Journal of Pediatric Surgery Case Reports 39 (2018) 48–52

Contents lists available at ScienceDirect

Journal of Pediatric Surgery Case Reports

journal homepage: www.elsevier.com/locate/epsc

Biliary atresia associated with pancreaticobiliary maljunction

Tatsuma Sakaguchi*, Yoshinori Hamada, Takeshi Shirai, Hiroshi Hamada, Takashi Doi

Division of Pediatric Surgery, Department of Surgery, Kansai Medical University, Hirakata, Osaka, Japan

ARTICLE INFO

Keywords:
Kasai type IIIa biliary atresia
Pancreaticobiliary maljunction
Infantile obstructive cholangiopathy

ABSTRACT

We report here two cases of Kasai type IIIa biliary atresia (BA), associated with pancreaticobiliary maljunction (PBM). Both cases were referred to our hospital because of hyperbilirubinemia and diagnosed with BA preoperatively by abdominal ultrasonography and biliary scintigraphy. They underwent Kasai portojejunostomy at 49 and 37 days old, respectively. Intraoperative cholangiographies performed from their gallbladders clarified Kasai type IIIa BA with PBM. Laboratory test of colorless fluid from the gallbladder of one patient showed high level of lipase (460 U/L). Reported cases of BA associated with PBM are rare and this is a second report showed elevated level of a pancreatic enzyme in bile. The findings in our cases indicate the pathogenesis of biliary anomaly in association with PBM.

1. Introduction

The etiology of biliary atresia (BA) remains unknown. Various evidence has shown that viruses, toxins, and gene sequence variations trigger inflammation and fibrosis in extrahepatic bile ducts [1]. However, anatomical aspects of the pathogenesis of BA have not well investigated. In 1974, Landing hypothesized the concept of “infantile obstructive cholangiopathy” as a common cause of neonatal hepatitis, choledochal cyst, and BA, which was based on the findings of pancreaticobiliary maljunction (PBM) [2]. In 1979, Miyano et al. reported the results of autopsied specimens from 28 BA cases in which 60% were associated with PBM [3]. They hypothesized that inflammation and fibrosis in extrahepatic bile ducts caused by reflux of pancreatic juice resulted in BA, although they failed to confirm pancreatic enzymes in bile. According to these authors, the comorbidity rate of PBM in BA cases did not appear to be low. However, reported cases of BA associated with PBM are rare [4–7].

In Kansai Medical University Hospital, a total of 50 patients who were diagnosed with BA have been surgically treated since 1963, including four cases of Kasai type IIIa BA (obstructed at the porta hepatis with patency of the common bile duct, Table 1) [8]. Among them, intraoperative cholangiography was successfully performed and PBM was clearly identified in two cases.

2. Case report

Case 1: A male neonate was born at 41 weeks of gestation. His body weight at birth was 3504 g. His serum total bilirubin level was 10.3 mg/dL at birth, and it gradually rose to 14.2 mg/dL at 12 days old, even after phototherapy. Abdominal ultrasonography (US) showed splenomegaly, but the gall bladder (GB) and intrahepatic bile ducts could not be well visualized. Biliary scintigraphy did not show biliary excretion. He underwent laparotomy at 49 days old with a diagnosis of BA. A hypoplastic GB was found and connective tissue was observed in the porta hepatis. Intraoperative cholangiography that was performed from the GB showed type III-a BA with PBM (5-mm length of the common channel) (Fig. 1). The GB and connective tissue on the hepatic hilum were dissected (Fig. 2), followed by Kasai portojejunostomy. His postoperative course was uneventful and he has been well after approximately 30 years of surgery.

Case 2: A male neonate was born at 40 weeks of gestation. His body weight at birth was 3280 g. An acholic stool was observed at 16 days old, and his family took him to a doctor at 29 days old. His general condition was good, except for his jaundice. Laboratory data showed an elevated serum total/direct bilirubin level (9.1/4.5 mg/dL). Abdominal US showed the GB and triangular cord sign (Fig. 3), but the intrahepatic bile ducts could not be well visualized. Biliary scintigraphy did not

* Corresponding author. 2-5-1 Shinmachi, Hirakata, Osaka, 573-1191, Japan.
E-mail address: sakaguct@hirakata.kmu.ac.jp (T. Sakaguchi).

https://doi.org/10.1016/j.epsc.2018.09.011
Received 27 September 2018; Accepted 30 September 2018
Available online 06 October 2018

Table 1
Kasai classification of biliary atresia.

1) Main Types	
Type I	Atresia of common bile duct
Type II	Atresia of hepatic duct
Type III	Atresia of bile duct at the porta hepatis
2) Subtypes	
a	patent common bile duct
b	fibrous common bile duct
c	aplasia of common bile duct
d	miscellaneous

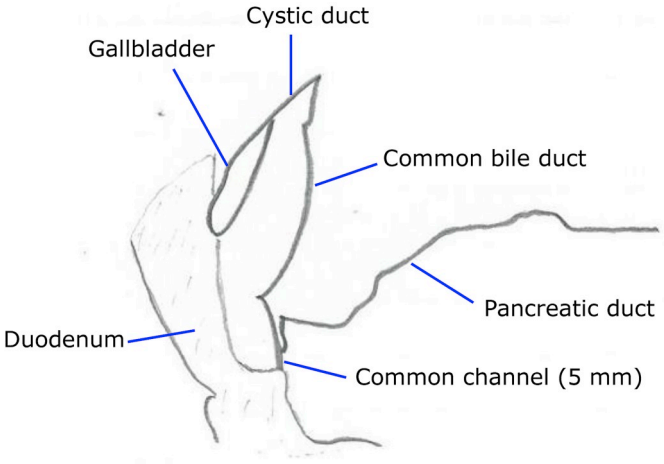

Fig. 1. Intraoperative cholangiography in case 1 (sketch).

show biliary excretion (Fig. 4). No bilirubin component was detected from duodenal fluid sampling. He underwent laparotomy at 37 days old with a diagnosis of BA. Intraoperative cholangiography that was performed from the GB showed type III-a BA with PBM (pancreatic duct type, 10-mm length of the common channel) (Fig. 5). The GB and connective tissue at the hepatic hilum were dissected (Fig. 6), followed by Kasai portojejunostomy with the anti-reflux valve. His postoperative course was uneventful and he has been well 20 months after surgery.

A laboratory test of colorless fluid from the GB showed a high lipase level (460 U/L), but a low amylase level (1 IU/L). Histopathologically, extremely small luminal structures with exfoliated epithelium were found in the connective tissue. Infiltration of lymphocytes and fibrosis was found in the distal side of the dissected connective tissue (Fig. 7).

3. Discussion

Few radiological studies have assessed the pancreaticobiliary junction in BA cases [4,5,9]. Theoretically, radiological assessment of the pancreaticobiliary junction with contrast medium is possible only in cases of subtype “a” (patency of the common bile duct) BA, which consists of only approximately 15% of all BA cases [10]. Preoperative assessment of PBM from various imaging modalities is insufficient. Saito et al. reported preoperative endoscopic retrograde cholangiopancreatography for BA that failed to visualize the common bile ducts

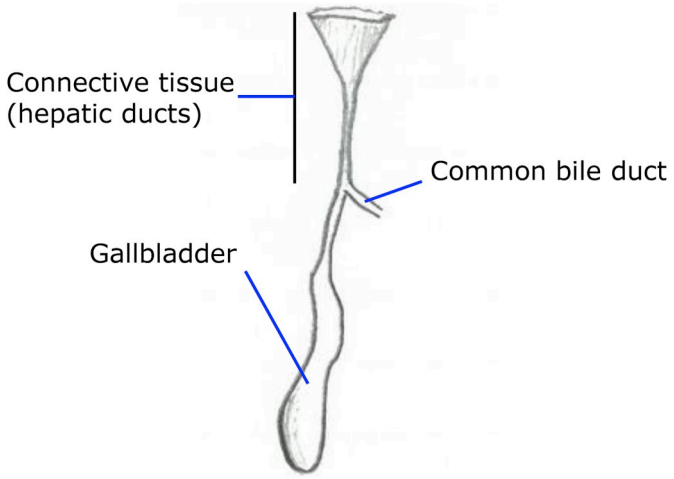

Fig. 2. Resected specimen in case 1 (sketch).

in 70% of all BA cases [9]. Chiba et al. reported the results of intraoperative cholangiography for 43 cases of subtype “a” in which the common channels of pancreaticobiliary ducts were obviously long in many cases of BA [4]. Deguchi et al. reported that 60% of five patients with BA who successfully had intraoperative cholangiography performed showed PBM [5].

Endo et al. reported the first case of BA with PBM with elevated lipase levels in bile [6]. They assumed that PBM might not be an etiological factor for atresia of the extrahepatic bile duct, but associated with patency of the common bile duct. Koshinaga et al. reported a perforated case of type III cystic biliary atresia associated with PBM [7]. They assumed that protein plugs or inspissated bile in the distal end of the bile duct were the possible cause of obstruction.

From our institutional retrospective clinical database, we found four cases of subtype “a” BA. Among them, intraoperative cholangiography was successfully performed in two cases. In case 1, the GB was hypoplastic and thus we could not obtain fluid from the GB. In case 2, the fluid that we obtained from the GB showed an elevated lipase level, which indicated reflux of pancreatic juice into the bile duct. Low amylase levels in the bile could be attributed to immaturity of the pancreas in infants [11]. Histologically, we found infiltration of lymphocytes and fibrosis in the distal side of the resected bile duct. This finding suggested that reflux of pancreatic juice through PBM was one of the factors for forming biliary inflammation and fibrosis in case 2.

We sometimes encounter cases in which BA or congenital biliary dilatation is not clearly distinguished. Congenital biliary dilatation is a congenital malformation involving both dilatation of the extrahepatic bile duct, including the common bile duct, and PBM. Various types of pathological conditions, such as flow disturbances of bile and pancreatic juice, reciprocal reflux between bile and pancreatic juice, and malignancy of biliary systems, can occur in the hepatobiliary system and pancreas secondary to bile duct dilatation and PBM [12]. In subtype “b” (obstruction of the porta hepatis and extrahepatic duct) BA, which is the most common subtype, intraoperative cholangiography is unable to be performed, and thus the presence of PBM is unknown. However, PBM may be one of the factors involved in the pathogenesis