CASE REPORT

Nagoya J. Med. Sci. **83**. 655–661, 2021
doi:10.18999/nagjms.83.3.655

Double common bile duct associated with pancreaticobiliary maljunction

Kenta Yamada[1], Takuya Ishikawa[1], Eizaburo Ohno[1], Tadashi Iida[1], Hirotaka Suzuki[1], Kota Uetsuki[1], Jun Yashika[1], Masakatsu Yoshikawa[1], Hideki Takami[2], Yoshikuni Inokawa[2], Hiroo Uchida[3], Hiroki Kawashima[4] and Mitsuhiro Fujishiro[1]

[1]*Department of Gastroenterology and Hepatology, Nagoya University Graduate School of Medicine, Nagoya, Japan*
[2]*Department of Gastroenterological Surgery (Surgery II), Nagoya University Graduate School of Medicine, Nagoya, Japan*
[3]*Department of Pediatric Surgery, Nagoya University Graduate School of Medicine, Nagoya, Japan*
[4]*Department of Endoscopy, Nagoya University Hospital, Nagoya, Japan*

ABSTRACT

A 45-year-old female was admitted to the hospital with a diagnosis of acute pancreatitis. A computed tomography scan showed two extrahepatic bile ducts. Magnetic resonance cholangiopancreatography suggested a stone in one of the bile ducts. Endoscopic retrograde cholangiopancreatography revealed two extrahepatic bile ducts joining at the hilum of the liver accompanied with pancreaticobiliary maljunction. Sphincterotomy was performed and a protein plug was drained from the bile duct. Several treatment options were discussed, and the patient was treated with laparoscopic cholecystectomy without extrahepatic bile duct resection and planned to be followed up considering the risk of carcinogenesis in the bile ducts.

Keywords: double common bile duct, pancreaticobiliary maljunction

Abbreviations:
PBM: pancreaticobiliary maljunction
CT: computed tomography
ERCP: endoscopic retrograde cholangiopancreatography
EST: endoscopic sphincterotomy
CBD: common bile duct
ACBD: accessory common bile duct

INTRODUCTION

Double common bile duct is defined as a congenital malformation in which two patency-preserved bile ducts open separately into the gastrointestinal tract. It is an extremely rare disease, with approximately two-thirds of patients being reported from Asia, and the majority are from

Received: November 18, 2020; accepted: December 25, 2020
Corresponding Author: Hiroki Kawashima, MD, PhD
Department of Gastroenterology and Hepatology, Nagoya University Graduate School of Medicine, 65 Tsurumai-cho, Showa-ku, Nagoya 466-8550, Japan
Tel: +81-52-744-2602 Fax: +81-52-744-2602, E-mail: h-kawa@med.nagoya-u.ac.jp

Japan. Pancreaticobiliary maljunction (PBM) is a congenital malformation in which the pancreatic and bile ducts join outside the duodenal wall and is a known risk factor for biliary cancer.

Double common bile duct associated with PBM is very rare, and there is no clear evidence for its treatment. We herein report a 45-year-old female who was diagnosed with double common bile duct associated with PBM and treated with laparoscopic cholecystectomy without extrahepatic bile duct resection.

CASE REPORT

A 45-year-old female with no previous medical history was admitted to the hospital with a diagnosis of acute pancreatitis after a visit to her family doctor for epigastric pain. Her alcohol intake was equivalent to approximately 40 g of pure ethanol, and alcoholic pancreatitis was initially suspected. Pancreatitis improved with conservative treatment. Magnetic resonance imaging (MRI) performed in search of a cause raised suspicion of a double bile duct and common bile duct stones. She was referred to our hospital for further investigation and treatment. She had no symptoms at that time. Her abdomen was flat and soft, with no tenderness. There was no abnormality in the blood test at the time of the patient's visit to our hospital. An abdominal contrast-enhanced computed tomography (CT) scan showed two extrahepatic bile ducts (Figure 1). No positive CT stones were noted. There were no anatomical anomalies in the arterial or portal systems. Magnetic resonance cholangiopancreatography (MRCP) also showed two extrahepatic bile ducts (Figure 2). One bile duct was suspected to have PBM. A low-intensity area was observed in this bile duct. Transabdominal ultrasound showed a hyperechoic structure in the bile duct (Figure 3). Two extrahepatic bile ducts were delineated, and the pancreatic duct joined one of the bile ducts outside the duodenal wall. Endoscopic ultrasound showed diffuse thickening of the gallbladder wall without an obvious mass. Endoscopic retrograde cholangiopancreatography (ERCP) showed the findings of PBM (Figure 4). Two extrahepatic bile ducts were delineated, and they joined at the hilum of the liver. The cystic duct branched from the bile duct and had no communication with the pancreatic duct. Both of the extrahepatic bile ducts opened into the major papilla, but there seemed to be two orifices, and we judged that the two ducts separately opened into the duodenum. Intraductal ultrasonography (IDUS) showed no thickening of the walls of the extrahepatic bile ducts. The pancreatic duct joined one of the extrahepatic bile ducts at two sites upstream of the sphincter of Oddi.

Based on the above exams, the patient was diagnosed with double common bile duct associated with PBM. Endoscopic sphincterotomy (EST) was performed at the time of ERCP, and a protein plug and mucus were drained. The cause of acute pancreatitis was thought to be the protein plug in the common bile duct. The gallbladder showed wall thickening suspected to be a hyperplastic change, but there was no evidence of wall thickening in the extrahepatic bile ducts. Several treatment options, including cholecystectomy and extrahepatic bile duct resection, were discussed, and consequently, the patient was treated with laparoscopic cholecystectomy without extrahepatic bile duct resection, which was performed at the patient's request. Postoperative pathology showed hyperplastic change in the gallbladder, with no malignancy. The patient was planned to be followed up with periodical imaging exams considering the risk of carcinogenesis in the bile ducts.

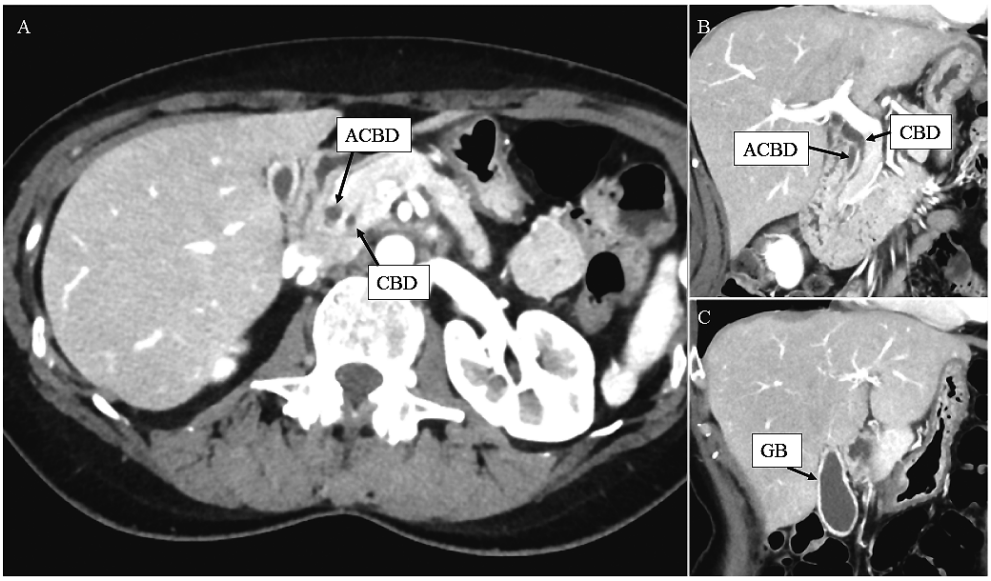

Fig. 1 Contrast-enhanced computed tomography (CT) findings
Fig. 1A: Axial abdominal contrast-enhanced CT findings showed two extrahepatic bile ducts. No positive CT stones were noted.
Fig. 1B: Coronal abdominal contrast-enhanced CT findings showed two extrahepatic bile ducts. No positive CT stones were noted.
Fig. 1C: The gallbladder wall was diffusely thickened.
CBD: common bile duct
ACBD: accessory common bile duct
GB: gallbladder

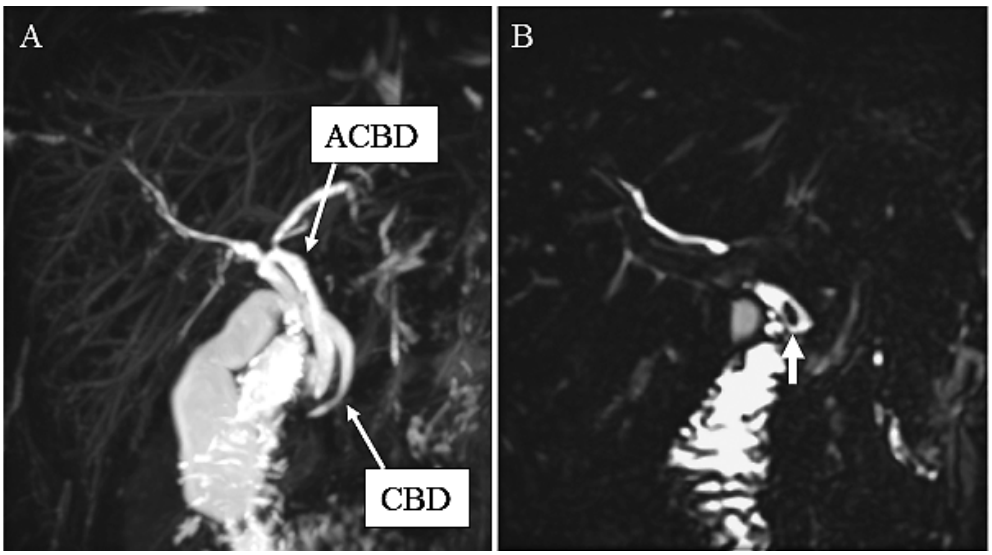

Fig. 2 Magnetic resonance cholangiopancreatography (MRCP) findings
Fig. 2A: MRCP findings showed two extrahepatic bile ducts.
Fig. 2B: A low-intensity area was observed in this bile duct (arrow).
CBD: common bile duct
ACBD: accessory common bile duct

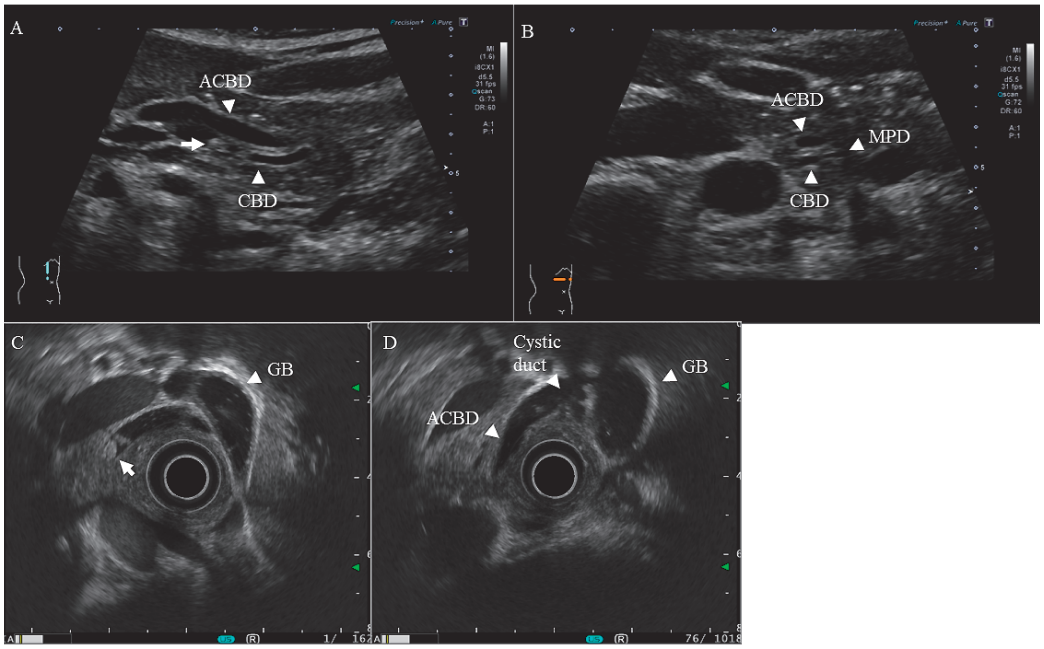

Fig. 3 Abdominal ultrasound (AUS) and endoscopic ultrasound (EUS) findings

Fig. 3A: AUS findings showed two extrahepatic bile ducts (arrowheads, common bile duct [CBD] and accessory common bile duct [ACBD]). A hyperechoic structure was observed in the CBD (arrow).

Fig. 3B: The main pancreatic duct (MPD) joined the CBD outside the duodenal wall.

Fig. 3C: EUS findings showed that the gallbladder wall was diffusely thickened. A hyperechoic structure was observed in the CBD (arrow).

Fig. 3D: The cystic duct branched from the accessory CBD.

CBD: common bile duct

ACBD: accessory common bile duct

MPD: main pancreatic duct

GB: gallbladder

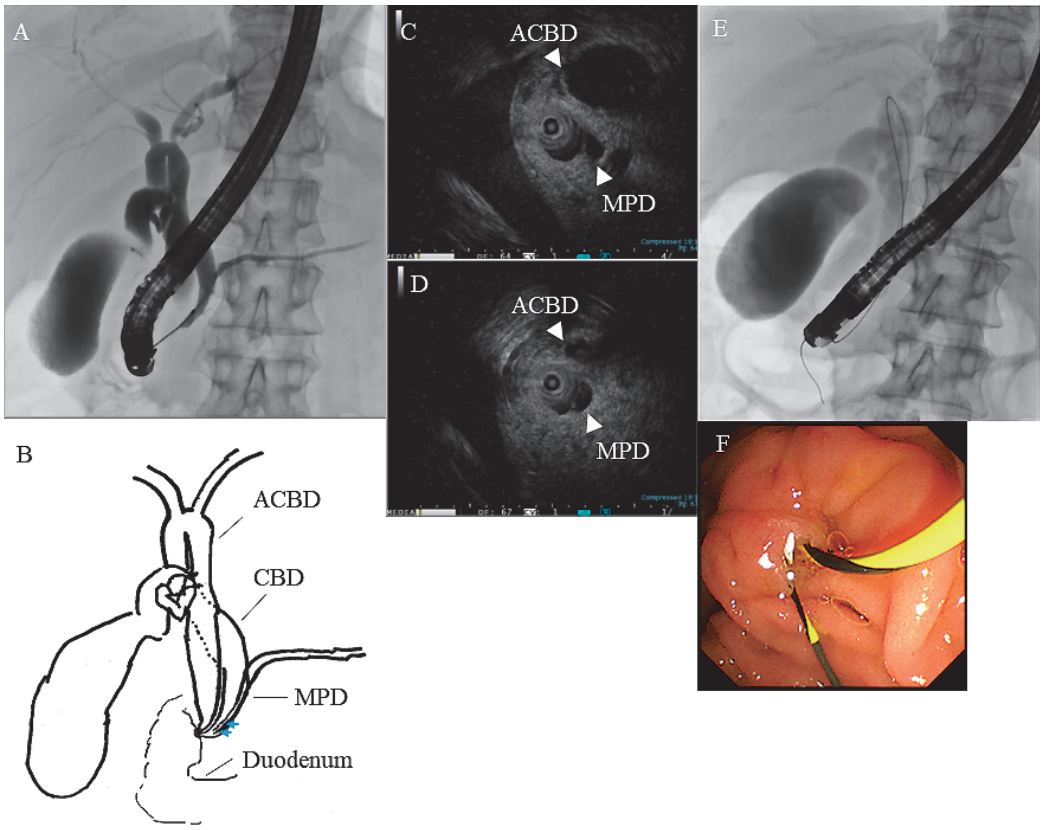

Fig. 4 Endoscopic retrograde cholangiopancreatography (ERCP) and intraductal ultrasonography (IDUS) findings

Fig. 4A: ERCP showed pancreaticobiliary maljunction. Two extrahepatic bile ducts were delineated, and they were joined at the hilum of the liver.

Fig. 4B: Schematic of Figure 4A. The MPD was connected at two sites (arrow).

Fig. 4C-4D: IDUS findings showed no thickening of the bile duct wall. The pancreatic duct communicated with the bile duct at two locations upstream of the sphincter of Oddi.

Fig. 4E-4F: The guidewire inserted into the CBD protruded from the major papilla passing through the accessory CBD.

CBD: common bile duct
ACBD: accessory common bile duct
MPD: main pancreatic duct

DISCUSSION

Double common bile duct was first described by Vesalius in 1543 and is defined as a congenital malformation in which two patency-preserved bile ducts open separately into the gastrointestinal tract.[1,2] It is an extremely rare disease, with approximately two-thirds of patients being reported from Asia, and the majority are from Japan.[3] The morphological classification of the disease is based on the classification made by Goor and Ebert[2] in 1972 and modified by Saito et al in 1988.[4] The present was classified as type II (common bile duct dichotomy).

PBM is a congenital malformation in which the pancreatic and bile ducts join outside the duodenal wall and is a known risk factor for biliary cancer. Yamashita et al reviewed 47 Japanese

patients with double common bile duct, 16 (34%) of whom had PBM concomitantly. Of the 16, gallbladder cancer was found in 7 (44%) patients; whereas, cholangiocarcinoma was not.[3] It is also known that PBM can produce protein plug in the main pancreatic duct, which potentially causes pancreatitis. Obviously, initial acute pancreatitis was a prologue of the subsequent clinical course in the present patient.

Regarding the treatment of double common bile duct, there are several reports on resection of the accessory bile ducts or extrahepatic bile ducts because of the suggested risk of recurrent cholangitis and carcinogenesis if the accessory bile ducts open ectopically,[3] but the long-term prognosis of this procedure is unknown. Extrahepatic bile duct resection is recommended for the treatment of PBM with dilated biliary tracts. On the other hand, prophylactic cholecystectomy is recommended for biliary nondilated PBMs, but there is no consensus on prophylactic extrahepatic bile duct resection.[5] 17.1% of adults after extrahepatic bile duct resection for PBM have complications such as cholangitis, intrahepatic stones, pancreatitis, pancreatic stones, and cancer, and 14.6% require hospitalization or cause life-threatening conditions. It has also been suggested that postoperative cholangiocarcinoma may be more common in patients after extrahepatic bile duct resection than in the general population.[6] Double common bile duct associated with PBM is very rare, and there is no available evidence supporting a specific surgical intervention.

In the present case, two bile ducts were present, and both ducts opened to the major papilla separately. The diameter of each bile duct was less than 10 mm, and there were no findings of bile duct dilatation. In addition, although wall thickening of the gallbladder was detected, suggesting chronic inflammation, which might be a risk factor for carcinogenesis, there was no wall thickening in the extrahepatic bile ducts. Although this case had double common bile duct, the pathogenesis was considered similar to that of PBM with a nondilated bile duct. Based on these reasons, laparoscopic cholecystectomy without extrahepatic bile duct resection was performed, and the patient continued follow-up.

The long-term prognosis of patients with double common bile duct associated with PBM is not clear, and there is no established follow-up strategy for the remaining extrahepatic bile ducts. Hyperplastic changes are reflected as wall thickening of the gallbladder and bile duct wall on ultrasound.[7] For carcinogenesis in PBM, a hyperplasia-dysplasia-carcinoma sequence has been suggested.[8] The gallbladder and bile ducts in the present case could be evaluated in detail by transabdominal ultrasound and endoscopic ultrasound, which are useful tools for follow-up. As EST prevents refractory pancreatitis associated with PBM, we performed EST prior to cholecystectomy.[9] In the present case, EST was preoperatively performed during ERCP, and we hope it will lead to the prevention of recurrent pancreatitis in the future. We previously reported another case of double common bile duct without PBM.[10] This case presented with acute cholecystitis, and the two nondilated extrahepatic bile ducts opened separately into the duodenum, but both were located in the second portion. The patient underwent cholecystectomy without extrahepatic bile duct resection and had an uneventful clinical course. A surgical decision should be made considering individual morphologic and pathophysiologic findings. Patients with the present rare abnormality should be followed-up and cumulated for a long collection period to determine the disease nature.

ACKNOWLEDGEMENTS

Not applicable.

CONFLICT OF INTEREST STATEMENT

There are no conflict of interest statement to declare.

REFERENCES

1 Vesalius A. De humani corporis fabrica liver. *Cap*. 1543;2:624.
2 Goor DA, Ebert PA. Anomalies of the biliary tree. Report of a repair of an accessory bile duct and review of the literature. *Arch Surg*. 1972;104(3):302–309. doi:10.1001/archsurg.1972.04180030050012.
3 Yamashita K, Oka Y, Urakami A, Iwamoto S, Tsunoda T, Eto T. Double common bile duct: a case report and a review of the Japanese literature. *Surgery*. 2002;131(6):676–681. doi:10.1067/msy.2002.124025.
4 Saito N, Nakano A, Arase M, Hiraoka T. A case of duplication of the common bile duct with anomaly of the intrahepatic bile duct [in Japanese]. *Nihon Geka Gakkai Zasshi*. 1988;89(8):1296–1301.
5 Kamisawa T, Ando H, Suyama M,et al. Japanese clinical practice guidelines for pancreaticobiliary maljunction. *J Gastroenterol*. 2012;47(7):731–759. doi:10.1007/s00535-012-0611-2.
6 Ando H, Horiguchi A. Remaining problems of a current Japanese clinical practice guidelines for pancreaticobiliary maljunction and congenital biliary dilatation; Proposal for next [in Japanese]. *J J B A*. 2019;33(4):713–717. doi:10.11210/tando.33.713.
7 Watanabe M, Shiozawa K, Tamura A, Nemoto T, Shibuya K, Sumino Y. A case of mucosal hyperplasia of gallbladder associated with double bile duct and pancreaticobiliary maljunction [in Japanese]. *Jpn J Med Ultrasonics*. 2011;38(3):297–300. doi:10.3179/jjmu.38.297.
8 Kamisawa T, Kuruma S, Chiba K, Tabata T, Koizumi S, Kikuyama M. Biliary carcinogenesis in pancreaticobiliary maljunction. *J Gastroenterol*. 2017;52(2):158–163. doi:10.1007/s00535-016-1268-z.
9 Terui K, Yoshida H, Kouchi K, et al. Endoscopic sphincterotomy is a useful preoperative management for refractory pancreatitis associated with pancreaticobiliary maljunction. *J Pediatr Surg*. 2008;43(3):495–499. doi:10.1016/j.jpedsurg.2007.10.071.
10 Ishikawa T, Hirooka Y, Itoh A, Kawashima H, Niwa Y, Goto H. A case of double common bile duct [in Japanese]. *J J B A*. 2009;23(4):709–712. doi:10.11210/tando.23.709.