Duodenal Duplication Cyst of the Ampulla of Vater

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Abstract

A 35-year-old man presented with the complaint of epigastric discomfort. Gastrointestinal endoscopy and endoscopic ultrasonography revealed a cystic lesion 20 mm in size at the ampulla of Vater. Endoscopic retrograde cholangiopancreatography (ERCP) revealed that the cystic lesion communicated with both the common bile duct and pancreatic duct via the common channel. Choledochocele was ruled out by close examination of the ERCP findings. The cystic lesion was surgically resected. Since histological findings revealed that the mucosa inside the lesion was duodenum-like and contained a layer of smooth muscle, the lesion was diagnosed as a congenital duplication cyst of the duodenum.

Key words: duplication cyst, ampulla of Vater, choledochocele

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Introduction

Duodenal duplication cysts (DDCs) are rare congenital malformations that occur most often in infants and children (1). Here, we present the case of an adult with a DDC of the ampulla of Vater that communicated with both the common bile duct and main pancreatic duct and was difficult to distinguish from choledochocele.

Case Report

A 35-year-old man was presented with the complaint of epigastric discomfort. He had no significant family or past histories. Physical examination demonstrated no abnormalities. All standard biochemical and blood tests were within normal ranges. Upper gastrointestinal endoscopy revealed a two-centimeter submucosal mass in the second part of the duodenum. Hypotonic duodenography revealed that the lesion was located at the ampulla of Vater. Endoscopic ultrasonography using a 20-MHz miniature probe revealed a cystic structure, but failed to define any anatomical association between the cystic lesion and the common bile duct and/or pancreatic duct. Computed tomography and magnetic resonance imaging of the abdomen did not reveal that the cystic lesion was located in the duodenal wall.

Using endoscopic retrograde cholangiopancreatography (ERCP), the location of the orifice of the papilla was confirmed to be at the anal side of the cystic lesion, allowing cannulation to be performed (Fig. 1a, b). The 2-cm cystic lesion was opacified first, after which the common bile duct and main pancreatic duct were opacified through the common channel (Fig. 2a-c). Amylase levels in the cystic fluid were high at 160,000 U/l (normal range: 37-120 U/l). The gallbladder, bile duct and pancreatic duct were of normal size and shape.

The original differential diagnosis was choledochocele (congenital bile duct dilatation, Todani type III), which is a cystic dilatation of the terminal end of the common bile duct. However, this diagnosis did not adequately describe the observed communication between the cyst and the common bile duct via the common channel (not the common bile duct). Surgical operation was performed based on a diagnosis of congenital malformations of the pancreatic obiliary system. During cholecystectomy, contrast fluid in the cystic duct showed no pancreaticobiliary maljunction, and the cystic lesion was subsequently resected. Amylase levels of the bile juice in the resected gallbladder were near nor-
Figure 1. Endoscopic images at ERCP. a) An elevated cystic lesion in the second portion of the duodenum. Arrow shows the location of the orifice of the papilla. b) The location of the orifice of the papilla was confirmed at the anal side of the cystic lesion, allowing cannulation to be performed.

Figure 2. ERCP findings. a) The 2-cm cystic lesion was opacified first. b) Subsequently, the common bile duct was opacified through the common channel. The cyst was not in direct communication with the common bile duct. c) At the end, the normal pancreatic duct was opacified.

normal limits at 123 U/l.

Histological findings from mucosa covering the inside of the resected cyst revealed duodenum-like mucosa that displayed a distinct layer of smooth muscle and lacked villi structure (Fig. 3a-c). Based on these findings, the cystic lesion was diagnosed as a congenital duplication cyst of the duodenum arising from the ampulla of Vater.

Discussion

Duplication cysts are spherical or tubular structures that are covered inside by gastrointestinal mucosa containing a distinct layer of smooth muscle. Although a duplication cyst may arise at any level of the alimentary tract from mouth to anus, they occur most frequently in the ileum and the posterior mediastinum. A duodenal location is less common and accounts for only 5% of all cases (2). Periampullary duplication cysts are extremely rare in duodenal cases, where discrimination from choledochocele demands particular attention.

The principle distinguishing features between periampullary DDC and choledochocele are histologic characteristics. Since the inner surface of the cyst was lined with duodenal and not bile duct mucosa, we classified it as a DDC and not as a choledochocele. Although reports of “choledochoceles” lined with duodenal mucosa exist, we believe that these choledochoceles would be better classified as DDC (3, 4). The possibility exists that cases similar to the present case were included among these reports of choledochoceles (3).

In 1998, Elton et al (5) proposed a new entity termed “dilated common channel syndrome”, which was distinguishable from a choledochocele by its small size. On ERCP, dilated common channel syndrome is defined by separate drainage of the common bile duct and the pancreatic duct into a common channel with a diameter of less than 1 cm. In discrimination of periampullary DDC from di-
Figure 3. Histopathological findings. a) The mucosa inside the cyst and the duodenal mucosa share the smooth muscle layer via rough connective tissue (Hematoxylin and Eosin staining, ×2). Arrow shows the mucosa inside the cyst. b) The cavity of the cyst was covered inside by duodenum-like mucosa that lacked villi structure. Brunner’s gland was also present in the duodenum-like mucosa (Hematoxylin and Eosin staining, ×10). c) Normal duodenal mucosa (villi) from the side of the duodenum is shown (Hematoxylin and Eosin staining, ×10).

Related common channel syndrome, none these findings were detected on ERCP in the present case.

To date, there are several reported cases of periampullary DDC (1, 3, 4, 6-9). Among these, communication with the pancreaticobiliary system was confirmed by ERCP in three cases (1, 8, 9). Luckmann et al (1) and Sezgin et al (8) reported a case involving a large periampullary DDC that contained gallstones and communicated with the bile duct. Niehues et al (9) reported a case of DDC that led to recurrent pancreatitis due to intermittent obstruction of both the bile and pancreatic ducts. This case was first diagnosed preoperatively as a double gallbladder with compression of the duodenum by the accessory gallbladder. In these cases, close examination of ERCP images was not reported.

In the present case, ERCP serial images were closely analyzed, but because the cyst communicated with the bile duct via the common channel, we could not interpret the lesion as a part of the bile duct. This feature discounted a diagnosis of choledochocele, since this condition is defined by cystic dilatation of the terminal end of the common bile duct. The cyst was resected surgically and histological examination of the cyst interior revealed duodenum-like mucosa that lacked villi structure but contained a distinct layer of smooth muscle. The cystic lesion was ultimately diagnosed as a congenital duplication cyst of the duodenum arising from the ampulla of Vater.

DDCs, including those arising from the ampulla of Vater, have traditionally been managed with complete surgical resection. More recently, cases of endoscopic treatment of DDCs have also been reported (8, 10-13). Using endoscopic incision by sphincterotome as an alternative to surgical intervention, Sezgin et al (8) accomplished safe clearance of gallstones in a periampullary DDC. DDCs are generally benign lesions, but two cases of malignant transformation have been reported to date (14, 15). Orr and Edwards (16) have also reported cases in adults in which duplication cysts presented with a potentially malignant nature. For this reason, en bloc resection is preferable, even in the absence of malignant symptoms.

A DDC arising from the ampulla of Vater and communicating with the bile duct is difficult to distinguish from choledochocele. In the present case, ERCP findings refuted any diagnosis of choledochocele. Ultimately, histological findings of resected specimens are the only evidence that can provide a clear distinction between DDC and choledochocele. Based on our experience with this case, discrimination between DDC and choledochocele may be possible in the future by application of a thorough pre-operative practice and close examination of ERCP images.

References

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