Ectopic pancreatic tissue in the wall of the small intestine
Two rare case reports
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Abstract
Rationale: Ectopic pancreas, which is a kind of rare congenital disease, forms during embryonic development. It can occur throughout the whole gastrointestinal tract, but has a low tendency to develop in the wall of the small intestine. It is easy for patients with ectopic pancreases to be misdiagnosed because the symptoms are atypical and can vary.

Patient concerns: In the present study, we reported two rare cases of ectopic pancreatic tissue in the wall of the small intestine, which presented with obvious abdominal pain and distention.

Diagnosis: The laboratory tests and computed tomography (CT) scans didn’t reveal any evidence of ectopic pancreas.

Interventions: The two patients received small intestine masses resection and intestinal anastomosis.

Outcomes: During surgery, an intestinal mass with a diameter of 4.0 cm was found in the first patient. An intestinal mass with a diameter of 0.8 cm, jejunum perforation, and diffuse peritonitis were found in the second patient. Histological analyses of the dissected intestinal masses confirmed them as ectopic pancreatic tissue. Interestingly, for the second patient, the intestinal perforation and diffuse peritonitis were not induced by the ectopic pancreas, but by a jujube pit that was found in the perforated site of the intestine.

Lessons: Our study demonstrated that an ectopic pancreas should be considered in cases of atypical abdominal symptoms with intestinal masses.

Abbreviations: CT = computed tomography, WBC = white blood cell.

Keywords: ectopic pancreas, perforation, small intestine

1. Introduction
Ectopic pancreas, which is a kind of congenital malformation of the pancreas, is very rare in clinic. It lacks vascular or anatomical communication with normal pancreatic tissue. Theoretically, it can locate in many organs, but is found mainly in the upper gastrointestinal tract.[1] Ectopic pancreatic tissue always remains asymptomatic throughout the patients’ life, and it is often detected incidentally by histological examination during surgery.[2] Depending on its location, symptoms can originate from the tumor mass effect, such as perforation-related peritonitis, gastrointestinal bleeding, abdominal pain, and distention.[2,3] In addition, adenocarcinoma of ectopic pancreas occurs sometimes, and this can also seriously endanger the patients’ life.[4] As a result, early diagnosis and proper treatment of ectopic pancreas are very important in clinic.

In the present study, we reported 2 ectopic pancreas cases that were diagnosed incidentally. Furthermore, the clinical types, examination, and treatment methods for each ectopic pancreas case were also discussed.

2. Case report
This study was approved by the Ethics Committee and institutional Review Board of the second hospital of Jilin University, Changchun, China.

2.1. Case 1
A 53-year-old man attended our hospital with intermittent abdominal pain and distention. The patient denied any relevant medical history. Physical examination only revealed epigastric tenderness. Laboratory tests indicated a white blood cell (WBC) count of 1.5 × 10^9/L, a neutrophil percentage of 90.0%, amylase at 256.0 U/L, and lipase at 373.0 U/L. Computed tomography (CT) scan showed a mixed density mass in the left abdomen. The mass was 64 mm × 39 mm in size, and was associated closely with the intestines (Fig. 1).

Under careful consideration, the patient received a surgery via exploratory laparoscopy. A 4.0 × 3.0 × 3.0 cm mass in the
proximal jejunum, which was 20 cm from the ligament of Treitz, was found (Fig. 2A). Small intestine mass resection, partial intestinal resection, and small intestine anastomosis were then performed. Post-surgical pathological examination demonstrated a heterotopic pancreas, with patchy necrosis and acute inflammatory exudation within the pancreatic tissue (Fig. 2B).

2.2. Case 2

A 77-year-old woman was admitted to our hospital due to acute abdominal pain, distention, nausea, and vomiting. As in case 1, no relevant medical history was presented. Physical examination demonstrated severe tenderness and muscle tension in the lower left abdomen. Laboratory tests revealed a WBC count of $1.6 \times 10^9/L$. CT examination showed intestinal perforation and acute peritonitis. During the exploratory laparoscopy, a 0.8 cm protruding intestinal mass, which was 20 cm from the ligament of Treitz, was identified. Perforations of the intestine, at 70 cm and 130 cm from the ligament of Treitz, were also found. In the second perforation site, which was 130 cm from the ligament of Treitz, a sharp object blocked the damaged intestinal canal. Partial small intestine adjacent to the second perforated site was resected (Fig. 3A), the first perforated site was repaired, and the intestinal mass was removed (Fig. 3B). Subsequent pathology indicated an ectopic pancreas, with pancreatic duct dilation and ductal epithelial hyperplasia (Fig. 3C). In addition, as confirmed with the patient, the sharp object that induced the intestinal perforation was a jujube pit.

### 3. Discussion

An ectopic pancreas, representing isolated pancreatic tissue from the normal body of the pancreas, is more common in 30- to 50-year-old men. Its incidence ranges from 0.5 to 13.7%. Most of the ectopic pancreases locate in the upper esophagus, duodenum, and colon. It is rare to find an ectopic pancreas in the small intestine.

An ectopic pancreas is usually small and always remains asymptomatic. Only a few patients with ectopic pancreas show clinical symptoms, and these symptoms are often nonspecific. When symptoms are present, pain is the most common, with other symptoms, such as intestinal obstruction and perforation, peritonitis, and gastrointestinal bleeding, also occurring occasionally. In addition, malignant transformation was also reported in patients with ectopic pancreas. Ectopic pancreases can be divided into several types, such as bleeding type, inflammation type, ulcer type, obstruction type, tumor type, and occult type. The 2 patients in our report presented with obvious abdominal pain. Although intestinal perforation and peritonitis were found in the second case, they were induced by a sharp jujube pit, not the ectopic pancreas. As a result, both the ectopic pancreases in these 2 cases were identified incidentally. In addition, they belonged to the tumor type, because only protruding ectopic pancreases and no other associated symptom were found.

The diagnosis of heterotopic pancreatic tissue is very difficult, for the following reasons:

1. As mentioned above, nonspecific symptoms are one of the most important reasons. Furthermore, the symptoms of an ectopic pancreas are similar to those of, for example, gastrointestinal stromal tumors, carcinoid, or gastrointestinal carcinoma.

2. An ectopic pancreas is usually small and may be missed by imaging.

3. An ectopic pancreas always locates in the deep submucosal layer of the intestine, which causes difficulties for pathological biopsy. As a result, the pathological biopsy usually captures the intestinal mucosa, not the ectopic pancreas.

For the 2 patients in our report, the symptoms were nonspecific. Imaging revealed no valuable result in Case 2, whose ectopic pancreas was small. For Case 1, the CT scan revealed a mixed

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**Figure 1.** CT findings of the abdominal mass in Case 1. The red arrow indicates the mixed density mass in the left abdomen. CT = computed tomography.

**Figure 2.** Macroscopic findings (A) and pathological examination (hematoxylin and eosin staining, × 20) (B) of the ectopic pancreas in Case 1. The red arrow in A indicates the ectopic pancreas.
density mass. However, because of the non-specific symptoms of this patient, our clinical team did not consider an ectopic pancreas initially. Therefore, exploratory laparoscopies were performed, which revealed the ectopic pancreases.

With the development of modern medical technologies, the followings should be noted during the diagnosis of an ectopic pancreas:

1. Digestive endoscopy always reveals a hemispherical bulge in the submucosal layer of the intestine, usually with a clear boundary and normal surface mucosa. Pathological biopsy is needed to confirm the ectopic pancreatic tissues.

2. Ultrasound endoscopy always demonstrates an uneven echo, with a low echo being more common.

3. Multislice spiral CT can reveal the lesion with similar enhancement to a normal pancreas.

4. As in the present study, laboratory tests can reveal the increased levels of amylase and lipase, which, combined with the imaging findings, point to a possible ectopic pancreas.

5. Pathology is always the gold standard to confirm the diagnosis of an ectopic pancreas.

In the reported cases, the 2 patients all presented with untypical symptoms. Laboratory tests and CT examinations did not reveal any evidence of ectopic pancreas. Local resections were prompt in the 2 cases, and postsurgical histopathologies confirmed the ectopic pancreases. As a result, for cases presenting with unexplained abdominal pain and distention, intestinal obstruction and bleeding, gastrointestinal ulcers, and increased levels of amylase and lipase, an ectopic pancreas should be considered. Local resection of the ectopic pancreas is still the most appropriate therapy to avoid any secondary changes.

References

[1] Straatman J, Meester RJ, van Grieken NCT, et al. Clinical picture: multiple sites of ectopic pancreatic tissue. Springerplus 2015;4:293.

[2] Liu C-Y, Chang W-H, Lin S-C, et al. Analysis of clinical manifestations of symptomatic acquired jejunoileal diverticular disease. World J Gastroenterol 2005;11:5557–60.

[3] Shiratori H, Nishikawa T, Shintani Y, et al. Perforation of jejunal diverticulum with ectopic pancreas. Clin J Gastroenterol 2017;10:137–41.

[4] Mulholland KC, Wallace WD, Epanomeritakis E, et al. Pseudocyst formation in gastric ectopic pancreas. JOP 2004;5:498–501.

[5] Christodoulides G, Zacharoulis D, Barbanis S, et al. Heterotopic pancreas in the stomach: a case report and literature review. World J Gastroenterol 2007;13:6098–100.

[6] 2017;Beralier A, Mees ST, Pump J, et al. Clinical impact of duodenal pancreatic heterotopias—is there a need for surgical treatment? 17:53.

Figure 3. Macroscopic findings and pathological examination (hematoxylin and eosin staining, × 20) of the ectopic pancreas in Case 2. (A) Resected part of the intestines with perforations (red arrows). (B) The removed ectopic pancreas. (C) Hematoxylin and eosin staining of the ectopic pancreas.