

# Heterotopic Pancreatic Tissue Found in the Esophagus of a 14-year-old Girl

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**H**eterotopic pancreatic tissue, otherwise known as pancreatic rest, is pancreatic tissue that lacks anatomic and vascular continuity with the main body of the pancreas. Common locations for this tissue include the stomach, duodenum, jejunum, Meckel diverticulum, and ileum.<sup>1,2</sup> We report a case of heterotopic pancreatic tissue found in the esophagus, which is an exceedingly rare finding.

## Case Report

A 14-year-old girl presented with complaints of peri-umbilical abdominal pain. Episodes of pain tended to occur at least once daily, typically lasting for 1 hour. The patient had no history of constipation, vomiting, fever, melena, or weight loss. Her physical examination revealed a healthy-appearing female patient who was at nearly the 75th percentile for weight. There was no abdominal tenderness or appreciable mass. Complete blood count with differential, chemistries, liver function tests, erythrocyte sedimentation rate, amylase levels, and lipase levels were all within normal limits. Serology tested negative for *Helicobacter pylori* and celiac disease. Computed tomographic scan of the abdomen and pelvis, along with upper gastrointestinal series with small bowel follow-through, were both unremarkable.

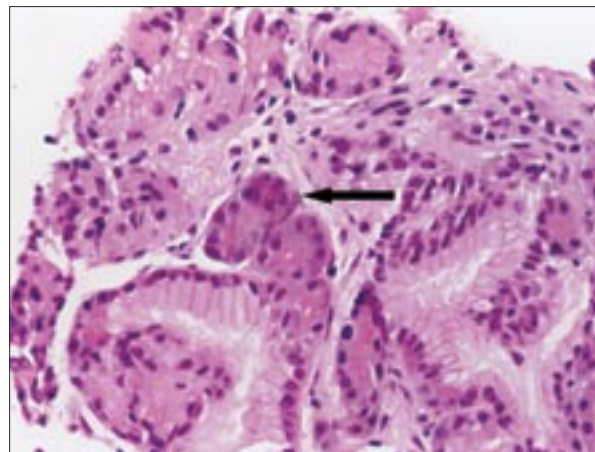
A circular patch of raised shiny yellowish mucosa, approximately 2 cm in diameter, was seen in the distal third of the esophagus during esophagogastroduodenoscopy (Figure 1). Hematoxylin and eosin staining of the biopsies from this area revealed ectopic pancreatic tissue in the submucosa (Figure 2).

In order to determine the extent of the lesion, endoscopic ultrasound was performed. The pancreatic

rest did not appear to extend beyond the submucosa. A small tubular anechoic structure thought to represent a pancreatic ductule was seen within the ectopic tissue (Figure 3). Endoscopic biopsies of the abnormal-appearing esophageal mucosa once again confirmed the presence of pancreatic acinar cells in the submucosa.



**Figure 1.** Pancreatic rest of the distal esophagus. Arrow points to pancreatic tissue.



**Figure 2.** Hematoxylin and eosin stain of esophageal biopsy. Arrow points to pancreatic tissue in the submucosa (original magnification,  $\times 200$ ).

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**Figure 3.** Endoscopic ultrasound image of distal esophagus. Outline of ectopic pancreatic tissue demarcated by thin yellow arrows. Short white arrow points to suspected pancreatic ductule.

## Discussion

Heterotopic pancreatic tissue, which occurs most commonly in the stomach and duodenum, appears endoscopically as a submucosal lesion that usually contains a central umbilication. Unless the pancreatic rest is large, upper gastrointestinal series is usually normal. Histologically, the tissue may contain acini, islets, and ducts. Patients do not usually develop symptoms from this entity, which is typically found during an evaluation for unrelated complaints. If symptoms develop, they are most likely secondary to a mass effect. Large lesions may cause obstruction, ulceration, hemorrhage, or intussusception. Other complications include pancreatitis, pseudocyst formation, carcinomas, islet-cell tumors, or inflammatory pseudotumors. Surgical excision is the only cure.<sup>1-3</sup>

Ectopic pancreatic tissue in the esophagus has been reported in the literature only 10 times. Only 3 of the patients with this lesion were younger than 18 years old. In these children, the ectopic tissue was associated with one or more anatomic abnormalities of the esophagus.<sup>2</sup> One patient had a tracheoesophageal fistula, esophageal atresia, and esophageal duplication.<sup>4</sup> Another patient had a congenital diverticulum,<sup>5</sup> and the third had a congenital cyst.<sup>6</sup>

Pancreatic rest was found in the distal third of the esophagus in 6 of the 10 patients. In 2 adult patients, the lesion was found to be malignant. Nine patients underwent resection or enucleation of the lesion, and 1 patient was observed.<sup>2</sup>

The prevalence of malignant transformation of heterotopic pancreatic tissue is unknown. Guillou and associates proposed that a carcinoma should be stated to originate from heterotopic pancreatic tissue only when the following three conditions are met: the tumor is found within or close to the ectopic pancreatic tissue; a direct transition between the pancreatic structures and the carcinoma can be shown (eg, duct cell dysplasia); and the nonneoplastic pancreatic tissue contains fully developed acini and ductal structures.<sup>7</sup>

## Conclusion

The patient described above has no known anatomic abnormalities. Currently, the lesion does not appear to extend beyond the submucosa, and there is no endoscopic or histologic evidence of neoplasia. Routine follow-up of the lesion is planned.

## References

1. Feldman M, Friedman LS, Brandt LJ, eds. *Sleisenger and Fordtran's Gastrointestinal and Liver Disease*. 7th ed. Philadelphia, Pa: Saunders; 2002:883-884.
2. Temes RT, Menen MJ, Davis MS, Pett SB Jr, Wernly JA. Heterotopic pancreas of the esophagus masquerading as Boerhaave's syndrome. *Ann Thorac Surg*. 2000;69:259-261.
3. Noffsinger AE, Hyams DM, Fenoglio-Preiser CM. Esophageal heterotopic pancreas presenting as an inflammatory mass. *Dig Dis Sci*. 1995;40:2373-2379.
4. Ishikawa O, Ishiguro S, Ohhigashi H, Sasaki Y, Yasuda T, et al. Solid and papillary neoplasm arising from an ectopic pancreas in the mesocolon. *Am J Gastroenterol*. 1990;85:597-601.
5. Chatterjee PK, Chatterjee SN, Dastidar N, et al. Heterotopic gastric mucosa and pancreatic tissue in congenital diverticulum of oesophagus. *Indian J Surg*. 1982;44:139-141.
6. Roshe J, Del Buono E, Domenico D, Colturi TJ. Anaplastic carcinoma arising in ectopic pancreas located in the distal esophagus. *J Clin Gastroenterol*. 1996;22:242-247.
7. Guillou L, Nordback P, Gerber C, Schneider RP. Ductal adenocarcinoma arising in a heterotopic pancreas situated in a hiatal hernia. *Arch Pathol Lab Med*. 1994;118:568-571.

# Review

## The Cantankerous Pancreas

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The case reported by Qualia and colleagues is a valuable contribution to the sparse esophageal heterotopic pancreas (HP) literature.<sup>1</sup> This entity arises from disturbances in normal embryologic development that create deposits of pancreas throughout the gastrointestinal tract. Consequently, esophageal HP is frequently associated with other foregut abnormalities, including esophageal atresia, tracheoesophageal fistula, duplication cyst, diverticulum, and sequestration. The tumors are submucosal and contain exocrine tissue, endocrine tissue, or a combination of both histologic types. Although nonesophageal HP is found in 0.2–0.5% of patients who have abdominal operations, esophageal HP is extremely rare, with only approximately one dozen cases reported.<sup>2–8</sup>

In addition to the sites discussed by Qualia and associates, HP has also been found in the mediastinum, lung, liver, gallbladder, bile duct, spleen, fallopian tube, large bowel, mesentery, omentum, umbilicus, tongue, lymph node, and other sites. Although only approximately one third of patients with nonesophageal HP have symptoms, all patients with esophageal HP have been symptomatic.<sup>8–11</sup> The presence of symptoms in nonesophageal HP has been related to tumor size, location, and mucosal involvement.<sup>9</sup>

Diagnostic evaluation traditionally includes both radiographic and endoscopic studies. Computed tomographic scans of gastric HP have not been useful.<sup>12–14</sup> Contrast radiographs reveal an intramural lesion with central umbilication in most cases.<sup>15</sup> Branching of the central duct within the lesion is sometimes seen.

The characteristic endoscopic appearance of HP is a submucosal mass with an intact mucosa and a ductal orifice. In some patients, biopsies are diagnostic. However, the submucosal location often hampers routine biopsy

techniques. More recently, esophageal ultrasound (EUS) has been helpful, especially when combined with needle biopsy.<sup>4,16–19</sup> This approach allows both anatomic and histologic delineation of the mass and could become the diagnostic modality of choice in the future.

Qualia and coworkers describe many possible complications that can occasionally occur with HP. The most important of these is the risk of neoplastic transformation. Adenocarcinoma, solid and papillary tumors, anaplastic cancer, islet-cell adenomas, cystadenocarcinomas, and other cancers have been associated with nonesophageal HP. There have been reports of cancers at most nonesophageal sites, but the frequency of these malignancies is quite low.<sup>7,11,20–22</sup> In contrast, the sparse available reports suggest that the risk of malignancy in esophageal HP is approximately 15–20%.<sup>3,5,7,8</sup> Although esophageal HP has been found in all age groups, cancers have only occurred in adults.

The management of nonesophageal HP is hampered by low clinical suspicion and inaccurate diagnosis.<sup>10,13,23,24</sup> EUS with needle biopsy in elective settings should improve the chances of correct diagnosis and treatment. Minimally invasive resection techniques allow simultaneous diagnosis and management of nonesophageal HP.<sup>11,18,19,25–28</sup> Therefore, symptomatic patients and patients without a definite diagnosis should undergo limited resection using endoscopic or minimally invasive surgical techniques when possible.<sup>11,22,25,27,28</sup> Asymptomatic patients with incidentally found, definitively diagnosed, benign, nonesophageal HP can sometimes be observed.<sup>11</sup> In emergent situations where diagnosis is not known, operation should be guided by frozen section to avoid unnecessarily radical resections.<sup>11,15</sup>

To date, all patients with esophageal HP have been symptomatic. Based upon the limited available data, adult patients with esophageal HP may have a significant risk for malignancy. Consequently, surgical resection should be considered in adults with esophageal HP. If observation is chosen, surveillance endoscopy and endoscopic ultrasound should be undertaken frequently to evaluate for progression. Serial biopsy or EUS needle aspirations should be performed to enhance imaging and facilitate early identification of malignant degeneration. Whether future applications of minimally invasive endoscopic or surgical techniques will be beneficial in esophageal HP remains to be determined.

## References

1. Qualia CM, Rossi TM, Ullah A. Heterotopic pancreatic tissue found in the esophagus of a 14-year-old girl. *Gastroenterol Hepatol*. 2007;3:939-940.
2. Gananadha S, Hunt DR. A unique case of pancreatitis and retention cyst in esophageal heterotopic pancreas. *Surg Laparosc Endosc Percutan Tech*. 2005;15:345-347.

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3. Guillou L, Nordback P, Gerber C, Schneider RP. Ductal adenocarcinoma arising in a heterotopic pancreas situated in a hiatal hernia. *Arch Pathol Lab Med.* 1994;118:568-571.
4. Rodriguez FJ, Abraham SC, Allen MS, Sebo TJ. Fine-needle aspiration cytology findings from a case of pancreatic heterotopia at the gastroesophageal junction. *Diagn Cytopathol.* 2004;31:175-179.
5. Roshe J, Del Buono E, Domenico D, Colturi TJ. Anaplastic carcinoma arising in ectopic pancreas located in the distal esophagus. *J Clin Gastroenterol.* 1996;22:242-247.
6. Shalaby M, Kochman ML, Lichtenstein GR. Heterotopic pancreas presenting as dysphagia. *Am J Gastroenterol.* 2002;97:1046-1049.
7. Tanaka K, Tsunoda T, Eto T, Yamada M, Tajima Y, et al. Diagnosis and management of heterotopic pancreas. *Int Surg.* 1993;78:32-35.
8. Temes RT, Menen MJ, Davis MS, Pett SB Jr, Wernly JA. Heterotopic pancreas of the esophagus masquerading as Boerhaave's syndrome. *Ann Thorac Surg.* 2000;69:259-261.
9. Armstrong CR, King PM, Dixon JM, Macleod IB. The clinical significance of heterotopic pancreas in the gastrointestinal tract. *Br J Surg.* 1981;68:384-387.
10. Eisenberger CF, Gocht A, Knoefel WT, Busch CB, Peiper M, et al. Heterotopic pancreas—clinical presentation and pathology with review of the literature. *Hepatogastroenterology.* 2004;51:854-858.
11. Neupert G, Appel P, Braun S, Tonus C. Heterotopic pancreas in the gallbladder: diagnosis, therapy, and course of a rare developmental anomaly of the pancreas [in German]. *Chirurg.* 2007;78:261-264.
12. Cho JS, Shin KS, Kwon ST, Kim JW, Song CJ, et al. Heterotopic pancreas in the stomach: CT findings. *Radiology.* 2000;217:139-144.
13. Hsia CY, Wu CW, Lui WY. Heterotopic pancreas: a difficult diagnosis. *J Clin Gastroenterol.* 1999;28:144-147.
14. Park SH, Han JK, Choi BI, Kim M, Kim YI, et al. Heterotopic pancreas of the stomach; CT findings correlated with pathologic findings in six patients. *Abdom Imaging.* 2000;25:119-123.
15. Lai EC, Tompkins RK. Heterotopic pancreas: review of a 26-year experience. *Am J Surg.* 1986;151:697-700.
16. Arantes V, Logrono R, Faruqi S, Ahmed I, Waxman I, Bhutani MS. Endoscopic sonographically guided fine-needle aspiration yield in submucosal tumors of the gastrointestinal tract. *J Ultrasound Med.* 2004;23:1141-1150.
17. Goto J, Ohashi S, Okamura S, Urano F, Hosoi T, et al. Heterotopic pancreas in the esophagus diagnosed by EUS-guided FNA. *Gastrointest Endosc.* 2005;62:812-814.
18. Kojima T, Takahashi H, Parra-Blanco A, Kohsen K, Fujita R. Diagnosis of submucosal tumor of the upper GI tract by endoscopic resection. *Gastrointest Endosc.* 1999;50:516-522.
19. Ormarsson OT, Gudmundsdottir I, Marvik R. Diagnosis and treatment of gastric heterotopic pancreas. *World J Surg.* 2006;30:1682-1689.
20. Makhlof HR, Almeida JL, Sobin LH. Carcinoma in jejunal pancreatic heterotopia. *Arch Pathol Lab Med.* 1999;123:707-711.
21. Matsuki M, Gouda Y, Ando T, Matsuoka H, Morita T, et al. Adenocarcinoma arising from aberrant pancreas in the stomach. *J Gastroenterol.* 2005;40:652-656.
22. Rimal D, Thapa SR, Munasinghe N, Chitre VV. Symptomatic gastric heterotopic pancreas: clinical presentation and review of the literature. *Int J Surg.* 2007;[Epub ahead of print].
23. Shi HQ, Zhang QY, Teng HL, Chen JC. Heterotopic pancreas: report of 7 patients. *Hepatobiliary Pancreat Dis Int.* 2002;1:299-301.
24. Ayantunde AA, Pinder E, Heath DI. Symptomatic pyloric pancreatic heterotopia: report of three cases and review of the literature. *Med Sci Monit.* 2006;12:CS49-CS52.
25. Hackett TR, Memon MA, Fitzgibbons RJ Jr, Mixter CG. Laparoscopic resection of heterotopic gastric pancreatic tissue. *J Laparoendosc Adv Surg.* 1997;7:307-312.
26. Hsu SD, Wu HS, Kuo CL, Lee YT. Robotic-assisted laparoscopic resection of ectopic pancreas in the posterior wall of gastric high body: case report and review of the literature. *World J Gastroenterol.* 2005;11:7694-7696.
27. Lee TH, Wang HP, Huang SF, Wang TH, Lin JT. Endoscopic mucosal resection for heterotopic pancreas in the stomach. *J Formos Med Assoc.* 1999;98:643-645.
28. Lucena JF, Alvarez OA, Gross GW. Endoscopic resection of heterotopic pancreas of the minor duodenal papilla: case report and review of the literature. *Gastrointest Endosc.* 1997;46:69-72.