CASE REPORT

AN UNUSUAL CAUSE OF OBSTRUCTIVE JAUNDICE

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ABSTRACT: **Compression of the common bile duct by a stone impacted in the cystic duct is an uncommon cause of obstructive jaundice. We present a case study and review of the literature pertaining to the presentation, diagnosis, and surgical treatment of Mirizzi syndrome.** ***(JUMMEC 1996 1(1): 41-43)***

Introduction

External compression of the common hepatic duct by a stone impacted in the neck of the gallbladder or in the cystic duct is a rare cause of obstructive jaundice.

Mirizzi first reported this phenomenon in 1948 (1) when he described the syndrome that now bears his name. The Mirizzi syndrome includes 1) anatomic arrangement of the hepatic duct such that it is parallel to the common hepatic duct, 2) impaction of a stone in the cystic duct or neck of the gallbladder, 3) mechanical obstruction of the common hepatic duct by the stone itself or by secondary inflammation, and 4) recurrent cholangitis (2,3,4.)

Case Study

A 55-year-old female presented to the University Hospital Kuala Lumpur with a 5-day history of right upper quadrant pain, nausea, vomiting and diarrhoea. The patient was noted to be jaundiced but afebrile. Initial laboratory data included a normal white blood cell count, total bilirubin of 123, alkaline phosphatase of 507, alanine transaminase of 124 and an lactate dehydrogenese of 137.

The patient was admitted and underwent an urgent ultrasound of his right upper quadrant. The ultrasound revealed a probable stone in the neck of the gallbladder and dilated intrahepatic biliary radicals, suggesting obstruction. The gallbladder was distended, and there was no definite shadowing seen in the region of the distal common bile duct.

ERCP was performed *(Figures 1 and 2)* two days later. The common bile duct appeared to have a large obstructing stone within its lumen; however, repeated attempts at retrieving the stone with a dormia basket failed. At this time Mirizzi syndrome was suspected, and the patient was scheduled for laparotomy.

At laparotomy, a large stone was found to be impacted in the cystic duct at its junction with the common bile duct. There was significant inflammation and oedema of the porta hepatic tissues and evidence of severe cholecystitis. The operative procedure involved first mobilisation of the fundus of the gallbladder down towards the porta hepatis. The gallbladder was then transected at approximately 5 to 6 mm from the insertion of the cystic duct to the common bile duct. A large stone was present in the cystic duct at its insertion into the common bile duct, and was found to be eroding into the common duct. The common duct was opened over the stone, and the stone was removed, at which time free bile flow was established. Choledochoscopy showed a normal appearing proximal duct. A T-tube was placed, and an intra-operative cholangiogram was performed that showed excellent filling of the distal bile duct with evidence of severe chronic inflammation. The T-tube was left in place to be removed on the 10th post-operative day.

The patient's post-operative course was complicated by fevers, and blood cultures obtained were positive for *Enterococcus* spp. The patient was placed on intravenous antibiotics with resolution of her symptoms, and she was discharged on the 12th post-operative day. The T-tube was removed 2 days prior to discharge (after a normal T-tube cholangiogram).

Diagnosis

Pre-operative diagnosis of the Mirizzi syndrome is important in order to avoid complications of unrecognised cholecystobiliary or cholecystoenteric fistulas. The symptoms of Mirizzi syndrome are nonspecific and include those of obstructive jaundice. For this reason, the preoperative diagnosis often requires the use of ultrasonography or CT scan, and ERCP.

The significant features of the Mirizzi syndrome on ul-

trasonography are 1) dilatation of the biliary system above the level of the gallbladder neck, 2) the presence of a stone impacted in the gallbladder neck and 3) an abrupt change in the normal width of the common bile duct below the level of the stone (5).

ERCP demonstrates actual obstruction of the common hepatic duct due to either compression by the stone, or the actual presence of the eroded stone in the common hepatic duct. Pre-operative fistula demonstration is only possible by ERCP. However, it does not reveal the fistula 100 percent of the time. The role of CT scan in the diagnosis of Mirizzi syndrome is primarily to exclude the presence of malignancy demonstrated by a mass in the porta hepatis, or by the presence of liver metastases (5).

Fig. 1 ERCP showing complete obstruction of the common bile duct by a stone impacted at the junction of the cystic duct.

While CT scan can be used to delineate dilated ductal systems and stones, there is no evidence that this adds significantly to ultrasonographic findings (5,6)

Classification

McSherry (7) proposed a classification of Mirizzi syndrome in 1982. In his classification, *type I* involves external compression of the common hepatic duct by a stone in the gallbladder neck or cystic duct. *Type II* involves erosion of the stone into the common hepatic duct with fistula formation. Csendes *et al* (8) in 1989 proposed a more elaborate classification based on the degree to which the stone has eroded into the common hepatic duct. In this scheme, *type I* involves external compression alone. *Type II* involves erosion of the stone to only one-third of the circumference of the common bile duct. *Type III* involved erosion of the stone up to two-thirds of the circumference of the duct, and *type IV* involves complete destruction of the bile duct wall. In Csendes' study of 219 patients with Mirizzi syndrome, he reported that 10.5 percent presented as type I; 41.1 percent as type II; 44.3 percent as type III; and 4.1 percent as type IV.

Surgical Management

Successful treatment of Mirizzi syndrome requires a high index of suspicion at the time of laparotomy. Dense adhesions and oedematous inflammatory tissue, causing distortion of the normal anatomy, lead to an increased risk of bile duct injury.

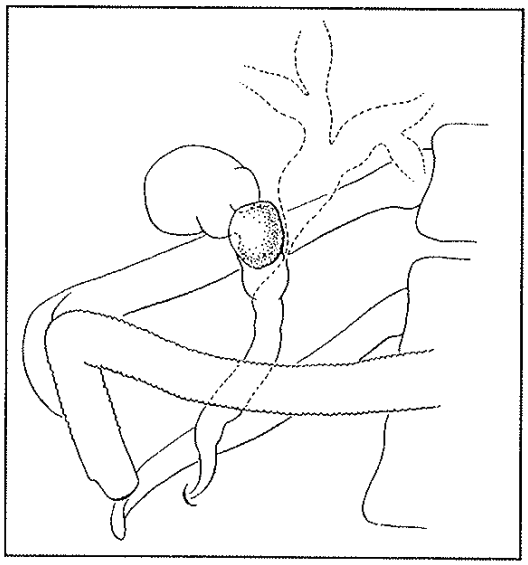

Fig. 2 Artist's rendition of ECRP

Surgical management centres around the often difficult task of protecting the common bile duct. Baer (9) and McSherry (7) both propose that the initial step in the operative procedure should involve opening the gallbladder at its fundus and removing the stone. If a gush of bile into the gallbladder is noted as the stone is removed, then a fistula should be suspected (9). At this point, a Foley catheter inserted into the gallbladder neck may be used to confirm the presence of a fistula. In the case of Mirizzi syndrome *type I* (compression without fistula formation), Baer advocates only partial cholecystectomy leaving the gallbladder neck in place, as the cystic duct is always occluded secondary to long standing inflammation and fibrosis. The risk to the common bile duct is thereby avoided. For Mirizzi *type II* as defined by McSherry, or in the presence of fistula, as was present in the case we present here, it becomes necessary to open the common bile duct via a longitudinal choledochostomy performed directly over the stone.

The cholecystectomy is then performed with the gallbladder being divided one to two centimetres from the common duct, and the divided cystic duct is then sutured closed over a T-tube (9,10).

Simple closure of the fistula defect can sometimes be technically difficult and necessitate a flap from the gallbladder wall. For this reason, it is important to identify the "type" of Mirizzi syndrome before cholecystectomy, as a portion of the gallbladder wall may need to be used for the purpose of closing the defect. An alternative approach advocated by Baer is the use of a choledochoduodenotomy or Roux-en-Y choledochojejunostomy. The basis of this recommendation lies in the high morbidity and mortality associated with common bile duct stricture (6).

Conclusion

Mirizzi syndrome is a rare but well described cause of obstructive jaundice. Surgical exercise to correct this pathophysiology can be quite challenging,and the precise anatomic definition is very helpful pre-operatively. Chronic inflammation and scarring, compounded by acute inflammation at the time of surgical presentation, combine to challenge the surgeon intra-operatively. Mirizzi syndrome occurs in less than 1 percent (8)of patients presenting for cholecystectomy; however, the anatomic anomaly that predisposes to the syndrome is more common. Deitrich (11) reported that as many as 18 percent of his patients undergoing operative cholangiograms had a low insertion of the cystic duct (2). The low insertion of the cystic duct into the common bile duct appears to be a critical predisposing factor. The continuing pathophysiology involves compression of the common hepatic duct externally by a gallbladder neck or cystic duct stone, causing inflammation and external penetration into the common bile duct. The result is cholobiliary fistula. Indeed, the Mirizzi syndrome has been described as simply one stage in the disease process of cholobiliary fistula (8)

Knowledge of the anatomy and pathophysiology of this syndrome should facilitate more accurate and successful management of the problem.

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