

Duodenal Mass from Heterotopic Pancreas: A Unique Cause of Septic Shock

Dear Editor,

Heterotopic pancreas is defined as pancreatic tissue found outside the pancreas without any anatomical or vascular connection with the main pancreas.¹ Heterotopic pancreas is also known as the ectopic pancreas, aberrant pancreas and accessory pancreas.² It can occur at any part of the gastrointestinal tract with 90% encountered in the stomach, duodenum and the jejunum.³⁻⁵ The entity is usually found as incidental finding.^{6,7}

Case Report

A 62-year-old Siamese lady presented with abdominal pain associated with early satiety, loss of appetite, loss of weight and repeated vomiting after a meal for 1 week. She was lethargic, afebrile but with no signs of peritonism. Full blood count, liver and renal functions were within normal range. There was a slight raise in the serum amylase level. Serum carcinoembryonic antigen (3.8 U/mL), alpha-fetoprotein (4.4 ng/mL) and cancer antigen-19-9 (38 U/mL) were all within normal ranges. Ultrasound showed the presence of fluid collection in the abdomen. Computed tomography (CT) scan of the abdomen revealed thickening at the first part of the duodenal wall (D1) and normal pancreas (Fig. 1). Subsequent oesophagogastroduodenoscopy (OGDS) revealed circumferential mass at D1 with few nodules and ulcer that caused narrowing of the lumen (Fig. 2). She showed some improvement and was discharged home.

However, she presented again several days later due to fatigue and poor oral intake. Physical examination revealed



Fig. 1. Radiological finding: Contrast-enhanced CT scan at the level of pancreas shows normal homogenous pancreas enhancement (+) with adjacent thickened duodenal wall (*).

that she was lethargic with impaired conscious level, having a blood pressure of 90/70 mmHg and pulse rate of 120/min. A biopsy that was taken from previous OGDS site showed an inflamed heterotopic pancreas (Fig. 3). It is composed of lobules of acinar cells, pancreatic ducts and a few small foci of the islet of Langerhans cells that could be highlighted by neuroendocrine markers (chromogranin and synaptophysin). In areas, there was marked neutrophilic activity noted for attacking the glands. Her condition then deteriorated and showed clinical evidence of disseminated intravascular coagulation (high level of D-dimer and deranged clotting profiles). She was admitted to intensive care unit, intubated and developed hospital-acquired pneumonia with type I respiratory failure. Unfortunately, she died after several days in intensive care unit due to multi-organ failure. An autopsy was not performed.

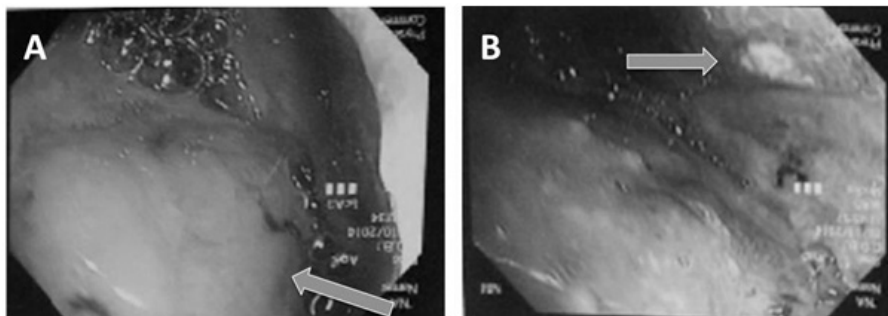


Fig. 2. Oesophagogastroduodenoscopy (OGDS) findings: In A) a circumferential pushing mass at D1 (arrow) is seen, and in B), an ulcer (arrow) is seen.

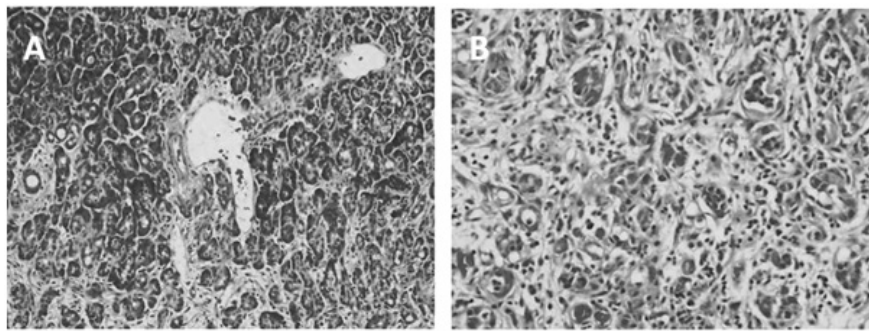


Fig. 3. Histological finding: A) shows lobules of acinar cells with occasional pancreatic duct typical of a pancreas, haematoxylin & eosin x 40, and in B), marked neutrophilic infiltration is also noted, haematoxylin & eosin x 200.

Discussion

In the literature, symptoms related to heterotopic pancreas are usually related to complications including intestinal or common bile duct obstruction, mucosal ulcer with haemorrhage, intussusception, gastrointestinal bleeding and abdominal mass. Rarely, the symptoms related to pathological conditions of the ectopic pancreas itself, such as acute or chronic pancreatitis,⁸⁻¹⁰ pancreatic cyst⁴ or neoplasm.¹¹ Non-specific gastrointestinal symptoms mainly occur with heterotopic pancreas in the upper gastrointestinal tract such as abdominal pain, nausea, vomiting, regurgitation, and anorexia or weight loss, etc.⁷ The current patient is the first case reported of an inflamed heterotopic pancreas complicated with shock and multiple organ failures.

Diagnosis of heterotopic pancreas is difficult even with the advancement in imaging, endoscopy and routine biopsy procedure. It is prone to misdiagnosis and missing diagnosis due to its scarceness, variations of anatomical location, small size and non-specific clinical symptoms.⁷ All these contributed to the rarity of preoperative or clinical diagnosis of heterotopic pancreas.^{3,7}

On OGDS, ectopic pancreas could be hinted by the presence of a firm, round, sub-epithelial mass having a central depression that represents the opening of the pancreatic duct. In the present case, the central depression was not observed, but an ulcer was noted.¹⁰

The radiological findings were also not much help due to several limitations. On CT scan /MRI (magnetic resonance imaging), small ectopic pancreas could be differentiated from other gastrointestinal submucosal tumour by their location, prominent enhancement, ill-defined border, endoluminal growth pattern and the longest/shortest diameter ratio (LD/SD).² However, these characteristic features may not be present. Specifically, in this case, the contrast CT scan showed only focal thickening of duodenal

mucosa with the adjacent normal pancreas. In contrast, the presence of multiple nodules detected on endoscopic examination as well as the clinical presentation gave the false impression of malignancy.

In recent years, endoscopic ultrasound (EUS), a combination of endoscopy and ultrasonographic images of high resolution together with fine needle aspiration biopsy (FNAB) are of increasing use for detecting gastrointestinal and peri-gastrointestinal diseases. Ectopic pancreas in stomach has been reported to be diagnosed by EUS-FNAB.^{12,13}

Management of heterotopic pancreas remains controversial. Some recommended no further investigation and management; others recommended local resection to avoid any future complication. Surgical resection is the best choice when dealing with symptomatic patients.^{6,11} Benign asymptomatic lesions generally do not require surgical intervention.⁶ To improve diagnosis rate and to avoid misdiagnosis or unnecessary extensive operations, endoscopic resection with intraoperative frozen section could be considered.⁵

Overall, the rarity of the presentation of heterotopic pancreas makes the diagnosis a great challenge. Though the majority of the cases are asymptomatic, there is potential for lethal complications when acute inflammation is noted in the heterotopic pancreas.

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