A case of proximal jejunal ectopic pancreas causing sporadic vomiting

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SUMMARY: Olguner M, Özdemir T, Ateş O, Akgür FM, Aktuğ T, Özer E. A case of proximal jejunal ectopic pancreas causing sporadic vomiting. Turk J Pediatr 2003; 45: 161-164.

Aberrant rests of pancreatic tissue can be found throughout the gastrointestinal system and are known as pancreatic heterotopia or ectopic pancreas (EP). Authors report a 12-year-old girl with jejunal EP with a long-lasting history of sporadic bilious vomiting. Upper gastrointestinal (GI) study showed delayed passage beyond duodeno-jejunal junction. During laparotomy a 2x2 cm mass was encountered on the mesenteric border of the jejunum, 3 cm distal to the ligament of Treitz. Histopathologic examination revealed pancreatic tissue. The mass was excised and end-to-end anastomosis was performed. Postoperative course of the patient was uneventful and she is doing well after 10 months. Intestinal obstruction due to EP has been reported to occur only if it causes intussusception. Intestinal obstruction without intussusception due to jejunal EP has not been reported. In our case, the EP tissue was located just beneath the mucosa and involved the muscular layer. The foreign body effect of the EP tissue involving the muscular layer may cause dysmotility and/or local spasm, which we think were responsible for the long-lasting sporadic bilious vomiting in our patient.

Key words: ectopic pancreas, pancreatic heterotopia, heterotopic pancreas, intestinal obstruction.

Aberrant rests of pancreatic tissue can be found throughout the gastrointestinal (GI) system and are known as pancreatic heterotopia or ectopic pancreas (EP). Despite many cases having been reported, EP causes diagnostic difficulties for clinicians. We herein report a 12-year-old girl with jejunal EP that caused sporadic vomiting.

Case Report

A 12-year-old girl with a long-lasting history of sporadic bilious vomiting was transferred from another hospital. There she had been treated conservatively with nasogastric decompression and had tolerated oral feedings for about a week but unfortunately her symptoms reappeared. Afterwards she was transferred on her parents' demand. Her weight and height were within appropriate limits for her age when she arrived. Plain abdominal radiograph showed a huge stomach extending to pelvic bones, and no air-fluid levels were detected. Upper GI study showed delayed passage beyond duodeno-jejunal junction (Fig. 1). During laparotomy a 2x2 cm mass was encountered on the mesenteric border of the jejunum, 3 cm distal to the ligament of Treitz. When the jejunal lumen was opened through a longitudinal incision on the antimesenteric side, the mass was seen to be lying just beneath the mucosa but was not obstructing the lumen. Macroscopically misinterpreting the mass as a lymph node and not being able to obtain frozen section examination due to the operation being performed late in the evening, the jejunal incision was closed transversely and incision biopsy was done. Histopathologic examination revealed pancreatic tissue. After the operation nasogastric drainage of the patient did not diminish so she was reoperated. During the second laparotomy the mass and surrounding bowel were inflamed. The jejunal segment with the mass was excised and end-to-end

anastomosis was performed. Histopathologic examination of the mass and adjacent intestine showed pancreatic acini with lobular configuration in the jejunal submucosa, which was invading muscularis propria, and muscle fibers were dispersed within the pancreatic acinar lobules (Figs. 2, 3). Postoperative course of the patient was uneventful and she is doing well after 10 months.

Discussion

Ectopic pancreas is a relatively rare entity,

Fig. 1. Huge distended stomach with the delayed passage in the upper gastrointestinal system series.

Fig. 2. Pancreatic acini with lobular configuration; muscle fibers (arrow) are dispersed within the pancreatic lobules (H&E x100).

Fig. 3. Pancreatic acini are seen adjacent to muscular layer in the jejunal submucosa (H&E x40).

usually of no clinical importance and found incidentally during laparotomy for another condition or during autopsy[1,2]. The most common locations for EP are stomach, duodenum and jejunum[3]. Most patients with EP are symptom-free. If present, symptoms are nonspecific. Most commonly reported nonspecific symptoms are abdominal pain, epigastric discomfort, nausea, vomiting and gastrointestinal bleeding[4,5]. Specific symptoms are usually due to EP tissue itself leading to intussusception and pyloric obstruction[4]. Additionally, pancreatic cyst, inflammation and carcinoma may develop in the EP tissue[4].

The mechanism of the nonspecific symptoms of EP has been attributed to the local effects of the pancreatic tissue. According to this proposal, foreign body effect and secretions of the pancreatic tissue may cause local spasm and chemical irritation in the surrounding tissue[2]. However, Dolan et al.[3] disagree with this mechanism in that they believe the mass of the EP tissue is usually too small to cause foreign body effect and also that it is incapable of producing a sufficient amount of enzymes. They have also questioned why a congenital anomaly would not have caused symptoms from the beginning. Actually, the significance of the nonspecific symptoms in EP has been found to be related to the size and mucosal relation of the EP tissue[6]. The EP greater than 1.5 cm and close to the mucosa is more likely to produce symptoms[6]. Additionally, EP tissues just beneath the mucosa have more tendency to involve the muscular layer[7]. This muscular involvement seems to be related to the significance of the symptoms in EP close to the mucosa. Adult and infant autopsy series have shown that the EP tissue may enlarge with age[2]. It seems highly probable that the EP tissue reaches sufficient size with age to produce symptoms.

Specific symptoms are directly related to the EP tissue itself. Pancreatic cyst, inflammation, and carcinoma may develop in the EP tissue and may be the source of ectopic hormone secretion that may cause hypoglycemia, Zollinger-Ellison syndrome or acromegaly[8-13]. The obstructive symptoms of the EP are mainly related to the localization of the EP tissue. While prepyloric EP may cause pyloric obstruction, EP near the sphincter of Oddi may lead to obstructive jaundice[14-16]. Prepyloric EP may cause pyloric obstruction by occluding the pyloric lumen or by inducing pylorospasm[5,14,16]. Obstructive symptoms may easily occur by these two mechanisms in an anatomically narrow area like the pylorus. However, obstruction resulting from luminal occlusion by the EP tissue seems unlikely in intestines where the lumen is wider and more flexible than the pylorus.

Actually, intestinal obstruction due to EP has been reported to occur only if it causes intussusception[15,17]. Intestinal obstruction without intussusception due to jejunal EP has not been reported. In our case, the EP tissue was located just beneath the mucosa and invaded the muscular layer. The foreign body effect of the EP tissue, disrupting the muscular continuity, may cause dysmotility and/or local spasm, which we think were responsible for the long-lasting sporadic bilious vomiting in our patient.

Ectopic pancreas may cause symptoms with one or more mechanisms, as described above. The radiological features are sometimes characteristic: the lesions appear well defined, with a dome-shaped filling defect with a central umbilication[2,18]. Similar findings can be observed during the endoscopic examination[18]. The best treatment for EP is the complete excision of the tissue with wedge resection[19]. Partial excision or biopsy can cause insistence of the symptoms by the ongoing foreign body effect.

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