Ectopic Opening of the Common Bile Duct Accompanied by Choledochocele and Pancreas Divisum

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Abstract

A 32-year-old woman was referred due to abdominal pain and elevated liver enzymes. Computed tomography and magnetic resonance imaging showed ectopic opening of the common bile duct (CBD) into the duodenal bulb. Esophagogastroduodenoscopy showed a hemispheric bulge in the duodenal bulb. Endoscopic retrograde cholangiopancreatography (ERCP) revealed the bulge to be cystic dilatation of the CBD. ERCP also showed no communication between the ventral and dorsal pancreatic ducts. We diagnosed the patient with ectopic opening of the CBD accompanied by choledochocele and pancreas divisum. Endoscopic incision was performed for the treatment of the choledochocele. The patient’s symptoms and elevated liver enzymes improved after treatment.

Key words: common bile duct, choledochocele, ectopic opening, pancreas, pancreas divisum, pancreaticobiliary abnormalities

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Introduction

Ectopic opening of the common bile duct (CBD) is a rare congenital biliary anomaly, with the CBD emptying into anomalous sites such as the third or fourth portion of the duodenum, stomach, or duodenal bulb (1-17). Choledochocele is also a biliary anomaly, with cystic dilatation of the intraduodenal segment of the CBD (18-20). Pancreas divisum (PD) is the most common pancreatic duct anomaly, which presents as failed fusion of the dorsal and ventral ducts (21, 22). To the best of our knowledge, no cases of ectopic opening of the CBD with choledochocele and PD have been reported. We herein report a case of ectopic opening of the CBD into the duodenal bulb accompanied by choledochocele and PD.

Case Report

A 32-year-old woman was referred to our hospital with upper abdominal pain and elevated liver enzymes. She had suffered from upper abdominal pain intermittently for six months. On admission, her subjective symptom had disappeared. Laboratory data showed elevated liver enzymes: alanine aminotransferase was 70 IU/L (normal <43 IU/L), alkaline phosphatase was 767 IU/L (normal <359 IU/L), and gamma-glutamyl transpeptidase was 290 IU/L (normal <47 IU/L). Other laboratory data were normal. Dilatation of the CBD was detected by transabdominal ultrasonography. Computed tomography and magnetic resonance imaging (MRI) showed dilatation and severe angulation of the CBD without communication between the CBD and major papilla (Fig. 1). Esophagogastroduodenoscopy revealed a hemispheric bulge with an orifice of the bile duct similar to a submucosal tumor in the duodenal bulb opposite the superior duodenal angle (Fig. 2A and B). Endoscopic ultrasonography (EUS) revealed a communication between the dilated CBD and the hemispheric bulge (Fig. 2C). We assumed that the patient had an ectopic opening of the CBD into the duodenal bulb accompanied by choledochocele,
Figure 1. MRI image. The MRI showed dilatation of the CBD and an ectopic opening of the lower CBD into the duodenal bulb with acute angulation. No communication was observed between the CBD and the major papilla.

Discussion

As shown in the schematic illustration (Fig. 4), our case had three pancreaticobiliary abnormalities as follows: ectopic opening of the CBD, choledochocele, and PD. Ectopic opening of the CBD is a rare congenital biliary condition. Openings of the CBD are usually located at the posteromedial wall of the second portion of the duodenum; the CBD rarely opens at anomalous sites such as the third or fourth portion of the duodenum, stomach, or duodenal bulb (1-17). Sezgin et al. (12) reported that 1.05% of patients undergoing ERCP are diagnosed with ectopic opening of the CBD into various sites. Choledochoceles are biliary anomalies with cystic dilatation of the intraduodenal segment of the CBD. They are often classified as type III choledochal cysts (23, 24). Previous reports have shown that 1-19% of patients with a choledochal cyst are diagnosed with a choledochocele (18-20). PD is the most common pancreatic duct anomaly, and involves failed fusion of the dorsal and ventral pancreatic ducts. Previous reports have suggested that the incidence of PD in autopsy cases is about 9% (21, 22). To our knowledge, this is the first report of an ectopic opening of the CBD into the duodenal bulb accompanied by choledochocele and PD (1-17). Choledochocele concomitant with PD is also rare, with only three case reports having been published (25-27); therefore, our case is extremely rare.

The etiology of the ectopic opening of the CBD and PD is considered to involve developmental errors during embryogenesis. The hepatic diverticulum (or ventral diverticulum) develops at the end of the foregut at four weeks of gestation, which divides into the pars hepatica, pars cystica, and ventral pancreatic bud. The liver, intrahepatic duct, and common hepatic duct develop from the pars hepatica, and the gallbladder, cystic duct, and CBD develop from the pars cystica. Furthermore, the dorsal pancreatic bud develops on the opposite side of the hepatic diverticulum. At five to seven weeks of gestation, the ventral pancreatic bud and bile ducts rotate from right to left, although the duodenum rotates clockwise, and the ventral and dorsal pancreatic buds fuse together (9, 21, 28, 29). PD occurs because of a failure of the dorsal and ventral pancreatic ducts to fuse during this embryogenetic process (21). Ectopic opening of the CBD is also considered to occur during the development of the biliary tree. In most reports about ectopic opening of the CBD, Boyden’s theory on double CBD is adopted (30). He hypothesized that early subdivision of the hepatic diverticulum into the pars hepatica and pars cystica gives rise to an aberrant bile duct apart from an anatomically normal bile duct. If the subdivision occurs very early, leaving the pars hepatica above the zone of growth that separates the stomach from the duodenum, the pars hepatica empties into the stomach. If subdivision occurs below the zone of growth, the pars hepatica empties into the duodenum (30). We assume that atresia of the normal bile ducts occurs after the embryonic event of double CBD, resulting in an ectopic opening of the CBD. Some previous reports have also made the same hypothesis (1, 6).

Choledochoceles are classified as type III choledochal cysts (23, 24); however, recent research has suggested that choledochoceles should not be included as choledochal cysts because of differences in their characteristics, such as the age, gender, and complications of affected subjects (20). With regard to etiology, choledochal cysts are considered to be a congenital condition, whereas choledochoceles are affected by innate and acquired factors (20, 31). Kagiyama et al. (32) reported that increased sphincter pressure due to sphincter of Oddi dysfunction or stenosis may cause chole-
cholechocele. In our case, although the sphincter of Oddi was absent at the ectopic opening site, there were two factors in the obstruction of bile drainage. One was an abnormal orifice of the bulge due to the ectopic opening, while the other was stenosis between the bulge and the lower CBD (Fig. 3C). We assume that the abnormal orifice and the stenosis increased the pressure between the two sites, resulting in the cholechocele. We speculated that the main factor for cholestasis was the abnormal orifice and the stenosis between the bulge and the lower CBD was not so tight because only an incision of the abnormal orifice caused an adequate outflow of bile juice.

The diagnostic criteria of ectopic openings of the CBD have not yet been established. In previous reports, such a diagnosis was often described as being made based on the following findings: 1) an orifice was observed in the stomach, bulb, or third portion of the duodenum by duodenoscopy or upper endoscopy, and the bile duct and/or the pancreatic duct were directly visualized radiographically when contrast medium was injected via this opening; 2) there was no evidence of any other drainage into the digestive tract on cholangiography; and 3) there was no evidence of a papilla-like structure in the second portion of the duodenum on duodenoscopic examination (3, 4, 8, 12, 15). In our case, the major papilla was observed at the normal location in the second portion of the duodenum, and the ventral pancreatic duct was joined to the major papilla. These findings do not meet the third of the above criteria. Some previous reports described the presence of the major papilla in the second portion of the duodenum (1, 6, 7). In other reports, the pancreatic duct was visualized in fewer than half of the total cases (2-5, 8-17), which suggests the possibility of another orifice joining with the pancreatic duct. Moreover, from the viewpoint of the embryonic process, the presence of the major papilla is consistent with Boyden’s theory (30), as described above. We propose that the ectopic openings of the CBD should be classified into two different types based on the communication between the bile and pancreatic ducts: a) both the CBD and the pancreatic duct empty into the ectopic opening (confluent type); and b) the CBD empties into the ectopic opening, whereas the pancreatic duct empties into the other opening, including the major papilla (separated type).

In general, an endoscopic incision should not be performed on patients with an ectopic opening of the CBD because the risk of perforation is very high owing to the ab-
Figure 3. ERCP images. (A) The major papilla (white arrow) and the minor papilla (white arrowhead) were observed at the normal locations. (B) Injection of contrast medium through the major papilla. The ventral pancreatic duct was short, without communication to the dorsal pancreatic duct. Cholangiography was not performed. (C) Cholangiography was performed through the orifice of the hemispheric bulge, which was observed as cystic dilatation at the terminus of the CBD. Stenosis was observed between the lower CBD and the bulge (white arrow). (D) After the endoscopic treatment.

Figure 4. Schema of the pancreaticobiliary system. Our patient had three anomalies: ectopic opening of the CBD, choledochocele, and pancreas divisum.

sence of a sphincteric structure (4, 8, 11, 15, 17). However, in our patient, the ectopic opening was accompanied by choledochocele. Previously, patients with choledochocele underwent surgery to prevent biliary malignancy. Recently, endoscopic sphincterotomy has become a standard therapy because the risk of malignancy in patients with choledochocele is extremely low, in contrast to that in the other types of choledochal cyst (20, 31). This low risk of malignancy is
considered to result from the absence of pancreatic juice reflux into bile duct (31). In our case, the biliary system and pancreatic system were separated, and there was no possibility of pancreatic juice reflux into the bile duct. Therefore, we performed an endoscopic incision for the choledochocoele by a method similar to that for endoscopic sphincterotomy. The suitability of an endoscopic incision should be carefully considered.

We report the first case of an ectopic opening of the CBD into the duodenal bulb accompanied by choledochocoele and PD. Ectopic openings of the CBD are rare congenital biliary anomalies, and the diagnostic criteria have not yet been established. Previous studies have reported contradictory information about “the presence of a papilla-like structure apart from the ectopic opening.” We propose that ectopic openings of the CBD should be classified into the two types described above. In patients with an ectopic opening of the CBD, careful examination with various modalities should be performed not only for the biliary system, but also for the pancreatic system, in order to make a correct diagnosis and provide appropriate and effective treatment.

The authors state that they have no Conflict of Interest (COI).

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References
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