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MIRIZZI'S SYNDROME: A DIAGNOSTIC DIALEMMA

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ABSTRACT

Mirizzi's syndrome is characterized by external compression of extra hepatic biliary system by impacted gall stone in cystic duct or infundibulum of gall bladder leading to obstructive jaundice and development of cholecystobiliary fistula in advanced cases. Csendes has described 4 types of the syndrome. In spite of modern investigations like MRCP/CTS available, preoperative diagnosis is not always obtained and it may present as surprise during cholecystectomy. We have done a retrospective study of 7 cases of Mirizzi's syndrome, in which preoperative diagnosis was obtained only in 3 cases. Cholecystectomy was performed in 2 cases, while 3 cases needed choledochoplasty in addition. Hepaticojejunostomy was needed in addition to cholecystectomy in 2 cases of TypeIII /IV respectively. Utmost care must be exerted during difficult cholecystectomy cases keeping possibility of Mirizzi's syndrome as culprit and appropriate surgical procedure performed for the treatment.

KEY WORDS Mirizzi, Hepatic duct, Cholecystobiliary fistula, Gall stone, Cholecystoplasty, Hepaticojejunostomy.

INTRODUCTION

Mirizzi's syndrome (MS) was first described in 1948 by Pablo Mirrizi as an external compression of the extrahepatic biliary system by an impacted stone in the gallbladder neck or the cystic duct leading to obstructive jaundice [(1)]. It is an unusual complication of prolonged cholelithiasis, with prevalence from 0.05% to 2.7% among patients with symptomatic gall stones [(2, 3)]. It presents a spectrum that varies from extrinsic compression of the common hepatic duct to

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the presence of cholecystobiliary or cholecystoduodenal fistula. Therefore, there may be high risk of complications involving common hepatic duct or vascular injury during cholecystectomy. [4, 5, 6].

Csendes et al [7] has classified Mirizzi's syndrome in 4 types. Type 1 has extrinsic compression of common hepatic duct (CHD) by gall stone impacted in cystic duct/ infundibulum. Type 2 has cholecystobiliary fistula with 1/3 diameter of circumference of CHD. Type 3 is present when the diameter of cholecystobiliary fistula is 2/3 of diameter of CHD while Type 4 is seen when the diameter involves entire circumference of CHD. Simple cholecystectomy may be sufficient in Type 1 while cholecystoplasty will be necessary in addition in Type 2 cases. Type 3 & 4 cases usually need cholecystectomy and hepaticojejunostomy.

There is always challenge to surgeon due to illusive anatomy due to chronic inflammatory process of the disease and an eventual communication between gallbladder and common hepatic duct [8]. A pre-operative diagnosis is crucial to determine the appropriate surgical management. In spite of advances in diagnostic modalities like CT scan, MRI, EUS, diagnosis may not be perfectly present in each case preoperatively and it may come as a surprise on operation table. This study presents a retrospective study of the diagnostic work-up and outcome of 7 cases of Mirizzi's syndrome treated within a single institution over last three years.

MATERIALS AND METHODS

This is a retrospective study of 7 patients who presented and treated in our institution from July 2017 to August 2020. Detailed history regarding presenting symptoms, history of jaundice and pain in abdomen, medical treatment and follow-up information was recorded from clinical records. Ultrasonography of abdomen with pelvis was performed in all patients. MRCP was performed in 5 patients and CT scan of abdomen in 3 patients. Preoperative diagnosis was present in 3 patients by presence of cholelithiasis and dilatation of CHD above level of biliary obstruction and normal duct diameter below the obstruction. All patients underwent cholecystectomy with additional treatment depending on type of Mirizzi's syndrome and were followed up from 3 months to 24 months.

OBSERVATIONS AND RESULTS

Seven patients who were diagnosed postoperatively with Mirizzi's syndrome and had complete follow-up data were evaluated and included in this study.

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The mean age was 47 years, ranging from 37 to 72 years. All patients had a history of jaundice and pain in abdomen. Duration of symptoms in all cases was more than two years. All patients had obstructive jaundice and raised serum alkaline phosphatase level.

TABLE 1- Ultrasound showing calculus size for different patient.

Patient	Calculus size on USG in cm
i	2.4
ii	2.2
iii	1.9
iv	2.1
v	1.8
vi	2.4
vii	2.3

Table 2 - Two patients were with Csendes type I while three patients were with type II respectively. One patient each was of Csendes type III and type IV respectively. One patient underwent Laparoscopic cholecystectomy and one underwent open cholecystectomy, both being Type 1 cases. Open cholecystectomy with choledochoplasty was done in three patients of Type II. One case of Type III underwent lap converted open cholecystectomy with hepaticojejunostomy and one case of Type IV underwent open cholecystectomy with hepaticojejunostomy. Postoperative recovery was uneventful in all patients.

Sr no.	Age	Sex	Clinical features and duration	Investigations	Surgery	Preop diagnosis present	Type of Mirizzi's syndrome	Complications
1	33	M	Painless jaundice	CT Scan MRCP	Open cholecystectomy with choledochoplasty	No	II	Nil
2	37	M	Pain in abdomen and itching	CT Scan	Open cholecystectomy	No	I	Nil
3	44	M	Painless jaundice	MRCP	Laparoscopic cholecystectomy	No	I	Nil

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4	40	F	Pain in abdomen and jaundice	MRCP	Open Cholecystectomy with choledochoplasty	Yes	II	Nil
5	72	F	Painless jaundice and itching	MRCP	Lap converted cholecystectomy with hepaticojejunostomy	Yes	III	Nil
6	44	M	Painless jaundice and itching	MRCP	Open cholecystectomy with hepaticojejunostomy	Yes	IV	Nil
7	52	M	Painless jaundice and itching	CT scan	Open Cholecystectomy with choledochoplasty	No	II	Nil

DISCUSSION

Mirizzi's syndrome has been an intriguing condition, presenting as a diagnostic dilemma from time to time, occurring in about 1 – 2% of cases of gall stone disease. [9] It is characterized by painless obstructive jaundice, caused by extrinsic compression of extra hepatic biliary ducts by impacted gall stone in cystic duct/ infundibulum and physical examination may be inconclusive. It may lead to cholecystobiliary fistula in advanced cases. The disease can mimic gall bladder cancer causing considerable diagnostic difficulties. [10] Indeed incidence of gall bladder cancer is around 1% of cases of MS. [9]

Despite advances in radiological modalities, diagnosis of MS is still difficult and is often made intraoperatively. [9] There is usually evidence, at ultrasonography, of a gallstone impacted in the cystic duct/gallbladder infundibulum causing external obstruction of the CBD with consequent dilation of the intrahepatic biliary tree [1]. The Common bile duct below the obstruction is usually not dilated. The diagnosis needs to be confirmed preoperatively by MRCP/CTS. MRCP provides equivalent information to ERCP without inherent risk of complications and should be the investigation of choice. [9] Clemente et al [10] has suggested preoperative ERCP with placement of stent which allows reducing the pressure in CBD before operative intervention [10].

However the diagnosis may not be apparent in all cases and MR may present as diagnostic surprise on operation table, especially during difficult cholecystectomy. One must keep Mirizzi's syndrome as possible culprit in all such cases and proceed with extreme caution during

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surgery, as there is always a chance of bile duct injury during cholecystectomy in cases of Mirizzi's syndrome. Various open surgical techniques for MS include partial cholecystectomy with primary closure, Roux-en-Y Hepaticojejunostomy, cholecystoduodenostomy and choledochoplasty with gall bladder flap [(9).] In our series, two patients of Type I syndrome underwent cholecystectomy, one laparoscopic and one open. Open cholecystectomy was to be supplemented with choledochoplasty in three cases of Type II. All these three cases had dense adhesions and needed careful dissection and the diagnosis was evident only on operation table in 2 cases. One case of Type III subjected to laparoscopic cholecystectomy needed conversion to open cholecystectomy and hepaticojejunostomy in addition. One case of Type IV was subjected to open cholecystectomy and required hepaticojejunostomy in addition. Both these cases had cholecystobiliary fistulas, concealed due to fibrous adhesions, which were revealed only after delicate dissection of the Calot's triangle and due to large size needed hepaticojejunostomy. All cases of choledochoplasty and hepaticojejunostomy (Figure 1.) were supplemented by stent insertion across the repair/ anastomosis.

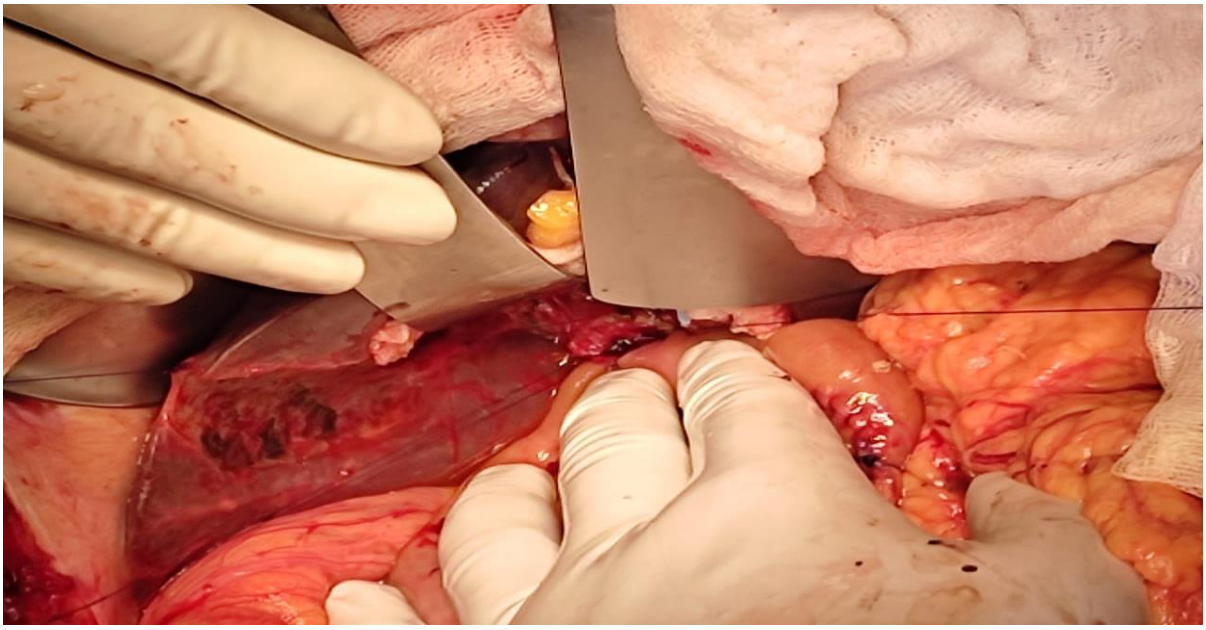

Figure 1 – Showing Hepatico – Jejunostomy Anastomosis.

Thus, it is important to realize that Mirizzi's syndrome may not be evident after all preoperative investigations. It may constitute diagnostic dilemma and spring surprise as operative diagnosis.

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Hence, in any case of difficult cholecystectomy, one must proceed cautiously with the dissection and Mirizzi's syndrome must be kept at back of mind as a possible underlying condition. Laparotomy allows better evaluation of biliary anatomy, avoiding any risk of bile duct injury and with the advantage of more accurate surgical procedure.[10]. Choice between cholecystectomy, choledochoplasty and hepaticojejunostomy should be evaluated carefully by the surgeon taking into account the extent of CBD involvement [10].

SUMMARY

This is a retrospective study of 7 cases of Mirizzi's syndrome which were operated in a tertiary care institution over a period of 3 years.

The mean age was 47 years, ranging from 37 to 72 years. All patients had a history of jaundice and pain in abdomen. All patients had obstructive jaundice and raised serum alkaline phosphatase level.

Ultrasonography of all patients showed calculus of size of minimum 1.8 cm in neck region of gall bladder [Table 1].

MRCP was performed in 5 patients preoperatively which confirmed the diagnostic findings of dilated bile duct above the level of obstruction and normal caliber of CBD below the obstruction.

One patient underwent Laparoscopic cholecystectomy and one underwent open cholecystectomy, both being Type 1 cases. Open cholecystectomy with choledochoplasty was done in three patients of Type II. One case of Type III underwent lap converted open cholecystectomy with hepaticojejunostomy and one case of Type IV underwent open cholecystectomy with hepaticojejunostomy [Table 2].

Mirizzi's syndrome may not be diagnosed preoperatively in spite of all appropriate investigations. It may constitute diagnostic dilemma and spring surprise as operative diagnosis. Hence, one must proceed cautiously with the dissection in any case of difficult cholecystectomy and Mirizzi's syndrome must be at back of mind as a possible underlying condition in all such cases.

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