Singapore Med J 1999; Vol 40(3):171-173

Mirizzi Syndrome – A Report of 3 Cases with a Review of the Present Classifications

T F Toufeeq Khan, S Muniandy, F Z Hayat, Z A Sherazi, M H Nawaz

ABSTRACT

We report three cases of Mirizzi syndrome, two with external compression of the common hepatic duct and another with cystobiliary fistula. All patients presented with jaundice. The diagnosis was suggested by ultrasonography and confirmed by endoscopic retrograde cholangiography (ERC). All three had the stones removed surgically, one through a choledochotomy, another through an opening in the gall bladder and the third at the time of subtotal cholecystectomy.

We would like to propose a simple classification of Mirizzi syndrome, based on surgical procedures necessary for the correction of the pathological anatomy. If it involves the removal of calculi with some form of cholecystectomy, we consider it as Type I, whereas Type II involves the construction of a hepaticojejunostomy apart from the removal of calculi.

Keywords: cholelithiasis, chronic inflammation, cystobiliary fistula, cystolithotomy, hepaticojejunostomy

INTRODUCTION

Mirizzi syndrome is an uncommon cause of obstructive jaundice which follows repeated attacks of cholecystitis and fibrosis. This results in external compression of the common hepatic duct (CHD), or in extreme cases, cholecystocholedochal or cystobiliary fistula[1]. Direct cholangiography is essential for confirmation and helps to prevent common duct injury during surgery[2]. The present classsifications[3,4] are complicated and difficult to understand because similar stages of the disease have been separated into different types, but are however, managed by the same type of procedure and also have a similar prognosis. We are proposing a simple classification based on the need for a hepaticojejunostomy. Type I includes cases of Mirizzi syndrome, with simple external compression or with a cystobiliary fistula, which can be managed by the removal of calculi alone or with some form of cholecystectomy. Type II includes all cases in whom an additional hepaticojejunostomy becomes necessary because of complete destruction of the bile duct.

Department of Surgery
School of Medical Sciences
University Sains Malaysia
16150 Kubang Kerian
Kelantan, Malaysia

T F Toufeeq Khan, MD, FRCS
Associate Professor

S Muniandy, MD

Department of Radiology
University Sains Malaysia

F Z Hayat, MD

Z A Sherazi, DMRD

Department of Medicine
Allama Iqbal Medical College
Lahore, Pakistan

M H Nawaz, FCPS, FRCP

Correspondence to:
Dr T F Toufeeq Khan

Department of Surgery
Division of Liver/GI
Transplantation
P O Box 015809
Miami, Florida 33101

METHODS

Over a two-year period (June 1992 – June 1994), three patients diagnosed as Type I Mirizzi syndrome were managed at the Hospital University Sains Malaysia. Two patients were female and one was male with a mean age of 55 (range 49 – 59). Jaundice was obvious in two patients, while the third was mildly icteric, but all three had a long history suggestive of gall bladder disease. Ultrasonography was the initial imaging modality followed by endoscopic retrograde cholangiography (ERC). All patients were followed up with liver function tests every month for the first three months and tri-monthly thereafter. The classification proposed by us is used in this report unless mentioned otherwise.

Case 1

A 49-year-old female was admitted with a 5-day history of upper abdominal pain and jaundice. She had been having similar attacks for the last ten years. Ultrasonography revealed a 2 cm calculus in the gall bladder with dilated intra- and extrahepatic ducts proximal to the stone. The common bile duct (CBD) measured 1.4 cm and the duct distal to the stone was normal. ERC demonstrated a cystic duct filling through the fistula, running parallel to the CBD and inserting into the hepatic duct confluence (Fig 1). At the time of surgery, the gall bladder (GB) was firmly

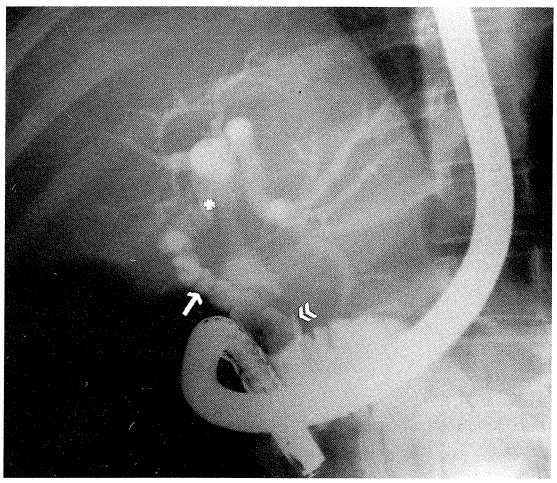

Fig 1 – Endoscopic retrograde cholangiogram (ERC). Early film revealing antegrade filling of the tortuous cystic duct (arrow) through the cholecystocholedochal fistula (arrowheads). The cystic duct is inserted into the confluence of the hepatic ducts (asterix) with partial filling of the CBD from above.

attached to the CBD and the cystic duct could not be identified. A large stone was palpable in the CBD adjacent to the GB. This was removed through a choledochotomy, which was then closed over a T-tube. She remains well at 33 months, with normal liver function.

Case 2

A 58-year-old male presented with painless jaundice of two weeks' duration. There was a 6-year history of recurrent right upper abdominal pain but noted jaundice only during the present admission. A plain abdominal film showed radio-opaque calculi in the region of the GB and ultrasound revealed dilated intrahepatic ducts. ERC demonstrated compression of the common hepatic duct by a large filling defect, suggestive of Type I Mirizzi syndrome (Figs 2a & 2b). At the time of surgery, the small gall bladder containing the stones was adherent to the common hepatic duct. The GB was opened by cutting down on the stones, and the absence of bile after stone removal confirmed the absence of a fistula. The opening in the GB was closed and the jaundice resolved over the following three weeks. He remains well at 26 months after the surgery with normal liver function.

Case 3

A 59-year-old, obese female presented with a two-year history of recurrent right upper quadrant pain and mild jaundice. Ultrasound revealed a few large calculi filling the gall bladder with very little fluid. ERC showed a minimally dilated intrahepatic biliary tree and multiple large stones in the GB causing external compression of the common hepatic duct.

At the time of surgery, the GB was found to be packed with large calculi which compressed the common hepatic duct. A subtotal cholecystectomy was carried out with removal of all calculi. She remains well 17 months post-operatively with normal liver function.

Fig 2a – Endoscopic cholangiography. Early film clearly showing external compression of the common hepatic duct (arrows) caused by radio-opaque gall bladder stones (arrowheads).

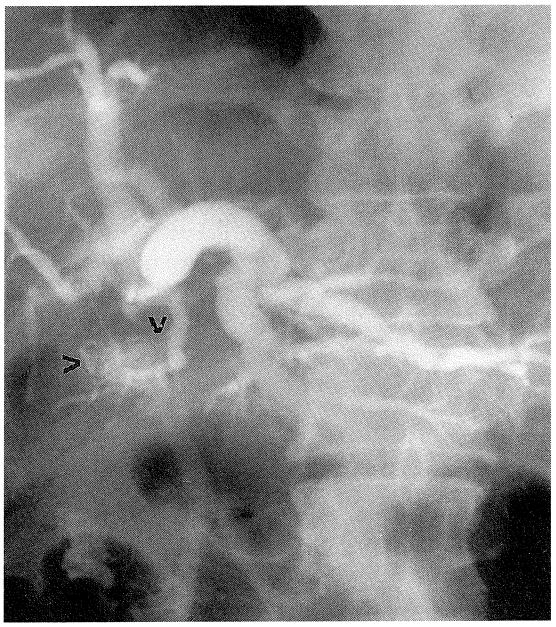

Fig 2b – Endoscopic cholangiography. Late film showing dilated intrahepatic ducts with some radio-opaque shadows in the region of the gall bladder (arrowheads).

DISCUSSION

Several classifications of Mirizzi syndrome[3-6] have been described, based on the pathological anatomy and some overlap is unavoidable which may result in some confusion. This is shown by the fact that in one classification, Types II and III are based on the amount of ductal circumference involved, but are both treated by subtotal cholecystectomy and choledochoplasty using the GB cuff. If both types are managed by similar procedures, we wonder what benefit can be derived by classifying them differently. Similarly in another report[4], both Types I and II were also managed by the same procedure and are perhaps only of some radiological interest, because its surgical significance appears questionable.

We have attempted to simplify the classification by categorising this condition by the type of surgical procedure neccessary for the relief of the symptoms. Since over 95% of cases in the larger series were external compression and cystobilary fistula (Types I, II, and III)[3], these should all be grouped together because they can all be managed by stone removal only, without the need for a bilioenteric procedure. The more complex and rare cases with complete destruction of the bile duct, however, need a hepaticojejunostomy. Pre-operative diagnosis of Mirizzi syndrome is difficult. We used ultrasonography as the primary imaging modality and it confirmed the presence of calculi and proximal intrahepatic ductal dilatation. Since the obstruction usually affects the common hepatic duct region, it has ominous implications because this benign condition may be confused with two malignancies – GB carcinoma and hilar cholangiocarcinoma. This is an important reason

for pre-operative diagnosis because in all these conditions, the area of Calots triangle is obliterated and an inexperienced surgeon may not attempt anything further and thus fail to treat a benign condition. The offending stone can easily be mistaken for a CBD calculus and cholangiography becomes essential to define its position and confirm the presence of a fistular as in one of our patients.

The objective of surgery in Type I is to remove the calculi causing the external compression. If a subtotal cholecystectomy is not possible because of a small adherent gall bladder, the opening in the gall bladder is simply closed. The presence of a fistular is included in our classification as Type I also, because the basic surgical procedure is not altered, only if a subtotal cholecystectomy is performed enough gall bladder is left in situ to affect a sound closure without narrowing the bile duct (choledochoplasty). If a subtotal cholecystectomy is not carried out, the opening in the gall bladder is closed without attempting to close the fistulous opening for fear of narrowing the bile duct. At times, the stone may have to be removed through a choledochotomy, which is then closed over a T-tube.

During surgery, we first attempt to identify the anatomy, and palpation of the calculi can help in proper orientation. In our first case, the stone was palpable in the CBD, adjacent to the adherent gall bladder and since the fistula was at such a low level, the appearance in certain ERC films was similar to that of a CBD stone. This stone was removed by a choledochotomy and no attempt was made to identify the fistula. Cholecystectomy and the bile duct was simply closed over a T-tube with good results. In the second patient, the gall bladder was identified by palpating the calculi, which were then removed by opening the gall bladder. The interior of the gall bladder was then inspected for the presence of bile and since no bile flow was observed, it confirmed the absence of a fistula. All three patients were followed up regularly in the clinic and were all asymptomatic with normal liver function.

Although we have reported only three cases of Mirizzi syndrome, we are proposing a simple classification based on surgical procedures, in an attempt to take away the complexity of the present classifications. Physician awareness, confirmation by cholangiography and removal of all calculi, remain the mainstay of the management of Mirizzi syndrome.

REFERENCES

1. Yip AWC, Chow WC, Chan J, Lam K. Mirizzi syndrome with cholecystocholedochal fistula: Preoperative diagnosis and management. Surgery 1992; 111:335-8.
2. Yip AWC, Ng WS, Suen WS, Cheng CS. Type II Mirizzi syndrome: Diagnosis by endoscopic cholangiography. J R Coll Edinb 1992; 37:49-50.
3. Csendes A, Diaz JC, Burdiles P et al. Mirizi syndrome and cholecystobiliary fistula: A unifying classification. Br J Surg 1989; 76:1139-43.
4. Mentes A, Cavusoglu H. The Mirizi syndrome: A clinical controversy. Dig Surg 1992; 9:61-5.
5. Bower TC, Nagorney DM. Mirizi syndrome. HPB Surgery 1988; :67-76.
6. McSherry CK, Festenberg H, Virshup M. The Mirizzi syndrome: Suggested classification and therapy. Surg Gastroenterol 1982; 1:219-25.