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Case Report

HETEROTOPIC PANCREAS OF THE GIT: TWO CASE STUDIES AND LITERATURE REVIEW

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ABSTRACT

Heterotopic pancreas is pancreatic tissue that is found outside its anatomical location. It is asymptomatic and is detected accidently. The risk of malignant transformation is low. It can be found in all organs of the GIT. **Purpose:** We present two clinical cases of heterotopic pancreas. The first patient is a 61-year-old man who presented with pain in the epigastrium, the upper right quadrant, weight loss and history of gallstone disease, gastritis and peptic ulcer. The second case is a 79-year-old woman, hospitalized with complaints of abdominal pain, nausea, and vomiting. **Results:** The patients were operated. The intraoperative finding was chronically inflamed gallbladder and tumour formation in the pyloro-antral part of the stomach. Cholecystectomy and resection of the tumour formation were performed, followed by pyloroplasty in first patient. Internal incarceration of the ileum with necrosis and Meckel's diverticulum was established. Resection of the small intestine was performed with anastomosis and resection of the Meckel's diverticulum in the second case. Histological examination of both specimens revealed ectopic pancreatic tissue in the stomach and in the Meckel's diverticulum. **Conclusions:** Heterotopic pancreas is a rare pathology. Surgical removal of the tumour is the only radical treatment.

Key words: heterotopic pancreas, ectopic pancreas, carcinoid syndrome, ultrasonography, gastroscopy, Mekel's diverticulum.

INTRODUCTION

Heterotopic pancreas is pancreatic tissue located outside the normal anatomical location of the pancreas. It is rare and can be found anywhere in the GIT. The stomach and small intestine are the most commonly affected organs. Clinically it can manifest itself with pain, bleeding, inflammatory processes and obstruction. It is discovered accidently. Most cases are asymptomatic and are diagnosed post mortem. (1)

The heterotopic pancreas was first described in 1727, discovered in a diverticulum of the ileum. Histologically it was proved by Klob in 1859 (2). It is rare and is defined as "extrapancreatic tissue", which has no anatomical or vascular connection with the pancreas and is a congenital pathology. Frequency of autopsy material varies from 0.11 - 0.21% and ratio men - women is 3:1 (3) There are two theories about its occurrence: "migration" of pancreatic tissue during embryonic development during the rotation of the anterior part of the primary intestine, when fragments of the pancreas separate and migrate to various organs of the GIT or penetration of immature gastric mucosa into the submucosa, with subsequent pancreatic metaplasia (4).

In the wall of Meckel's diverticulum are found colon, stomach and duodenal glands, as well as ectopic pancreatic tissue, the frequency of the latter varies from 5 to 16% of cases. (5, 6)

Case 1

A 61-year-old man was admitted to A&E with the following complaints: heaviness and pain in the epigastrium and right hypochondrium, accompanied by nausea, no vomiting, loss of appetite and weight loss - about 20 kg in the last few months. A few months earlier, he had been hospitalized in another hospital with the same

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complaints. He was treated conservatively. Upper endoscopy was performed - no pathological processes were detected. On examination, the patient's abdomen was at the level of the chest, respiratorily active, soft, palpably painful in the epigastrium and right hypochondrium, with physiological peristalsis. From the laboratory tests – elevated leukocytes were observed - $12.98x10^9$. Ultrasound examination of the abdomen and computed tomography established calculosis of the gallbladder and bilateral cysts of the kidneys. The patient underwent surgery. Intraoperatively, a chronic inflammation of the gall bladder which was filled with stones, nondilated extrahepatic bile ducts and a tumour formation located in the pyloro-antral part of the stomach with approximately 4/5 cm dimensions were found. Pylorotomy. The described tumour formation was dissected and material for frozen section was taken - data for ectopic pancreatic tissue in the stomach**.** Tumour resection was performed with pyloroplasty. The specimen is shown on **Figures 1 and 2.**

Figure 1. Heterotopic pancreas

Figure 2. Heterotopic pancreas

Normal postoperative period. No complaints at follow-up. Histological examination of the specimen revealed an encapsulated formation of serous acini of the pancreas and single islets of Langerhans among cystically dilated and deformed pancreatic ducts, without cellular atypism of the epithelium. Conclusion: tumorous heterotopic pancreatic tissue in the stomach wall (choristoma) at the background of chronic antral gastritis. **Figures 3 and 4** show pancreatic tissue in the gastric wall magnification HE x 100

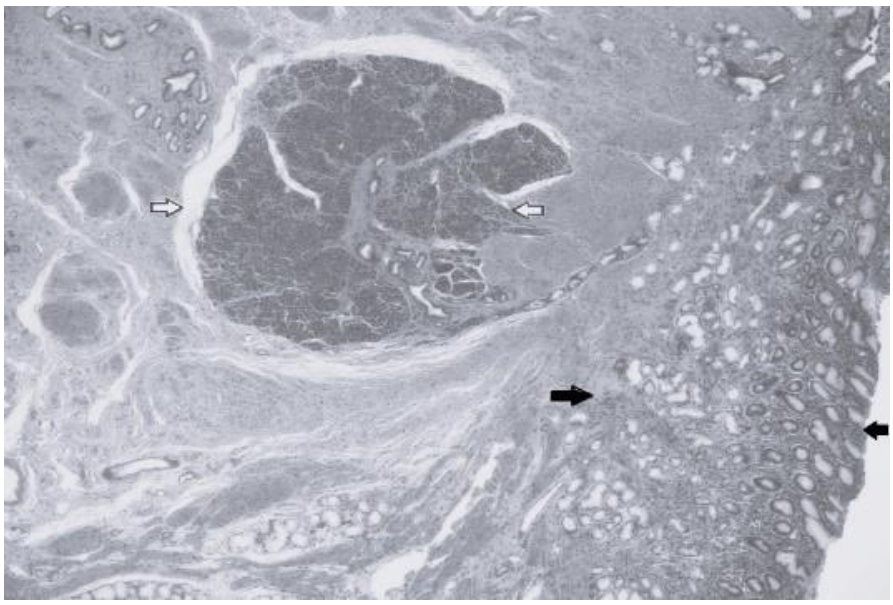


Figure 3. Pancreatic tissue in muscular layer of the gastric wall - white arrows

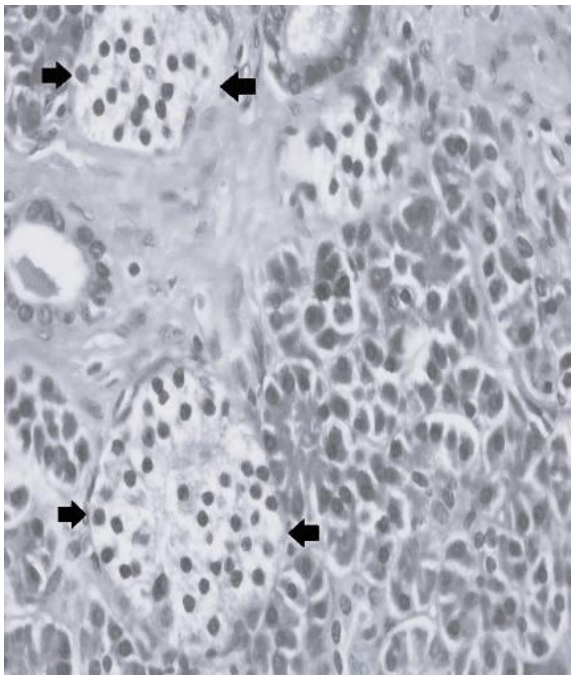


Figure 4. Langerhans islets Black arrows – antral mucosa

Case 2

A 79-year-old woman was admitted with complaints of abdominal pain, nausea and repeated vomiting. The physical examination revealed severe general condition, pain in the hypogastrium, bloating of the abdomen, lack of peristalsis. From the laboratory tests - increase of leukocytes up to 13.1 x10^9, increased urea - 29.7 mmol / l and creatinine - 455.0 mcmol / l. CT- dilated small bowel, internal incarceration of the distal ileum and Meckel's diverticulum (**Figures 5 and 6).**

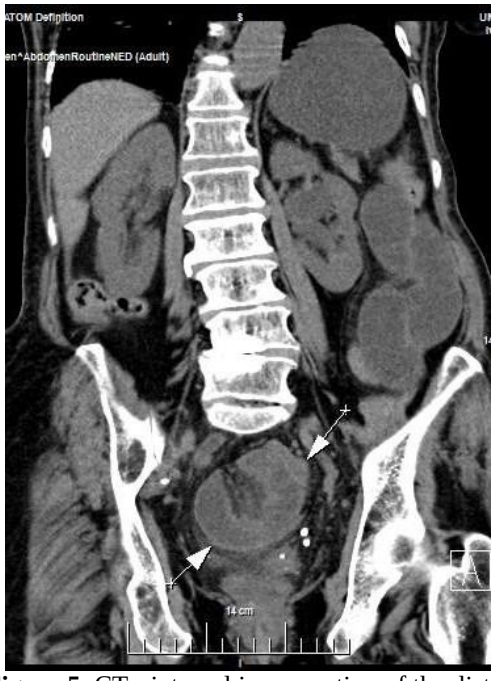
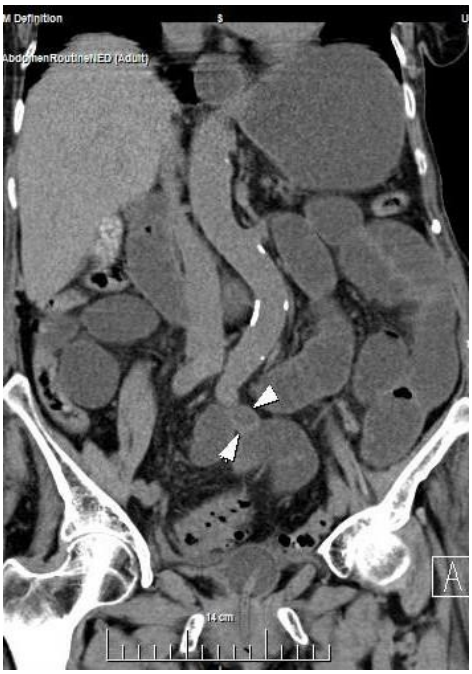


Figure 5. CT - internal incarceration of the distal ileum



Figute 6. Meckel's diverticulum.

Intraoperatively, a dilated ileum was found, starting about 30-40 cm from the ileocecal valve, where an internal incarceration with necrosis of the involved area and Meckel's diverticulum was established **(Figures 7 and 8).** Resection of the affected area was performed with subsequent anastomosis and resection of the Meckel diverticulum.

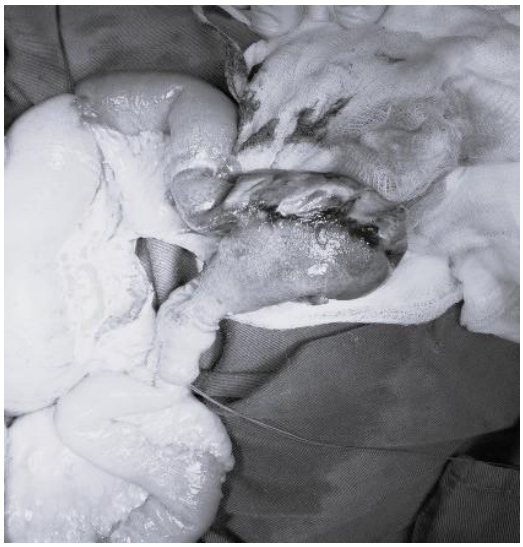


Figure 7. Necrotic terminal ileum

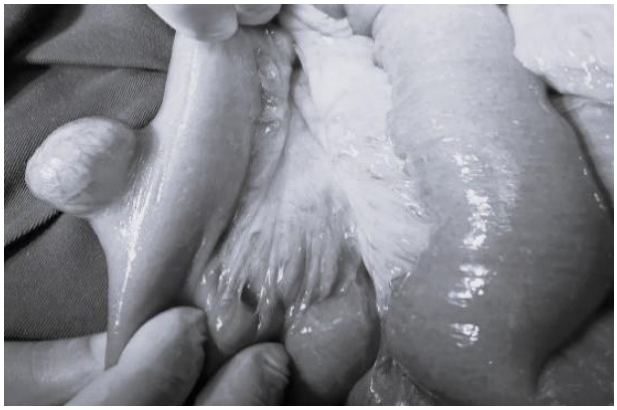
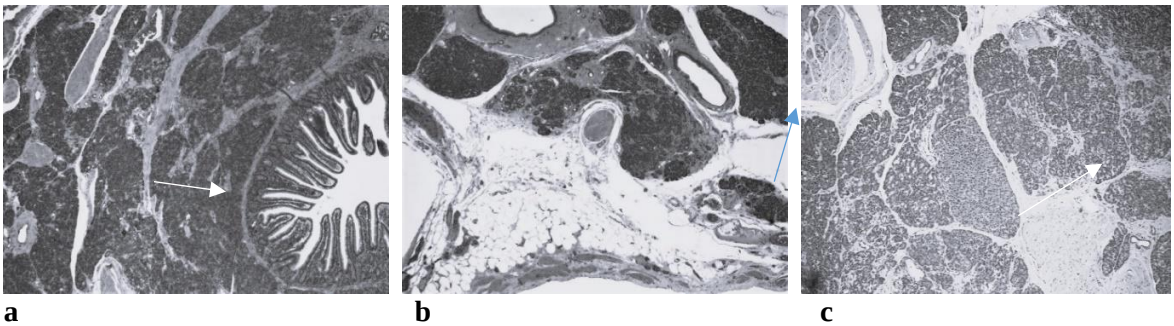


Figure 8. Meckel diverticulum

The patients was discharged in good general condition. No complaints at follow-up. Histological finding of Meckel's diverticulum showed pancreatic tissue involving submucosis, muscularis propria and subserosis of its wall. **(Figure 9 a, b and c).**



a **b** **c**

Figure 9. Ectopic pancreatic tissue in submucosis, muscularis propria and subserosis.

DISCUSSION

The heterotopic pancreas was first described in 1727 from Schultz in the ileal diverticulum. In adult patients it occurs mainly in men between the fourth and sixth decade of their life. In 1909, Heinrich created the first classification of the heterotopic pancreas, into three main types, which underwent several modifications, while in 1973 Gaspar-Fuentes completed it so that it acquired the following types:

Type I - typical pancreatic tissue with acini, ducts and islet cells that occur in the normal pancreas.
Type II - only pancreatic ducts in different variants
Type III - acinar tissue only (exocrine pancreas)
Type IV - islet cells (endocrine pancreas) only (7).

The non-specific symptoms are explained by the secretion of enzymes caused by the ectopic pancreas, which determines the occurrence of spasms, chemical irritation of the surrounding tissue and inflammatory processes (8). The formation of cysts in the ectopic tissue is a result of the inability of the exocrine secretion to drain into the cavity of the organ (retention cysts). The formation of pseudocysts is unusual (9). Malignant transformation of the heterotopic pancreas is rare - 0,7% - 1,8% of cases (10).

Heterotopic pancreas can be found in all parts of the GIT. The most common sites are the stomach-25.5%, duodenum-27.7%, jejunum-15.9%, Meckel's diverticulum-5.3% and ileum-2.8%. (11) Atypical are the colon, spleen or liver. (12, 13)

Park et al. in 2005 (Mayo Clinic) reports the experience of treating 1476 patients with Meckel's diverticulum, from whom 180 were operated. In 43%, heterotopic pancreatic tissue was detected. (14) According to the localization of heterotopic pancreatic tissue in the wall of Meckel's diverticulum - 54% - in submucosis, 23% - submucosis and muscularis propria, 8% - muscularis propria, 11% - subserosis and only in 4% of all cases the whole diverticulum wall was affected. (4)

The clinical symptoms of pancreatic ectopic tissue are determined by the location, size and / or secretion of pancreatic enzymes, which are expressed in local inflammation. When hormones are secreted, they can affect the function of the endocrine glands. Lesions smaller than 15 mm are asymptomatic, until they inflammation or obstruction appears. Usually, they are discovered accidently. Last but not least, it is possible for the ectopic pancreas to become malignant - turning into an adenocarcinoma or a neuroendocrine tumour (15). According to Osanai et al., the classification of carcinomas originating from ectopic pancreatic tissue is determined by the following criteria:

- The tumor should be located in or near the aberrant pancreatic tissue.
- Clearly demarcated area between pancreatic structures and carcinoma tissue (tubular dysplasia and, or carcinoma in situ).
- Non-malignant ectopic tissue must contain fully developed acini or ductal structures. (16).

In our study both of the patients had a different clinical symptomatics which was not indicative of ectopic pancreas – the first had symptoms of cholelithiasis and pyloric stenosis, while the second had internal incarceration of the small intestine. This confirms the statement that in lesions bigger than 15mm, the clinical symptoms are dictated by signs of inflammation and obstruction.

In children, the clinical picture is different. It is manifested by obstructive syndrome, intussusception, as well as some congenital diseases such as granulosa pancreas, esophageal atresia, Meckel's diverticulum, malrotation, choledochal cysts and extrahepatic biliary atresia (17).

Imaging methods, such as computed tomography, magnetic resonance imaging and endoscopic ultrasound with fine-needle biopsy are needed for choosing the best surgical intervention. (18)

The sensitivity of this study varies from 80 to 100%. (19, 20). Contrast examination of the upper GIT may show a typical image of a round defect with a centrally located recess. Nikolau et al. reported a sensitivity of 87.5% and a specificity of 71%. (21)

CONCLUSIONS

Heterotopic pancreas is a rare pathology. Surgical removal of the tumour is the only radical treatment. Treatment of the symptomatic Meckel's diverticulum with ectopic pancreas is a definitive surgery, including diverticulectomy, partial and segmental resection, performed by open or laparoscopic approach. Intraoperative biopsy distinguishes it from malignant diseases of the gastrointestinal tract.

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