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An extremely rare combination of choledochocoele and bile duct duplication escalating severe acute pancreatitis and cholangitis: A case report

Hirotaka Okamoto*, Shugo Shiba, Hiroyuki Wakana, Kenji Kawashima, Toshio Fukasawa

Department of Surgery, Tsuru-municipal Hospital, Yamanashi, Japan

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ABSTRACT

BACKGROUND: Combination of choledochocoele and extra-hepatic duct duplication is an extremely rare congenital abnormality.

CASE PRESENTATION: The patient was an 81-year-old Japanese man. He visited the emergency room for severe abdominal colic pain. He was diagnosed with severe pancreatitis with cholelithiasis and treated conservatively by percutaneous trans-hepatic gallbladder drainage (PTGBD) for 4 months. Thereafter, he was transferred to our institute and cholangiography was performed via the PTGBD tube, revealing cholecysto- and choledocholithiasis. The cystic-duct joined the right hepatic duct with extra-hepatic bile duct duplication and the terminal bile duct flowed into the cystic papilla of Vater. The main pancreatic duct also joined into the cystic papilla. These observations confirmed choledochocoele with extra-hepatic bile duct duplication. Surgical exploration was performed, and hepatico-jejunostomy with hepatic-ductoplasty and cholecystectomy with choledocholithotomy were carried out. He was discharged and his course was uneventful.

CONCLUSION: A very rare combined case of choledochocoele with bile duct duplication, which would escalate the pancreatitis and cholangitis, was successfully treated. Their pathogeneses in relation to pancreaticobiliary maljunction is discussed.

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1. Introduction

Choledochocoele was first reported by Wheeler [1] as a rare type of congenital choledochal dilatation (choledochal cyst). Choledochal cysts were previously classified by Alonso-Lej [2]. He classified choledochal cysts into 3 categories: Type I, represents segmental dilatation or a fusiform common bile duct; Type II, exhibits a diverticulum of the common bile duct; and Type III involves choledochocoele, dilatation of the distal portion of the common bile duct, and bulging into the duodenal lumen. Todani also further classified 2 types of choledochal cysts; Type IV, a combination of cysts of the common bile duct and intrahepatic ducts and Type V, a variation of dilatation of the common bile duct with Caroli's disease [3]. This disorder is associated with pancreaticobiliary maljunction (PBMJ), but their exact relationship is unclear, especially in cases of choledochocoele.

Extra-hepatic bile duct duplication is also a rare abnormality of the biliary system, which can be divided into 5 types [4,5]. These

biliary tree abnormalities were described in association with biliary disorders or malignancy.

There are no reports of combined case of choledochocoele with bile duct duplication. We first report an extremely rare case of an 81-year-old Japanese male with repeated severe pancreatitis and cholangitis escalated by combined choledochocoele (Type III choledochal cyst) and bile duct duplication. Informed consent was obtained from the patient for the publication of this report. This work is reported in line with the SCARE criteria [6] for case report publication.

2. Case presentation

An 81-year-old Japanese male was admitted to the emergency department with a chief complaint of severe upper abdominal pain. On admission, he was diagnosed with acute cholecystitis with cholecystolithiasis. Therefore, percutaneous trans-hepatic gallbladder drainage (PTGBD) was performed first. Surgery was planned, but severe pancreatitis caused by a biliary stone developed. Conservative treatment of the pancreatitis was performed, resulting in recovery with subsequent ileus. The paralytic ileus was released by decompression treatment of the ileus tube 4 months

* Corresponding author at: 5-1-55, Tsuru, Tsuru-city, Yamanashi 402-0056, Japan.

E-mail address: hirotakaoka@hotmail.com (H. Okamoto).

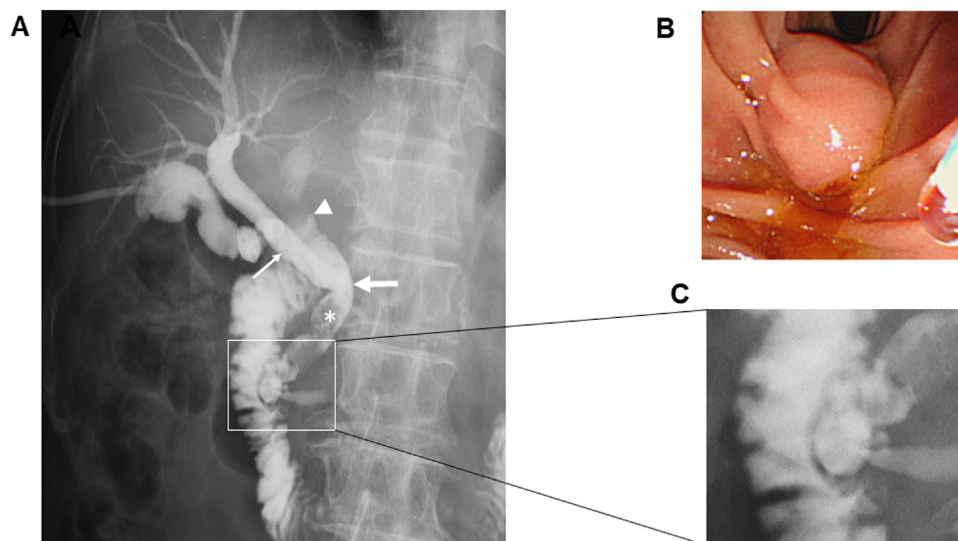


Fig. 1. A. Cholangiography from the PTGBD tube. Asterisk indicates choledocholithiasis. The large white arrow indicates the bifurcation between the right and left bile ducts. The small arrow indicates the junction of the cystic duct of the gallbladder to the right bile duct. The arrowhead indicates the left bile duct. B. Endoscopic view of the papilla of Vater during ERCP. The papilla of Vater was swollen with a single orifice, from which biliary fluid was observed. C. Magnified view of the duodenal papilla on cholangiography. Contrast medium was pooling in the terminal bile duct at the junction of the main pancreatic duct, indicating choledochoceale.

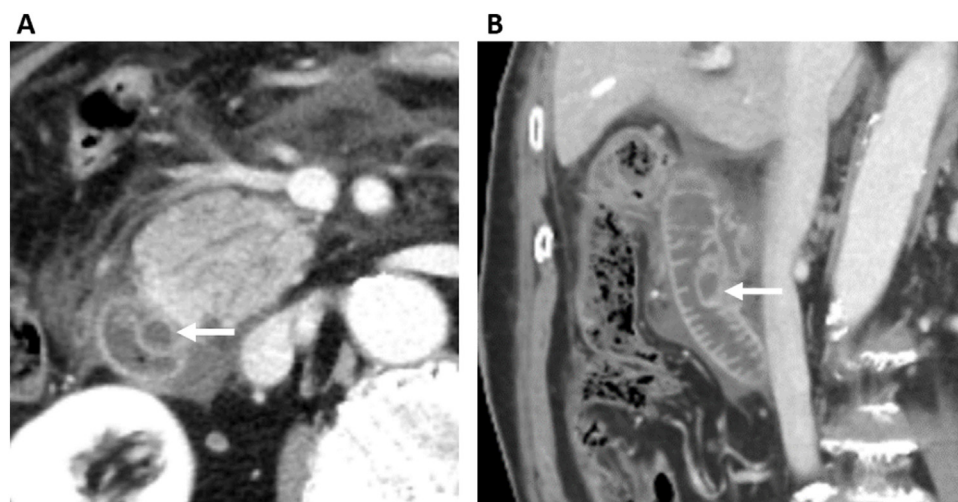


Fig. 2. Abdominal CT view. The white arrow indicates a small intraduodenal cystic lesion adjacent to the pancreas head (A, Transverse view, B, Coronal view), indicating choledochoceale.

after the initial GTGBD treatment. He was then transferred to our institute for further treatment.

Cholangiography was performed from the PTGBD tube, revealing the filling defect of stone (asterisk in Fig. 1A), the junction of the cystic duct to the right hepatic duct (thin white arrow in Fig. 1A), and the lower junction of the right and left hepatic ducts (thick white arrow in Fig. 1A). These observations demonstrated bile duct duplication. Furthermore, the terminal bile duct flowed into the cystic papilla of Vater and the main pancreatic duct joined into the cystic papilla (Fig. 1C). The distal portion of the common bile duct was dilated and bulging into the duodenal lumen. Duodenal endoscopy also revealed swelling of the papilla of Vater with a normal orifice (Fig. 1B). On abdominal CT, a cystic lesion with similar wall characteristics to those of the duodenal wall at the papilla of Vater of the descending portion of duodenum was noted (white arrow in Fig. 2A, B), suggesting choledochoceale.

The patient had lost weight and was exhausted due to long-term fasting from severe pancreatitis with subsequent ileus. Surgical exploration was not planned until he recovered, including the ileus. After starting per-oral intake, his body weight gradually increased and his course was uneventful for a few months. However, he was re-admitted due to acute cholangitis. Thereafter, exploratory surgery was conducted. Dense fibrous adhesion was extensive, especially at the upper abdominal portion with minimal ascites. After removing the fibrous attachment between the transverse colon and duodenum, the hepatoduodenal ligament was exposed. The hepatic ducts were identified superior to the duodenal bulb, and junction of the right and left hepatic ducts was observed just cranial to the pancreas head. Intraoperative findings included duplication of the extrahepatic bile ducts (Fig. 3). The right bile duct was isolated by taping. Bifurcation was noted above the duodenal bulb. Direct identification of choledochoceale by duodenotomy

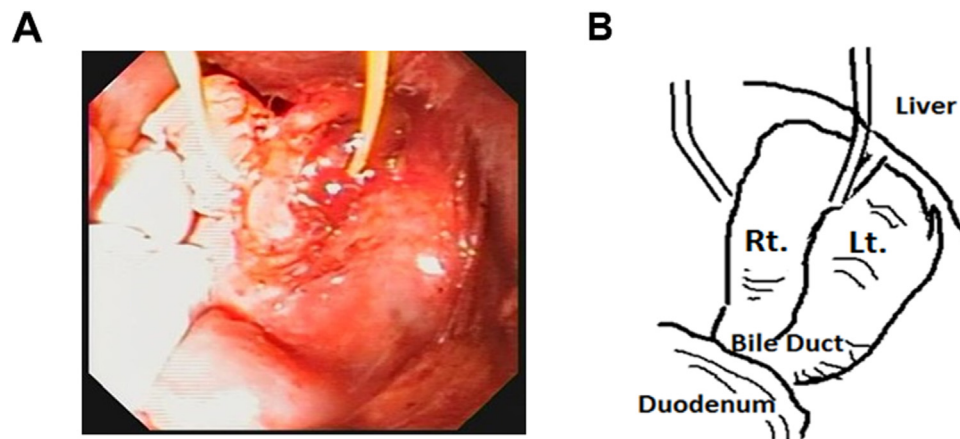


Fig. 3. A. Intraoperative findings. The right extra-hepatic bile duct was isolated by taping. Bifurcation was noted just above the duodenal bulb (B, schematic view).

was not carried out because it was previously confirmed by pre-operative imaging. Cholecystectomy and choledochotomy with choledocholithotomy was performed. Additionally, hepatic duct-plasty and biliary reconstruction of hepatico-jejunostomy were done. The hepatic duct-plasty was carried out as follows: To create a single lumen, the right and left hepatic ducts were isolated at the hepatic hilus, and dissected down to their fusion in the distal hepatic ligament. Thus, two hepatic lumens were conjoined by suturing the closed side wall to each other, resulting in one lumen. Hepatico-jejunostomy was performed via the ante-colic route in an end-to-side manner. Separation of the bile and pancreatic flows may prevent the formation of biliary stones and pancreatitis. The postoperative course was uneventful and the patient was discharged 2 weeks later. He remained asymptomatic at the 2-year follow-up.

3. Discussion

There are few reports of combined case of congenital choledochal dilatation (choledochal cyst) with bile duct duplication [7]. In their report, all cases of choledochal dilatation were cystic or fusiform type (Type I). However, there are no reports of the combined case of choledochoceles (Type III) with bile duct duplication. We first documented an extremely rare combination of choledochoceles and bile duct duplication manifesting severe pancreatitis and cholangitis.

Choledochocoele is rare, accounting for less than 2% of all cases of choledochal cysts [8]. The rarity of this disease makes diagnosis difficult. Its etiology also remains unclear, although there are two theories, congenital and acquired. Schwegler et al. proposed the congenital theory in which two orifices of the ampulla existed during embryogenesis. Development of the two orifices may have been impaired, resulting in the formation of choledochoceles and pancreatic-biliary maljunction [9]. On the other hand, Sterling proposed the acquired theory that obstruction of the bile flow by papillary stenosis or Oddi sphincter dysfunction leads to the increase in pressure in the distal bile duct, causing it to protrude into the duodenal wall [10]. In our case, it would be hard to diagnose a choledochocoele due to acute pancreatitis progression on initial treatment. First diagnosis at the initial hospital was choledocholithiasis with cholecystolithiasis, resulting in acute pancreatitis. Biliary stone might cause a pancreatitis. A choledochocoele had not been diagnosed until by cholangiography via PTGBD at our hospital. Furthermore, we pointed out the possibility of another reason of an acute pancreatitis progression. It was that not only the stone but also the choledochocoele might cause the pancreatitis progression.

We speculated that retention of the stone in the choledochocoele might lead to the further pancreatitis progression.

An intraluminal duodenal diverticulum (IDD) is also a rare abnormality and should be differentiated from choledochocoele [11,12]. The IDD was attached partly to the posterolateral wall of the duodenum, whose base is usually located around the papilla of Vater. Although CT cannot always precisely differentiate between IDD and choledochocoele, a double-lumen appearance with a small orifice of the diverticulum and large true lumen of the duodenum can be identified by endoscopy. In our case, no double-lumen was noted on endoscopy, thereby excluding IDD.

Duodenal duplication cysts should be considered in the differential diagnosis [13]. Duplication cysts are also a rare congenital abnormality presenting in young children. They are located near the papilla of Vater and the second region of the duodenum is similar to choledochocoele, but choledochocoele presents at an older age. Other than age, differential diagnosis between choledochocoele and duodenal duplication cyst depends on whether the epithelium is lining the duodenal mucosa or biliary tract mucosa according to histopathological examination [14]. Although, in our case, histopathological examination did not be performed, clinical findings of the cholangiography showed that the cystic papilla directly continued from the bile duct. These findings suggested the continuation of intra-cystic epithelium from the bile duct rather than the duodenal mucosa.

Choledochocoele can be morphologically classified into three groups [15]. Type A: cystic dilatation of the common channel, Type B: cystic dilatation of the intramural bile duct forming the common channel, and Type C: cystic dilatation of the intramural bile duct opening into the duodenum separately from the pancreatic duct. This classification reflects the therapeutic indications, including pancreaticobiliary maljunction. Briefly, in Type A and B, forming a common channel, biliary-pancreatic fluid reflux may develop, whereas in Type C, the fluid cannot be regurgitated. Our case was considered to be Type A because of cystic dilatation of the common channel. Theoretically, biliary pancreatic fluid reflux is possible; however, in our case, the bile amylase level in the gallbladder and bile duct was not high on clinical examination.

PBMJ is defined as a congenital malformation in which the pancreatic and bile ducts join anatomically outside the duodenal wall [16]. The sphincter of Oddi cannot influence the pancreaticobiliary junction due to the abnormally long common channel. This disorder may induce reciprocal reflux between pancreatic juice and bile, resulting in pathological conditions, including impairment of pancreatic and biliary fluid excretion, and biliary malignancy. In our case, choledochocoele was found inside the duodenal wall on CT (Fig. 2), demonstrating the absence of PBMJ. However, the exact

location of the sphincter of Oddi was unclear because microscopic examination was not performed in this case of choledochocoele. Regarding biliary malignancy, no cancer was micro- or macroscopically detected in the resected gallbladder.

Biliary tree anomalies are clinically common. Variations were reported in approximately 40% of the general population [17]. Among them, extrahepatic bile duct duplication is rare, with 5 proposed types [4,5]. Type I has a septum in the extrahepatic bile duct. Type II exhibits division of the duct in two separated junctions. Type III has double independent drainage. Type IV has double drainage and one or two intercommunications. Type V has double drainage joined at the lower region of the bile duct system. Our case corresponded to Type V. Direct cholangiography and intraoperative findings demonstrated extrahepatic bile ducts joining at the lower region of the bile duct near the cranial edge of the pancreas head. Cholangiography revealed the cystic duct joining the right extrahepatic bile duct (Figs. 1A and 3).

In the case of choledochocoele alone, malignant transformation was reported to be rare [18]. However, biliary malignancy may be more frequent in combination with PBMJ [19]. On the other hand, bile duct duplication variants alone may not be associated with malignancy. Similar to choledochocoele, when accompanied by PBMJ, malignancy may be more frequent [4]. In our case, biliary malignancy was absent, and the patient was diagnosed during the peri-operative and follow-up periods, thus long-term follow-up may be necessary.

Resection of choledochocoele, papillectomy, or pancreatic resection was not chosen in this case, because only choledochocoele itself could not cause a pancreatitis and a cholangitis. Biliary drainage was achieved by hepatico-jejunostomy with hepatic duct-plasty and cholecystectomy to prevent the cholangitis with the stone formation. The patient had an uneventful operative course and follow-up up to 2 years.

4. Conclusions

We reported the successful treatment of a very rare case of choledochocoele with bile duct duplication escalating pancreatitis and cholangitis.

Declaration of Competing Interest

The first author and the co-authors have no conflicts of interest to declare.

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Ethical approval

This is a case report with no identifiable information included in the manuscript, so ethical approval was not obtained. Our institution has exempted case reports from ethnical approval.

Consent

Written informed consent was obtained from the patient and is available upon request. No patient identifying material was used in this manuscript.

Author contribution

This research was designed by Hirotaka Okamoto.

Data collection, analysis and case management: Shugo Shiba, Hiroyuki Wakana.

Revising of the final version of the manuscript: Hirotaka Okamoto, Shugo Shiba, Hiroyuki Wakana, Kenji Kawashima, Toshio Fukasawa.

Registration of research studies

As this is not a 'first-in-man' case study, our paper is eligible to be registered.

Guarantor

Hirotaka Okamoto

Department of Surgery, Tsuru Municipal Hospital, 5-1-55, Tsuru, Tsuru-city, Yamanashi, 402-0056 Japan

Department of Gastrointestinal, Breast & Endocrine Surgery, Faculty of Medicine, University of Yamanashi, Shimokato, Chuo-city, Yamanashi, 409-3898 Japan

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References

- [1] W. Wheeler, An unusual case of obstruction of the common bile-duct (choledochocoele?), *Br. J. Surg.* 27 (1940) 446–448.
- [2] F. Alonso-Lej, W.B. Lej, D.J. Pessagno, Congenital choledochal cysts with a report of 2 and analysis of 94 cases, *Int. Abstr. Surg.* 108 (1959) 1–30.
- [3] T. Todani, Y. Watanabe, M. Naruse, K. Tabuchi, K. Okajima, Congenital bile duct cysts, *Am. J. Surg.* 134 (1977) 263–269.
- [4] K. Yamashita, Y. Oka, S. Urakami, T. Iwamoto, T. Tsunoda, T. Eto, Double common bile duct: a case report and a review of the Japanese literature, *Surgery* 131 (2002) 676–681.
- [5] E. Choi, J.H. Byun, B.J. Park, M.G. Lee, Duplication of the extrahepatic bile duct with anomalous union of the pancreaticobiliary duct system revealed by MR cholangiopancreatography, *Br. J. Radiol.* 80 (2007) e150–e154.
- [6] R.A. Agha, M.R. Borrelli, R. Farwana, K. Koshy, A. Fowler, D.P. Orgill, For the SCARE Group, The SCARE 2018 statement: updating Consensus Surgical Case Report (SCARE) guidelines, *Int. J. Surg.* 60 (2018) 132–136.
- [7] K. Tahara, Y. Ishimatsu, J. Fujino, M. Suzuki, M. Hatanaka, A. Igarashi, H. Ikeda, Association of extrahepatic bile duct duplication with pancreaticobiliary maljunction and congenital biliary dilatation in children: a case report and literature review, *Surg. Today* 43 (2013) 800–805.
- [8] M. Yamaguchi, Congenital choledochal cyst. Analysis of 1,433 patients in the Japanese literature, *Am. J. Surg.* 140 (1980) 653–657.
- [9] R.A. Schwegler, E.A. Boyden, The development of the pars intestinalis of the common bile duct in the human fetus, with the special reference to the origin of the ampulla of Vater and sphincter of Oddi, *Anat. Rec.* 67 (1937) 441–467.
- [10] J.A. Sterling, Diverticula in the terminal portion of the common bile duct, *Am. J. Pathol.* 25 (1948) 325–337.
- [11] S. Willemer, H. Dombrowski, G. Adler, J.F. Bussmann, R. Arnold, Recurrent acute pancreatitis and intraluminal duodenal diverticulum, *Pancreas* 7 (1992) 257–261.
- [12] R.H. Hartley, A.P. Barlow, J.O. Kilby, Intraluminal duodenal diverticulum: an unusual cause of acute pancreatitis, *Br. J. Surg.* 90 (1993) 488.
- [13] A. Al-Harake, A. Bassal, M. Ramadan, M. Chour, Duodenal duplication cyst in a 52-year-old man: a challenging diagnosis and management, *Int. J. Surg. Case Rep.* 4 (2013) 296–298.
- [14] G.E. Sarris, D. Tsang, Choledochocoele: case report, literature review, and a proposed classification, *Surgery* 105 (1989) 408–414.
- [15] T. Kamisawa, M. Yoshiike, N. Egawa, K. Tsuruta, A. Okamoto, M. Matsukawa, Classification of choledochocoele, *Hepatogastroenterology* 52 (2005) 29–32.

- [16] T. Kamisawa, H. Andoh, Y. Suyama, et al., Diagnostic criteria for pancreaticobiliary maljunction 2013, *J. Hepatobiliary Pancreat. Sci.* 21 (2014) 159–161.
- [17] S.G. Puente, G.C. Bannura, Radiological anatomy of the biliary tract: variations and congenital abnormalities, *World J. Surg.* 7 (1983) 271–276.
- [18] S.D. Ladas, I. Katsogridakis, P. Tassios, T. Tastemiroglou, T. Vrachliotis, S.A. Raptis, Choledochocoele, an overlooked diagnosis: report of 15 cases and review of 56 published reports from 1984 to 1992, *Endoscopy* 27 (1995) 233–239.
- [19] J. Horaguchi, N. Fujita, G. Kobayashi, Y. Noda, K. Ito, O. Takasawa, Clinical study of choledochocoele: is it a risk factor for biliary malignancies? *J. Gastroenterol.* 40 (2005) 396–401.

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