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Emergent biliary drainage for choledochal cyst

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Two patients with choledochal cyst who needed an emergent biliary drainage are presented. Case 1: Spontaneous rupture of the cyst was demonstrated by hepatobiliary scintigram in a 21-month-old girl and an emergent T-tube drainage relieved her symptom. Acute obstruction by protein plugs was considered to have caused rapid expansion of the cyst, which resulted in rupture eventually. Case 2: Marked hypoproteinemia was demonstrated in a 29-month-old boy with choledochal cyst. Bile drainage through Foley catheter promptly made the serum protein level elevated above normal range. Portal hypertension due to severely dilated cyst may have increased ascites amount, which caused extracellular fluid shift and protein to be lost into ascites. In both cases the excision of the extrahepatic bile duct and hepaticojejunostomy was carried out successfully later.

Treatment of 20 pediatric cases in our institute and the literature were reviewed. Bile drainage would be safer in emergency condition even though it has been suggested that reconstructive surgery may be tolerable. Definitive surgery should be regarded as a procedure with some risk of postoperative complications because asymptomatic cases operated electively had serious ones. Treatment strategy of patients with choledochal cyst is not straightforward and should be arranged based upon their conditions.

Key words: choledochal cyst, cyst rupture, hypoproteinemia, infant, surgery

INTRODUCTION

Choledochal cyst, cystic dilatation of the common bile duct, is an uncommon congenital disease and is definitively treated by excision of the extrahepatic bile duct and anastomosis of the hepatic duct with either jejunum or duodenum [12]. Infrequently, biliary drainage is necessary due to acute symptoms before definitive surgery [10]. Herein we report two pediatric patients who needed an emergent biliary drainage. One was due to spontaneous rupture of the cyst and the other was due to profound hypoproteinemia. These rare conditions associated with choledochal cyst are discussed in terms of their pathogenesis and our experience is reviewed and treatment strategy of pediatric choledochal cases is also discussed.

CASE REPORTS

Case 1: A 21-month-old girl visited a local hospital because of several times of vomiting. Dilatation of the common bile duct was detected by the abdominal ultrasonography and serum amylase level was found elevated. For further treatment she was referred to our institute under the diagnosis of pancreatitis due to choledochal cyst.

On admission, her temperature was 36.5˚C, BP 102/68 mmHg, PR 84/min, RR 24/min. She had distended abdomen but was neither jaundiced nor had palpable tumor. Laboratory tests demonstrated unremarkable findings other than elevated serum amylase level (Table 1). After treatment with 70 mg/kg/day of flomoxef and 3 mg/day of nafamostat mesilate, amylase level decreased but she became jaundiced and her AST, ALT, γ-GTP, and total bilirubin levels became

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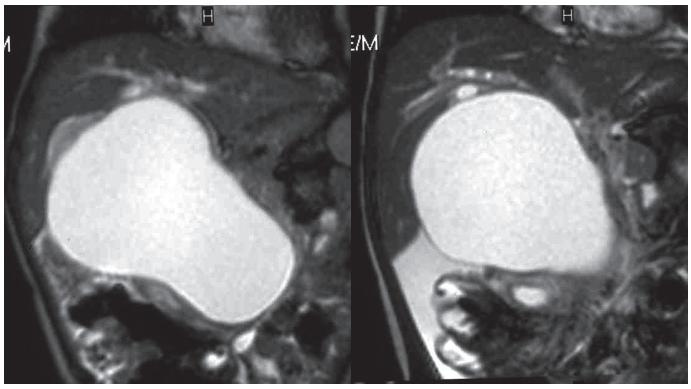

Fig. 1 MRI image one day before the rupture (Case 1). Remarkable expansion of the duct and accumulation of ascites is demonstrated.

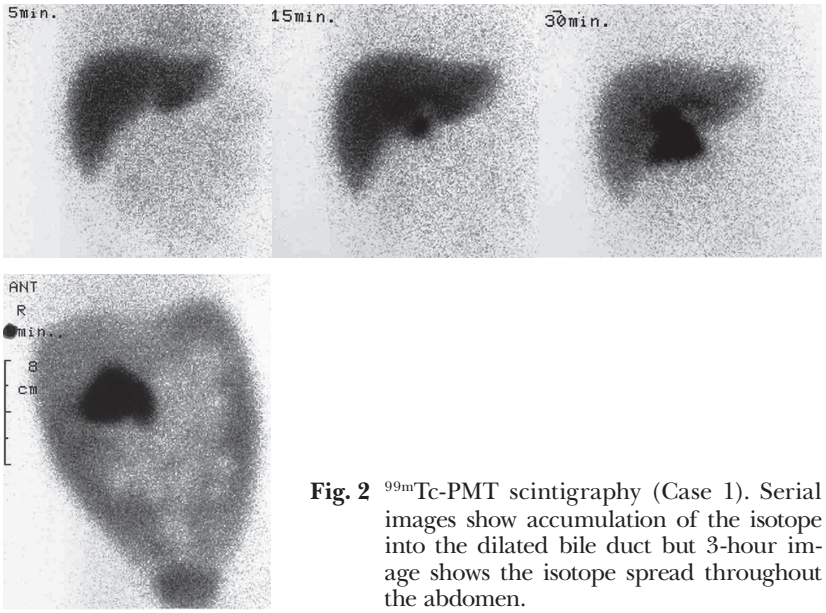

Fig. 2 ^{99m}Tc-PMT scintigraphy (Case 1). Serial images show accumulation of the isotope into the dilated bile duct but 3-hour image shows the isotope spread throughout the abdomen.

elevated. She continued to be irritable but her vital signs had been rather stable. While an elective surgery was planned serial imaging studies including CT and MRI for anatomical evaluation demonstrated the gradual increase of the cyst size and accumulation of ascites (Fig. 1). Hepatobiliary scintigram done on her seventh day after admission showed the isotope spread throughout the abdominal cavity and spontaneous rupture of the biliary tract was suspected (Fig. 2).

Emergency laparotomy revealed ruptured cyst with necrotic tissue surrounding the hole at the anterior wall of the cyst (Fig. 3) and greenish protein plugs were retrieved from the bile (Fig. 4). Peritoneal lavage and drainage through T-tube placed at the perforated site relieved her symptom. Contrast study through the T-tube confirmed the diagnosis of choledochal cyst Type Ia (Todani's classification [12]) and definitive surgery of hepatojejunostomy with Roux-Y anastomosis was carried out 7 weeks later and discharged on the 17-th postoperative day without any remarkable event.

Case 2: 29-month-old boy had been suffered from abdominal pain for two weeks before visiting a clinic. After referred to a local hospital he was demonstrated to have choledochal cyst by computed tomography and was transferred to our hospital for further treatment.

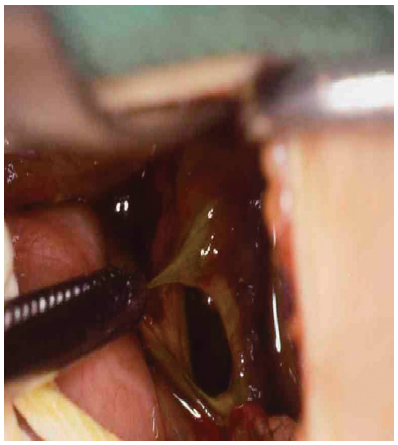

Fig. 3 Ruptured wall of the dilated bile duct had the necrotic rim (Case 1).

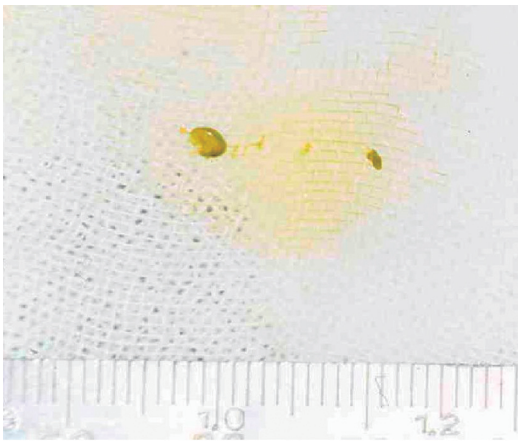

Fig. 4 Protein plug retrieved from the bile (Case 1).

Table 1 Laboratory test results on admission.

Item	(normal range)	Case 1	Case 2
WBC	4000-8000	8000	7900
RBC	4.10-5.30 x 10^4	421 x 10^4	421 x 10^4
Hb (g/dl)	13.5-17.5	11.5	8.8
Ht(%)	40.0-48.0	35.1	32.4
Plt	14.0-40.0	20.0 x 10^4	46.7 x 10^4
TP (g/dl)	6.5-8.0	6.6	3.7
Alb (g/dl)	4.1-5.0	4.4	2.2
AST (IU/dl)	<30	36	42
ALT (IU/dl)	<35	12	60
γ GTP (IU)	<80	13	13
T.Bil (mg/dl)	0.2-1.1	0.8	0.2
D.Bil (mg/dl)	0.0-0.3	0.2	
Serum-AMY (IU)	50-170	752	144
CRP (mg/dl)	<0.3	1.03	2.55

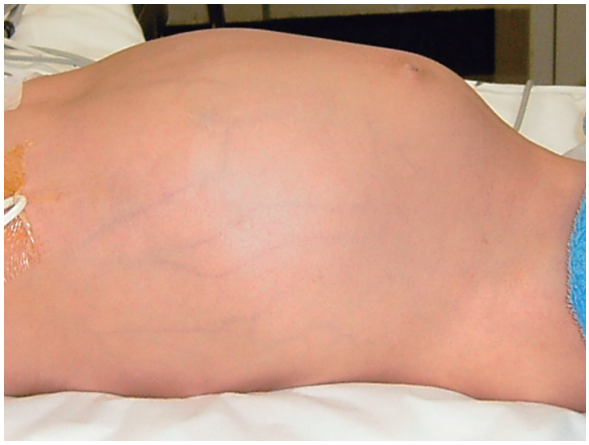

Fig. 5 Markedly distended abdomen (Case 2).

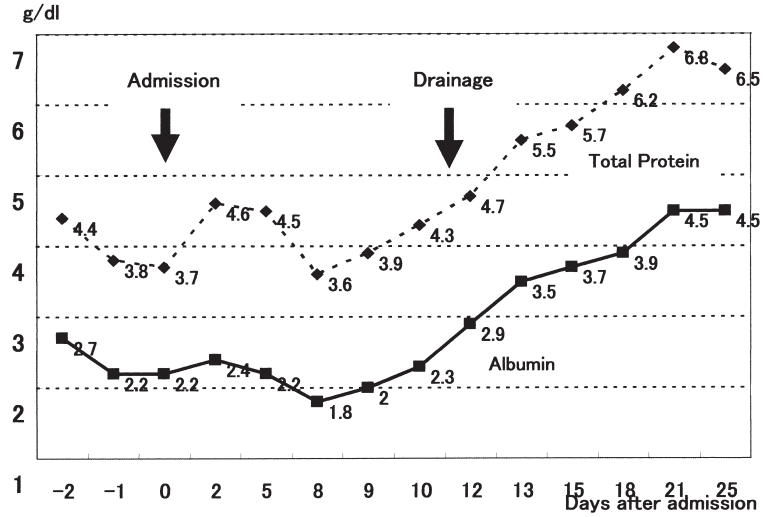

Fig. 6 Serial total protein and serum albumin levels in Case 2. Both promptly leveled up to normal ranges after drainage operation.

On admission, he was lethargic and his temperature was 37.2°C, BP 102/56 mmHg, PR 120/min, RR 24/min. He had distended abdomen but was neither jaundiced nor had palpable tumor. Laboratory tests demonstrated marked hypoproteinemia associated with mild anemia, hepatic dysfunction and hyperamylasemia (Tabel 1). Treatment with 100 mg/kg/day of cefotiam and 2.5 mg/day of nafamostat mesilate started immediately but fever and his abdominal pain with marked distension continued (Fig. 5) while associated hypoproteinemia and hypoalbuminemia became worsened (Fig. 6). Elective reconstructive surgery was abandoned and bile drainage through Foley catheter placed into the cyst was carried out on his 11-th day of admission and 700 ml of bile was aspirated at the operation. Serum protein concentration elevated above normal range promptly while his abdominal distension improved dramatically (Fig 6). Definitive surgery carried out 2 weeks later did not cause any postoperative complication and discharged on the 12-th postoperative day.

DISCUSSION

Choledochal cyst is treated by the excision of the extrahepatic bile duct and anastomosis of the hepatic duct with either jejunum or duodenum [12]. Although its treatment requires reconstructive surgery but its associated conditions sometimes necessitate emergency surgery resulting in delaying the definitive surgery [10].

Spontaneous rupture of choledochal cyst had been considered rare but recent reviews reported the rate of this condition to be 2 to 7% (Table 2) [2, 3, 5, 13]. Since it can sometimes be the initial manifestation of the disease it should be considered in the presence of bile-like fluid at the time of emergency laparotomy [4]. Bile peritonitis following spontaneous rupture may lack peritoneal signs and be difficult to be diagnosed but can be made by hepatobiliary scintigram demonstrating an extrahepatic bile leakage [1, 11]. In our case 1, not only scintigram but serial imaging studies along with blood biochemistry had happened to demonstrate the time course of the spontaneous rupture. Jaundice, rapid increase of the cyst size and ascites accumulation had developed before demonstrated bile leakage. The presence of protein plugs and the necrosis of the ruptured cyst wall would suggest that acute obstruction by protein plugs lead to develop jaundice and rapid expansion of the cyst, which resulted in cholangitis with wall necrosis and eventual rupture. This speculation is supported by Ando *et al.* who proposed the similar hypothesis [2].

Most reported cases have been managed with external drainage of the cyst followed

Table 2 Reported incidence of spontaneous rupture of the congenital dilatation of the common bile duct.

Author(s)(year reported)	Incidence of rupture	Reference No
Kim (1980)	2.1% (4/188)	5
Yamaguchi et al. (1981)	1.8% (26/1433)	13
Ando et al (1995)	4.1% (5/123)	2
Ando et al (1998)	7.0% (13/187)	3

Table 3 Choledochal cyst cases (1992-2004).

Case	Sex	Age at operation		Symptoms		Operative procedures	Postoperative complications
		Yr	Mo	At presentation	At operation		
1	M	3	10	Abdominal pain	None	Primary reconstruction	Anastomotic leakage
2	F	0	1	None	None	Primary reconstruction	None
3	F	12	4	Abdominal pain	None	Primary reconstruction	None
4	F	10	9	Abdominal pain	None	Primary reconstruction	None
5	F	6	2	Abdominal pain	None	Primary reconstruction	Intrahepatic stones
6	M	3	11	Abdominal pain	None	Primary reconstruction	Cholangitis?
7	F	3	4	Abdominal pain	None	Primary reconstruction	None
8	M	4	5	Abdominal pain	None	Primary reconstruction	None
9	F	0	6	None	None	Primary reconstruction	None
10	F	0	0	Vomiting/ Abdominal distension	Vomiting/ Abdominal distension	Primary reconstruction	None
11	F	12	2	Abdominal pain	None	Primary reconstruction	None
12	M	5	5	Abdominal pain	None	Primary reconstruction	None
13	M	2	0	Abdominal pain	None	Primary reconstruction	None
14	F	0	1	None	None	Primary reconstruction	None
15	F	1	10	Vomiting	Abdominal pain/ rupture	Drainage followed by reconstruction	None
16	F	1	11	Abdominal pain	None	Primary reconstruction	None
17	F	0	4	Acholic stool	Acholic stool	Primary reconstruction	Intracranial bleeding?
18	F	0	7	None	None	Primary reconstruction	Postoperative bleeding
19	F	2	4	Abdominal pain/Jaundice	Abdominal pain/Jaundice	Primary reconstruction	None
20	M	2	5	Abdominal pain/ hypoproteinemia	Abdominal pain/ hypoproteinemia	Drainage followed by reconstruction	None

by a second procedure to excise the cyst and reconstruct the biliary tract as in our case but some reported that ruptured cysts were treated with primary reconstructive surgery [7].

Hypoproteinemia associated with choledochal cyst was only reported in one Japanese case [8]. This condition can be extremely rare or may be missed unnoticed. In our case 2, this condition was promptly relieved after drainage, which would indicate this to be due to the disease.

Generally, hypoproteinemia with hypoalbuminemia is caused either by disturbance of synthesis, increased loss or changed distribution due to shift of the extracellular fluid. The reported case by Mokusawa *et al.* is concluded to be due to protein loosing enteropathy [8]. In case 2, there was no apparent severe liver dysfunction which can cause decrease of protein production and no sign of nephrotic syndrome or protein loosing enteropathy. Portal hypertension is a known complication of delayed presentation of choledochal cyst and to cause accumulated ascites [9]. Experimental bile duct ligation can cause increased capillary permeablity without compensatory albumin synthesis [6]. One can speculate the increased portal vein pressure due to severe bile duct dilatation

and long-term bile duct obstruction may increase ascite amount, which can lead extracellular fluid shift and protein loss into the ascites.

In our institute, 20 pediatric cases with choledocal cyst were encountered since 1992 and all were treated with definitive reconstructive surgery (Table 3). Fifteen of them had symptoms at presentation but they were initially treated conservatively. In 5 cases symptoms persisted in spite of treatment and presented two cases underwent emergent bile drainage followed by reconstruction afterwards. Reconstructive surgery may be tolerable even in emergency condition as proposed by the literature if a patient's preoperative condition is stable enough [7]. However, one of our three cases (case No. 17) who were treated by primary reconstructive surgery while their symptoms were persistent was lost by postoperative death because of possible intracranial hemorrhage, which suggests that an emergent biliary drainage would be safer for patients with acute symptoms.

Ten out of 15 cases had their symptoms improved and were operated electively after their symptoms had subsided. There were another group of patients with choledochal cyst who are asymptomatic when have it found incidentally such as routine antenatal ultrasonography and infantile health checkups. We experienced 5 such cases and were treated surgically within their infantile period (Table 3). Among these 15 cases treated electively without any symptom at operation 3 had serious postoperative complications including two (case No. 1, 6) who required a reoperation, which indicate that postoperative complications are not preventable even after symptoms subside or when patients are asymptomatic.

In conclusion, when they have acute symptom(s) an emergent biliary drainage should be considered as a safer procedure to relieve them. Reconstructive surgery may be tolerable even in emergency but it should be regarded as a procedure with a risk of postoperative complications even when they are asymptomatic. When considering a surgery for cases incidentally found without symptoms the complication risk should be informed and appropriate timing should be discussed based on treatment result of a large group of patients, which has not yet been reported. Thus, treatment strategy of patients with choledochal cyst is not straightforward and should be arranged based upon their clinical conditions.

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