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Heterotopic Pancreatic Pseudocyst Radiologically Mimicking Gastrointestinal Stromal Tumor

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Heterotopic pancreas is a relatively common variant of foregut embryologic dystopia that can be described as pancreatic tissue found outside the normal anatomic location, being independent from vascular supply of normal pancreas. Having all features of pancreatic tissue except for the major duct structures, this ectopic tissue may be clinically recognized when pathologic changes take place. Inflammation, hemorrhagic or obstructive states, and eventually malignancy-related problems may become a diagnostic challenge for clinician and finally lead to consequences of misdiagnosis. In this article we will discuss a case of heterotopic pancreatic tissue located in gastric cardia, which was diagnosed preoperatively as gastrointestinal stromal tumor.

Key words: Ectopic pancreas – Pseudocyst – Gastrointestinal stromal tumor – Misdiagnosis

Heterotopic (ectopic) pancreas is defined as the presence of pancreatic tissue in an aberrant site that is not anatomically and vascularly connected to the pancreas proper.[1,2] Ectopic pancreatic tissue is most commonly found in the duodenum (particularly the second portion), ampulla, stomach (prepyloric antrum), and upper jejunum; it can also occur in the liver, biliary tract, Meckel's diverticulum, and several other sites such as large bowel, spleen, omentum, mediastinum, lung, thyroid, and even brain.[1–6] The heterotopic pancreatic tissue is usually located in the submucosa and/or the muscularis or subserosa.[1,2,4]

Practically any pancreatic pathology can also occur in the heterotopic pancreatic tissue including acute pancreatitis and neoplastic transformation.[1,4,7,8] Episodes of acute pancreatitis may cause local inflammation and typical pancreatitis-related com-

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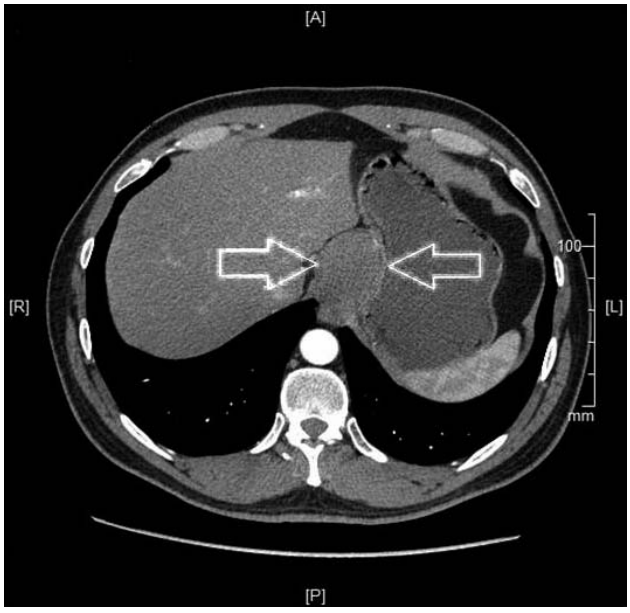

Fig. 1 Well circumscribed, hypodense lesion radiologically considered to be a gastrointestinal stromal tumor. Arrows delineate outer borders of the tumor.

plications in the host organ. The ectopic pancreatic tissue can lead to ulceration and bleeding in the cases located beneath a mucosa. Being a mass-forming lesion, it may also result in luminal obstruction (particularly in the prepyloric antrum).[1,4]

Although it is a relatively common congenital anomaly (0.5% to 13% in autopsy series), in most cases heterotopic pancreas remains clinically silent and is found incidentally during surgery or an endoscopic and/or radiological investigation.[1,9] Heterotopic pancreas may be confused with other solid tumors of the host organ.

Heterotopic pancreas in the stomach is usually located in the prepyloric area.[10] We report here an unusual case of heterotopic pancreas with a pseudocyst formation in the gastric cardia mimicking gastrointestinal stromal tumor.

Case Report

A 44-year-old male presented with a two month history of abdominal pain, nausea, and vomiting. The pain was intense in the mid-epigastric region and radiated to all over the abdomen and had no relation with meals. The patient denied any weight loss during this time period. Physical examination showed normal findings. Routine blood tests including serum alfa fetoprotein, CA 19-9, and carcinoembriyonic antigen levels were within normal limits. One week before admission to our hospital the patient undertook a computerized tomography with oral and intravenous contrast which showed a 4.8 × 4.9 × 4.3 cm well-defined mass lesion on the lesser curvature in the gastric cardia with no evidence of local invasion (Fig. 1). The lesion was in close proximity with the left lobe of liver. The patient was referred to us for further treatment. An endosonographic study at our hospital revealed a 4.3 cm diameter hypoechoic lesion located on the lesser curvature of the stomach just below the esophagogastric junction and a gastrointestinal stromal tumor was suspected. A decision was made to proceed with surgery. On exploration, an exophytic cystic lesion measuring 6 × 5 cm was identified on the anterior wall of the stomach in the cardia region. The pancreas was found to be normal and no other intra-abdominal pathology was detected. The lesion was surrounded by a mild inflammatory reaction which produced adherence to the hepatogastric ligament and the left lobe of liver. The lesion appeared to infiltrate serosa but not the muscularis propria and mucosa and was not infiltrating the surrounding tissue. During the dissection of the lesion the cyst cavity was opened and approximately 40 ml of creamy white fluid aspirated for further analysis. The lesion's anterior wall was excised completely. The gastric wall underneath the lesion appeared to be intact; there was no gross involvement of the mucosa. Since the lesion appeared benign and nonneoplastic we decided not to perform gastric resection. Unfortunately, the frozen section examination was not available at the time of the surgery. The patient did have an uneventful postoperative period and was discharged 5 days after surgery. He has remained free of symptoms with negative endoscopic and computerized tomographic findings since then.

Biochemical study of the diluted (1/10) aspirate revealed high levels of pancreatic amylase (1422 U/L). Histopathologic examination of the resected lesion showed pancreatic tissue with acinar cells and dilated ducts located subserosally (Fig. 2A and 2B). The ductal epithelia and acinar cells were positively stained with antibodies against cytokeratin 19 (Fig. 2C).[11] The pathologic diagnosis was heterotopic pancreatic tissue with pseudocyst formation.

Discussion

Pancreatic heterotopia is often an incidental finding.[1,12] It is generally asymptomatic, but it may cause symptoms when complicated by inflammation,[7] ulceration, hemorrhage,[13] obstruction,[4] or

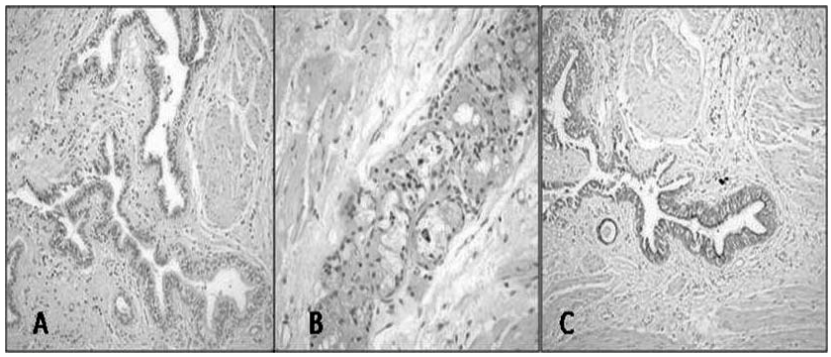

Fig. 2 Heterotopic pancreatic tissue in the muscularis propria [HE, (A) ×200, (B) ×400]. (C) Heterotopic pancreatic ducts showing CK19 expression (x200).

malignant transformation.[8] Ectopic pancreatic tissue in the stomach is usually found in the prepyloric area.[10] Ectopic pancreas in the gastric cardia has been rarely reported.[10,14,15] In majority of cases it is buried in submucosal layer of the gastric wall.[2] In the presented patient, the ectopic pancreatic tissue was located in the cardia and largely the subserosal layer and partially muscularis propria of the stomach were involved. The gastric ectopic pancreatic tissue may cause abdominal pain, nausea, and vomiting, as in our case.[12]

Cystic changes can be seen in the ectopic pancreatic tissue.[7,16–18] If there is no communication between the ectopic tissue and the gastric lumen, retention of the exocrine secretions may result in cyst formation. Retention cysts are usually small (less than 1–2 cm in size) and lined by a single layer of normal epithelium; whereas, pseudocysts are usually bigger and their wall lacks epithelial lining.[19] In our case, the cyst was devoid of epithelial lining and also mild inflammation was found around the cyst. Therefore, we thought that it was more likely a pseudocyst.

Endoscopic ultrasonography (EUS) and computerized tomography (CT) are frequently used for the diagnosis of gastric submucosal mass lesions. Because gastrointestinal stromal tumor (GIST) is the most common gastric submucosal mass lesion, heterotopic pancreas is usually confused with GIST at EUS or CT.[20] It is difficult to distinguish ectopic pancreas from gastric GIST at CT or EUS, because there are no characteristic findings of ectopic gastric pancreas.[15,21,22] However, recent studies[23,24] have suggested that combined use of some imaging features (endoluminal growth pattern, ill-defined border, and prepyloric location) of gastric ectopic pancreas can help distinguish it from GIST. In our case, both CT and EUS misdiagnosed the ectopic pancreatic tissue as a GIST that underwent degeneration. Presence of a well-defined border, exophytic growth pattern, and proximal gastric location of the lesion led to radiologist interpret the lesion as gastric GIST.

Asymptomatic ectopic pancreas does not require treatment in the presence of a histopathologic diagnosis. However, it is difficult to obtain a tissue diagnosis of heterotopic pancreas without an operative intervention and therefore in most cases a surgical removal of the lesion is unavoidable. Symptomatic patients should undergo surgery both to obtain tissue diagnosis and for symptom relief. Surgery can be performed by using open or minimally invasive techniques depending on the location and size of the lesion. In the present patient, due to proximal location and size of the lesion we decided to proceed with open surgery.

In conclusion, preoperative diagnosis of gastric ectopic pancreas remains challenging. In this case report we demonstrated a gastric ectopic pancreas with an atypical appearance and location.

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