INTRODUCTION

Heterotopic pancreas (HP) is typically an asymptomatic malformation that can present anywhere along the gastrointestinal tract. It is often found incidentally as a single focal mass in patients with abdominal complaints undergoing routine diagnostic testing. HP occasionally presents symptomatically, and the manifestations vary depending on the location of the lesion. We describe the first reported case of symptomatic jejunal HP which presents as a circumferential infiltrative lesion with multiple foci rather than a single focal mass. This is accompanied by a review of the current literature regarding the pathogenesis, clinical manifestations, and treatment of HP.

CASE REPORT

A 37-year-old woman with a history of systemic lupus erythematosus and cadaveric kidney transplant in 1997 was referred to our institution for recurrent nausea, vomiting, diarrhea, and abdominal pain of two years duration. Her symptoms occurred in discrete episodes requiring repeated hospitalizations for dehydration and abdominal pain. On physical exam, her abdomen was tender to palpation in the left upper quadrant and epigastrium, with voluntary guarding but no rebound tenderness.

Extensive evaluation including laboratory studies, abdominal ultrasound, upper endoscopy, and colonoscopy were unrevealing. An abdominal computed tomographic (CT) scan with oral and intravenous contrast demonstrated an area of abnormal small bowel localized to the proximal jejunum (Figure 1). Capsule endoscopy was subsequently performed to evaluate the small bowel and was unremarkable. Due to persistent symptoms, the patient underwent single bowel enteroscopy (SBE) to visualize directly and to biopsy the abnormal jejunal segment seen on CT.

Examination by SBE revealed a 6cm grossly edematous and circumferentially ulcerated segment of mucosa located approximately 100cm distal to the Ligament of Treitz (Figure 2). The lesion was biopsied; however, pathological examination only revealed mild vascular congestion. The patient was eventually taken for surgical resection. Intraoperatively, the lesion was visualized. In addition, a separate ulcerated lesion was seen in the small bowel 10cm proximal to the original lesion. Both lesions were resected, and histological examination revealed heterotopic pancreatic tissue involving the adventitia, muscularis propria, and submucosa extending to the muscularis mucosa (Figure 3). There was no evidence of vasculitis. The patient experienced symptoms of abdominal pain and nausea postoperatively but these symptoms resolved completely within one month.
DISCUSSION

Heterotopic pancreas (HP) is defined as pancreatic tissue that lacks an anatomic or vascular communication with the normal body of the pancreas. Although HP can occur throughout the entire gastrointestinal tract, it is most commonly found in the stomach (25%-38%), duodenum (17%-36%), and jejunum (15%-21%). Rare cases have described HP in the esophagus, biliary tract, gallbladder, spleen, and mesentery. HP can be pathologically subdivided into four subtypes. Type 1 heterotopia consists of typical pancreatic tissue. The remaining subtypes consist of specific components of normal pancreatic tissue such as pancreatic ducts only (type II), acinar tissue only (type III), or islet cells only (type IV). Heterotopic pancreatic tissue can be found in patients of any age and is slightly more common in men.

The development of heterotopic pancreatic tissue is unclear. The two predominant theories of origin of HP involve misplacement of pancreatic tissue during development versus tissue metaplasia. Embryologically, the pancreas develops from invaginations of endodermal tissue in the primitive duodenum. The ventral aspect forms the head of the pancreas and the dorsal aspect forms the body and tail. The misplacement theory proposes that, during rotation of the foregut, several elements of the primitive pancreas become separated and eventually form mature pancreatic tissue along the length of the gastrointestinal tract. The metaplasia theory states that pancreatic heterotopia arises from areas of pancreatic metaplasia of the endoderm which migrate to the submucosa during embryogenesis.

Heterotopic pancreatic tissue is a common incidental finding in patients undergoing laparotomy for other indications or at autopsy. The reported frequency of this finding during laparotomy is 0.5% and at autopsy is 1.7%. Despite the relatively frequent occurrence of HP, the vast majority of these cases are asymptomatic. When present, symptoms vary depending on the anatomical location and size of the lesion. Abdominal pain, nausea, vomiting, and gastrointestinal bleeding are the most commonly reported symptoms and are most likely to be seen with lesions greater than 1.5cm in diameter. Pain associated with HP may be related to the local secretion of hormones and enzymes resulting in tissue inflammation or chemical irritation. Pain may also be related to mechanical obstruction of the intestinal lumen, especially when associated with nausea or vomiting. Gastric lesions are the most likely to be symptomatic, presenting with either epigastric pain or symptoms of gastric outlet obstruction due to a pre-pyloric mass. Rarely, jejunal lesions may result in intestinal obstruction or intussusception. Even after finding an HP lesion in a patient with the above mentioned symptoms, other causes must be ruled out before the symptoms can be attributed to the HP. HP may also present with symptoms related to complications similar to those normally associated with the pancreas. Cases of pancreatitis due to gastric lesions have been reported. Pseudocyst formation complicating duodenal lesions is another known complication. Malignant transformation of heterotopic pancreas can rarely occur with up to 15 cases reported. In order to be identified as malignancy derived from HP, the malignant...
tissue must be within or in close proximity to the heterotopic focus. Additionally, a transition between the carcinoma and normal pancreatic structures must exist. Fully developed ducts and acini in the non-neoplastic heterotopic pancreatic tissue must also be seen. Overall, adenocarcinoma arising from HP has a better prognosis than malignancy which arises from the normally located pancreas.8

HP has several characteristic radiographic and endoscopic features that aid in its identification. Gastric HP classically presents as a rounded filling defect with a central indentation on barium studies or as a broad based, umbilicated, submucosal lesion on endoscopy.6 This central umbilication results from a subjacent pancreatic duct.21 However, these features are not always present, leading to a difficulty in diagnosis. In 1999, Hsia et al reported that only 3 of 17 cases of HP presented preoperatively with the endoscopic finding of a submucosal lesion with a central indentation.22 Various imaging tests can be used to identify heterotopic pancreas in the gastrointestinal tract. It is important to stress that these tests are almost always performed to evaluate more common causes of abdominal symptoms, and the finding of HP is usually incidental. Computed tomographic findings are usually nonspecific although heterotopic pancreatic tissue can enhance to the same degree as normal pancreas with intravenous contrast.6

Endoscopic ultrasound (EUS) is the standard test to evaluate submucosal lesions of the gastrointestinal tract. In the gastric antrum, EUS can diagnose submucosal heterotopic pancreas (pancreatic rests) ranging from 0.5-2cm in diameter, and when combined with fine needle aspiration the cytological evaluation has a sensitivity of 80%-100%.24

The management of HP is a controversial topic. Surgical resection of HP should be performed in symptomatic patients after more common causes of abdominal complaints such as peptic ulcer disease, gastro-esophageal reflux disease, and biliary disease have been ruled out. Benign asymptomatic lesions generally do not require surgical intervention. Ormarsson et al followed 32 patients with HP of the stomach or small bowel for 13 years and found that there was no malignant transformation in any of the patients over this time. The literature is divided on the management of asymptomatic and incidental lesions. Some state that management should include frequent endoscopy, while others suggest that there is no need for increased surveillance. In lesions with malignant changes or in which malignancy is uncertain, surgical resection is required.4,17,25

Although heterotopic pancreas is not uncommon, this is the first reported case of jejunal HP that presents as a circumferential mural infiltration with multiple foci rather than a single focal mass. This patient had two separate jejunal lesions which involved the adventitia, muscularis propria, and submucosa resulting in symptoms of recurrent intestinal obstruction. Additionally, this case highlights the difficulty in making a diagnosis of HP even with the aid of newer techniques such as capsule endoscopy and SBE. Although an area of gross inflammation was located endoscopically in the jejunum, a diagnosis of HP could not be made until the specimen was surgically excised.

REFERENCES