ECTOPIC PANCREAS IMITATING GASTROINTESTINAL STROMAL TUMOR (GIST) IN THE STOMACH

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Ectopic pancreas is a rare congenital disorder defined as pancreatic tissue lacking vascular or anatonic communication with the normal body of the pancreas. Most cases of ectopic pancreas are asymptomatic, but it may become clinically evident depending on the size, location and the pathological changes similar to those observed in case of the normal pancreas. It is often an incidental finding and can be located at different sites in the gastrointestinal tract. The most common locations are: the stomach, duodenum or the proximal part of small intestine. The risk of malignancy, bleeding and occlusion are the most serious complications. Despite the development in diagnostics, it still remains a challenge for the clinician to differentiate it from neoplasm.

In this report, we described a case of 28-years old woman who presented recurrent epigastric pain. The upper gastrointestinal endoscopy revealed gastrointestinal stromal tumor on the border of the body and antrum of the back wall of great curvature of the stomach. The histopathological examination after surgery showed heterotopic pancreatic tissue. Ectopic pancreas should be considered in the differential diagnosis of gastric mass lesions.

Key words: ectopic pancreas, heterotopic pancreas, gastrointestinal stromal tumor, stomach

Ectopic (heterotopic, accessory, aberrant) pancreas is defined as developmental anomaly which is characterized by the presence of pancreatic tissue outside its anatomical location in the pancreas. This tissue has no anatomic, neural and vascular connections of the organ from which it is derived (1). Usually ectopic pancreas is a solid structure, but often cystic lesions with mixed heterogeneous structure are observed. Heterotopic pancreatic tissue generally has all physiological functions of the pancreas and is characterized by the susceptibility of occurrence to various diseases (cysts, inflammation, tumors) similar to the organ from which it is derived. Ectopic pancreas may be present in all segments of the gastrointestinal tract, in the organs neighboring to the gastrointestinal tract, and also in the organs located outside the abdominal cavity. This disease is usually asymptomatic until the appearance of complications. It may give non-specific symptoms such as abdominal pain or discomfort, nausea and vomiting, and GI bleeding (2, 3). More rarely appear obstruction, intussusception and the symptoms associated with development of acute or chronic pancreatitis.

Ectopic pancreas is found incidentally at endoscopic or imaging examinations, surgery and autopsy. In autopsy studies in adult patients ectopic pancreas is diagnosed with an incidence of 0.5-3.7%, slightly more common in middle-aged men. In patients operated on this incidence it is slightly higher and estimated at 1:500 laparotomy. Causes of ectopic pancreas are not exactly known, probably it is formed as a result of disordered processes of embryogenesis and abnormal differentiation of the stem cell lines (4).

According to the Heinrich’s classification from 1909 and the subsequent Gaspar-Fuentes modification from 1973, there are four basic
types of ectopic pancreas: type I – composed of all elements of normal pancreas, type II consists of pancreatic ducts only, termed as canalicular variety. Type III is composed of acinar tissue only (exocrine pancreas) and type IV of islet cells only (endocrine pancreas) (5, 6). The differential diagnosis of ectopic pancreas should take into consideration other submucosal changes such as GISTs, leiomyomas, lipomas, fibromas or lymphoma. However, the final confirmation of the diagnosis of ectopic pancreas is possible only on the basis of histopathological examination of the biopsy or resective material (6, 7).

This report presented a rare case of ectopic pancreas present in the submucosa of gastric wall imitating gastrointestinal stromal tumor which was successfully treated by laparoscopic local resection of the stomach.

CASE REPORT

A 28-year old woman was admitted to the 2nd Department of General and Gastrointestinal Surgery on 18th May 2014 with suspicion of gastrointestinal stromal tumor in the stomach. The patient complained about non-characteristic abdominal pain lasting for one year with no relation to meals, without vomiting, diarrhea and weight loss. In the history of the disease, operations of varus foot were recorded-postoperative scars both, on lower leg and feet. The palpation of the abdomen showed no abnormalities. Laboratory parameters were in the normal range. Chest X-ray showed no changes. Abdominal ultrasonography showed no pathological conditions. Endoscopic examination of the upper gastrointestinal tract performed one year earlier demonstrated lesion located on the border of the body and antrum of the back wall of great curvature infiltrating into the lumen of the stomach, 1 cm in diameter, covered with a macroscopically unchanged mucous membrane. Helicobacter pylori test was negative. In biopsy specimens taken for histopathological examination, chronic low-grade inflammation of low activity has been revealed. In the second endoscopic examination performed on 5th May 2014 on the border of the body and antrum of the back wall of great curvature nodular change with diameter 1.5 cm and features of the stromal tumor has been found and thus, further diagnostics was abandoned.

On the basis of this diagnosis the patient was qualified to the surgery. The surgery performed on 19th May 2014 confirmed earlier diagnosis found in the gastroscopy. Laparoscopic wedge resection was performed maintaining a margin of healthy gastric wall. Postoperative material with suspected gastrointestinal stromal tumor was sent for histopathological examination. Microscopic examination of the lesion identified ectopic pancreas and showed pancreatic tissue with ducts, acini and islets distributed from submucosa to serosa (fig. 1, 2). There was no histologic evidence of acute or chronic pancreatitis in the mass. The postoperative course was uneventful. Six days after the operation the patient was discharged from the hospital in good general condition.

DISCUSSION

The first case of ectopic pancreas, developed in an ileal diverticulum, was published in 1729 by Schulz, but the first histological confirmation was described by Klob in 1859 (5). Ectopic

Fig. 1. Ectopic pancreatic tissue below the gastric mucosa (H+E staining, magnification 40x)

Fig. 2. Ectopic pancreatic tissue shows acini, duct and islets of Langerhans (H+E staining, magnification 200x)
pancreas is a rare congenital disorder, defined as pancreatic tissues lying outside its normal location without vascular or anatomic communication with the normal body of the pancreas, yet possessing histological features of pancreatic acinar formation, duct development, and islets of Langerhans with independent blood supply and ductal system (7). Heterotopic pancreatic tissue has the same genetic expression with normal exocrine and endocrine function. It occurs with 0.5-13% frequency in general population. Ectopic pancreas may be present at any age, but most often is discovered in the fifth and sixth decades of life, with a male predominance.

The exact causes of ectopic pancreas are still not recognized, however, several theories have been proposed to explain the pathogenesis and occurrence of pancreatic heterotopia. One of them implicates that ectopic pancreatic tissue originates from the pancreatic buds during embryonic development of the pancreas. According to Arey and Haffer’s theory, the pancreas is formed in the 4th week of embryonic development from three primitive endodermal buds of the anterior intestine. The right ventral evagination fuses with the dorsal one and become the body, tail and upper part of the pancreas head while the lower part of the head and processus uncinatus stems from the left ventral evagination. Next, there is a rotation of the ventral part of the pancreas. During this fusion the buds are in close contact with the wall of the stomach and duodenum, allowing implantation of pancreatic germinal cells; from which histological components of the pancreas may develop. The evaginations may also remain within the intestine wall and may be carried along with the longitudinal growth of the intestine which allows the formation of heterotopic pancreatic tissue far from the normally located pancreas (8, 9).

Another theory suggests that a possible cause of ectopic pancreas is metaplasia of primitive endoderm located in the membrane surrounding antral gastric submucosa. Microscopically, these pancreatic remnants may be similar to normal pancreatic parenchyma with organized acini and ducts. On the other hand pancreatic acini are not present and the lesion is composed of disorganized pancreatic ductal structures admixed with smooth muscle, so called “adenomyoma.” In the lesions referred to as pancreatic choristoma, pancreatic acini are present in a disorganized mass of ducts and smooth muscle (10). Ectopic pancreatic tissue may be found anywhere along the gastrointestinal tract. The most common locations are: the antral part of the stomach (25-38%), duodenum (17-36%) and jejunum (15-21%) but may also be present in the mucosa of the esophagus, gall-bladder, common bile duct, spleen, mesentery and extremely rare in the peritoneum, mediastinum, lung, adrenal glands, bones, cranial cavity and fallopian tube and lymph node. Gastric lesions are found in the antrum in 85%-95%, either on the posterior or anterior wall, being more common along the greater curvature (11, 12). Moreover, lesions in the stomach may include submucosal layer (73%), muscularis (17%) and subserosal layer (10%). In the presented case the pancreatic tissue involved only the submucosal layer.

The presence of ectopic pancreas in most cases is usually asymptomatic. The occurrence of clinical symptoms depends on the size of the lesion, anatomical location and development of pathological changes similar to that of the pancreas (acute or chronic pancreatitis, cyst formation, cystic degeneration, malignant changes). Gastric ectopic pancreas is located in the antrum in 85-95% of the cases and symptoms depending upon the anatomical location (e.g., gastric outlet obstruction in a pre-pyloric ectopic pancreas) originate from the mass effect of the tumor and its size (lesions larger than 1.5 cm in diameter are more likely to cause symptoms). The most common symptom is pain, which is caused by endocrine and exocrine function of the heterotopic pancreas, and relates to the secretion of hormones and enzymes, being responsible for inflammation or chemical irritation of the involved tissues. Symptoms less often observed include nausea, vomiting, bleeding, obstructive jaundice, ulceration, occlusion and presence of neoplastic lesions (13). In our patient pain was the only symptom.

The diagnosis of ectopic pancreas is difficult as there are no specific diagnostic methods. Endoscopic examination has become useful adjunct in the evaluation of submucosal lesions. The endoscopic picture of pancreatic rest usually reveals broad-based, umbilicated, firm, slightly irregular submucosal lesion in the stomach, or elsewhere in the gastrointestinal tract. In most cases, biopsies are superficial and non-diagnostic, however, positive biopsies can establish the diagnosis. Endoscopic ultrasonography has be-
come a useful adjunct in identification of pancreatic rests, localizing in the submucosa and ranging 0.5-2 cm. The combination of endoscopic ultrasonography with fine-needle aspiration is much efficient examination to confirm the diagnosis and its sensitivity is 80-100% (14). Barium swallow study may sometimes be helpful in the diagnosis by showing nonspecific fold thickening with rounded filling defects and a typical central indentation. Its sensitivity and specificity is 87.5% and 71.4%, respectively. Contrast-enhanced computed tomography scan may also help in the diagnosis and provide information regarding surgical resection (15).

The intraoperative diagnosis may be sometimes difficult due to the gross similarity of other submucosal changes such as: especially GIST (as it was in our patient) and GALT, leiomyoma, myoma, lipoma, fibroma, lymphoma and carcinoid tumors. It should be emphasized that the final confirmation of the diagnosis of ectopic pancreas is possible only on the basis of histopathological examination of biopsy or resectable material.

In conclusion, although ectopic pancreas is a rare entity, it should always be considered in the differential diagnosis of extramucosal gastric lesions.

REFERENCES