Title

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CLINICAL VIGNETTE

A Case of Heterotopic Pancreatic Tissue at the Gastroesophageal Junction

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Case Presentation

A 45-year-old male presented for a gastroenterology consultation for heartburn and irregular bowel habits. The patient reported heartburn around two days per week especially after spicy foods. His heartburn recently started after starting meloxicam daily 3 times a week. He also reported about 3 loose bowel movements per day for the last 10 years.

Laboratory evaluation included normal CBC, TSH and negative HIV. Stool for ova and parasites and pancreatic elastase was normal. Tissue transglutaminase (tTG) antibody was positive at 34.

Based on his positive celiac antibody, loose stools, and heartburn, an esophagogastroduodenoscopy (EGD) noted a 2cm nodule, distal to the gastroesophageal (GE) junction. *This was biopsied and removed by a hot snare.* There was gastritis in the antrum characterized by erythema, with normal duodenum. Pathology from the GE junction nodule was consistent with heterotopic pancreatic tissue. The gastric biopsies were consistent with chronic active gastritis and the duodenal biopsies were negative for celiac disease.

Discussion

This rare case of quite large and symptomatic heterotopic pancreas located at the GE junction, was incidentally discovered during EGD performed for heartburn, loose stools, and positive celiac antibody testing.

Heterotopic pancreas (HP) (pancreatic rest or ectopic pancreas) is a congenital anomaly in which there is ectopic pancreatic tissue, separate from the main pancreatic gland, without any continuity (anatomic, vascular, or ductal) with the main pancreatic gland. HP is generally found in the stomach, duodenum, or proximal jejunum. It has been reported in other locations including the esophagus, ileum, and Meckel's diverticulum.[1] The prevalence of HP reported as 0.55 to 13.7% on autopsy; 0.2% to 0.5% in abdominal operations, and 0.9% of gastrectomies.[1] It is more commonly reported in adult males with peak incidence between 46-60.[2] It is uncommon to find HP in the esophagus with only nineteen prior reports.[3]

Pancreatic rest is a submucosal nodule where pancreatic tissue is developed and commonly found in the upper small intestine or stomach. The pathogenesis for HP is not known. Different theories of origin include: misplacement theory, which suggests fragments get separated during foregut rotation; metaplasia theory which suggests that endodermal tissues migrate to the submucosa during embryogenesis; and the totipotent cell theory which suggests that endodermal cells lining the gastrointestinal tract differentiate into pancreatic tissue.[3]

Nearly all pancreatic rests have no symptoms. It can be incidentally found at surgery or endoscopy. HP may still function in a similar manner as the main pancreatic gland and may secrete enzyme rich serous fluid with proteolytic enzymes causing a local inflammatory response.

Symptomatic HP located at the GE junction is rare with only six prior case reports.[3-7] When present at the GE junction, symptoms include dysphagia due to mass effect, symptoms of heartburn, or epigastric pain. The endoscopic appearance is usually a well-circumscribed submucosal lesion commonly with central "umbilication" covered by normal mucosa. Surface biopsies may be non-diagnostic due to sampling of overlying mucosa. The final diagnosis is based on histology of the endoscopically or surgically resected specimen. Endoscopic ultrasound with fine needle aspiration of the lesion may also be utilized for diagnosis. The endoscopic differential diagnosis includes carcinoid tumor, lymphoma, or gastrointestinal stromal tumors. The management includes conservative management with observation, resection of the HP, or esophagectomy.

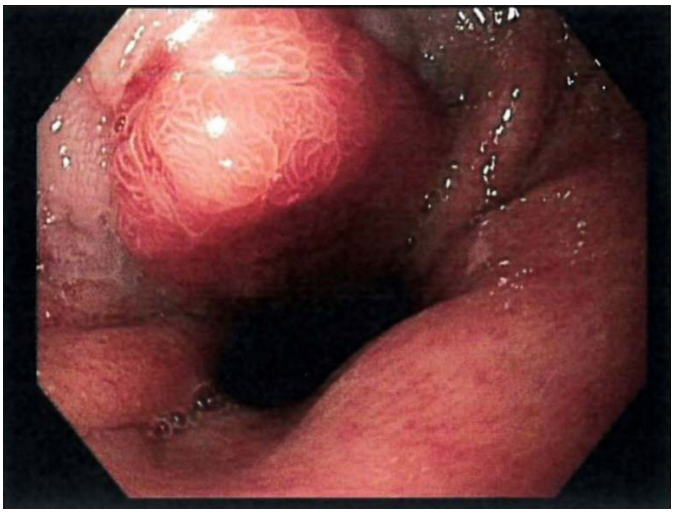

Figure 1: GE junction showing two-centimeter nodule. Pathology was consistent with a heterotopic pancreas

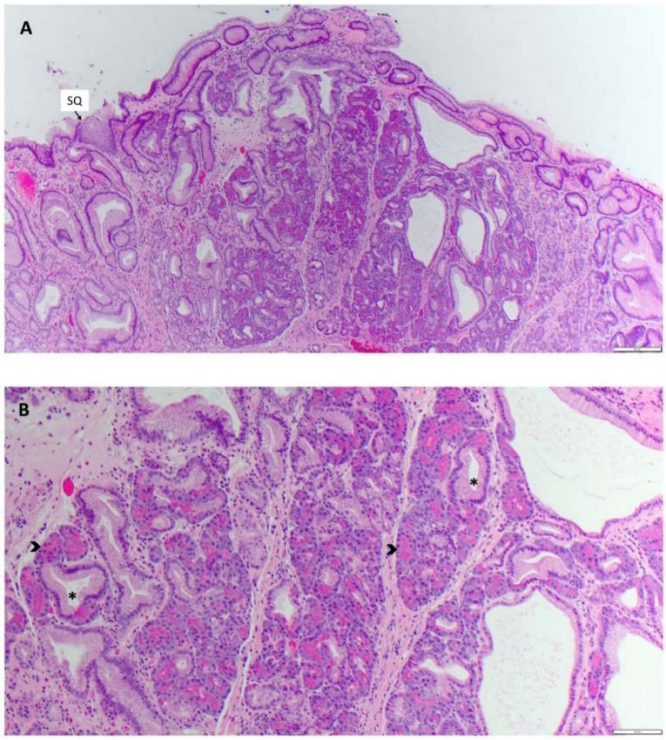

Figure 2: A: Pancreatic heterotopia/metaplasia is the lobular structure within the lamina propria in this biopsy from the gastroesophageal junction; a small focus of esophageal squamous epithelium (SQ) is seen (hematoxylin and eosin stain, x40). B: Pancreatic acinar cells (arrowhead) have deeply eosinophilic granules and ductal cells (*) have pale cytoplasm (x100).

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