

Intraductal Papillary Mucinous Neoplasia in Ectopic Pancreas Located in the Jejunum: Case Report

Jejunumda Yerleşmiş Ektopik Pankreas İçinde İntraduktal Papiller Müsinöz Neoplazm

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ABSTRACT Ectopic pancreas is a commonly seen congenital anomaly in gastrointestinal system, but jejunal pancreatic heterotopia is seen rarely. Although every pathological change that occurs in the pancreas can be seen in its heterotopic counterpart, neoplasia is an unusual complication. Intraductal papillary mucinous neoplasms (IPMNs) are tumors originate from ductal epithelial cells of the pancreas that constitute the main pancreatic duct or its major side branches and have a low incidence. Only two cases regarding the IPMN in heterotopic pancreas have been reported previously, but none of them were located in the jejunum. An IPMN in an ectopic pancreas tissue located in the jejunum in a 62-year-old male patient who presented with ileus is presented. It is important to recognize a tumor arising in heterotopic pancreas in order to prevent its misinterpretation as metastatic tumor or direct invasion of another tumor. To the best of our knowledge, this is the first reported IPMN case occurring in ectopic pancreas tissue located in the jejunum.

Key Words: Carcinoma, pancreatic ductal; jejunum

ÖZET Ektopik pankreas, gastrointestinal sistemin sık görülen bir konjenital anomalisidir ancak jejunal pankreatik heterotopiye nadiren rastlanır. Pankreasta görülen tüm patolojilerin heterotopik dokularda da görülebilmemesine karşın, neoplazi az rastlanan bir komplikasyondur. İntraduktal papiller müsinöz neoplaziler (IPMNler) ana pankreatik kanal ya da bu kanalın ana yan dallarını oluşturan pankreasın duktal epitel hücrelerinden köken alırlar ve düşük bir görülme sıklığına sahiptirler. Bundan önce heterotopik pankreas dokusunda IPMN ile ilgili sadece iki vaka bildirilmiştir ve bunların hiçbiri jejunumda yerleşmemiştir. Altmış iki yaşında bir erkek hastada ileus ile ortaya çıkan, jejunumda yerleşmiş ektopik pankreas dokusu içinde gelişmiş bir IPMN vakası sunacağız. Heterotopik pankreas içinde gelişen bir tümörün tanınması metastatik tümör ya da bir başka tümörün direkt invazyonu gibi yanlış yorumlamaları önlemek açısından önemlidir. Bildiğimiz kadarıyla, bu vaka jejunumda yerleşmiş ektopik pankreas dokusu içinde gelişen bildirilmiş ilk IPMN vakasıdır.

Anahtar Kelimeler: Karsinoma, pankreatik duktal; jejunum

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Ectopic pancreas, which is called as heterotopic or aberrant pancreas as well, is a common congenital anomaly defined as pancreatic tissue found outside its normal anatomic location. It is commonly located in the stomach and duodenum. Other rare sites are Meckel's diverticulum, biliary tract, mesocolon, spleen, liver, mesentery and lymph nodes.¹ Jejunal location and neoplastic transformation of ectopic pancreatic tissue have rarely been reported.¹⁻³

Most of the patients in whom the tissue is usually found in the stomach or duodenum are usually asymptomatic. It is usually diagnosed incidentally at laparotomy, executed for other disorders.^{1,3}

Intraductal papillary mucinous neoplasm (IPMN) has been recently described in the category of pancreatic exocrine tumors. It produces mucin and grows prominently in intraductal papillary pattern.^{4,5} This neoplasm has been described to occur within heterotopic pancreas in the Meckel's diverticulum⁶ and gastric wall⁷ in the case reports.

In this article, we report IPMN in an ectopic pancreas tissue located in the jejunum in a 62-year-old male patient who presented with ileus.

CASE REPORT

A 62-year-old male patient had admitted with increasing abdominal pain for two days. The abdomen was distended, tender and hard on examination and the bowel sounds were hyperactive. The spiral computed tomographic examination of abdomen revealed the distention in the stomach and through the small intestines, an air density without contrast, and an image sized 5 x 3 cm compatible with bezoars in the stomach and ileum. Pancreas was normal. An exploratory laparotomy was performed for ileus. Intraoperative findings were multiple adhesions among falciform ligament, stomach and anterior abdominal wall subsequent to prior gastric operation, as well as bezoars, cholelithiasis, incidental appendicitis and a subserosal jejunal, 2 x 3 cm sized mass located 40 cm away from Trietz ligament. The mass was not related to the lumen. Intestinal bezoar was removed manually and the one in the stomach was removed through gastrotomy. Cholecystectomy, appendectomy and intestinal wedge resection were done. No complications occurred following the operation.

PATHOLOGICAL FINDINGS

In the pathological examination of 5 x 2.5 x 1 cm small bowel segment, a tan-colored, firm, 1.2 x 1 x 0.7 cm nodule was seen within serosa and the mucosa was intact. The cut surface of the mass had solid and cystic areas filled with mucinous material.

The cysts were sized between 0.3 to 0.5 cm in diameter without any necrosis or hemorrhage.

Microscopic examination of the jejunal nodule revealed pancreatic ducts and acini embedded within the small bowel, involving the submucosa, muscularis propria and serosa (Figure 1 and 2). Langerhans' islets were not detected. Ducts were cystically dilated and lined by mucinous columnar epithelial cells. Areas of complex papillary structure associated with nuclear stratification and cytological atypia characterized by nuclear enlargement, elongation and hyperchromasia were seen (Figure 3-5). No mitoses were seen. There was no evidence of severe dysplasia, carcinoma in situ and/or infiltrative growth pattern. Our case was classified as borderline IPMN with those findings.

DISCUSSION

The pathogenesis of pancreatic heterotopia is not known clearly. It is thought to arise during rotation of the foregut when fragments of pancreas got separated and developed into mature elements, or due to pancreatic metaplasia of endodermal tissues that end up in the submucosa during embryonic life. It is defined as pancreatic tissue that is not normally situated, has no relation with the normal pancreas, and possesses its own duct system and vascular supply.¹ Microscopically, acini and ducts always exist, whereas islets of Langerhans' are only found in one third of patients.^{1,6} In our case there was no Langerhans' islet. Pancreatic heterotopia is found between 2 to 15% of all autopsies. The most frequent sites of ectopic pancreas are gastric antrum (30%), duodenum (30%), jejunum (20%) and Meckel's diverticulum (5%). Unusual locations are colon, spleen, liver, biliary tract, mesentery and lymph nodes.¹ It is usually an incidental finding, either at the time of laparotomy for another disease or during radiographic or endoscopic examination of the upper gut, but may present with peptic ulceration or intestinal obstruction.¹ Ectopic pancreas may undergo any one of the changes that may be seen in normal pancreas such as acute and chronic pancreatitis, pseudocyst formation, cystic dystrophy and hemorrhage. Neoplastic transformation has also been described, but is rare. There are

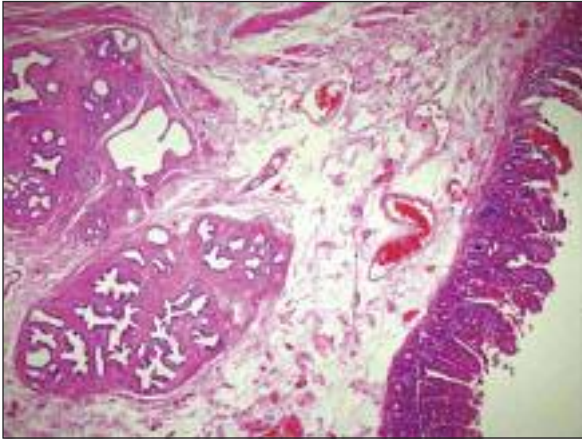


FIGURE 1: Low power view of ectopic pancreas in the submucosa of the jejunum (H&E, x40).

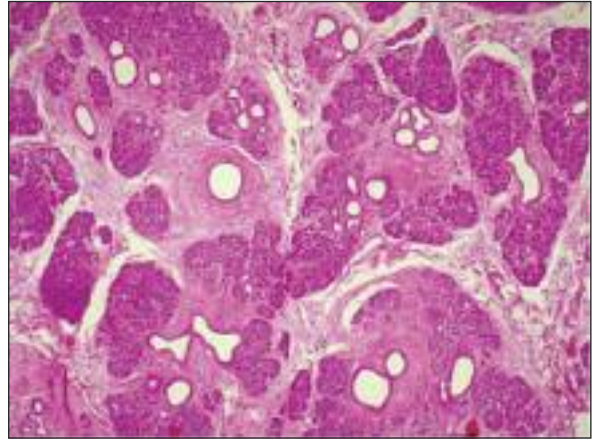


FIGURE 2: Ectopic pancreas with acini and ducts (H&E, x100).

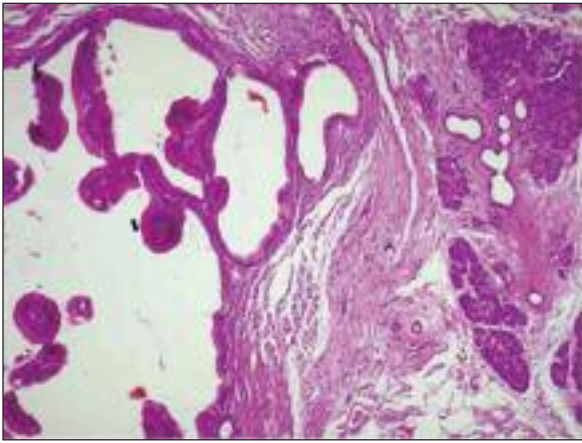


FIGURE 3: Ectopic pancreas with acini and ducts adjacent to intraductal papillary mucinous neoplasm (H&E, x100).

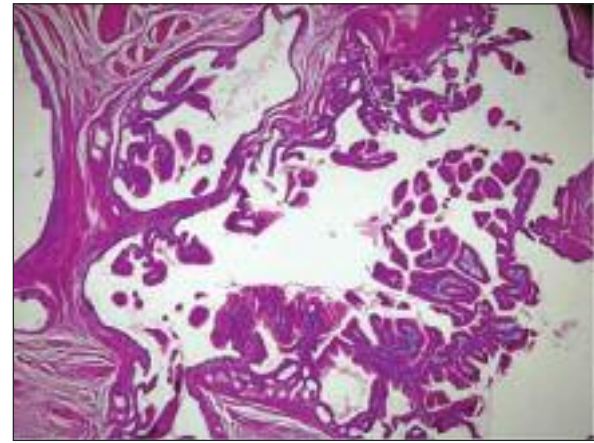


FIGURE 4: Areas of tumor with complex papillary growth pattern (H&E, x100).

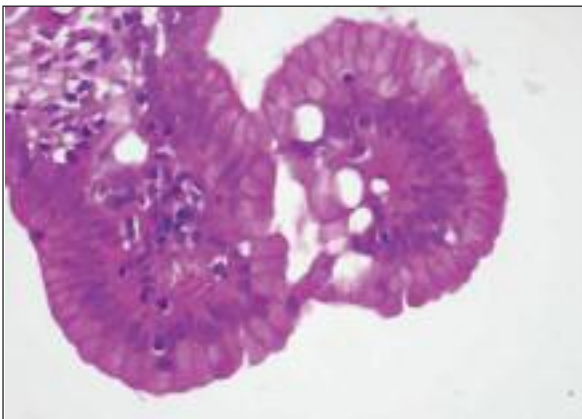


FIGURE 5: Complex papillary areas showing stratification, elongation and hyperchromatism of nuclei (H&E, x400).

only two reported cases of IPMN arising from in heterotopic pancreas^{6,7} and most of the reported cases of neoplastic transformation are ductal adenocarcinomas.^{1,2,8,9}

To document the neoplastic transformation of ectopic pancreatic tissue, the following criteria have been proposed: 1- The tumor should be found within or close to the ectopic pancreatic tissue, 2- Direct transition between pancreatic structures and tumor must be observed, 3- The non-neoplastic pancreatic tissue must comprise at least fully developed acini and ductal structures.⁹ Our case matched all of these three criteria.

Pancreas may be normal or may have similar pathologies that appear in the ectopic pancreas. In our case, the pancreas had no pathology in the computed tomography (CT) images.

IPMNs are a recently defined subset of pancreatic neoplasms, comprising approximately 0.5–9.8% of pancreatic exocrine tumors. Approximately one-third of IPMNs have an associated invasive carcinoma.^{4,5} IPMNs are relatively uncommon tumors of pancreatic ductal epithelial cells that line the main pancreatic duct or its major side branches.^{4,6} These tumors are primarily characterized by cystic dilatation of pancreatic ducts with variable degrees of mucin production, epithelial proliferation, papillary formations and cytological atypia.^{4,6}

IPMNs display a spectrum of cytoarchitectural atypia and have been divided into adenoma (low grade dysplasia), borderline (moderate dysplasia) and carcinoma (high-grade dysplasia) categories.^{4,5} Nuclear enlargement, elongation, hyperchromasia, loss of polarity, pseudostratification; prominent nucleoli; irregularity in papillary structures are the findings seen in increasing degrees from adenoma to carcinoma. Our case was classified as borderline due to the presence of complex papillary architecture associated with nuclear stratification and cytological atypia characterized by nuclear enlargement, elongation and hyperchromasia, besides absence of mitoses, severe dysplasia, carcinoma in situ and/or infiltrative growth pattern.

The differential diagnosis in this case includes mucinous cystic neoplasms (MCNs). MCNs and IPMNs are cystic neoplasms composed of mucin producing epithelial cells and papillary formations. MCNs of the pancreas are seen predominantly in younger women and are mostly found in the tail or body of pancreas. IPMNs of the pancreas are seen predominantly in elderly man and are mostly in the head of pancreas. MCNs are associated with an ovarian type of stroma, but IPMNs are not associated with hypercellular stroma. MCNs do not communicate with the larger pancreatic ducts. Although MCNs are almost always unifocal, 30% of IPMNs are grossly multifocal.³⁻⁵

It has been reported that the prognosis is excellent for borderline tumors with three to five years survivals approaching 100%.¹⁰

In conclusion, to the best of our knowledge, our case is the first IPMN located in jejunal ectopic pancreas, and the third IPMN case situated in ectopic pancreas, after the ones reported one in the gastric wall,⁷ and the other one located in a Meckel's diverticulum.⁶

Diagnosing these preinvasive pancreatic neoplasms is important because of their excellent prognosis. Therefore, pathologists should keep the possibility of IPMN in mind and be careful about applying the diagnostic criteria strictly.

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