Recurrent Acute Pancreatitis Caused by a Gastric Duplication Cyst Communicating with an Aberrant Pancreatic Duct

Satoshi Oeda¹, Taiga Otsuka¹, Takumi Akiyama¹, Keisuke Ario¹, Masanori Masuda², Shohei Taguchi³, Takeshi Shono³ and Seiji Kawazoe¹

Abstract

A 38-year-old woman was hospitalized in August 2007. This visit was her fifth episode of acute pancreatitis. Computed tomography revealed a cystic structure located near the antrum. Communication between this structure and the pancreatic duct was revealed by endoscopic retrograde cholangiopancreatography. Ultrasonography revealed that the cyst wall had a layered structure. Thus, we regarded it as a gastric duplication cyst. We thought that the gastric duplication cyst communicating with an aberrant pancreatic duct was responsible for the recurrent acute pancreatitis. In August 2008, a cyst gastrostomy was performed between the gastric duplication cyst and the stomach. No recurrence of acute pancreatitis has since occurred.

Key words: aberrant pancreatic duct, acute pancreatitis, congenital anomaly, cyst gastrostomy, gastric duplication cyst

(Inter Med 49: 1371-1375, 2010)
(DOI: 10.2169/internalmedicine.49.3392)

Introduction

Duplication of parts of the digestive system is a rare congenital abnormality (1). Gastric duplication cysts account for 3.8-4% of all duplications of the gastrointestinal tract (2-4). A gastric duplication cyst communicating with an aberrant pancreatic duct is rarer. Fifteen cases (5-18) have been reported, and several cases with recurrent acute pancreatitis have also been reported (Table 1). It has been suggested that the underlying cause involves obstruction of the pancreatic duct by viscous mucus secretions, ulcer bleeding or biliary sludge. Here, we report that cyst gastrostomy was effective in a case with recurrent acute pancreatitis caused by a gastric duplication communicating with an aberrant pancreatic duct.

Case Report

A 38-year-old woman was hospitalized in August 2007 to evaluate upper abdominal pain. She had previously been diagnosed at another hospital with idiopathic pancreatitis at the age of 22 years old (16 years previously) and she had previously been admitted four times. Although the latest admission was 4 years before the current admission, she had intermittently suffered from mild abdominal pain since then. On admission in August 2007, upper abdominal tenderness without peritoneal signs was noted. There were no Cullen or Grey-Turner signs. Blood tests showed an elevated white blood cell count of 18,300/μL, serum amylase concentration of 3,884 IU/L, and lipase concentration of 1,160 IU/L. We diagnosed acute pancreatitis and treated her with fluid replacement, antibiotics and protease inhibitors. She was cured and discharged on day 22 of hospitalization.

We examined the cause of recurrent pancreatitis during hospitalization. Computed tomography revealed pancreatic swelling and a cystic structure near the gallbladder and antrum (Fig. 1A, B). Magnetic resonance cholangiopancreatography revealed that the cystic structure communicated with the pancreatic duct. Endoscopic retrograde cholan-
Table 1. Cases of Gastric Duplication with an Aberrant Pancreatic Duct

<table>
<thead>
<tr>
<th>First author</th>
<th>Year</th>
<th>Age</th>
<th>Sex</th>
<th>Symptoms</th>
<th>Pancreatitis</th>
<th>Location</th>
<th>Operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bradbeer (5)</td>
<td>1959</td>
<td>17</td>
<td>M</td>
<td>Postprandial epigastralgia</td>
<td>+</td>
<td>Juxtapancreas</td>
<td>Resection of duplication</td>
</tr>
<tr>
<td>Case record of Massachusetts General Hospital (6)</td>
<td>1964</td>
<td>5</td>
<td>M</td>
<td>Abdominal pain</td>
<td>+</td>
<td>Intrapancreas</td>
<td>Resection of duplication and accessory pancreatic lobe</td>
</tr>
<tr>
<td>Akers (7)</td>
<td>1972</td>
<td>7</td>
<td>M</td>
<td>Abdominal pain, vomiting, fever</td>
<td>+</td>
<td>Juxtapancreas</td>
<td>Resection of duplication, pancreaticojejunostomy</td>
</tr>
<tr>
<td>Akers (7)</td>
<td>1972</td>
<td>21</td>
<td>F</td>
<td>Abdominal pain</td>
<td>+</td>
<td>Juxtapancreas</td>
<td>Resection of duplication, Roux-Y drainage of pancreas</td>
</tr>
<tr>
<td>Longmire (8)</td>
<td>1973</td>
<td>15</td>
<td>F</td>
<td>Gastrointestinal bleeding, epigastralgia</td>
<td>+</td>
<td>Posterior wall of the pylorus and antrum</td>
<td>Pancreaticoduodenectomy</td>
</tr>
<tr>
<td>Traverso (9)</td>
<td>1975</td>
<td>32</td>
<td>F</td>
<td>Abdominal pain, nausea, vomiting</td>
<td>+</td>
<td>Contiguous to the antrum wall</td>
<td>Distal pancreatectomy, resection of duplication</td>
</tr>
<tr>
<td>Hoffman (10)</td>
<td>1987</td>
<td>18</td>
<td>F</td>
<td>Epigastralgia, nausea, vomiting</td>
<td>+</td>
<td>Contiguous to the antrum wall</td>
<td>Resection of duplication and aberrant pancreatic lobe</td>
</tr>
<tr>
<td>Lavine (11)</td>
<td>1989</td>
<td>6</td>
<td>F</td>
<td>Epigastralgia, vomiting, dehydration, fever</td>
<td>+</td>
<td>Contiguous to the antrum wall</td>
<td>Resection of duplication and aberrant pancreatic lobe</td>
</tr>
<tr>
<td>Bearzi (12)</td>
<td>1990</td>
<td>53</td>
<td>F</td>
<td>Abdominal pain</td>
<td>+</td>
<td>Contiguous to the antrum wall</td>
<td>Distal gastrectomy, Billroth</td>
</tr>
<tr>
<td>Johnstone (13)</td>
<td>1991</td>
<td>41</td>
<td>F</td>
<td>Postprandial epigastralgia, vomiting</td>
<td>-</td>
<td>Juxtapancreas</td>
<td>Cyst gastrostomy</td>
</tr>
<tr>
<td>Moss (14)</td>
<td>1996</td>
<td>9</td>
<td>F</td>
<td>Epigastralgia, hyperamylasemia</td>
<td>-</td>
<td>Contiguous to the antrum wall</td>
<td>Resection of duplication, Roux-Y drainage of pancreas</td>
</tr>
<tr>
<td>Whiddon (15)</td>
<td>1999</td>
<td>24</td>
<td>F</td>
<td>Bouts</td>
<td>+</td>
<td>Contiguous to the duodenum</td>
<td>Resection of duplication and aberrant pancreatic lobe</td>
</tr>
<tr>
<td>Muraoka (16)</td>
<td>2002</td>
<td>46</td>
<td>F</td>
<td>Epigastralgia, nausea, vomiting</td>
<td>-</td>
<td>Contiguous to the stomach</td>
<td>Resection of duplication and aberrant pancreatic lobe</td>
</tr>
<tr>
<td>Gugig (17)</td>
<td>2004</td>
<td>7</td>
<td>F</td>
<td>Abdominal pain</td>
<td>+</td>
<td>Juxtapancreas</td>
<td>Distal pancreatectomy</td>
</tr>
<tr>
<td>Hishiki (18)</td>
<td>2008</td>
<td>1M</td>
<td>M</td>
<td>Hypertrophic pyloric stenosis</td>
<td>-</td>
<td>Anterior wall of the pylorus</td>
<td>Resection of duplication</td>
</tr>
<tr>
<td>Our case</td>
<td>2008</td>
<td>38</td>
<td>F</td>
<td>Epigastralgia</td>
<td>+</td>
<td>Contiguous to the antrum wall</td>
<td>Cyst gastrostomy</td>
</tr>
</tbody>
</table>

Figure 1. Computed tomography on admission. A) Pancreatic swelling and necrosis near the pancreas. B) Cystic structure (arrow) near the gallbladder and antrum.
Endoscopic retrograde cholangiopancreatography revealed that the cystic structure communicated with an aberrant pancreatic duct (arrow).

The wall of the cystic structure had a layered structure, similar to that of the digestive tract.

The gastric duplication cyst (black arrow) communicating with the pancreatic duct (black arrowhead) shared a wall with the stomach (white arrow). The white arrowhead indicates the border between the cyst and stomach.

Cyst gastrostomy was performed. The upper layer is the gastric mucosa and the lower layer is the cystic lesion.

Figure 4. The gastric duplication cyst (black arrow) communicating with the pancreatic duct (black arrowhead) shared a wall with the stomach (white arrow). The white arrowhead indicates the border between the cyst and stomach.

Figure 5. Cyst gastrostomy was performed. The upper layer is the gastric mucosa and the lower layer is the cystic lesion.

giopancreatography confirmed the communication between the cystic structure and the pancreatic duct (Fig. 2). Ultrasoneography revealed that the diameter of the cystic structure was about 40 mm and the wall had a layered structure (Fig. 3). Thus, we regarded this case as a duplication cyst and we believed that the gastric duplication cyst communicating with the aberrant pancreatic duct was responsible for the recurrent acute pancreatitis. As she then experienced a fifth episode of pancreatitis, we considered that surgery would offer a better approach to prevent recurrent acute pancreatitis.

In August 2008, the patient underwent celiotomy. We found a cystic prominence along the greater curvature, and the color of this cystic structure differed from that of the gastric wall. The cystic structure shared a wall with the stomach (Fig. 4). The cystic structure contained a large amount of serous fluid and a very high concentration of amylase ($4.1 \times 10^5$ IU/L). The fluid was transparent and there was no debris or bleeding in the cyst. We injected a contrast agent into the orifice of the duct in the cyst and confirmed that the duct is an aberrant pancreatic duct. There was no communication between stomach and cyst. Thus, we diagnosed this cystic structure as a gastric duplication cyst that communicated with an aberrant pancreatic duct, and considered that this malformation was responsible for the recurrent acute pancreatitis. The pancreatic juice was drained by performing a cyst gastrostomy between the duplication cyst and the stomach (Fig. 5).

Microscopically, the wall consisted of a mucosa, muscularis mucosae, submucosa and muscle layer (Fig. 6A). We could identify the gastric crypt epithelium, with cells similar to the pyloric gland, and intestinal metaplasia in the mucosa (Fig. 6B). There was extensive hyperplasia, congestion and edema in the muscularis mucosae. There were no malignant cells. In the 19 months since the time of the gastrostomy, she has not experienced any episodes of acute pancreatitis.

Discussion

Duplication of the gastrointestinal tract is a rare congenital anomaly. The ileum and ileocecum are the most com-
Gastric duplication cysts, typically along the greater curvature, account for 3.8–4% of all duplications of the alimentary tract (2–4). Moreover, gastric duplication cysts communicating with an aberrant pancreatic duct are extremely rare. Only 15 cases (5–18) have been reported to date, including 12 (80%) females and three (20%) males (Table 1). In seven patients (46.7%), this syndrome was diagnosed at less than 10 years of age due to congenital malformations. On the other hand, several cases in patients older than middle age have been found with differing locations or symptoms. Abdominal pain, nausea and vomiting have been reported as symptoms of this communicating cyst. Hishiki et al reported pyloric stenosis with a huge gastric duplication cyst (18). Recurrent pancreatitis with this disease has been reported in 10 cases (66%). There were no significant differences between the pancreatitis group and non-pancreatitis group in terms of either sex (pancreatitis vs. non-pancreatitis groups: M/F=3/7 vs. 1/4; p=0.6797) or age (18.4±4.7 vs. 19.6±9.9 years old; p=0.950). It was suggested that the underlying cause involves obstruction of the pancreatic duct by viscous mucus secretions, ulcer bleeding or biliary sludge. There are also some reports of an adenocarcinoma arising in the duplication (19, 20).

Although the etiology of gastric duplication cysts remains unknown, several theories have been proposed. The theory of McLetchie et al (21) and Bremer’s theory (22) are the most widely accepted. Bremer’s theory of errors of recanalization is based on the observation that, during the 5th-6th week of life, epithelial proliferation of the gut results in obliteration of the lumen. Secretory vacuoles form coalesce to restore the lumen. Failure of the vacuoles to fuse or coalesce into a single intestine could result in the formation of a duplication cyst. The theory of McLetchie et al is as follows. The primitive gut develops during the 3rd week of life. The notochord and the embryonic endoderm fuse to form the notochordal plates. After the formation of the notochord, the embryonic mesoderm forms a continuous layer separate from the notochord during the 4th week of life. Should anomalies occur in the separation of the notochord from the endoderm, this may result in a neurenteric canal if a hollow connection persists, or a neurenteric band giving rise to traction diverticula of the foregut.

Because the generation of the pancreas is complicated, congenital anomalies in the pancreas are possible. Visceral rotation begins in the 6th week of life. The ventral primordium rolls with the primitive common bile duct and adheres to the dorsal primordium. We believe that the developmental process (i.e., the etiology of gastric duplication cysts and visceral rotation in the pancreas) is responsible for gastric duplication cysts communicating with an aberrant pancreatic duct.

Treatment of gastric duplication cyst involves excision or drainage. In most of the 15 cases reported to date, surgical resection was preferred. Because cases with malignant transformation have been reported (19, 20), surgical resection might be the better option. However, we believe that cyst gastrostomy is a therapeutic option in terms of resolving symptoms. Transmural drainage of duodenal duplications is also well accepted. In our case, the duplication shared a wall with the stomach and the pancreatic duct connection was not obstructive based on the indentation of contrast material. Therefore, cyst gastrostomy was performed after considering the surgical invasiveness.

The present case shows that recurrent pancreatitis might occur in patients who have a gastrointestinal duplication cyst with a pancreatic duct connection. Adequate therapy could prevent future relapse of pancreatitis. Although this syndrome is very rare, we should be aware of this malformation as a cause of recurrent pancreatitis.

References

3. Koltun WA. Gastric duplication cyst. Endoscopic presentation as

© 2010 The Japanese Society of Internal Medicine
http://www.naika.or.jp/imindex.html