Journal of Pediatric Surgery (2011) **46**, 2296–2300

ELSEVIER

Journal of Pediatric Surgery

www.elsevier.com/locate/jpedsurg

Clinical characteristics of liver fibrosis in patients with choledochal cysts

Jun Fujishiro*, Yasuhisa Urita, Toko Shinkai, Chikashi Gotoh, Noriko Hoshino, Kentaro Ono, Hiroaki Komuro[1]

Department of Pediatric Surgery, University of Tsukuba, Tsukuba, Ibaraki, 305-8575, Japan

Received 24 August 2011; accepted 3 September 2011

Key words:
Choledochal cyst;
Congenital biliary dilatation;
Liver fibrosis;
Jaundice;
Complications

Abstract

Purpose: The aim of the study was to identify the clinical characteristics and outcome of patients with liver fibrosis in choledochal cyst (CC).

Methods: Forty patients with CC who underwent liver biopsy were included. Liver fibrosis was classified as follows: grade 0, no fibrosis; grade 1, mild fibrosis localized in the portal area; grade 2, moderate fibrosis with occasional bridging; and grade 3, severe fibrosis with diffuse bridging.

Results: Fourteen patients (35%) had liver fibrosis. Patients in the fibrosis group were significantly younger (1.2 vs 2.7 years) and had higher total bilirubin (5.3 vs 2.6 mg/dL). Severity of liver fibrosis was inversely correlated with age ($P = .044$). Amylase and lipase in bile were significantly lower in the fibrosis group (amylase, 531 vs 15,000 U/L; lipase, 783 vs 23,100 U/L). Postoperative serum analysis demonstrated no differences between the two groups. Most patients in both groups had normal aspartate aminotransferase, alanine aminotransferase, total bilirubin, and γ-glutamyl transpeptidase regardless of severity of fibrosis. Postoperative biliary complication or cholangiocarcinoma was not found in the fibrosis group.

Conclusions: Our data suggest that liver fibrosis is mainly influenced by obstructive cholangiopathy rather than refluxed pancreatic secretion. Prognosis of patients with CC and liver fibrosis was as good as that of patients without fibrosis.

Choledochal cyst (CC), or congenital biliary dilatation, is a rare anomaly of the biliary tree characterized by dilatation of the extrahepatic and/or intrahepatic bile ducts. Refluxed pancreatic secretion caused by pancreaticobiliary malunion is thought to be involved in the etiology of this anomaly [1]. The classical triad is jaundice, abdominal pain, and palpable mass, and patients with CC often present with pancreatitis or cholangitis. Although the long-term outcome after total cyst excision with Roux-en-Y hepaticojejunostomy is generally good, late complications such as cholangitis, biliary stones and stricture, pancreatitis, and malignancy have been recorded [2]. In rare cases, liver transplantation is necessary for patients with CC [3].

Abnormal levels of liver enzymes are generally observed in patients with CC, and varying degrees of liver fibrosis is found in some patients. It is known that liver fibrosis is more common in neonates and infants [4], and post-

Presented at the Pacific Association of Pediatric Surgeons 44th Annual Meeting, Cancun, Mexico, April 10-14, 2011.

* Corresponding author. Tel.: +81 29 853 3094; fax: +81 29 853-3149.
E-mail addresses: jfujishi@md.tsukuba.ac.jp (J. Fujishiro), hiro-kom@md.tsukuba.ac.jp (H. Komuro).
[1] Sponsor information: Hiroaki Komuro, MD, PhD, Department of Pediatric Surgery, University of Tsukuba, Tennodai 1-1-1, Tsukuba, Ibaraki, 305-8575, Japan.

0022-3468/$ – see front matter © 2011 Elsevier Inc. All rights reserved.
doi:10.1016/j.jpedsurg.2011.09.017

operative reversal of cirrhosis has been reported in some cases [5,6]. In this study, we compared the characteristics at presentation, types, and postoperative complications of patients with liver fibrosis and those without liver fibrosis.

1. Methods

Patients with CC who underwent definitive operation and wedge liver biopsy at our department from 1980 to 2010 were included in the study. The patients were classified into fibrosis and nonfibrosis groups, according to the presence or absence of liver fibrosis. Demographic data, serum and bile laboratory data, as well as operative and postoperative clinical data, were obtained using retrospective chart review. The severity of liver fibrosis was graded as follows: grade 0, no fibrosis; grade 1, mild fibrosis localized in the portal area; grade 2, moderate fibrosis with occasional bridging; and grade 3, severe fibrosis with diffuse bridging [7], which was diagnosed by pathologists at our institution. The type of CC was defined using the Todani classification system [8].

Statistical analysis was performed using Fisher's exact test, the Mann-Whitney U test, and Spearman's rank correlation analysis, and a P value less than .05 was considered significant. Continuous variables are expressed as the median (range).

This study was approved by our institutional ethics committee (No. H22-642).

2. Results

Forty patients with CC underwent primary operation and liver biopsy at our department. All patients received cyst excision with Roux-en-Y hepaticojejunostomy. Of the 40 patients included in the study, 14 (35%) with liver fibrosis were defined as the fibrosis group and 26 (65%) without liver fibrosis were defined as the nonfibrosis group. Table 1 summarizes patient demographics, the severity of liver fibrosis, and symptoms for both groups. The sex distribution in both groups was similar, with a predominance of women observed. The ages at presentation and at operation were significantly younger in the fibrosis group than in the nonfibrosis group (1.2 vs 2.7 years, P = .049; and 1.45 vs 2.92 years, P = .039; respectively). Patients in the fibrosis group were significantly less likely to present with abdominal pain (31.5% vs 76.9%, P = .006) (Table 1). Jaundice was more common at the time of presentation in the fibrosis group (78.6% vs 53.8%), but the difference was not significant (P = .177). Preoperative serum analysis demonstrated that the total bilirubin (T. Bil.), direct bilirubin (D. Bil.), and γ-glutamyl transpeptidase (γ-GTP) levels were significantly higher, and the serum amylase was significantly lower, in the fibrosis group (Table 2). These trends concerning presenting symptoms and laboratory data were similar when we compared only older patients (>2 years old; data not shown). There were no differences in levels of preoperative serum aspartate aminotransferase (AST) and alanine aminotransferase (ALT) between the fibrosis and non-fibrosis groups.

Table 1 Patient demographics, grade of fibrosis, and symptoms of patients with or without liver fibrosis in CC

	Fibrosis (n = 14)		Nonfibrosis (n = 26)		P
	Median/n	Range/%	Median/n	Range/%	
Sex					1.000 [a]
Female	11	78.6%	19	73.1%	
Male	3	21.4%	7	26.9%	
Age (y)					
At presentation	1.2	0.00-7.87	2.7	0.00-10.27	.049 [b]
At operation	1.45	0.03-7.94	2.92	0.05-10.44	.039 [b]
Liver fibrosis					
0: none			26	100%	
1: mild	7	50%			
2: moderate	4	28.6%			
3: severe	3	21.4%			
Symptoms					
Abdominal pain	4	31.5%	20	76.9%	.006 [a]
Jaundice	11	78.6%	14	53.8%	.177 [a]
Palpable mass	3	21.4%	4	15.4%	.679 [a]
Acholic stool	3	21.4%	6	23.1%	1.000 [a]
Fever	3	21.4%	3	11.5%	.646 [a]
Vomiting	6	42.9%	15	57.7%	.510 [a]
Prenatal diagnosis	2	14.3%	2	7.7%	.602 [a]

[a] Fisher's exact test.
[b] Mann-Whitney U test.