ACUTE ECTOPIC PANCREATITIS WITH ILEUS OCCURRING IN GASTRIC ECTOPIC PANCREATIC TISSUE

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Abstract. Ectopic pancreatic tissue is most often an incidental finding of imaging, surgery, or autopsy. Image-guided diagnosis is difficult and definitive diagnosis usually relies on histological analysis. A case of ectopic pancreatic tissue located near the lesser curvature of the stomach complicated with acute ectopic pancreatitis is presented followed by a brief discussion of the clinical management of heterotopic pancreas.

Keywords: pancreatitis, heterotopic pancreas, ileus

Introduction

Ectopic (heterotopic) pancreatic tissue, often referred to as a pancreatic rest is the presence of pancreatic tissue found outside the normal anatomic position and lacking vascular connection with the body of the pancreas[1] occurring in 0.55% to 13.7% of the population[2]. In most cases, the tissue is functional and is commonly found along the greater curvature of the stomach, duodenum, proximal jejunum and ileum [3.4.5]. Less frequently heterotopic pancreatic tissue can be found in the umbilicus, common bile duct, gallbladder, and Meckel's diverticulum. Despite its rare clinically significance ectopic pancreatic tissue has been associated with ulceration and bleeding, intussusceptions, and even pancreatitis and adenocarcinoma. When the tissue is symptomatic, treatment consists in surgical resection, but the removal of ectopic pancreatic tissue that is found incidentally remains a controversial matter.

Pathophysiology

The pathophysiology of heterotopic pancreas is not fully understood and multiple theories exist. One explanation proposes that during embryogenesis, pancreatic metaplasia of the endodermal tissues localized in the gastric mucosa occurs [5,6]. Another theory suggests that heterotopic pancreas could be caused by the inhibition of normal cellular signaling during development; inhibiting hedgehog signaling in chick embryos leads to ectopic budding of pancreatic structures in the stomach and duodenum [7].

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Presentation

Most patients are asymptomatic, but symptoms can include nausea, vomiting, epigastric pain, dyspepsia, abdominal fullness, and melena [1,3]. The most common symptom is epigastric pain[7]. About a third of symptomatic patients report clinical symptoms that mimic disease related to the organ in which the tissue resides[5]. More serious complications can ensue such as massive gastrointestinal bleeding, gastric outlet obstruction, gastric or duodenal ulceration, pancreatitis, and malignant degeneration[3,8].

Diagnosis

Diagnosis of heterotopic pancreas can be very difficult and is dependent on presentation (aici nu inteleg sensul- depinde de prezentare?). The physical examination rarely provides clues to the diagnosis of pancreatic rests as they are rarely, if ever, large enough to be detected by palpation[2]. CT can sometimes be helpful; the findings usually depict a small, round or oval sharply margined broad-based mass[3]. Occasionally, it may appear as a mass with an irregular surface resembling an adenomatous polyp or polypoid carcinoma[3]. Five CT criteria have been used with good sensitivity and specificity to help differentiate between ectopic pancreas and gastrointestinal stromal tumor (GIST)[9]. These criteria are as follows: pre-pyloric antrum or duodenum in location, an ill-defined border, an endoluminal growth pattern, a long diameter/short diameter ratio larger than 1.4, and prominent mucosal enhancement. When two or more criteria are met, the sensitivity and specificity for diagnosing ectopic pancreas approaches 100% and 82.5%, respectively[9]. EGD can be useful and findings can be described as a small (around 2 cm) nodular, submucosal mass
covered by normal mucosa with or without central umbilication[4,1]. Endoscopic ultrasonography can be helpful for determining the nature of the mass. Findings of indistinct borders, anechoic duct-like structures, mural growth pattern, presence in more than one layer, and indistinct borders suggest ectopic pancreatic tissue (and less likely other tumors) in the stomach[10]. Barium swallow studies reveal a similar appearance. Biopsies done during EGD are frequently non-diagnosed due to the superficial sample; they are usually reported as normal gastric mucosa. Definitive diagnosis is always made histopathologically[3,1]. Specimens can be sent for frozen section[1].

**Treatment**

Treatment of heterotopic pancreas is specific for the patient with obvious symptomatology. In the asymptomatic patient, maintaining medical supervision with periodic reviews is recommended[5]. The symptomatic patient usually experiences relief when the lesion is removed [1,3,4]. When encountered incidentally during surgery, lesions can be excised to prevent further operations or complications.

**Prognosis**

The prognosis in patients with surgically treated pancreatic rests is usually excellent[3,1]. Although most of these lesions remain asymptomatic, it is important to remember that they are still susceptible to the same pathologic conditions as normal pancreatic tissue such as pancreatitis and malignant change[3,1].

**Case report**

A 38-year-old woman presented to the emergency room (ER) with moderate, colicky abdominal epigastric pain, nausea and vomiting, absence of stool and gas passage. The symptoms started three days prior to presentation and evolved progressively. The patient had a surgical history for a right oophorectomy due to an extrauterine pregnancy. On presentation the patient denied fever, diarrhea, constipation, black stools, hematochezia, hematemesis, dysuria, or abnormal vaginal bleeding. On physical examination, vital signs were within normal limits. The abdomen was soft, non-distended, but mildly tender in the epigastric quadrant. No rigidity, rebound, or guarding was present. Her white blood cell count increased to 11,300/ml with a left shift. Comprehensive metabolic profile showed hypercholesterolemia and hypertriglyceridemia, amylase and lipase level 10 xUNL; her urinalysis was within normal limits. Her pregnancy test was negative. Conservative management was implemented for a presumptive diagnosis of pancreatitis. The abdominal ultrasound revealed slow bowel movements and free liquid in the pelvic floor that measures 9mm. The plain radiography described air-fluid levels in the mesogastric quadrant, without free gas in the abdominal cavity (figure 1).

Esophagogastroduodenoscopy (EGD) was advised to confirm there were no changes on the wall of the stomach. The EGD showed an extrinsic/submucosal mass with antral localization, with central depression, without free gas in the abdominal cavity (figure 1).

The follow-up CT found the mass to be essentially unchanged. On the external face of the head of the pancreas, in contact with the structure of the pancreas, we describe a round structure with the highest diameter 23/16mm, with the same density with the pancreas, structure that impress the duodenum between the second and the third portion with irregular contour and heterogeneous enhancement(figure 4).

Free fluid in the Douglas pouch measuring 19mm. The diagnosis of acute pancreatitis on ectopic pancreas and functional ileus was established.

The patient improved with pain management and intravenous fluids; she was then discharged home without pain; the patient was counseled regarding
treatment; given the risk of the recurrence and the rare risk for malignant transformation and the fact that she elected conservative management and observation.

Discussions

Heterotypic pancreas is an infrequent diagnosis and rarely makes its way into differential diagnoses. Symptoms can be variable and the incidence is low enough so that many physicians have never seen a case. In addition, it can manifest clinically in some rare diseases of the pancreas including pancreatitis, islet cell tumor, pancreatic carcinoma, and pancreatic cyst. To date, there have been a few reports describing pancreatitis due to heterotopic pancreas. Therefore, the present case was considered to be a very rare case of this disorder; the incidence is reported to be 0.4%.

Although many modalities are available for workup, definitive diagnosis is always histological. Specific CT findings described above can be helpful for differentiating between ectopic pancreas and GIST. However, the tumor met a part of the criteria (pre-pyloric antrum location, an ill-defined border, an endoluminal growth pattern and prominent mucosal enhancement), and therefore the diagnosis could be made with CT alone. Frozen section can allow for immediate local excision without further operations but in our case it was recommended that the patient should not undergo invasive procedures because of the risk of pancreatitis recurrence. However, for our patient, it was felt the risk of complication with excision outweighed the benefit and it was also the patient’s desire for conservative management. In this circumstance, we felt that monitoring was a more appropriate option.

Conclusions

The importance of this case is emphasized by the low frequency of acute pancreatitis on ectopic pancreatic tissue which in our opinion should be actively assessed and treated appropriately; also, it is important for all the specialists involved in the medical management of these patients to distinguish their high potential for relapsing symptoms and very rare life threatening complications like gastric perforation. Ectopic pancreas with its unusual clinical manifestations, locations or complications is of clinical interest. To conclude, heterotopic pancreas and its complications should always be considered in the differential diagnosis of acute abdomen.

References