Diagnosis and treatment of congenital choledochal cyst: 20 years’ experience in China

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Abstract

AIM  To summarize the experience of diagnosis and treatment of congenital choledochal cyst in the past 20 years (1980–2000).

METHODS The clinical data of 108 patients admitted from 1980 to 2000 were analyzed retrospectively.

RESULTS Abdominal pain, jaundice and abdominal mass were presented in most child cases. Clinical symptoms in adult cases were non-specific, resulting in delayed diagnosis frequently. Fifty-seven patients (52.7%) had coexistent pancreaticobiliary disease. Carcinoma of the biliary duct occurred in 18 patients (16.6%). Ultrasonic examination was undertaken in 94 cases, ERCP performed in 46 cases and CT in 71 cases. All of the cases were correctly diagnosed before operation. Abnormal pancreaticobiliary duct junction was found in 39 patients. Before 1985 the diagnosis and classification of congenital choledochal cyst were established by ultrasonography preoperatively and confirmed during operation, the main procedures were internal drainage by cyst enterostomy. After 1985, the diagnosis was established by ERCP and CT, and cystectomy with Roux-en-Y hepaticojejunostomy was the conventional procedures. In 1994, we reported a new and simplified operative procedure in order to reduce the risk of choledochal cyst malignancy. Postoperative complication was mainly retrograde infection of biliary tract, which could be controlled by the administration of antibiotics, there was no perioperative mortality.

CONCLUSION The concept in diagnosis and treatment of congenital choledochal cyst has obviously been changed greatly. CT and ERCP were of great help in the classification of the disease. Currently, cystectomy with Roux-en-Y hepaticojejunostomy is strongly recommended as the choice for patients with type I and type IV cysts. Piggyback orthotopic liver transplantation is indicated in type V cysts (Caroli’s disease) with frequently recurrent cholangitis.

Subject headings  choledochal cyst/surgery; choledochal cyst/radiography; choledochal cyst/diagnosis; biliary tract/abnormalities; choledochal cyst/therapy; Caroli’s disease/diagnosis; Caroli’s/surgery

INTRODUCTION

Choledochal cyst, or congenital cystic dilatation of the common bile duct, is a rare entity in western countries. Most of reported cases in the world come from Asia, about two-thirds of cases reported from Japan[1]. In recent years, cases of choledochal cyst are reported increasingly in China[2-5]. We analyzed retrospectively the clinical data of 108 patients admitted from 1980 to 2000 in our two hospitals in order to summarize the Chinese experience of diagnosis and treatment of this congenital choledochal cyst.

MATERIAL AND METHODS

Patients

From October 1980 to February 2001, a total of 108 patients with choledochal cyst were treated in the Department of Surgery, the Second Affiliated Hospital of Zhejiang University School of Medicine and Zhongda Hospital of Southeast University. There were a total of 85 females and 23 males. The mean age was 27.8 years, with a range from 3 to 68 years. Among them, 91 cases were adults. According to Todani and colleagues’ classification of congenital choledochal cyst[6]. Seventy-five patients belonged to type I (solitary extrahepatic cyst), 19 type IV A (extrahepatic and intrahepatic cysts), 5 type IVa (multiple extrahepatic cysts), 1 type III (choledochocele) and 6 type V (only intrahepatic segmental ductal dilatation, also known as Caroli’s disease); two cases were unclassifiable. In 23 patients their condition was associated with biliary tract stones. Abdominal pain, jaundice and abdominal mass were presented in most child cases, and clinical symptoms in adult cases were non-specific, resulting in delayed diagnosis frequently. Fifty-seven patients (52.7%) had coexistent pancreaticobiliary disease, 7 patients had synchronous and 11 had metachronous carcinoma lesions arising from the biliary duct cyst. Three patients appeared in pregnant period. Among complications at the time of admission for care there were jaundice in 77 patients, cholangitis in 61 patients. Ultrasonic examination was undertaken in 94 cases, ERCP performed in 46 cases and CT in 71 cases.

Surgical procedures

From 1980 to 1985, Roux-en-Y cystojejunostomy was performed in 19 patients, side to side cystoduodenostomy in 15 patients and exploratory laparotomy in 4 patients. From 1986 to 1994, 24 cases underwent cystectomy plus cholecystectomy with Roux-en-Y hepatointerostomy, 6 underwent partial cystectomy plus cholecystectomy with Roux-en-Y hepatointerostomy, 4 underwent cystectomy or cyst transection plus duodenocholedochotomy by jejunal transection plus duodenocholedochotomy by jejunal
interposition, and 3 underwent left hepatectomy. After 1995, we also performed cyst excision plus cholecystectomy with Roux-en-Y hepatoplasty in 19 cases. Meanwhile, we reported a new and simplified operative procedure (cyst wall resection plus Roux-en-Y hepatoduodenostomy and doudenoejunostomy) in order to reduce the risk of choledochal cyst malignancy. This procedure was performed in 9 patients. Two cases underwent piggyback orthotopic liver transplantation.

RESULTS

There was no operative death in this series. Twenty-eight patients developed early postoperative complications including cholangitis, bile leakage and wound infection. All of them recovered with conservative therapy. Late complications occurred in twenty-three patients, who suffered from ascending cholangitis, hepatolithiasis, pancreatitis and adhesive intestinal obstruction six months to five years after operation. Twenty-two cases underwent reoperation. Among them, 11 cases owing to bile duct malignant change resulting from previous internal drainage procedures (including cystoduodenostomy and cystojejunoanastomosis); 6 cases owing to supplicative cholangitis or pancreatitis; 3 cases owing to postoperative biliary stricture accompanied stones; and 2 cases owing to calculus of bile duct. The reoperative procedures included resection of tumor or plastic repair of stricture. Sixty-two patients were under follow-up for an average period of ten years (6 months to 18 years).

DISCUSSION

Diagnosis

Choledochal cysts are recognized as a disease of childhood but in our study, 91 (84.2%) of 108 patients were younger than 14 years at the time of operation. Thereafter, the surgeon should be aware of choledochal cysts even in the adult patients. Abdominal pain, jaundice and abdominal mass were present in most child cases. In this series, 12 patients (70.5%) had these symptoms. Clinical symptoms in adult cases were non-specific. In addition, the increased likelihood of associated hepatobiliary disease, as well as previous surgery makes management in adults more complex. Although the classical clinical triad of abdominal pain, jaundice and right hypochondriac mass has been reported in children, most patients (71 cases) described had symptoms that were chronic and intermittent, often resulting in delayed diagnosis. Furthermore, secondary hepatobiliary disease in adults may obscure the primary problem and compound the complexities of subsequent surgery. Twenty-three patients with intra- or extra hepatic stones were found in this series, it deserved attention. In 1959, Alonso-Lej and in 1977, Todani classified choledochal cysts based on the location of the cyst. From that time, they were diagnosed more frequently with the aid of improved diagnostic techniques, for example, Ultrasound, CT and direct cholangiography such as PTC and ERCP. US is the first imaging modality of choice in establishing the diagnosis of choledochal cyst, and to recognize the presence of an anomalous pancreaticobiliary duct junction (APBDJ) recommended by us. In our series, of 46 patients examined with ERCP, 39 patients (93.4%) had APBDJ. Komi also reported a 92.2% association between choledochal cysts and APBDJ in 645 cases, which is similar to our results. There were many theories about the etiology of the choledochal cysts. Babbitt suggested that in these patients there is an abnormal pancreaticobiliary duct junction that allows reflux of pancreatic secretions into the biliary system during a critical stage of its development. Consequent chemical and enzymatic destruction of the duct wall leads to cystic dilation. In 1978, Motoc and other authors found that the choledochal dilation has a close association with abnormal choledochopancreatic duct junction, whether direct or indirect. CT can clearly visualize cyst location, number, scope, stone and relationships with surrounding structures. After 1994, CT was routinely performed with CT examination, all of them were diagnose definitely. In recent 5 years, CT cholangiography and MR cholangiography have been used in most advanced countries. Our center also want to practice this new METHODS in the future.

Operative procedure

Choledochal cyst is a congenital abnormality that requires surgical intervention to prevent hepatobiliary and pancreatic complications. A lot of reports have demonstrated that cystenterostomy, an internal drainage procedure without resection, carries a high morbidity rate and often requires subsequent reparative operation. Todani analyzed carcinoma arising from retained cysts with internal drainage procedures, and suggested that enteric drainage tend to create a cul-de-sac in the choledochal cyst and to activate pancreatic juice when intestinal juice with enteroxin influxes into the cyst through an anastomotic stoma. As a result, inflammation of the bile duct wall is accelerated, possibly resulting in carcinoma because of the long-standing irritation of the biliary epithelium. Kobayashi points out that the incidence of bile duct carcinoma is still high, even after excision of extrahepatic bile ducts in APBDJ patients with choledochal dilatation. For these patients, careful long-term follow-up is necessary, especially after operations. In the present series, 11 of 108 patients who had previous cyst enterostomy developed carcinoma. However, all cases who developed carcinoma in our study were over 30 years of age, which supports the previous report that the risk of carcinoma increases with age. After 1985, cyst internal drainage was abandoned and total excision of the cyst and Roux-en-Y hepatojjunostomy had been recommended as the first choice by us. In 1994, we reported a new and simplified operative procedure to separate the biliary and pancreatic flow so as to reduce the risk of malignancy of choledochal cyst, which was clinically applied in 9 cases, and proved effective.

Total excision of type I and type IV choledochal cyst and Roux-en-Y hepatojjunostomy has been recommended by many investigators because of the lower incidence of postoperative complications, especially in Japan, and has been
increasingly popularized worldwide[24,31,32], we agree with their opinions. Treatment for type V cysts is still controversial. At present, hepatic resection is safe and effective for some of type V cysts (Caroli’s disease)[33]. In our series, three left lobectomy and four nonanatomical hepatic resections were performed and symptomatic relief was obtained completely and permanently. About type V cysts with frequently recurrent cholangitis, resulting in biliary liver cirrhosis, liver resection is seldom feasible because of associated congenital hepatic fibrosis.

In this setting, liver transplantation may represent the only effective and durable form of treatment and offers the only hope for such patients. In this study, nearly two years ago we performed piggyback orthotopic liver transplantation for two cases of Caroli’s disease and both patients recovered smoothly with satisfactory results after operation and are healthily surviving[34]. In conclusion, the surgical strategy should be selected based on the type of cyst. For those who had internal drainage operation before, careful long-term follow-up is required. We recommended total excision of type I and type IV choledochal cyst and Roux-en-Y hepaticojejunostomy. It is an important topic to actively explore new procedure in preventing malignancy of congenital cyst of common bile duct, and it deserves paying attention to. About type V cysts with frequently recurrent cholangitis resulting biliary liver cirrhosis, liver transplantation should be considered.

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