Answer

Heterotopic Pancreas

Heterotopic pancreas (HP) was first described in 1727 when it was discovered in an ileal diverticulum.[1] It was defined as we currently understand it in 1921 as aberrant localization of pancreatic tissue with no direct or vascular connection to the normal pancreas that results from a developmental anomaly.[2]

Because HP is overwhelmingly asymptomatic or confused with other common gastrointestinal pathology such as peptic ulcer disease, its true incidence is challenging to determine. A variable incidence has been recorded in the literature with a range of 0.55% to 13.7% in autopsy and 0.17% to 0.83% in laparotomy.[3] It appears to be slightly more common in men.[4] An early classification scheme of HP was proposed in 1909 but replaced in 1973 by that which is currently accepted. Type I consists of acini, ducts, and islet cells similar to those seen in normal pancreas. Type II heterotopia is composed of pancreatic ducts only, referred to as *canalicular type*. Type III comprises acinar tissue only (exocrine pancreas), and type IV consists of only islet cell tissue (endocrine pancreas).[5]

The embryologic origin of HP is unclear; however, the predominant theory suggests that the ectopic tissue separates from the normal pancreas during embryonic rotation and fusion of the dorsal and ventral pancreatic buds. The pancreas develops from invaginations of endoderm in the primitive duodenum. The ventral aspect forms the head and the dorsal aspect forms the body and tail. According to the theory, ectopic pancreatic tissue in the stomach and duodenum is derived from the dorsal segment, and tissue in the jejunum and ileum originates from the ventral segment.[6]

Heterotopic pancreas can occur anywhere along the gastrointestinal tract. It is found in the stomach in 25% to 38% of cases, duodenum in 17% to 36%, and jejunum in 15% to 22%. It has been reported to occur in the esophagus, gallbladder, common bile duct, spleen, mesentery, mediastinum, and fallopian tubes.[7] Overwhelmingly asymptomatic, HP can cause symptoms varying with size of lesion or anatomic location. The most commonly reported symptoms include abdominal pain, nausea, vomiting, and melena. Pain may be associated with mechanical luminal obstruction or due to autodigestion of surrounding tissue by pancreatic enzymes. Gastric lesions are most likely to cause symptoms, presenting with epigastric pain or outlet obstruction.[8] Heterotopic pancreas can also cause symptoms that would be associated with normally localized pancreas. These include pancreatitis, pseudocyst, and even malignant transformation, with only 15 cases reported thus far.[9]

The diagnosis of HP as demonstrated by our case can prove to be exceedingly challenging. Gastric lesions can be identified in barium studies as having rounded filling defects with a central indentation or as broad, umbilicated submucosal lesions on endoscopy. These features are nonspecific, however, and often absent. Computed tomographic studies are typically nonspecific, although heterotopic pancreatic tissue can enhance to the same degree as normal pancreas with intravenous contrast. Endoscopic ultrasonography of the upper gastrointestinal tract along with fine-needle aspirate has been increasingly used for the diagnosis of heterotopic pancreas. However, endoscopic removal of submucosal lesions is regarded as technically difficult and one of the more dangerous endoscopic procedures with regard to the risks of bleeding and bowel perforation.[4] In our case, computed tomography enterography was used as an adjunct to endoscopy to aid in the diagnosis of the offending lesion. An alternative to small-bowel follow-through with fluoroscopy, this technique involves traditional abdominal computed tomography with the addition of barium-based contrast material given orally to enhance the lumen, mucosa, and extraluminal structures of the small bowel.

Heterotopic pancreatic tissue is a rare cause of occult gastrointestinal pain, obstruction, and bleeding. Asymptomatic lesions do not require intervention or surveillance. The preferred treatment for symptomatic lesions is surgical resection.

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Submissions

Due to the overwhelmingly positive response to the Image of the Month, the *JAMA Surgery* has temporarily discontinued accepting submissions for this feature. Requests for submissions will resume in April 2013. Thank you.