Mirizzi's Syndrome

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Citation

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Abstract

The Mirizzi Syndrome is a rare cause of obstructive jaundice caused by an impacted gallstone either in the cystic duct or the gall bladder resulting in narrowing of the extrahepatic bile duct accompanied in severe cases by cholecystocholedochal fistula. Clinical features are not diagnostic. Laboratory investigations are supportive. A preoperative ERCP is usually mandatory to establish the diagnosis. Definitive surgical therapy requires an operation which is tailored for each individual presentation, ranging from cholecystectomy to bilio-enteric anastomoses and choledochoplasties.

INTRODUCTION

Mirizzi's Syndrome was first described by P. L. Mirizzi in 1948. [1] It was described as an unusual presentation of gallstones that, when lodged in either the cystic duct or the Hartmann pouch of the gall bladder, externally compressed the common hepatic duct causing symptoms of obstructive jaundice.

SURGICAL ANATOMY

A brief review of surgical anatomy is essential for proper interpretation of ERCP pictures as well as intraoperative orientation. The gall bladder usually comprises the fundus, body, infundibulum and neck. From the neck, the cystic duct runs until it converges with the CHD to form the CBD which eventually joins the descending duodenum via the ampulla of Vater. The cystic artery which supplies the gall blader usually runs parallel to the cystic duct. Calot's triangle is formed by the inferior border of the liver, the CHD and the cystic duct. The right hepatic or the cystic artery is seen in Calot's triangle.

PATHOPHYSIOLOGY

Multiple large gallstones can reside chronically in Hartmann's pouch of the gall bladder, causing inflammation, necrosis, scarring and ultimately fistula formation. As a result the CBD becomes obstructed by either scar or stone, leading to jaundice. These sequelae are not seen as distinct and separate steps but as part of a continuum. [2, 3].

McSherry et al. proposed a 2-stage classification based on the results of ERCP and PTC [4].

Type I is simple external compression of the CHD, whereas type II involves the presence of a cholecystocholedocal fistula.

Type I is simple external compression of the CHD. It does not have a fistula and can be further classified into

Type 1 A – Presence of the cystic duct and

Type 1B – Obliteration of the cystic duct.

Type II – which has a fistula is further classified as

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

Csendes reported that 11% of their patients with Mirizzi Syndrome had type I, 41% had type II, 44% had type III and 4% had type IV lesions [5].

CLINICAL FEATURES

Mirizzi Syndrome has no consistent or unique clinical features that distinguish it from other common forms of obstructive jaundice. Symptoms may be recurrent cholangitis, jaundice, right upper quadrant pain and abnormal hepatic serum biochemical findings and may or may not be present [6]. Co-existence of Mirizzi Syndrome with adenomyomatosis of the gall bladder and carcinoma of the gall bladder has been reported [7,8,9].

DIAGNOSIS

Diagnosis of Mirizzi Syndrome prior to surgery is critical

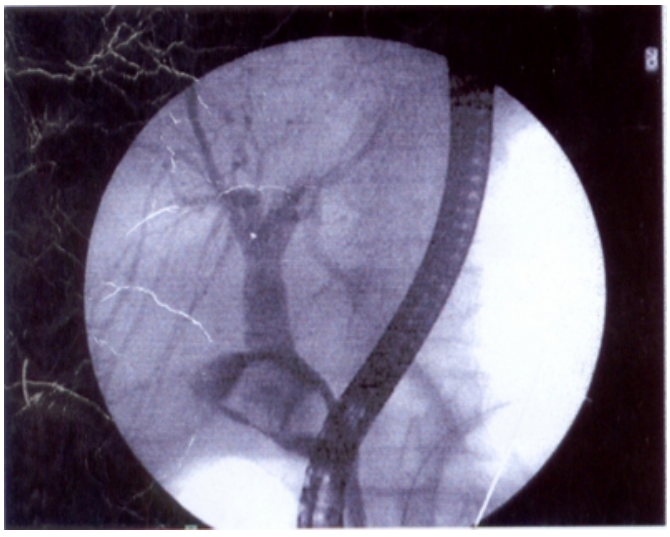
because awareness can prevent intraoperative complications. Dense adhesions and fibrosis in the Calot's triangle make the dissection very complicated. It is always preferable to convert a lapasoscopic procedure into an open procedure. Various imaging modalities have been utilized to make a preoperative diagnosis of Mirizzi Syndrome.

Ultrasound findings include (1) an impacted calculus in the Hartmann pouch, (2) dilatation of the CHD above the level of the stone, (3) narrowing of the CHD at the level of impaction and (4) normal caliber of the CBD below the impaction. Though ultrasound is easy to perform and noninvasive, it does not provide confirmatory evidence as it is performer-dependent. [10]

ERCP is the investigation of choice for this condition. This will reveal (1) the impacted stone, (2) dilatation of the bile duct above the level of the stone, (3) normal diameter of the bile duct below the level of the stone and (4) compression of the CHD (Figure 1). [2] CT scan helps in ruling out a malignant etiology for jaundice. [11] Typical CT scan findings are (1) dilatation of the biliary system including the CHD, proximal to the level of the gall bladder neck, (2) an impacted calculus in the neck of the gall bladder, (3) a contracted gall bladder and (4) a normal diameter of the CBD below the level of the stone [10]. MRCP reveals the same findings as an ERCP [2]

Figure 1

Figure 1



ERCP showing a cholecystocholedochal fistula with the stones in the distal part of the CBD and dilatation of the CHD

TREATMENT

The treatment of choice for Mirizzi's Syndrome is surgical. Complete removal of the gall bladder in patients suffering from Mirizzi Syndrome may be difficult. Partial cholecystectomy is advisable in very adherent gall bladders. This leaves the neck of the gall bladder in place which due to long standing inflammation and fibrosis always leads to occlusion of the cystic duct [12]. The stenosis of the CBD generally resolves as the inflammation subsides [2].

For Mirizzi Syndrome types II-IV (Fistula present) more complex surgical interventions are indicated. Type II defects are treated with either cholecystectomy and closure around a T-tube or partial cholecystectomy with in situ T-tube placement [2].

Other therapeutic options for type III & IV are incision of the CBD directly over the gallstone, followed by cholecystectomy and subsequent suturing of the remaining gall bladder flaps around a T tube [4, 11].

If the fistula is quite large then a choledochoplasty with a well vascularized and bile tolerant gall bladder flap is done [5, 16].

Roux-en-Y choledochojejunostomy or choledochoduodenostomy can also be performed as it reduces the risks of biliary fistulae [2, 12].

Laparascopic stapled cholecystofistulectomy which avoids contamination of the peritoneal cavity may be performed in well equipped centers if a cholecystoenteric fistula is present [13].

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