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Case Report

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Incidental Neuroendocrine Tumor in Mesocolonic Ectopic Pancreas Discovered During Colorectal Cancer Surgery

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Abstract

Background: Ectopic pancreas is a rare congenital anomaly, most commonly found in the stomach and duodenum, with mesocolonic involvement being exceptionally rare. While neoplastic transformation can occur, a neuroendocrine tumor (NET) arising in mesocolonic EP had not been previously reported.

Case Description: We describe the case of a 51-year-old female who presented with nonspecific abdominal symptoms and was diagnosed with a moderately differentiated pT4a tubular adenocarcinoma of the descending colon. She underwent surgical resection, and histopathological examination incidentally revealed ectopic pancreatic tissue in the mesocolic fat, exhibiting an area of epithelioid proliferation. Immunohistochemistry showed strong positivity for cytokeratin AE1/AE3, chromogranin A, and synaptophysin, confirming neuroendocrine differentiation. Notably, the adjacent ectopic pancreatic islets also showed synaptophysin positivity, highlighting the relationship between the tumor and the heterotopic pancreatic tissue. Ki-67 proliferation index was <1%, classifying it as a grade 1 NET. Postoperative 68Ga-DOTATATE PET/CT ruled out distant metastasis, and the patient underwent adjuvant chemotherapy for the colorectal cancer, with ongoing surveillance.

Conclusions: NETs can arise in ectopic pancreatic tissue, but their occurrence in the mesocolon had never been documented before. The case underscores the need for thorough histopathological evaluation of incidental findings during oncologic resections, and highlights the importance of considering ectopic pancreas as a potential site for neuroendocrine tumorigenesis.

Keywords: Neuroendocrine tumor; Ectopic pancreas; Mesentery; Mesocolon; Incidental finding

Introduction

Ectopic pancreas (EP), also known as heterotopic or aberrant pancreas, refers to the relatively rare occurrence of pancreatic tissue found outside its usual anatomical location, without any connection to the normal pancreas, having its own vascular and ductal systems. This condition is typically asymptomatic and only infrequently results in complications such as pancreatitis or malignant transformation [1]. Ectopic pancreatic tissue is identified in 0.25% of abdominal surgeries and 1.2% of gastrectomy procedures. During autopsies, its occurrence rate has been reported to range from 0.55% to 13.7% [2]. EP tissue has been described in both abdominal and extra-abdominal areas, most commonly in the stomach (24-38%), the

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duodenum (9-36%) and the jejunum (0.5-27%) [3]. Less frequent sites include Meckel's diverticulum, biliary tract, liver, spleen, mesentery and retroperitoneum [4-8].

The origin of EP tissue remains unclear, but there are two main hypotheses [9]. The "poor position" theory suggests that during embryonic development, fragments of the primitive pancreas may become separated during intestinal rotation and get trapped in other tissues [10]. Meanwhile, the "metaplasia" theory posits that EP arises from pancreatic metaplasia foci that migrate to ectopic locations during embryogenesis [11].

Modern classification of pancreatic heterotopias [12,13] identifies four types: Type I (complete pancreatic heterotopia with all pancreatic cell types), Type II (pancreatic ducts only), Type III (acinar tissue only), and Type IV (islet cells only).

There are some reports of mesocolonic EP giving rise to pseudopapillary [14], papillary [4,5] and acinar [8] neoplasms, but a neuroendocrine tumor (NET) in mesocolonic EP tissue had not been documented before, which we will present here.

Case Presentation

The patient, a 51-year-old female teacher from a small town in Brazil with no significant past medical history but a notable family history of gastrointestinal cancers, presented with nonspecific abdominal symptoms, including pain and constipation. Initial diagnostic workup, including computed tomography (CT) scans, revealed multiple hepatic lesions suggestive of hemangiomas and a collection adjacent to the colon, initially interpreted as diverticulitis. Further imaging with magnetic resonance imaging (MRI) corroborated the hypothesis of hepatic hemangiomas but also revealed focal parietal thickening in the descending colon adjacent to the collection, raising suspicion for perforated neoplasia. Colonoscopy identified an obstructive lesion in the descending colon, and biopsy confirmed a moderately differentiated tubular adenocarcinoma. Surgical resection of the tumor was performed.

Pathological examination revealed a pT4a moderately differentiated tubular adenocarcinoma with perforation of the visceral peritoneum. No vascular invasion was detected. The proximal, distal, and circumferential surgical margins were clear, and no metastasis was found in the 27 dissected lymph nodes. The omentum showed no neoplastic involvement. Examination of the spleen, Gerota's fascia, and diaphragm revealed a chronic exudative abscessed inflammatory process with fistulization associated with a foreign body-type giant cell reaction, without evidence of neoplasia. Ectopic pancreatic tissue was identified in the mesocolic fat, exhibiting epithelioid proliferation arranged in plaques, trabeculae, and cords (Figure 1).

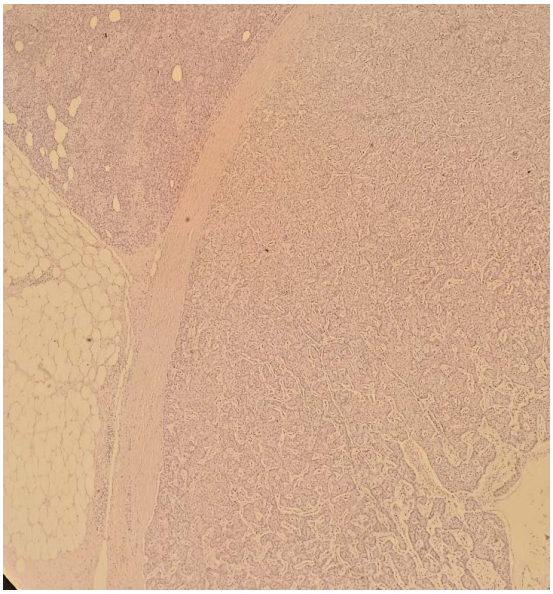

Figure 1: Hematoxylin & eosin (H&E) staining showing a well-circumscribed neuroendocrine tumor (NET) arising within mesocolonic ectopic pancreatic tissue. The upper left corner displays normal ectopic pancreatic tissue, with acinar and ductal structures. The lower left portion consists of mesocolic adipose tissue, indicating the tumor's ectopic location. The right side of the image is occupied by a solid, well-demarcated neuroendocrine neoplasm, composed of uniform epithelioid cells in trabecular and nested arrangements.

Further immunohistochemical staining was positive for cytokeratin AE1/AE3, chromogranin A, and synaptophysin, and negative for S100 protein, with Ki67 positivity in less than 1% of cells. This confirmed a grade 1 well-differentiated neuroendocrine tumor within the ectopic pancreatic tissue (Figure 2). Positron Emission Tomography (PET)/CT with 68Ga-labeled DOTATATE ruled out distant metastasis of the pancreatic neuroendocrine tumor (pNET).

Given the presence of perforated pT4a adenocarcinoma, the patient was recommended to receive adjuvant chemotherapy with the FOLFOX regimen. Adjuvant chemotherapy was initiated, and the patient is under close follow-up to monitor for recurrence of either neoplasm.

Discussion

EP tissue in the mesocolon is an extremely rare occurrence where pancreatic tissue is found in the mesentery associated with the colon. Neuroendocrine tumors (NETs) can occasionally arise in EP tissue, although this is also an uncommon phenomenon. Their prognosis depends on various factors, including the tumor grade, stage at diagnosis, and completeness of surgical resection. In this case report, we described the first patient to develop a NET in an EP located in the mesocolon, which was an incidental finding in a surgery performed for another different neoplasia. Given

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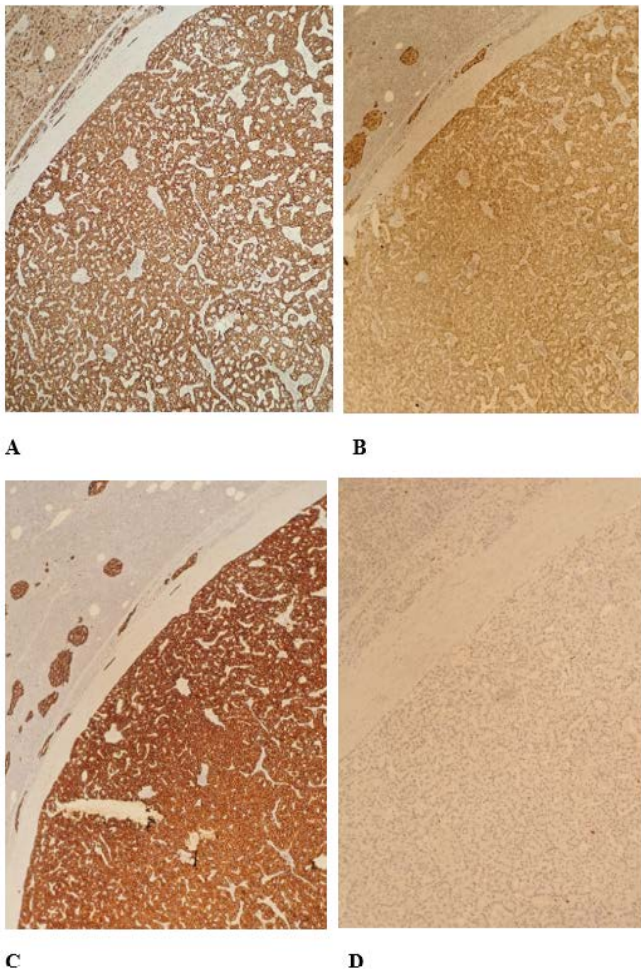

Figure 2: A: Cytokeratin AE1/AE3 immunostaining, highlighting **epithelial differentiation** of the neoplastic cells. B: Chromogranin A immunostaining confirming neuroendocrine differentiation, with uniform and intense positivity throughout the tumor. C: Synaptophysin immunostaining demonstrating **strong, diffuse positivity** in the **neuroendocrine tumor (NET)**, confirming its neuroendocrine differentiation. Notably, in the **upper left corner**, the **ectopic pancreatic tissue is also marked**, with **well-defined pancreatic islets staining positive** for Synaptophysin (as well as for Chromogranin A). This finding visually reinforces the **neuroendocrine nature of both the tumor and the native pancreatic islets**, highlighting the ectopic pancreatic origin of the lesion. **D:** Ki-67 immunostaining demonstrating a proliferation index **below 1%**, confirming the **low-grade** nature of the neuroendocrine tumor (WHO Grade 1).

the small size and its well-differentiated nature, we believe that surgical resection was curative.

We performed a PubMed search for ("ectopic pancreas" OR "heterotopic pancreas" OR "aberrant pancreas") AND ("neuroendocrine tumor" OR "neuroendocrine tumour") AND ("mesocolon") on May 23, 2024, which retrieved 20 results, whose titles were manually checked and none corresponded to a case of a NET arising from an EP in the mesocolon. We then repeated the search without the mesocolon restriction ("ectopic pancreas" OR "heterotopic pancreas" OR "aberrant pancreas") AND ("neuroendocrine tumor" OR "neuroendocrine tumour"), which yielded 230 results, again individually scrutinized.

We found 3 case reports of pNETs arising from EP: 2 in the stomach [15,16] and 1 in the retroperitoneum [7]. In this search we did note 7 reports of ectopic/extrapancreatic insulinomas, only one of which had a description of EP [17].

In conclusion, NETs in EP tissue located in the mesocolon, while rare, present unique diagnostic and therapeutic challenges. Early detection and appropriate management are essential to ensure favorable outcomes for patients with this uncommon condition. Surgical resection remains the mainstay of treatment, especially in symptomatic cases or when malignancy is suspected. Regular follow-up is essential to monitor for recurrence or potential complications.

Statements and Declarations

Competing Interests

The authors declare that they have no financial or non-financial competing interests.

Funding

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Ethics Approval

Ethical approval was not required for this case report, as it is a retrospective description of a clinical case without direct patient intervention. The study was conducted in accordance with institutional and ethical guidelines.

Consent for Publication

Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

Availability of Data and Materials

All relevant data generated or analyzed during this study are included in this published article. Additional data can be made available upon reasonable request.

Author Contributions

R.D.P. was responsible for conception and design. Pathology images were curated and interpreted by M.G.F. The first draft of the manuscript was written by M.Z.C., under mentorship from both R.D.P. and M.D.S.D. All authors contributed to the manuscript's revision, read, and approved the final version.

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