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An unusual cause of recurrent pancreatitis: A gastric duplication cyst with an accessory pancreatic lobe

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ABSTRACT

Congenital anomalies of pancreas and its ductal drainage are uncommon but in general surgically correctable causes of recurrent pancreatitis. A gastric duplication cyst communicated with an accessory pancreatic lobe is an extremely rare cause of recurrent pancreatitis, but an early and accurate diagnosis of this anomaly is important because suitable surgical treatment may lead to a satisfactory outcome. Herein, we presented multidetector computed tomography and magnetic resonance imaging findings of a gastric duplication cyst communicating with an accessory pancreatic lobe via an aberrant duct in a 29-year-old woman with recurrent acute pancreatitis and also reviewed other similar cases reported in the literature.

Keywords: Aberrant pancreatic duct, accessory pancreatic lobe, acute pancreatitis, gastric duplication cyst, multidetector computed tomography, magnetic resonance imaging

Case Report

INTRODUCTION

Congenital causes of recurrent pancreatitis include anomalies of the biliary or pancreatic ducts, especially pancreas divisum. A gastric duplication cyst communicating with an aberrant pancreatic duct is an extremely rare but curable cause of recurrent pancreatitis in children and young adults. Only 18 cases that have this unusual anomaly have been reported in the literature (1-17), and in minority of them an associated accessory pancreatic lobe was described (6,10-13,16). In the patients with this anomaly, a definitive diagnosis was rarely made preoperatively and several operative explorations were required in some cases (11,12). The aberrant pancreatic ductal system was revealed by endoscopic retrograde cholangiopancreaticography (ERCP) in majority of the cases diagnosed preoperatively and the usefulness of ERCP was emphasized (6,13). To our knowledge, there are only a few reports of spiral computed tomography (CT) (13) and no previous report of multidetector CT and magnetic resonance imaging (MRI) findings of this unusual anomaly.

Herein, we presented multidetector CT and MRI findings of a gastric duplication cyst communicating with an accessory pancreatic lobe via an aberrant duct in a 29-year-old woman with recurrent acute pancreatitis and also reviewed other similar cases reported in the literature.

CASE PRESENTATION

A 29-year-old woman with a 6 year history of recurrent acute pancreatitis was admitted to our hospital due to severe epigastric pain radiating to the back, nausea and vomiting. Physical examination revealed mild epigastric tenderness. On blood tests, serum amylase level (451 IU/L, normal<100) and lipase level (736 IU/L, normal<60) were increased. Serum alanine aminotransferase (ALT) and aspartate aminotransferase (AST) levels were normal. On ultrasonograpy (US) the size and echogenity of the pancreatic parenchyma were normal. There were multiple stones in the gallbladder. No dilatation of the common bile or intrahepatic ducts was detected. US revealed a 4.7x5.3 cm cystic lesion adjacent to the gastric antrum. The cyst had a hyperechoic

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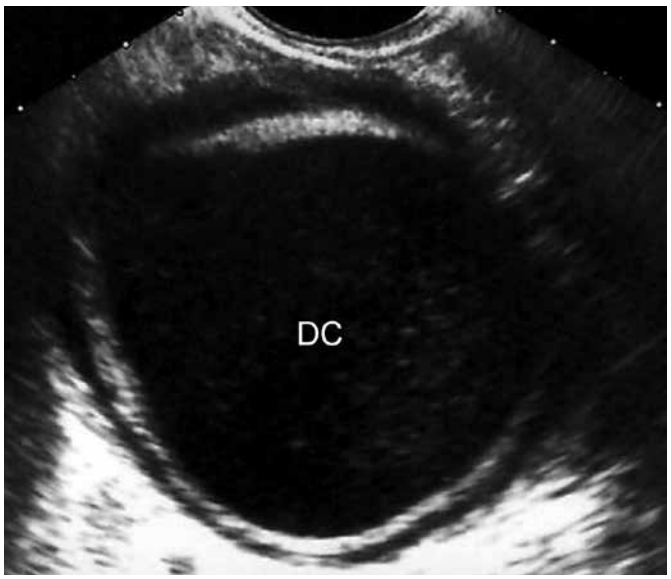

Figure 1. Endoscopic ultrasound image shows a cystic lesion with a hyperechoic inner layer representing the submucosa and a hypoechoic outer layer representing the smooth muscle, suggestive of a duplication cyst (DC).

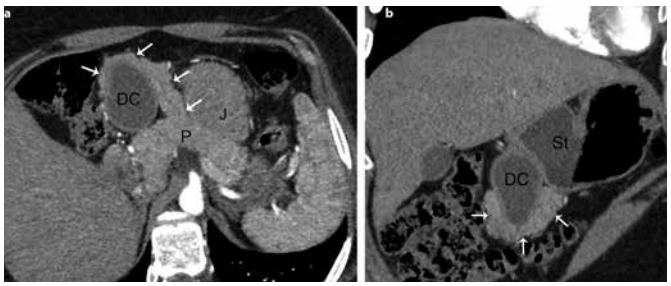

Figure 2. a, b. Oblique axial **(a)** and coronal multiplanar reformatting **(b)** multidetector CT images show a long segment of accessory pancreatic lobe (arrows), arising from the pancreatic (P) neck, projecting anteriorly and attaching to the gastric duplication cyst (DC) adjacent to the gastric antrum (J: jejenum, St: stomach).

inner layer representing the submucosa and a hypoechoic outer layer representing the smooth muscle, suggestive of a duplication cyst (Figure 1). 64-slice multidetector CT (Aquilion; Toshiba Medical Systems; Tokyo, Japan) revealed an unusually long segment of accessory pancreatic lobe, arising from the pancreatic neck, projecting anteriorly and attaching to the gastric duplication cyst (Figure 2). MRI (3T; Magnetom Verio, Siemens, Erlangen, Germany) confirmed the diagnosis of a gastric duplication cyst and an associated accessory pancreatic lobe (Figure 3). But, magnetic resonance cholangiopancreaticography (MRCP) failed to demonstrate the aberrant pancreatic duct and cyst communication (Figure 4). Endoscopic US guided aspiration of the cyst fluid revealed a high amylase ($4.3x10^5$ IU/L) and high carcinoembryogenic antigen (CEA) level (611.58 ng/ml, normal<5 ng/ml), but no malignant cells were seen. At laparotomy, the gastric duplication cyst and the accessory pancreatic lobe were resected. The surgical specimen revealed a 5 cm round shaped duplication cyst, which was attached to a 2.5 cm width and 12 cm long accessory pancreatic lobe with the aberrant ductal system (Figure 5). The histopathologic examination

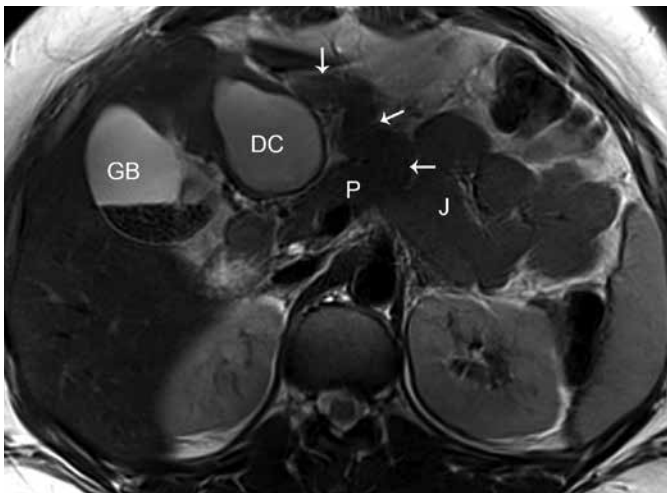

Figure 3. Axial T2-weighted MR image shows an accessory pancreatic lobe (arrows) which is isointense with pancreatic tissue (P), arising from the pancreatic neck, projecting anteriorly and attaching to the gastric duplication cyst (DC). Note mulltiple tiny bile stones in the gallbladder (GB) (J: jejenum).

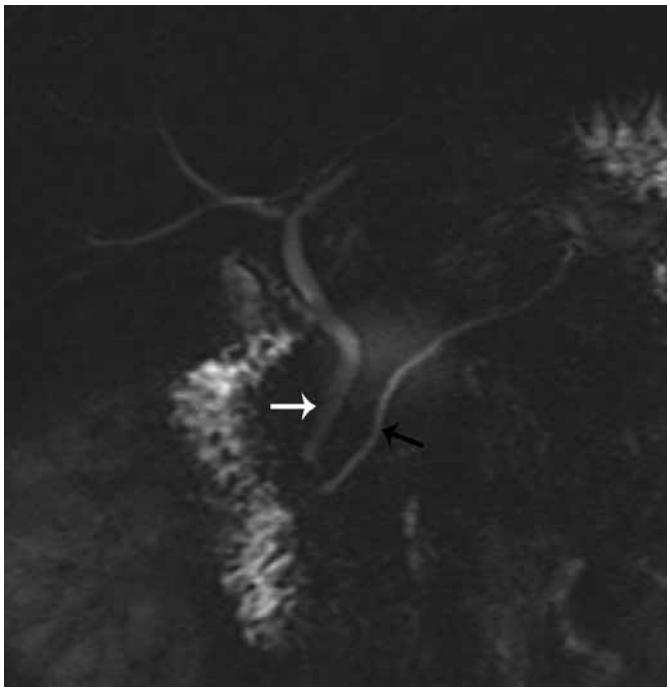

Figure 4. MRCP image shows biliary tree and main pancreatic duct (black arrow) (white arrow: common bile duct). The aberrant duct of the accessory pancreatic lobe is not seen.

confirmed a gastric duplication cyst with well formed layers of gastric type epithelium communicating with an aberrant pancreatic duct. No comminucation between the stomach and the cyst was present. There were ectopic islands of pancreatic tissue in the wall of the cyst. No malignancy was detected in the duplication cyst. The patient made an uneventful recovery and remains well 6 months after surgery.

DISCUSSION

Duplications of the gastrointestinal tract are uncommon congenital anomalies and most commonly occur the ileum and

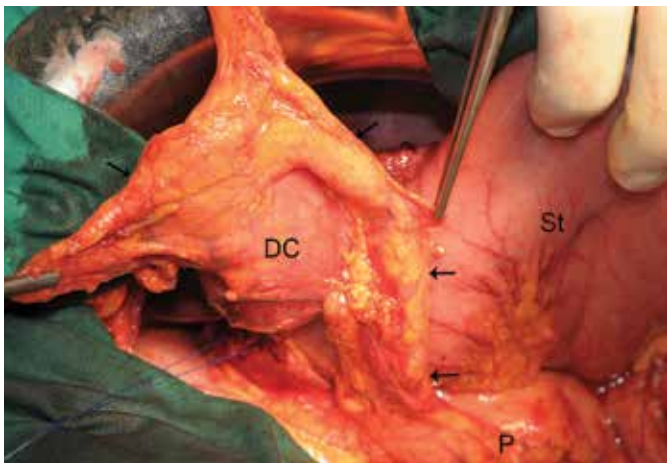

Figure 5. Intraoperative photograph shows an aberrant pancreatic lob (arrows) attaching to the gastric duplication cyst (DC) (P: pancreas, St: stomach).

ileocecum. Gastric duplications are the least common (3.8%) type of intestinal duplications (18). Approximately 50% of the patients with gastric duplication cysts are associated with other congenital anomalies, such as oesophageal and duodenal diverticula, duplication cysts elswhere in the digestive tract, annular and ectopic pancreas, and spinal anomalies (18). However, a gastric duplication cyst associated with accessory pancreatic lobe is extremely rare. Our review of the literature revealed only 18 cases that have been reported to date (1-17), since Bradbeer described the first case in 1959 (1). 14 cases (78%) were female and four cases (22%) were male, and the mean age was 20 years, ranging from 1 months to 53 years. In eight patients, this anomaly was diagnosed at less than 10 years of age (2,5,7,10,11,14,15). Anatomically, gastric duplication cysts were contiguous with the antrum wall of the stomach in most of the reported cases (3,4,6-8,10-13,17). They can communicate with the gastric lumen or not.

No more information is known about the embryogenesis of gastrointestinal duplications; further, in such cases associated with pancreatic anomaly, the mechanism is thought to be more complicated. Abnormal foregut development is believed to be responsible for this congenital anomaly. McLetchie et al. (19) suggested a neuroenteric hypothesis, in which an embryonic entoectodermal adhesion gives rise to a neuroenteric band, which may not elongate as quickly as its surrounding structures, thereby causing traction diverticula leading to gut cyst formation. This hypothesis may fit this anomaly since traction on a pancreatic duct by a neuroenteric band is known to produce both gastric and pancreatic abnormalities.

Abdominal pain, nausea and vomiting have been reported as symptoms of gastric duplication cysts (1,2,4). Rarely, gastrointestinal bleeding, peritonitis, malignancy, or even acute abdomen may be seen (3,10,20). Also, pyloric stenosis due to compression of a huge gastric duplication cyst have been reported by Hishiki et al. (15). In patients with gastric duplication cysts communicating with aberrant pancreatic duct, abdominal pain is often related to acute pancreatitis. Recurrent acute pancreatitis caused by this anomaly have been reported in 12 cases (66%) (4-9,11-14,16,17). Oeda et al. (17) reported that there were not significant differences between the patients with pancreatitis and without pancreatitis in terms of either sex or age. The underlying cause of recurrent acute pancreatitis was hypothesized to be obstruction of the pancreatic duct by viscous mucus secretions, ulcer bleeding or biliary sludge (3,4). In our case, despite the existence of cholelithiasis, it was thought that acute pancreatitis was caused by gastric duplication cysts with aberrant pancreatic duct rather than gallstones due to non-increased liver enzyme levels during acute pancreatitis attack and the absence of dilatation in the bile ducts.

Anatomic variations of the pancreas usually imply variations of duct drainage, fusion anomalies (pancreas divisum), pancreatic agenesis or hypoplasia, annular pancreas, ectopic pancreas, and duplication anomalies. The accessory pancreatic lobe is an extremely rare congenital anomaly of the pancreas (6,10-13,16). This anomaly is defined as an accessory lobe of pancreatic tissue originating from the main pancreatic gland and it contained an aberrant duct. The aberrant duct of the accessory pancreatic lobe communicates with the main pancreatic duct and gastric duplication cyst. The accessory pancreatic lobe may be short or long, with a wide or narrow connection to the main portion of the pancreas. In our patient, a long segment (12 cm) of accessory pancreatic lobe arose from the pancreatic neck and projected anteriorly and attached to the gastric duplication cyst.

With improving imaging tools, gastrointestinal duplication cysts may be recognized more accurately. US may be used as an initial study to distinguish a duplication cyst from a pseudocyst. The former is characterized by its 'gut signature' which refers to the hyperechoic inner layer representing the submucosa surrounded by a hypoechoic outer layer representing the smooth muscle. Peristalsis of the cyst wall has also beeen noted on ultrasound and is strongly suggestive of a duplication cyst (13). CT and MRG may help identify the exact size and location of the duplication cyst and clearly reveals the associated accessory pancreatic lobe. MRCP and ERCP are useful to identify the ductal anatomy and demonstrate any communication between the duplication cyst and the main pancreatic duct. Although ERCP is considered the gold standart for evaluating pancreaticobiliary anomalies, the procedure is invasive and not always easy to perform in patients. In our case, a definitive diagnosis of gastric duplication cysts with accessory pancreatic lobe had been made preoperatively by using a combination of non-invasive imaging modalities consisting of US, CT and MRI.

Treatments of gastric duplication cysts communicating with accessory pancreatic lobe include surgery and endoscopic drainage. Since cases with malign transformation have been reported, surgical resection may be the better option (20). In most of

the cases reported to date, surgical resection was preferred. The optimal surgical treatment is probably a local excision of the gastric duplication cyst with a margin of normal stomach followed by primary closure, and ligation of the accessory pancreatic lobe with transection at its origin (13). In our study we preferred surgical resection due to high level of CEA (611.58 ng/mL, normal<5 ng/mL) in the cyst fluid. But no malignancy was detected in the cyst wall on histopathologic examination.

In conclusion; in patients with recurrent acute pancreatitis, imaging studies should be performed to detect possible complications and to exclude anatomic variations that are surgically correctable. A gastric duplication cyst communicating with an accessory pancreatic lobe via a aberrrant duct is an unusual cause of recurrent pancreatitis, but an early and accurate diagnosis of this anomaly is important because suitable surgical treatment may lead to a satisfactory outcome. A combination of US, multidetector CT and MRI findings enables a definitive diagnosis of this anomaly non-invasively.

Conflict of Interest: No conflict of interest was declared by the authors.

Case Report

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