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Case Report

INTESTINAL OBSTRUCTION AS A RESULT OF PANCREATIC ECTOPY IN JEJUNUM: A CASE REPORT AND LITERATURE REVIEW

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SUMMARY – Pancreatic heterotopia is a relatively rare condition that implies pancreatic tissue anatomically separated from the main gland, showing no vascular or ductal continuity. It is known to occur in many sites of gastrointestinal tract, with stomach being the most common one. This case is of great interest because ectopic pancreatic tissue in jejunal wall presented as bowel obstruction. The aim of this article is to provide a review of clinical, histopathologic and immunohistochemical features of ectopic pancreatic tissue in jejunum, emphasizing the possible diagnostic pitfalls in gastrointestinal tract, especially in its upper parts. Knowing the most common locations, imaging features and histopathologic criteria, the diagnosis of pancreatic heterotopia could be successfully set preoperatively. Associated complications are various and sometimes difficult to handle, so accurate and precise diagnosis, as well as surgical resection, is often needed.

Key words: *Ectopic; Pancreas; Heterotopia; Jejunum*

Introduction

One of the relatively rare developmental anomalies of gastrointestinal (GI) tract is ectopic pancreas, which represents pancreatic tissue anatomically separated from the main gland[1]. Also known as aberrant or accessory pancreas, choristoma or adenomyoma[2], the incidence of this heterotopic tissue is difficult to determine as most patients are asymptomatic. This type of displacement usually occurs within different sites in the upper GI tract, with the stomach being the most common one. Other, more distant sites as spleen, appendix, omentum, gallbladder, fallopian tubes, lungs or mediastinum have been reported as case reports due to rare involvement of the listed sites[3]. Ectopic pancreas has the same genotypic and phenotypic characteristics as the orthotopic pancreatic tissue, with no vascular or ductal continuity[4]. Growing outside pancreatic frame, this congenital disorder has clinical significance due to its asymptomatic appearance in most cases. In the majority of patients, this anomaly is found upon undergoing routine imaging examination, endoscopy or surgery for other reasons[3]. Furthermore, autopsy findings in general population show an incidence ranging from 0.5% to 13.7%[5]. As

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this ectopy can occur in all ages and usually measures a few millimeters to few centimeters without causing any symptoms, it represents a diagnostic challenge as computed tomography (CT) has little diagnostic value and endoscopy is often inconclusive[4]. Depending on its localization, the existence of ectopic tissue could be complicated most likely by inflammation, obstruction, bleeding or malignant transformation[2]. Definitive diagnosis depends on postoperative histopathologic examination of surgical specimen.

Heterotopic pancreas as a jejunal mass lesion is a rare incidental finding. We report a case in which hematoxylin and eosin (H&E), as well as immunohistochemical staining showed pancreatic ectopy in jejunal wall, which extended through the mucosal and submucosal layer.

Case Report

The excised specimen from a surgically treated patient presenting with ileus was sent for histopathologic analysis. On gross examination, jejunal mass was described as irregular bowel tissue fragment measuring 2.8x0.8x0.5 cm, with partly present serosa at the surface measuring 1x0.9 cm. The ill-defined mass was white with interspersed yellowish areas and firm consistency. Careful serial sectioning showed relatively homogeneous appearance with areas of cystic change measuring 1.2x1 cm. After fixation in formaldehyde, grossly described nodules with surrounding jejunal wall were processed for microscopic examination as 2 tissue specimens stained with hematoxylin and eosin (H&E). Histopathologic examination of the lesion revealed well-defined tissue in jejunal mucosa, submucosa and muscularis propria (Fig. 1). The tissue contained dilated and cyst-like spaces and tubular structures with mucous acini around these structures. Cystic and tubular structures were lined with regular columnar, mucus-producing epithelium without any atypia. Smooth muscle cells arranged in bundles were present around cystic and tubular spaces (Fig. 2). Immunohistochemical analysis with CA 19-9 (Dako, Glostrup, Denmark) showed strong positivity of epithelial cells, which pointed to the epithelium of pancreaticobiliary ducts (Fig. 3). The elements of endocrine and exocrine pancreas were not present in histologic sections.

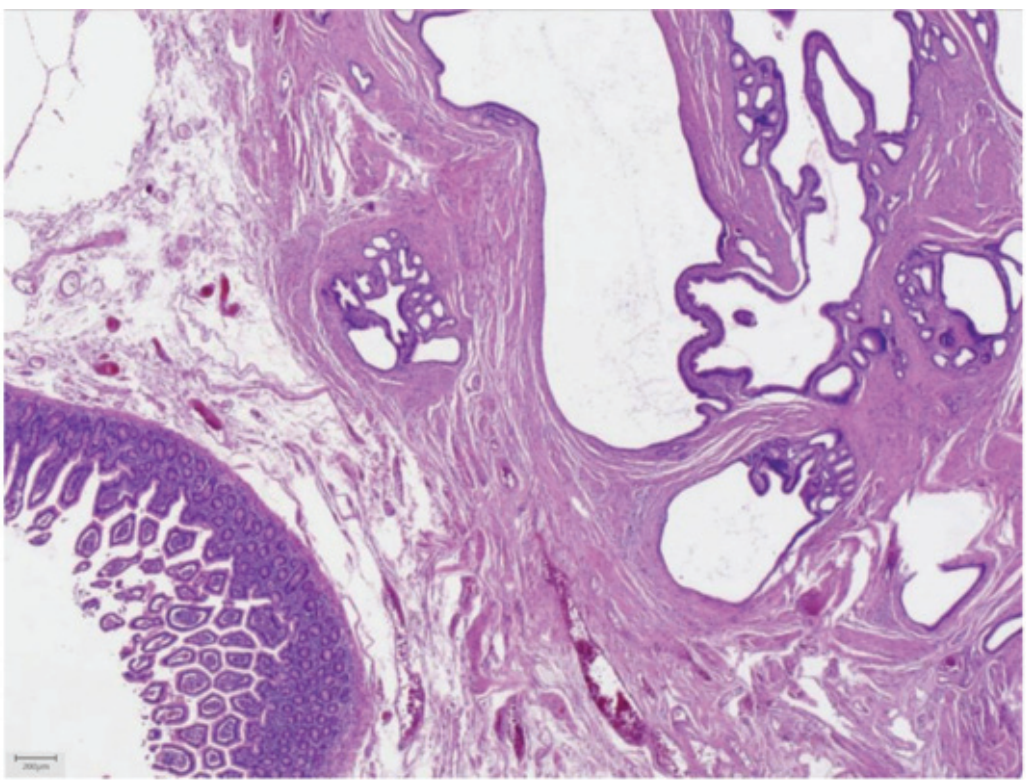

Fig. 1. Histologic finding of well-defined heterotopic pancreatic tissue in jejunal wall (H&E, x10).

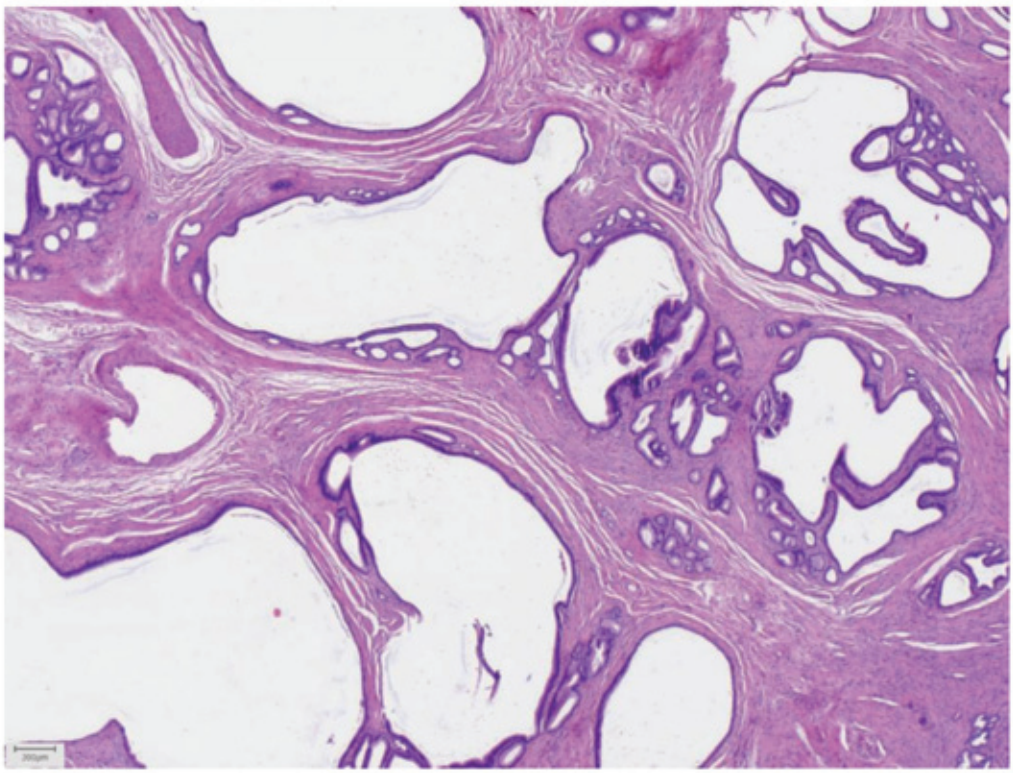

Fig. 2. Histologic finding of cyst-like spaces and tubular structures surrounded with mucous acini (H&E, x10).

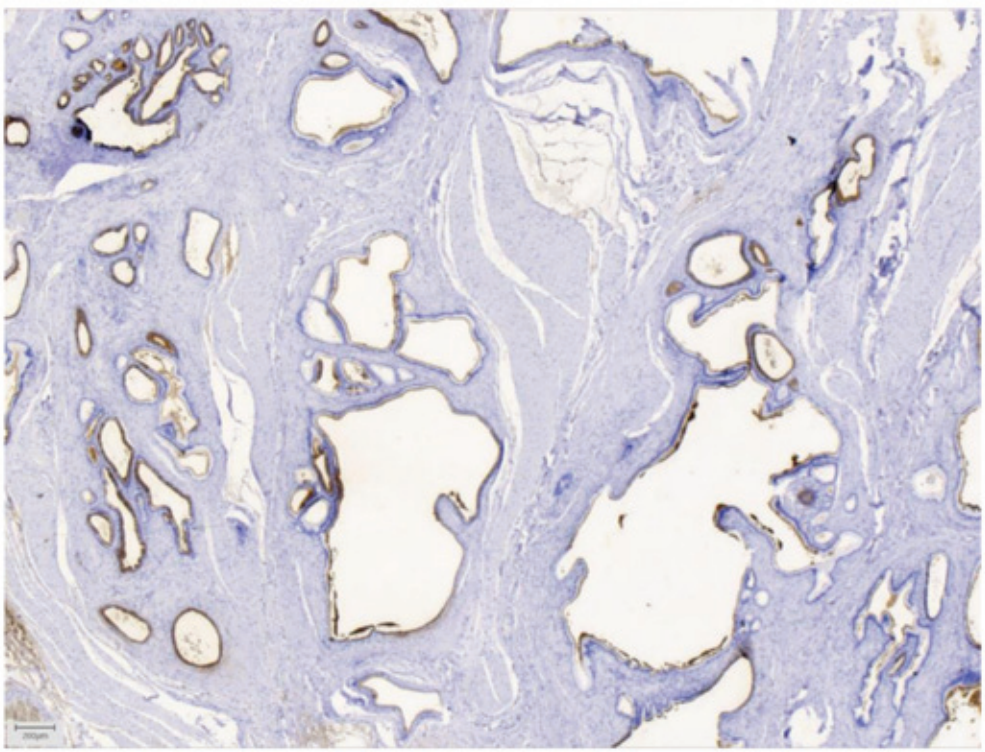

Fig. 3. Immunohistochemical analysis with CA 19-9 showing strong positivity of epithelial cells (x10).

Based on histologic classification, these characteristics corresponded to type II pancreatic ectopy (heterotopia of canalicular type), and due to CA 19-9 immunohistochemical positivity of the epithelium, definitive diagnosis of ectopic pancreatic tissue in jejunal wall was reported.

Discussion

There are two currently accepted theories trying to explain pancreatic ectopy. Pancreatic tissue develops from endodermal tissue invaginations in primitive duodenum. According to the misplacement theory, the elements of primitive pancreatic tissue separate during foregut rotation and form independent and mature pancreatic tissue throughout entire GI tract. In the metaplasia theory, pancreatic ectopy arises from endodermal pancreatic metaplasia and migrates to the submucosal layer during the process of embryogenesis[3]. Histopathologic classification of pancreatic heterotopia into four types by Heinrich and later by Gaspar-Fuentes is helpful in the evaluation of pathologic differences of this anomaly. Type I is predominated by typical pancreatic tissue elements, including acini, ducts and islet cells; type II is a canalicular variant with pancreatic ducts; type III consists of acinar tissue only (exocrine pancreas, adenomyoma); and type IV is composed of islet cells only (endocrine pancreas)[1]. In our case, the microscopic characteristics matched type II. However, this classification does not have an effect on clinical appearance or treatment of the patient.

Symptomatic cases of pancreatic ectopy can develop various clinical features depending on the location, size and association with other disorders. Abdominal pain, nausea, vomiting, weight loss, melena, and consequently anemia and/or anorexia could be explained by the endocrine and exocrine function of heterotopic pancreatic tissue that secretes enzymes and hormones related to chemical irritation, inflammation and spasm[6,7]. Local spasm, dysmotility and persistent vomiting could be expected if muscular layer is involved[8]. These examples of clinical presentation are possible if any mass is present in the GI tract, including ectopic pancreas, which leads to a more challenging differential diagnosis.

Complications that are specific to pancreatic tissue include pancreatitis, formation of pseudocysts, benign and malignant neoplasms, GI bleeding and intussusception[3]. Acute pancreatitis is rather discernable at histopathologic analysis, with elevated enzyme levels in laboratory results, and thickening of the involved bowel wall with surrounding inflammation on imaging[9]. Pseudocysts can form either secondary to pancreatitis or as a result of duct obstruction[3]. Malignant transformation in ectopic pancreas is uncommon and it can arise from ductal components or acini[10]. Most neoplasms of the ectopic pancreatic tissue occur in the stomach, followed by stenotic or ulcerated appearance and with adenocarcinoma being the most frequent histologic type. In about 30 documented cases of malignant transformation of ectopic pancreas, other types of neoplasia were reported, arising mostly within type I heterotopia[11]. Given the rarity of these conditions, these malignant tumors could be an important pitfall in differential diagnosis of GI tract tumors[12,13].

Different diagnostic tests are used to identify pancreatic heterotopia in various sites. These tests are usually done for the purpose of more common causes of abdominal symptoms, thus the finding of ectopic pancreatic tissue is usually incidental. CT and endoscopic ultrasound are considered to be standard tests for detecting submucosal lesions, but often unreliable[4]. Thus, surgical resection is desirable so the clinically relevant diseases such as GI stromal or neuroendocrine tumors could be distinguished from pancreatic heterotopia[2].

We conclude that this rare condition remains a diagnostic challenge due to its asymptomatic and unspecific clinical appearance, many possible associated complications, and wide differential diagnosis in GI tract. This article could help clinicians better recognize cases in which pancreatic heterotopia and its complications are considered to be diagnostic possibilities.

References

1. Eldruki S, Lawgaly SA. Heterotopic pancreas in the jejunum: a case report and literature review. Libyan J Med Sci. 2019;3(2):71-2. DOI: 10.4103/LJMS.LJMS_70_18.
2. Rezvani M, Menias C, Sandrasegaran K, Olpin JD, Elsayes KM, Shaaban AM. Heterotopic pancreas: histopathologic features, imaging findings, and complications. Radiographics. 2017;37(2):484-99. DOI: 10.1148/rg.2017160091.
3. Kim DU, Lubner MG, Mellnick VM, Joshi G, Pickhardt PJ. Heterotopic pancreatic rests: imaging features, complications, and unifying concepts. Abdom Radiol. 2016;42(1):216-25. DOI: 10.1007/s00261-016-0874-9.
4. Swarnkar M, Pandey P. Heterotopic subserosal pancreatic tissue in jejunum. Formos J Surg. 2018;51(4):167-70. DOI: 10.4103/fjs.fjs_88_17.
5. So H, Cross TJ, Zonta M. A case report of incidental ectopic pancreatic tissue during laparoscopic appendicectomy. Int J Surg Case Rep. 2018;45:77-8. DOI: 10.1016/j.ijscr.2018.03.023.
6. Eisenberger CF, Gocht A, Knoefel WT, Busch CB, Peiper M, Kutup A, *et al*. Heterotopic pancreas - clinical presentation and pathology with review of the literature. Hepatogastroenterology. 2004;51(57):854-8. PMID: 15143933.
7. Rimal D, Thapa SR, Munasinghe N, Chitre VV. Symptomatic gastric heterotopic pancreas: clinical presentation and review of the literature. Int J Surg. 2008;6(6):52-4. DOI: 10.1016/j.ijsu.2007.02.002.
8. Jyothi AR, Sharmila PS, Rajaram T. Pancreatic heterotopia in jejunum presenting as small bowel obstruction. Med Inn. 2013;2(1):101-4.
9. Kim DW, Kim JH, Park SH, Lee JS, Hong S, Kim M, *et al*. Heterotopic pancreas of the jejunum: associations between CT and pathology features. Abdom Imaging. 2014;40(1):38-45. DOI: 10.1007/s00261-014-0177-y.
10. Yamaoka Y, Yamaguchi T, Kinugasa Y, Shiomi A, Kagawa H, Yamakawa Y, *et al*. Adenocarcinoma arising from jejunal ectopic pancreas mimicking peritoneal metastasis from colon cancer: a case report and literature review. Surg Case Reports. 2015;1:114. DOI: 10.1186/s40792-015-0118-1.
11. Cazacu IM, Alexandra A, Chavez L, Nogueras GM, Adrian G, Bhutani MS. Malignant transformation of ectopic pancreas. Dig Dis Sci. 2018;64(3):655-68. DOI: 10.1007/s10620-018-5366-z.
12. Goodarzi M, Rashid A, Maru D. Invasive ductal adenocarcinoma arising from pancreatic heterotopia in rectum: case report and review of literature. Hum Pathol. 2010;41(12):1809-13. DOI: 10.1016/j.humpath.2010.06.005.
13. Khan MA, Maken RN, Nisar H, Fatima I, Khan IU, Masood M, *et al*. The role of preoperative carcinoembryonic antigen in recurrence of resectable colorectal carcinoma. Acta Clin Croat. 2020;59:216-22. DOI: 10.20471/acc.2020.59.02.03.

Sažetak

CRIJEVNA OPSTRUKCIJA KAO REZULTAT PANKREATIČNE EKTOPIJE U JEJUNUMU: PRIKAZ SLUČAJA I PREGLED LITERATURE

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Pankreatična heterotopija je relativno rijedak nalaz koji podrazumijeva prisustvo pankreatičnog tkiva koje je anatomski odvojeno od glavne žlijezde i ne pokazuje vaskularni i duktalni kontinuitet s njom. Može se javiti na različitim lokalizacijama u gastrointestinalnom traktu, pri čemu je želudac najučestalija lokalizacija. Ovo je slučaj od većeg dijagnostičko-terapijskog interesa, jer se pankreatična ektopija u zidu jejunuma prezentirala kao crijevna opstrukcija. Cilj rada je pregled kliničkih, histopatoloških i imunohistokemijskih osobina ektopičnog pankreatičnog tkiva u jejunumu, čime se naglašavaju potencijalne diferencijalno dijagnostičke dileme u gastrointestinalnom traktu, osobito u njegovim gornjim segmentima. U skladu s literaturno poznatim lokalizacijama, obilježjima snimaka i histopatološkim kriterijima, dijagnoza pankreatične heterotopije se može uspješno postaviti prijeoperacijski. Udružene komplikacije su različite i ponekad teške za saniranje pa je potrebna ažurna i precizna multidisciplinarna dijagnostika, kao i operativno liječenje.

Ključne riječi: *Ektopično; Gušterača; Heterotopija; Jejunum*