Original Article

Adult Choledochal Cyst

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OBJECTIVE: Choledochal cyst is a rare disease in adults. Excellent results have been achieved with proper management. However, malignancy complicating choledochal cyst still carries a dismal prognosis despite radical surgery. The aim of this study was to review the clinical course and operative results of the disease in adulthood, with emphasis on the occurrence of malignancy.

METHODS: A retrospective review of adult patients who underwent surgery for choledochal cysts in a 12-year period was performed.

RESULTS: Of the 25 adults, 80% were female and the median age was 30 years. A total of 32% of patients had previous drainage procedures for their choledochal cyst disease. Malignancy was noted in five patients (20%). There were significantly more males among the patients with malignancy than among those with benign disease (60% vs 10%), a greater number of older patients (median age, 39 vs 27 years) and more impaired liver function tests, but there was no difference in terms of incidence of previous drainage procedures between the two groups. There was no operative mortality; operative morbidity was 36%. No significant long-term complications were noted in the benign group but three of the five patients with malignancy died or developed recurrence within a few months after surgery.

CONCLUSION: The prognosis for patients with malignancy complicating choledochal cyst remains poor despite aggressive radical surgery such as Whipple's operation or additional hepatectomy. Malignancy should be suspected in older and male patients who present with jaundice or impaired liver function tests. Total cyst excision and hepaticojejunostomy is an effective and safe treatment for patients with the common type I choledochal cyst in order to reduce the chance of subsequent development of malignancy. [*Asian J Surg* 2005;28(1):29–33]

Key Words: choledochal cyst, adult, excision, malignancy

Introduction

Choledochal cyst refers to a group of diseases manifested by cystic dilatation of the biliary ductal system, which can be extrahepatic, intrahepatic or both. The disease usually manifests in the paediatric age group, but a significant portion of patients present in adulthood. The purpose of the present study was to determine the clinical course of adult choledochal cyst, with emphasis on the occurrence of malignancy and treatment options.

Patients and methods

This was a retrospective review of adult patients who underwent surgery for choledochal cyst in a 12-year period from 1991 to 2002 at the Department of Surgery, Prince of Wales Hospital, Hong Kong. Clinical presentations, investigation results, operations and follow-up outcomes were analysed. Different parameters between patients with or without malignancy complicating choledochal cyst were compared. Fisher's exact test and the unpaired *t* test were used for statistical

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analysis. All p values less than 0.05 were considered to be statistically significant.

Results

Twenty-five patients were diagnosed with choledochal cyst and underwent surgery in the study period. There were 20 females and five males. The age of patients ranged from 15 to 81 years (median, 30 years).

About half of the patients (13) presented with epigastric pain as the only symptom. Eight patients presented with acute cholangitis, three with acute pancreatitis and one with obstructive jaundice. Physical examinations were unremarkable except for four patients who had clinical jaundice. Most patients had mildly deranged liver function tests. The mean serum bilirubin was 29 μmol/L, alkaline phosphatase (ALP) was 177 IU/L, alanine transaminase (ALT) was 115 IU/L, and amylase was 153 U/L. Diagnosis of choledochal cyst was usually made by ultrasound and endoscopic retrograde cholangiopancreatography. Some patients also underwent investigations including computed tomography (CT), magnetic resonance cholangiopancreatography or endoscopic ultrasound.

According to Todani's classification, most patients had a type I choledochal cyst (Figure). No patient in the present series had a type III, IVb or V choledochal cyst.

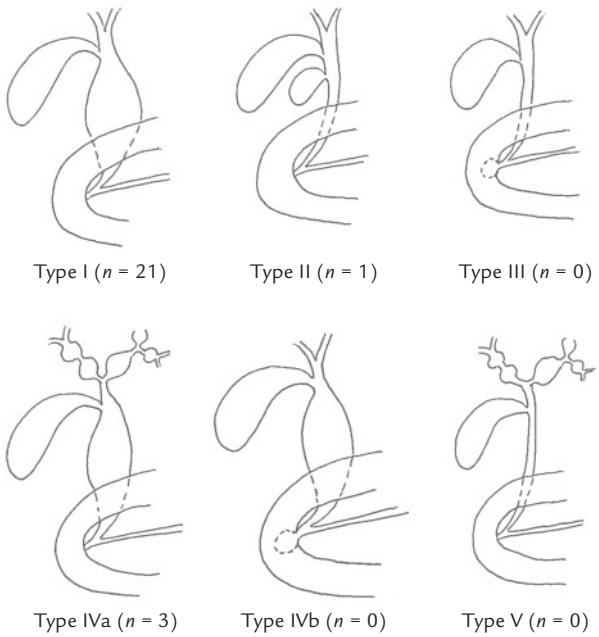

Figure. Types of choledochal cysts in the 25 patients according to Todani's classification.

Eight of the 25 patients (32%) had undergone previous surgery for choledochal cysts (Table 1). It should be noted that in a significant number of patients, the exact nature of previous surgery was only known during subsequent operations.

The types of operations performed for the 25 patients are shown in Table 2. One patient underwent two operations as the final pathology of the specimen revealed malignancy and Whipple's operation was subsequently performed.

Malignancy of the biliary tract was revealed in five patients (20%). Three patients had carcinoma of the cyst wall, one had carcinoma of the gallbladder and the other had carcinoma of the intrahepatic duct. Frozen sections were used in three of these patients and confirmed malignancy in two patients; the third patient had a false-negative frozen-section result. In comparison to their benign counterparts, patients with malignancy were older, more likely to be male, more likely to present with jaundice, and have higher mean serum bilirubin and ALP levels. However, there was no significant difference between

Table 1. Types of previous surgery for choledochal cysts

Type of operation	*n*
Cystoduodenostomy	3
Roux-en-Y cystojejunostomy	2
Cholecystectomy + T-tube drainage	1
Cholecystectomy + T-tube drainage + pancreatic necrosectomy	1
Excision of choledochal cyst + Roux-en-Y hepaticojejunostomy	1
Total	8

Table 2. Types of operations performed for choledochal cysts

Type of operation	*n*
Excision of choledochal cyst + Roux-en-Y hepaticojejunostomy	20
Pylorus-preserving pancreatoduodenectomy*	2
Whipple's operation*	1
Bisegmentectomy 4 & 5 + excision of choledochal cyst + hilar dissection*	1
Left hepatectomy + excision of choledochal cyst + removal of right hepatic duct stones	1
Left hepatectomy + pylorus-preserving pancreatoduodenectomy*	1
Total	26

*Operation for choledochal cyst with malignancy.

the two groups in terms of Todani's type of choledochal cyst, presence of stone disease, serum amylase, serum ALT and incidence of previous surgery or drainage procedures for choledochal cyst (Table 3).

Seven patients harboured stones in their choledochal cysts: three had gallstones and four had intrahepatic ductal stones.

There was no operative mortality but complications occurred in nine patients (36%) (Table 4). No postoperative complication was noted in the five patients with malignancy in the choledochal cyst.

The duration of follow-up ranged from 6 months to 11 years (median, 3 years). Of the five patients with malignancy, one patient died 2 months after surgery due to recurrent disease and two patients developed metastatic disease 7 months after surgery. Among patients with benign pathology, two patients developed complications related to the biliary tract. One patient had an attack of acute pancreatitis for which no underlying cause was found despite thorough investigation. Another patient developed liver abscess 8 months after surgery, for which no biliary tract stricture or stone was found.

Discussion

Choledochal cysts are congenital dilatations of the biliary tree. They represent a rare group of diseases with an estimated incidence of 1 in 13,000 to 1 in 2 million live births.[1,2] Choledochal cysts were first described by Vater and Ezler in 1723.[3] However, it was not until 1959 that Alonzo-Lej et al gave a thorough description of the disease and classified choledochal cysts into three types.[4] This classification was later modified by Todani et al into five types.[2] Currently, Todani's classification is most commonly used.

Table 3. Comparison of patients with or without malignancy complicating choledochal cysts

	Malignancy present (*n* = 5)	Malignancy absent (*n* = 20)	*p*
Male gender, *n*	3	2	0.04
Age*	39 (25–81)	27 (15–57)	0.02
Cyst type, *n*			
I	4	17	NS
II	0	1	NS
IVa	1	2	NS
Previous surgery, *n*	1	7	NS
Previous drainage procedure, *n*	1	4	NS
Clinical jaundice, *n*	3	1	0.02
Bilirubin (μmol/L)*	74 (10–85)	13 (3–71)	< 0.001
ALP (IU/L)*	46 (37–455)	120 (39–837)	0.02
ALT (IU/L)*	109 (15–662)	38 (9–669)	NS
Amylase (U/L)*	87 (48–105)	225 (40–1,657)	NS
Stone in choledochal cyst, *n*	1	6	NS
Gallstone, *n*	0	3	NS
Intrahepatic stone, *n*	0	4	NS

*Median (range). ALP = alkaline phosphatase; ALT = alanine transaminase.

Table 4. Operative complications

Complication	*n*	Treatment
Bile leak	3	2 resolved with percutaneous drainage, 1 underwent revision hepaticojejunostomy
Postoperative bleeding	2	1 needed re-exploration, the other received transfusion
Intra-abdominal collection	1	Percutaneous drainage
Wound dehiscence	1	Resuture of wound
Wound infection	1	
Minor wound problem	1	

Radiologically, a type I choledochal cyst may resemble a grossly dilated common bile duct secondary to obstruction. However, histologically, the wall of the choledochal cyst is a thick structure of very dense connective tissue interlaced with strands of smooth muscle. In most instances, some degree of inflammatory reaction is noted. Typical biliary mucosal lining is not seen, although in some instances, sparse islands of columnar epithelium and microscopic bile ducts within the wall of the cyst are noted.[5]

As it is a congenital condition, choledochal cyst commonly presents in infancy and childhood: 25% of cases are diagnosed within the first year of life and 60% before the age of 10 years.[4,6] Adult presentation is less common. Some of these adult patients have known choledochal cysts and underwent surgery in childhood. In our series, 32% of the patients had undergone previous operations for choledochal cysts. Together with the increased incidence of associated biliary tract stone disease, stricture formation and superimposed malignancy, previous operations may lead to greater challenges in the management of adult choledochal cysts.

The incidence of malignancy in choledochal cyst in the present series was 20%. The reported incidence of malignancy in the literature ranges from 2.5% to 30%.[7–11] The risk is age-related: 0.7% in children below the age of 10, 6.8% between the ages of 11 and 20, and 14.3% above the age of 20.[12] Furthermore, the rate increases to 50% in patients with a prior internal drainage procedure and occurs at a mean of 10 years postoperatively.[13] A similar phenomenon was observed by Liu et al.[10] However, we did not find an increased incidence of malignancy in patients with a history of internal drainage procedures for choledochal cyst, but older age, male gender, presence of impaired liver function and jaundice were associated with superimposed malignancy. Thus, a higher degree of suspicion should be entertained in the older male patient and in the presence of impaired liver function.

Total cyst excision remains the gold standard treatment for choledochal cysts.[5,10,14] Cholecystectomy should be done at the same time, as the gallbladder usually arises from or adheres to the cyst wall and occasionally contains stones or even tumour, to facilitate bilioenteric reconstruction. Superimposed malignancy necessitates more extensive surgery such as Whipple's operation for lower common bile duct cancer or hepatectomy for proximal bile duct cancer or gallbladder cancer. Restoration of biliary drainage is usually achieved by Roux-en-Y hepaticojejunostomy or, less commonly, by hepaticoduodenostomy.

Simple cystenterostomy, common in the past, should no longer be used. The remaining cyst can lead to biliary stasis, stone formation, cholangitis, pancreatitis and, worst of all, malignant transformation.[14] Such a simple drainage procedure accounted for 20% of patients in this series, reflecting the inadequacy of this procedure alone. Furthermore, subsequent surgery is more difficult due to adhesion and sometimes due to lack of information about the nature of the previous surgery.

Sometimes, type I choledochal cyst extends almost to the common bile duct and pancreatic duct junction. Incomplete excision of the cyst results if one stops prematurely when dissecting distally due to the worry of damaging the pancreatic duct. The residual cyst can give rise to stone formation, sepsis and malignancy.[9,14,15] Thus, the strategy of complete excision of a choledochal cyst should be strictly adhered to.[9,10,15] We believe that this can be done by careful dissection around the cyst towards the lowermost non-dilated portion with the help of traction, while at the same time safeguarding the pancreatic duct. Diagnosis and excision of a residual cyst can be very difficult, as seen in one patient in the present series. The diagnosis was only made after CT scan in the patient who presented with stenosis of hepaticojejunostomy after previous excision of a choledochal cyst. Excision of the residual cyst inside the pancreas proved to be extremely difficult. Similarly to the lower end, we advocate resection of the ductal confluence if this is involved, and reconstruction by separate left and right hepaticojejunostomies. For a type IVa cyst, the extrahepatic component should be completely resected as for a type I cyst. The management of the intrahepatic component depends on the severity of disease and extent of involvement. Treatments range from percutaneous drainage through stricture dilatation and stone removal to partial hepatectomy or liver transplantation.[9]

The use of frozen sections during surgery is recommended to exclude malignancy. In suspicious cases, malignancy should be confirmed by frozen section so that major radical resection can be done in a single session. Of course, the probable extent of surgery should be discussed with patients before surgery. The limitation of frozen section should also be mentioned as false-negative or, much more rarely, false-positive results may occur. One patient in the present series who had a false-negative frozen-section result needed to undergo a second operation as malignancy was confirmed by paraffin section.

Though there was no operative mortality, complications occurred in 32% of patients undergoing surgery for choledochal cysts. Reported complication rates vary from as low as 14%[10] to as high as 64%.[15] Operations for adult choledochal

cyst are expected to be more difficult than their paediatric counterparts due to the additional problem of inflammatory adhesions, infection, stone disease, malignancy and previous surgery.

A limitation of the present study was the relatively short follow-up period. However, of the five patients with malignancy complicating choledochal cysts, one patient died from recurrent disease and two others developed metastases within a few months after surgery. Among patients with benign disease, no significant long-term complications were observed. This study showed that patients who have malignancy complicating choledochal cysts have a dismal prognosis despite aggressive surgery, while total cyst excision with drainage is safe and effective for those with benign disease. In view of the risk of developing malignancy, it is advocated that patients who have undergone internal drainage for choledochal cysts in the past should undergo resection of the cyst.[16]

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