A Rare Combination of Choledochal Cyst Type III and Iva

—A Case Report

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Abstract

Choledochal cyst is a rare disease of the biliary tree. A combination of two types of choledochal cysts is extremely rare. We report a 69-year-old female who had a combination of choledochal cysts type III and IVa, which was demonstrated by endoscopic retrograde cholangiopancreatography and percutaneous transhepatic cholangiography, and finally proved after total excision of extrahepatic choledochal cyst with Roux-en-Y hepaticojejunostomy.

Key words: choledochal cyst, choledochocele, endoscopic retrograde cholangiopancreatography (ERCP), percutaneous transhepatic cholangiography (PTC), ultrasonography

Introduction: Choledochal cyst is a rare anomaly of the biliary tree; it consists of cystic or cylindrical dilatation of intra- and/or extrahepatic bile ducts(1). The first classification of choledochal cyst was made by Alonzo-Lej et al. in 1959(2), and the most widely accepted

classification was modified by Todani et al. in 1977(3). However, there are very few reports on a combination of more than two types of choledochal cysts in a patient. Herein, we report on an adult female with a combination of choledochal cyst type III and IVa, and review the literature.

Case report: A 69-year-old female had suffered from right upper quadrant pain intermittently for 1 month and had a fever (38.7^{o} C) at admission. She was found to have had gallbladder stones and diffuse biliary tree dilatation 5 years prior to admission. She received cholecystectomy for acute cholecystitis at another hospital 4 years ago. She had been hospitalized 5 times due to cholangitis and sepsis in the past 3 years. Endoscopic retrograde cholangiopancreatography (ERCP) performed 2 years ago demonstrated choledochocele but failed to show the entire biliary tree. Percutaneous transhepatic cholangiography (PTC) was carried out, and a diagnosis of type IVa choledochal cyst was entertained (Fig. 1). No stones were found on ERCP or PTC. Surgical intervention was suggested but the patient refused at that time.

The laboratory data at this admission revealed a white blood cell count of 13,900/mm^3, and hemoglobin level of 11.9 g/dl. Serum amylase was 83 u/l, total bilirubin level 0.7 mg/dl, alkaline phosphatase 136 u/l, aspartate transaminase 36 u/l, alanine transaminase 60 u/l, blood urea nitrogen 25 mg/dl, and creatinine 0.7 mg/dl. Abdominal ultrasonography showed

diffuse biliary tree dilatation, and the common bile duct (CBD) measured up to 4.5 cm in diameter. Escherichia coli was recovered from blood cultures. ERCP showed a soft glistening nodule protruding into the duodenal lumen associated with a markedly dilatated CBD and intrahepatic ducts (Fig. 2). A combination of choledochal cyst type III and IVa was considered. Because of recurrent cholangitis and sepsis, excision of extrahepatic choledochal cyst and Roux-en-Y hepaticojejunostomy were performed and the choledochocele was left intact. Grossly the common bile duct was a saccular cystic mass measuring 9 x 4 x 4 cm. Histological findings revealed a fibrotic cystic wall lined by columnar epithelium with focal round cell infiltration, and no evidence of malignant change. Unfortunately, she expired on the 35^{th} postoperative day due to a perforated duodenal ulcer and multiple organ failure.

Discussion: Choledochal cyst is a relatively rare disease with predominance in children and females. Type I choledochal cyst is the most common and comprises of 80%-90% of reported cases (4). In adult patients, type IV comprises about 40%, and type III (choledochocele) is one of the rarest types, accounting for only 1.4%-5.0% of cases (1, 4, 5). The combination of more than two types of choledochal cyst in a patient is extremely rare. Kimura et al. found three cases of multiple extrahepatic diverticula in combination with Caroli's disease (6), while Gupta et al. reported a case of choledochocele (A3 type) associated with Caroli's disease and diverticula of the CBD(7). In Todani's original classification, he incorporated a

case of choledochocele with diffuse dilatation of the choledochus as a type IVb, but he also questioned whether it might be induced by the choledochocele. Longmire et al. called type IVb choledochal cyst an “extrahepatic multiple cystic dilatation”(8). In this case, we prefer type IVa plus choledochocele (A2 type) rather than type IVb choledochal cyst since the etiology of a choledochocele might be different from other types of choledochal cysts.

The etiology of choledochal cysts is still unknown; it is considered to be a congenital anomaly because most cases are infants and young adults. Babbit first suggested that malformation of the pancreaticobiliary system is an important cause of this disease (9), because “long common channel” with anomalous insertion was found in 10.5%-76% of cases (1, 10, 11). The reflux of pancreatic enzymes into the bile duct produces inflammation, weakening of the wall, and then cystic dilatation of the biliary tree(11). However, this common channel theory seems unlikely to explain isolated intrahepatic duct dilatation (type V) and choledochoceles (type III). Sphincter of Oddi dysfunction had been reported as a possible etiology of choledochocele (12). Some authors had questioned whether choledochoceles were actually not choledochal cysts at all (13). According to Gupta’s report and our case, a combination of two or more types of choledochal cysts usually has a choledochocele with another type of choledochal cyst. We suggest that the etiology of choledochal cysts and choledochoceles may be different, but this combination is extremely rare, and more cases must be collected.

Complete excision of the cyst and reconstruction with Roux-en-Y hepaticojejunostomy is the treatment of choice because many complications may occur such as cholangitis, liver abscess, portal hypertension, biliary cirrhosis, cholelithiasis, pancreatitis, and malignant change(1, 4, 5, 14, 15). The risk of malignant change is high and appears to be related to age, especially those who have undergone enteric drainage previously. Our patient was a 69-year-old female with repeated attacks of cholangitis and sepsis, and although she had no drainage procedure or any sign of malignant change, we suggested surgical intervention for her. Unfortunately, she expired.

Reference:

1. Crittenden SL, Mackinley MJ. Choledochal cyst. Clinical features and classification. Am J Gastroenterol 1985; 80: 643-649.
2. Alonso-Lej F, Rever WB Jr, Pessagno DJ. Congenital choledochal cysts, with a report of 2, and an analysis of 94 cases. Surg Gynecol Obstet 1959; 108: 1-30.
3. Todani T, Watanabe Y, Narusue M, et al. Congenital bile duct cysts: classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. Am J Surg 1977; 134: 263-269.
4. Yamaguchi M. Congenital choledochal cyst. Analysis of 1,433 patients in the Japanese literature. Am J Surg 1980; 140: 653-657.
5. Chang-Chien CS, Wu CS, Lin DY. Two cases of choledochal cyst: Todani type II and

type IVB. J Formosan Med Assoc 1987; 86: 560-563.

6. Kimura K, Ohto M, Ono T, et al. Congenital cystic dilatation of the common bile duct: Relationship to anomalous pancreaticobiliary ductal union. Am J Roentgenol 1977; 128: 571-577.
7. Gupta R, Pruthi HS, Baijal SS, et al. An unusual choledochal cyst. J Gastroenterol Hepatol 1997; 12: 771-774.
8. Longmire WP Jr, Mandiola SA, Gordon HE. Congenital cystic disease of the liver and biliary system. Am Surg 1971; 174: 711-725.
9. Babbit DP. Congenital choledochal cyst: new etiological concept based on anomalous relationships of common bile duct and pancreatic bulb. Ann Radiol 1969; 12: 231-240.
10. Savader SJ, Benenati JF, Venbrux AC, et al. Choledochal cysts: classification and cholangiographic appearance. Am J Radiol 1991; 156: 327-331.
11. Iwai N, Yanagihara J, Tokiwa K, et al. Congenital cohledochal dilatation with emphasis on pathophysiology of the biliary tract. Ann Surg 1992; 215: 27-30.
12. Ladas SD, Katsogridakis I, Tassios P, et al. Choledochocele, an overlooked diagnosis: Report of 15 cases and review of 56 published reports from 1984 to 1992. Endoscopy 1995; 27: 233-239.
13. Spier LN, Crystal K, Kase DJ, et al. Choledochocele: newer concepts of origin and diagnosis. Surgery 1995; 117: 476-478.

14. Stain SC, Guthrie CR, Yellin AE, et al. Choledochal cyst in the adult. Ann Surg 1995; 222: 128-133.

15. Todani T, Watanabe Y, Toki A, et al. Carcinoma related to choledochal cysts with internal drainage operations. Surg Gynecol Obstet 1987; 164: 61-64.

Legends of figures:

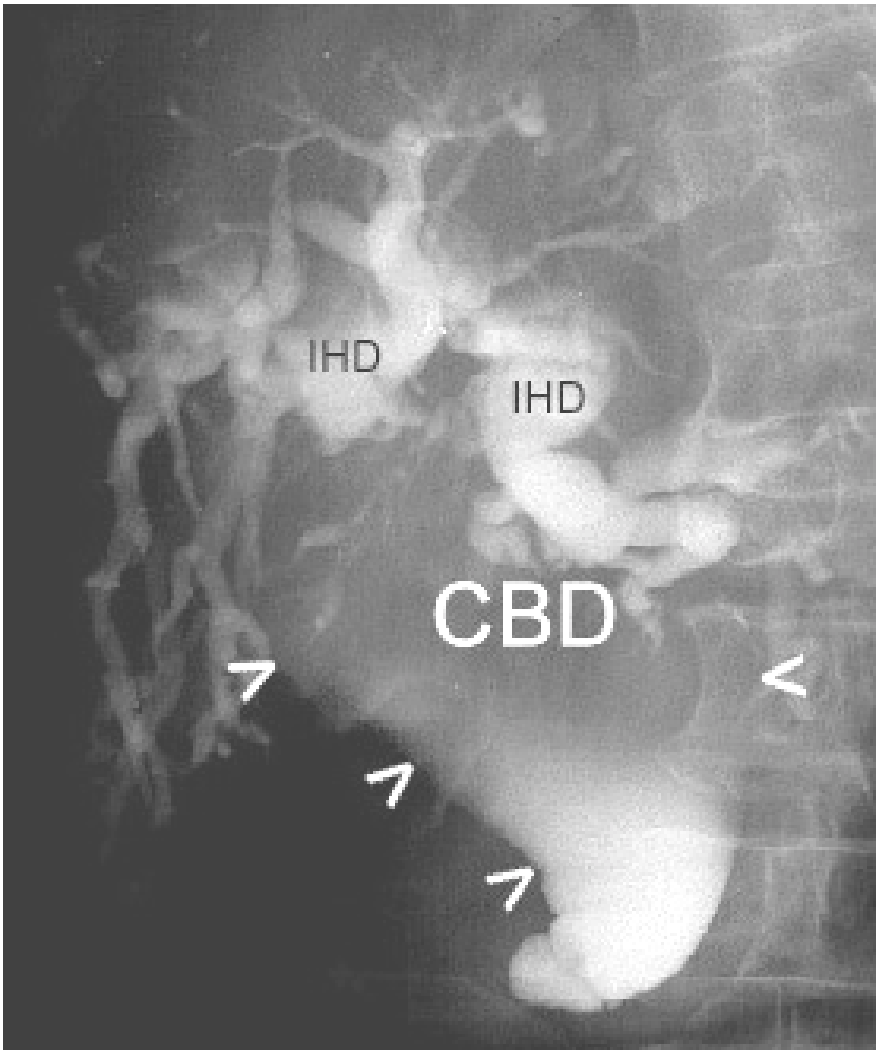

Figure 1. Percutaneous transhepatic cholangiography showing diffuse biliary tree dilatation, CBD: common bile duct; IHD: intrahepatic duct; arrowheads mark the margin of CBD.

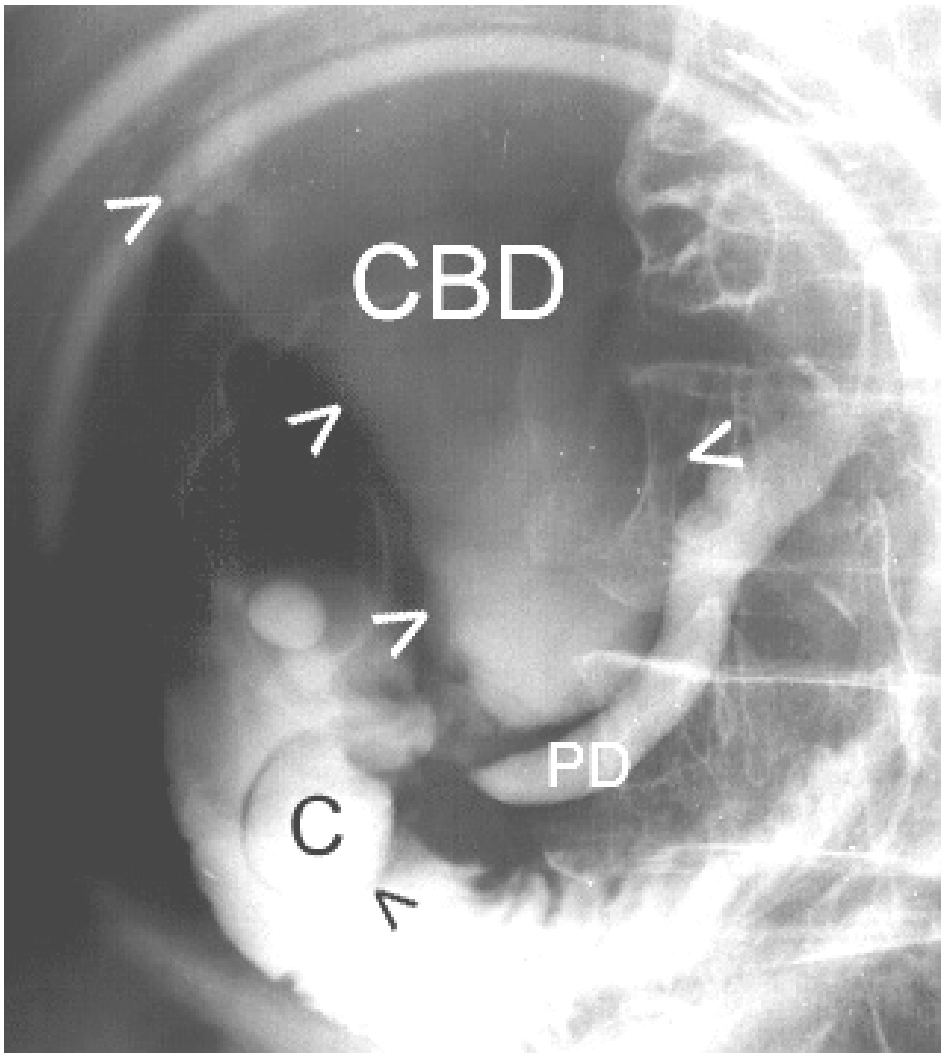

Figure 2. Endoscopic retrograde cholangiopancreatography showing choledochocele associated with a type IVa choledochal cyst.

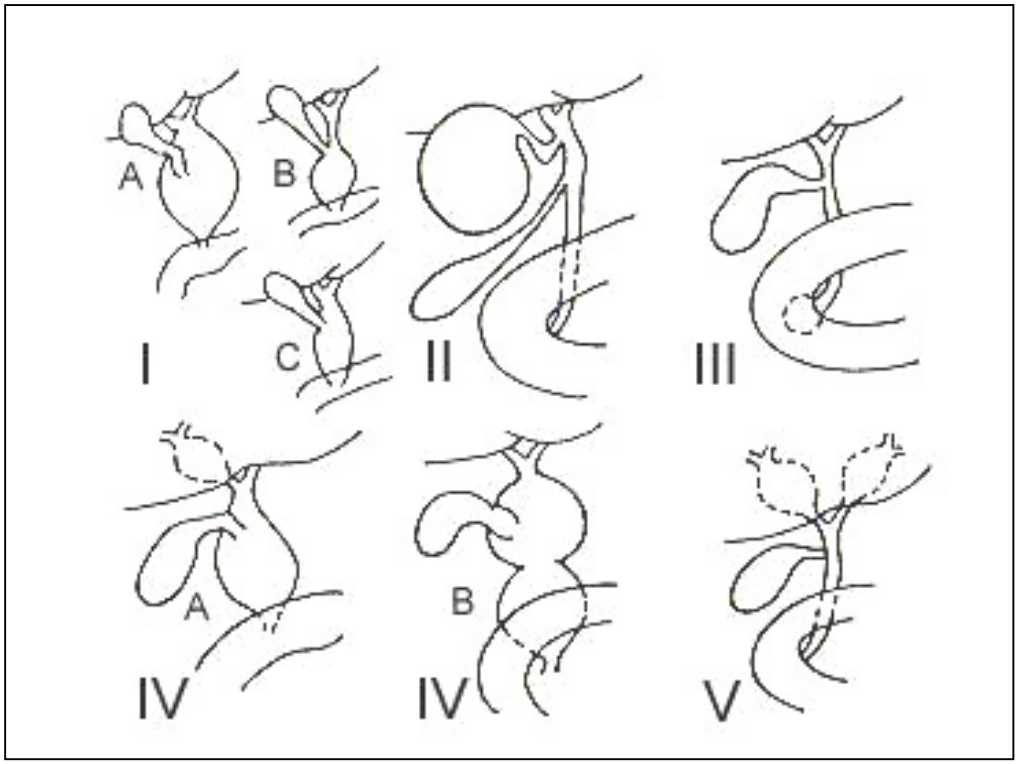

Figure 3. Todani’s classification of choledochal cyst(5).