Management of adult choledochal cyst

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ABSTRACT

Introduction: Choledochal cyst is a rare benign biliary disease mostly presenting during childhood. Adult presentation is rare and associated diseases and complications are common. This paper aims to review the management of adult patients who presented to our institution with choledochal cyst, focusing on their presentation, pre-operative investigations, surgical treatment given and postoperative course.

Methods: A retrospective review of all our choledochal cyst patients from January 2000 to August 2004 was performed. Data collected included demographics and clinical information.

Results: There were ten patients, eight female (80 percent) and two male (20 percent). The average age at presentation was 38.6 (range 16–81) years. The commonest presenting complaints were obstructive biliary disease (nine out of ten, 90 percent). There were seven Type I (70 percent), one type IVA (10 percent), one type IVB (10 percent) and one (10 percent) with Caroli's disease. Two patients had concomitant cholangiocarcinoma (20 percent). Three patients had associated cystolithiasis and one patient had pancreatitis. One patient had early cirrhosis due to her disease. Six patients underwent total cyst excision with a Roux-en-Y hepaticojejunostomy. One patient who previously had a biliary bypass underwent further resection of her cyst and Whipple's operation because of development of cholangiocarcinoma in the distal remnant cyst. They are currently well with no surgical complications. The average length of follow-up was 16 months (range six months to three years).

Conclusion: Adult patients with choledochal cyst have associated biliary problems such as the presence of cholangiocarcinoma, cystolithiasis, cholecystitis and liver cirrhosis with portal hypertension. They tend to present similar to obstructive biliary disease. The best surgical option for these patients is a total cyst excision together with a Roux-en-Y hepaticojejunostomy.

Keywords: adult choledochal cyst, biliary disease, biliary dilatation, obstructive biliary disease

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INTRODUCTION

Choledochal cyst is a rare benign cyst of the biliary tree, with estimates of actual clinical incidence ranging from one in 13,000 to one in two million patients.[1] More than 60% of them present during the first year of life.[2] A proportion of them present in adulthood and their presentations differ compared to childhood cases. Many theories have been proposed to explain the origin of the choledochal cyst. The most widely-accepted hypothesis is that cystic dilatation of the bile ducts is related to an anomalous arrangement of the pancreaticobiliary ductal junction. The pancreatiocobiliary junction is proximal to the sphincter of Oddi. The long common channel allows reflux of pancreatic juice into the biliary tree, which can cause inflammation, ectasia and finally, dilatation.[3,4] We reviewed our own series of adult patients with choledochal cysts seen in the past four years.

METHODS

The clinical and laboratory findings of all patients who were treated for choledochal cysts from January 2000 to August 2004 were analysed for demographics and clinical information. The preoperative data included the patient's age at diagnosis, presenting complaints, the results of the diagnostic tests, and their management. Preoperative radiological imaging findings were reviewed to verify the morphological condition of the biliary abnormalities.

RESULTS

There were ten patients diagnosed with choledochal cysts in the study period. Both endoscopic retrograde cholangiopancreaticogram (ERCP) and magnetic resonance cholangiopancreaticogram (MRCP) were done. There were eight female patients and two male patients. The average age at presentation was 38.6 (range 16–81) years. Nine out of ten patients presented with epigastric or right hypochondrium pain, with our oldest patient being discovered incidentally with absolutely no symptoms.

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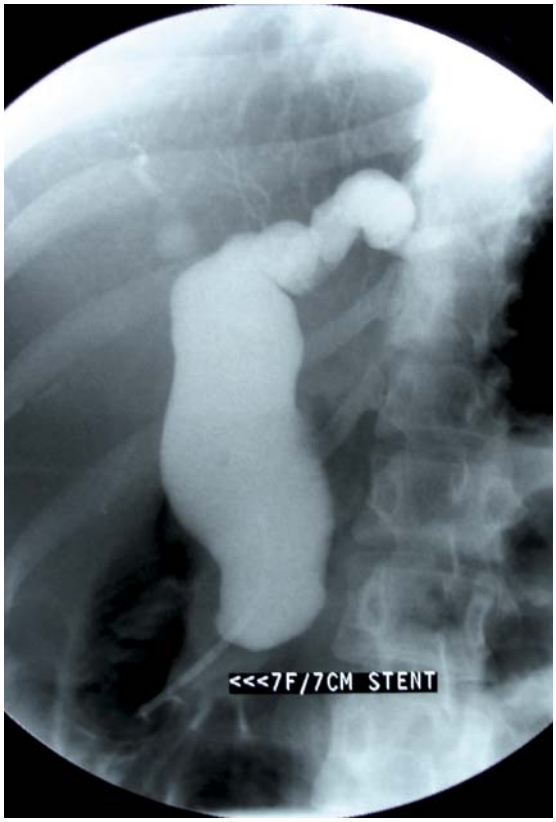

Fig. 1 ERCP image shows a type I choledochal cyst.

Eight patients had hyperbilirubinaemia and increased alkaline phosphatase at presentation. One patient also had raised amylase levels.

Seven patients had type I choledochal cysts (Fig. 1), one had type IVa, one had type IVb and one had Caroli's (type V) disease. Six patients had resection of their cyst with a hepaticojejunostomy while the patient with Caroli's disease was on the transplant waiting list. Five of them had type I cysts, while one had type IVb cyst with cholangiocarcinoma. The type I cysts were resected down to the distal limit of the cyst (intra-pancreatic portion) to ensure that no cyst wall was left behind. The distal common bile duct (CBD) stump was then closed with interrupted sutures, and a Roux-en-Y, end-to-side hepaticojejunostomy was fashioned with single layer of interrupted 4/0 PDS sutures, with extra-mucosal sutures on the jejunal side. Cholecystectomy was also performed concomitantly. A suction drain was routinely placed in the subhepatic space, and remained in place for three to five days whereupon it would be removed before the patients were discharged.

Patients who underwent surgery suffered no major complications such as bile leaks or intra-abdominal abscesses. However, one patient was re-admitted to hospital on the tenth postoperative day with superficial wound infection which resolved with wound dressings and oral antibiotics. One patient who had a biliary bypass in the early 1980s for his disease refused further surgery and is still on follow-up with our unit. Another also refused surgery as she was asymptomatic and elderly. One patient who had a biliary bypass in the early 1980s presented with a cholangiocarcinoma in the distal cyst remnant. A Whipple's operation including resection of her remnant cyst was done. The average follow-up period was one year and three months (range six months to three years). Our patients' findings are summarised in Table I.

DISCUSSION

Choledochal cyst is a rare congenital benign disease of the biliary tree. They account for approximately 1% of all benign biliary diseases.[5] It is reported to be more common in Asian populations and in females.[6,7] Most series show that the most common cyst is the Todani type I,[3,4,10] which is also found in our series where we have seven patients with this type of cyst. Similarly, we have a preponderance of female patients in our series (80% patients). Neonates or children with choledochal cysts usually present with an abdominal mass or abdominal pain.[8] Initial presentation in adults is rare.[2] They usually present with non-specific upper abdominal pain associated with obstructive jaundice, cholangitis or pancreatitis.[2] Spontaneous perforation of the cysts is seen in childhood choledochal cysts. But this is not so for their adult counterparts.[9]

In our series, most of our patients presented with non-specific upper abdominal pain together with obstructive jaundice, with one patient presenting as pancreatitis and biliary obstruction, and another being totally asymptomatic. This reflects a wide variety of presentations but the commonest symptoms were similar to that of a biliary obstructive disease. In adult patients with choledochal cyst, there are associated biliary problems such as the presence of cholangiocarcinoma, cystolithiasis, cholecystitis and liver cirrhosis with portal hypertension. Indeed, studies in adults have shown that nearly 80% of them present with one of these conditions.[2,10] In this series, two patients had cholangiocarcinoma, one patient has early cirrhosis, three patients had associated cystolithiasis, and one had associated pancreatitis. Interestingly, our oldest patient, an 81-year-old woman, was totally asymptomatic. Her cyst was discovered during a routine screening for her breast cancer. A MRCP showed that she had a type I cyst. As she is asymptomatic, she has refused surgery and is well to date. The wide variety of presentation among adults is probably secondary to the presence of complications of the cyst, such as stones, cholangiocarcinoma, pancreatitis and cirrhosis.[10,11] This was seen in 60% of our patients.

Preoperative imaging often involved both ERCP and MRCP in our patients. MRCP has the advantage of being non-invasive and is able to visualise the biliary cyst as well as ERCP (but without the complications of

Table I. Summary of patient findings.

Patient	Age	Gender	Symptoms	Type	Size (mm)	Management	Follow-up
1	46	F	Epigastric discomfort with raised bilirubin and alkaline phosphatase (ALP).	IVB	120 × 60	Resection with hepaticojejunostomy.	Well. Histology revealed a small cholangiocarcinoma: T1 NO MO. Followed-up for three years.
2	16	M	RHC discomfort with raised bilirubin and ALP.	I	105 × 56	Resection with hepaticojejunostomy.	Well. Followed-up for six months.
3	27	F	Epigastric pain with raised bilirubin and ALP. Found to have cystolithiasis.	I	85 × 40	Resection with hepaticojejunostomy.	Well. Followed-up for three years.
4	53	F	Epigastric pain with raised bilirubin and ALP.	I	75 × 45	Resection with hepaticojejunostomy.	Well. Followed-up for one year.
5	37	F	RHC pain with raised bilirubin and ALP. Early cirrhosis.	V	NA	Supportive management. Placed on transplant waiting list.	Well. Followed-up for one year.
6	31	F	Epigastric pain with raised bilirubin and ALP.	I	68 × 35	Resection with hepaticojejunostomy.	Well. Followed-up for four months.
7	81	F	Asymptomatic. Discovered during screening.	IVA	NA	Refuses surgery.	Well. Followed-up for one year.
8	31	M	Epigastric pain with raised bilirubin and ALP. Found to have cystolithiasis.	I	NA	Had biliary bypass in early 80's. Refuses further surgery.	Repeated admissions for cholangitis. Followed-up for two years.
9	18	F	Pancreatitis with raised bilirubin and ALP.	I	55 × 35	Resection with hepaticojejunostomy.	Well. Followed-up for one year
10	46	F	Epigastric pain with raised bilirubin and ALP. Found to have cystolithiasis.	I	NA	Had biliary bypass in early 1980s. Distal remnant cyst had cholangiocarcinoma. Had resection of cysts and Whipple's.	Well. Histology:T1 NO MO Followed-up for six months.

ERCP). It is hence widely held as the imaging modality of choice for choledochal cyst. However, ERCP was done for most of our patients for its therapeutic role. Our patients commonly present with right hypochondrial pain associated with an obstructive picture on the liver function test, raising the suspicion of choledocholithiasis which may be dealt with by ERCP.

It was once believed that the internal drainage of choledochal cyst alone without any resection was a sufficient and appropriate treatment. However, results subsequently showed that 30%–50% of patients with biliary bypass had late complications such as cholangitis, secondary biliary cirrhosis and development of cholangiocarcinoma.[12-14] This was seen in two of our patients who had their surgery in the early 1980s. One had a cholangiocarcinoma in the distal remnant cyst and had to undergo a Whipple's operation and resection of remnant cyst. She is doing well. Another patient has had repeated admission for cholangitis but unfortunately refused surgery.

Six of our patients have had resection of their cysts with a Roux-en-Y hepaticojejunostomy. A wide-calibre hepaticojejunostomy must be done to the normal bile duct using a meticulous surgical technique to avoid leaks. All this will ensure a patent biliary bypass. All these patients are currently doing well. Hepaticoduodenostomy after excision of the cyst is practised in some centres because of its simplicity with one less anastomosis (the jejuno-jejunostomy of the Roux-en-Y hepaticojejunostomy). It also offers the possibility of continued postoperative

surveillance of the biliary tree with ERCP. However, incidence of reflux cholangitis is higher and it is therefore not routinely done. From our experience, Roux-en-Y hepaticojejunostomy is the reconstruction method of choice.

Biliary tract malignancy has been reported to occur in 2.5%–28% of patients with choledochal cyst, representing a risk at least 20 times greater than that of the normal population.[15,16] The risk is age-related and has been reported to be 14.5% in patients older than 20 years.[17] Possible aetiologies included chronic inflammation, bile stagnation with possible developments of carcinogens, and a spare distribution of the protective mucin-secreting glands of the bile duct.[2] Two of our patients had associated cholangiocarcinoma. One was discovered histologically after resection of the cysts. One had cholagiocarcinoma in the distal remnant cysts.

In conclusion, though initial presentation of choledochal cyst in adults is rare, it does occur. The presentations are variable and are due to presence of associated diseases. It can be totally asymptomatic. The correct surgical option would be total excision of the cyst and reconstruction with a Roux-en-Y hepaticojejunostomy.

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