clinical presentation of intermittent or constant jaundice.² Pathophysiologically either the chronic and/or acute inflammatory changes lead to contraction of the gallbladder, which then fuses with; and causes secondary stenosis of the common hepatic duct or large impacted stones lead to cholecystocholedochal fistula formation secondary to direct pressure necrosis of the adjacent duct walls. Based on pathophysiology, findings of endoscopic retrograde cholangiopancreatography (ERCP) and percutaneous transhepatic cholangiography (PTC) McSherry et al³ suggested a simple two stage classification. Type I is simple external compression of the CHD, whereas type II involves the presence of a cholecystocholedochal fistula. These are further sub-classified (Csendes classification)⁴ into the following groups to aid surgical treatment of patients: Type I-No fistula present; Type IA-Presence of the cystic duct, Type IB-Obliteration of the cystic duct; Types II-IV-Fistula present, Type II-Defect

smaller than 33% of the CBD diameter, Type III-Defect 33-66% of the CBD diameter and Type IV-Defect larger than 66% of the CBD diameter. The aim of this study was evaluate diagnostic features, treatment strategies and post operative complications in Mirizzi Syndrome at our institution.

PATIENTS AND METHODS

During a period of 10 years, 1995-2005, 800 patients underwent cholecystectomy for cholelithiasis in surgical Unit II Liaquat University of Medical and Health Sciences, Jamshoro. Twelve cases were identified as having Mirizzi syndrome. The records of these patients were then reviewed carefully for clinical symptoms, diagnostic methods, surgical procedures performed, complications and follow up. Every patient has had ultrasound done from sonologist of our institute. All these patients were operated by consultant having clinical experience of at least 5 years.

RESULTS

Out of 12 patients, 5 (42%) were male and 7 (58%) were female with mean age of 42 years.

Review showed that the presenting features were in no way distinct and were as we see in cholelithiasis/cholecystitis and includes recurrent pain or persistent dull ache at right hypochondrium (100%) associated with nausea and vomiting in about 60% of cases, frequent bouts of swinging temperature (66.66%), jaundice or past history of jaundice (50%). Intraoperatively, to categorize the patients into various sub types of MS, we followed Csendes' classification (Table). Patients with type III and IV having large fistulous opening after extraction of the gall stones, we performed Roux-en-Y choledochojejunostomy. Common finding in all cases includes dense adhesion at Calot triangle that required careful dissection. Intraperitoneal drainage was done in all cases.

Post operatively, for patients with type I MS, intra-peritoneal drain was removed on 3rd post operative day, while for patients with T-tube drainage, removal of T-tube precede by the intraperitoneal drain on 5th and 7th day respectively. For type III and IV, intraperitoneal drain was removed on 5th postoperative day.

Table 1. Details of patients.

S/N	Age/sex	Symptoms	Bil (mg %)	Alp (mg %)	Imaging	Pr-op diagnosis	Op findings	Procedure	Complications
1	36/F	J,P	4	450	U/S	Cholelithiasis	MS type I	Par-Chole	BL
2	32/F	P,F, N, V	1.2	210	U/S	Cholecystitis	MS type II	Chole+T-Tube	CH
3	33/F	J,P,F	6.5	700	U/S	Cholecystitis	MS type III	Chole+CJ	WI
4	36/F	P,N,V,F	2.7	570	U/S	Cholecystitis	MS type IV	Chole+CJ	CH/Expired
5	41/F	P,N,V,F	2.9	450	U/S	Cholecystitis	MS type I	Par-Chole	-
6	46/F	P,N,V	2.5	650	U/S	Cholelithiasis	MS type I	Chole	WI
7	46/F	J,P,F	6	575	U/S	Cholecystitis	MS type I	Par-Chole	
8	63/M	P,F,J,N,V	4	525	U/S	Cholecystitis	MS type II	Chole+T-Tube	CH
9	51/M	P,N,V	3.75	480	U/S	Cholelithiasis	MS type I	Par-Chole	-
10	43/M	J,P,F	4	570	U/S	Cholecystitis	MS type II	Chole+T-Tube	-
11	40/M	J,P,F	4.2	345	U/S	Cholecystitis	MS type II	Chole+T-Tube	-
12	29/M	P,N,V	1.4	230	U/S	Cholelithiasis	MS type I	Chole	B

Bil=Bilirubin mg%, Alp= Alkaline phosphatase, J=jaundice, P=pain, N=nausea, V=vomitting, F=fever. BL=biliary leak, CH=cholangitis, WI=wound infection, U/S=ultrasonography

Two patients developed biliary leakage, and in one case re-exploration was done. This patient died of multiple organ failure. Overall morbidity was 58.35% and mortality 8.35%. Histopathology of resected specimen showed chronic cholecystitis in all cases (Table 1).

DISCUSSION

Pablo Luis Mirizzi (1893-1964), the father of operative cholangiography, described Mirizzi Syndrome (MS) in 1948 as gall stone impacted in the cystic duct or Hartmann pouch, compressing the common hepatic duct.² Mcsherry in 1982 sub-classified this clinical entity into two sub-types.³ In our study, the majority of patients were of type I in contrast to Csendes series⁴ where majority were of type3 and this may simply be attributed to less number of patients in our study.

Clinical diagnosis of MS is difficult, as classical findings of a contracted GB dilated intra hepatic ducts but normal caliber CBD was only seen in 8.3%.⁵ However, in this series ultrasound (U/S) demonstrated contracted gall bladder and normal caliber CBD, and normal intrahepatic ducts. In spite of all modern diagnostic modalities, diagnosis is made only during operation.⁶ Intra operatively, the presence of MS can be suggested by the finding of intense adhesions between the GB and the CHD in the area of Calots' triangle.⁷ It is also postulated that a long parallel cystic duct or its low insertion into CBD predispose to MS.^{8,9} However, we found it difficult to demonstrate this anatomical variant because of severe inflammation in Calots' triangle.

Abdominal U/S had 0 % sensitivity in diagnosing MS in our series. This is in keeping with Csendes series, where U/S revealed bile duct dilatation in 81% of patients and raised suspicion of MS in 27%.⁴ Some authors consider preoperative diagnosis essential in avoiding CBD injuries.¹⁰⁻¹² Others have concluded that with a cautious intra operative approach to

periductal inflammation and judicious dissection, it is not necessary for successful management.^{13,14} Additional imaging is often needed to obtain details of the biliary pathology because of lack of sensitivity of U/S. However, we could not proceed for further imaging diagnostic modalities such as ERCP and MRCP due their unavailability.

There have been reports of association with GB carcinoma.^{15,16} Although we did not find GB carcinoma in any of our cases, but where this happens, a more complex surgical procedure should be avoided.^{17,18} Favorable outcome is also reported in roux-en-y cholecystocholeodochojunostomy with type III and type IV.¹⁸ We achieved satisfactory results with partial/complete cholecystectomy for type I and type II. We performed bypass procedure for type III and IV and obtained almost identical results with other series.¹⁸

Role of laparoscopic approach in the treatment of MS remains controversial. Condition is not suitable for laparoscopic surgery because the inflammatory tissue in the area of Calots' triangle carries a high operative risk during dissection.^{19,20} However, laparoscopic treatment of MS is technically feasible and safe only in MS type I²¹ and that is why some still propose the laparoscopic surgery.²² For high risk patients, ERCP can be used for biliary drainage by endoscopic sphincterotomy and placement of a stent in the CBD.²³ Moreover, placement of a nasobiliary catheter in conjunction with cholangioscopy for electro hydraulic lithotripsy has been reported.²⁴

CONCLUSION

Even when state of the art diagnostic facilities are available, preoperative diagnosis of MS is not an easy task and continues to be a challenge for the surgeons. A high index of suspicion must be maintained during dissection of obscured Calots' triangle. Vigilance to the presence of MS and use of intra operative cholangiogram help in appreciating the full extent of the disease and selection of an appropriate procedure.

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