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Periampullary Neuroendocrine Tumor as a Cause of Acute Pancreatitis

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Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Funds Collection G

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Conflict of interest: None declared

Patient: Male, 60
Final Diagnosis: Periampullary neuroendocrine tumor
Symptoms: Abdominal discomfort
Medication: —
Clinical Procedure: —
Specialty: Surgery

Objective: Rare disease

Background: Duodenal and ampullary carcinoids are very rare tumors accounting respectively for 2% and 0.03% of all carcinoid tumors. Clinical findings vary according to the location of the tumor within the periampullary region; with epigastric pain being the most common presenting symptom in duodenal carcinoids and jaundice the most common clinical finding in ampullary carcinoids. Treatment options include pancreaticoduodenectomy, local excision, and endoscopic excision.

Case Report: In this case report, we present a 60-year-old male who presented with a one-week history of intractable epigastric pain. He was diagnosed with duodenal periampullary carcinoid tumor and treated with local excision.

Conclusions: Although duodenal and ampullary carcinoid tumors may have different clinical presentations, as well as histochemistry characteristics and metastatic potential, they appear to benefit from the same surgical treatment.

MeSH Keywords: Carcinoid Tumor • General Surgery • Pancreatic Neoplasms

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Background

Small bowel tumors account for about 2% of all gastrointestinal neoplasms and only 0.3% of all neoplasms. These tumors are usually either misdiagnosed upon the first presentation or diagnosed late [1]. It has been reported that carcinoid tumors account for approximately one-third of malignant small bowel tumors [2], however, due to the advancement in diagnostic modalities, their incidence seems to be increasing. According to the Connecticut Tumor Registry study, held between 1980 and 2000, carcinoid tumors were found to be the most common type of small bowel neoplasms, followed by adenocarcinoma [1]. As for the location within the small bowel, 50% of carcinoid tumors reside in the ileum and only 10% in the duodenum [3].

Periampullary tumors are defined as tumors arising within 2 cm of the major papilla in the duodenum; and are divided into 4 different types: pancreatic, duodenal, biliary, and ampullary [4].

Although the term carcinoid has been reserved for serotonin secreting tumors, mainly midgut neuroendocrine tumors (NETs), it seems that in the literature the term is still used for foregut NETs such as those localized to the ampulla or the duodenum.

Carcinoid tumors consist of neuroendocrine cells called Kulchitsky cells, scattered throughout the crypts of Lieberkuhn. These cells differ amongst each other by the type of secreted amino acid. The ones that secrete serotonin are called argentaffin cells due to their ability to fix and reduce silver substrate, whereas the ones that secrete other substances are called argyrophils as they only fix the silver substrate after being treated with reducing agents [5]. Finally, the type of secretion for periampullary carcinoid tumors varies with the location of the tumor within the periampullary region [6].

Case Report

A 60-year-old male, presented to the Emergency Department complaining of a one-week history of intolerable epigastric pain, irradiating to the back, burning in quality, not related to meals and with no relieving factors. Past surgical history was positive for a left hemicolectomy at 6 months prior to presentation for colonic adenocarcinoma. During a previous metastatic workup post-hemicolectomy, a computed tomography (CT) scanning of the abdomen revealed an incidental duodenal mass for which the patient refused further investigation.

At presentation, the patient was cooperative and stable. He was looking ill but had no signs of dehydration or malnutrition. On physical examination, the scleare were anicteric and the conjunctiva well injected. The abdominal examination was positive for epigastric tenderness upon deep palpation but negative for any palpable mass or hepatosplenomegaly. An ultrasound was done and showed a distended gall bladder with no evidence of gallstones. The laboratory workup revealed disturbance in hepatic and pancreatic enzymes (Table 1).

Table 1. Laboratory values upon presentation showing disturbances in pancreatic and hepatic enzymes.

White blood cells	9500/mm^3
Hematocrit	41%
SGOT	114 U/L
SGPT	217 U/L
Alkaline phosphatase	345 UI/L
Total bilirubin	1.11 mg/dL
Direct bilirubin	0.2 mg/dL
Amylase	167 U/L
Lipase	648 U/L
C-reactive protein	66 mg/L

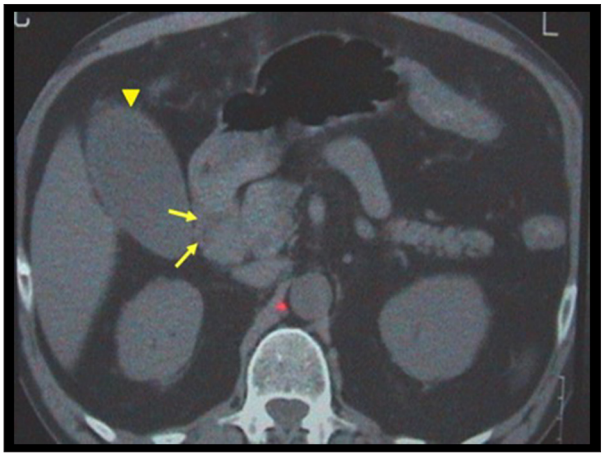

Figure 1. Computed tomography showing a hypodense polypoid duodenal mass about 2 cm in diameter near the ampulla (arrows) with distended gallbladder (arrowhead).

An abdominal CT-scan was performed next, and findings were consistent with a duodenal periampullary hypodense mass, about 2 cm in diameter, resulting in a distended acalculous gallbladder (Figure 1). No evidence of metastasis was noted on the CT scan. The patient then underwent an upper gastrointestinal endoscopy that showed a peripapillary, pedunculated, ulcerated mass, about 5 cm in diameter (Figure 2). An attempt was made to biopsy the mass but failed due to bleeding upon minor contact. Following catheterization of the duodenal papilla, an endoscopic retrograde cholangiopancreatography (ERCP) was performed, and revealed an irregular mass

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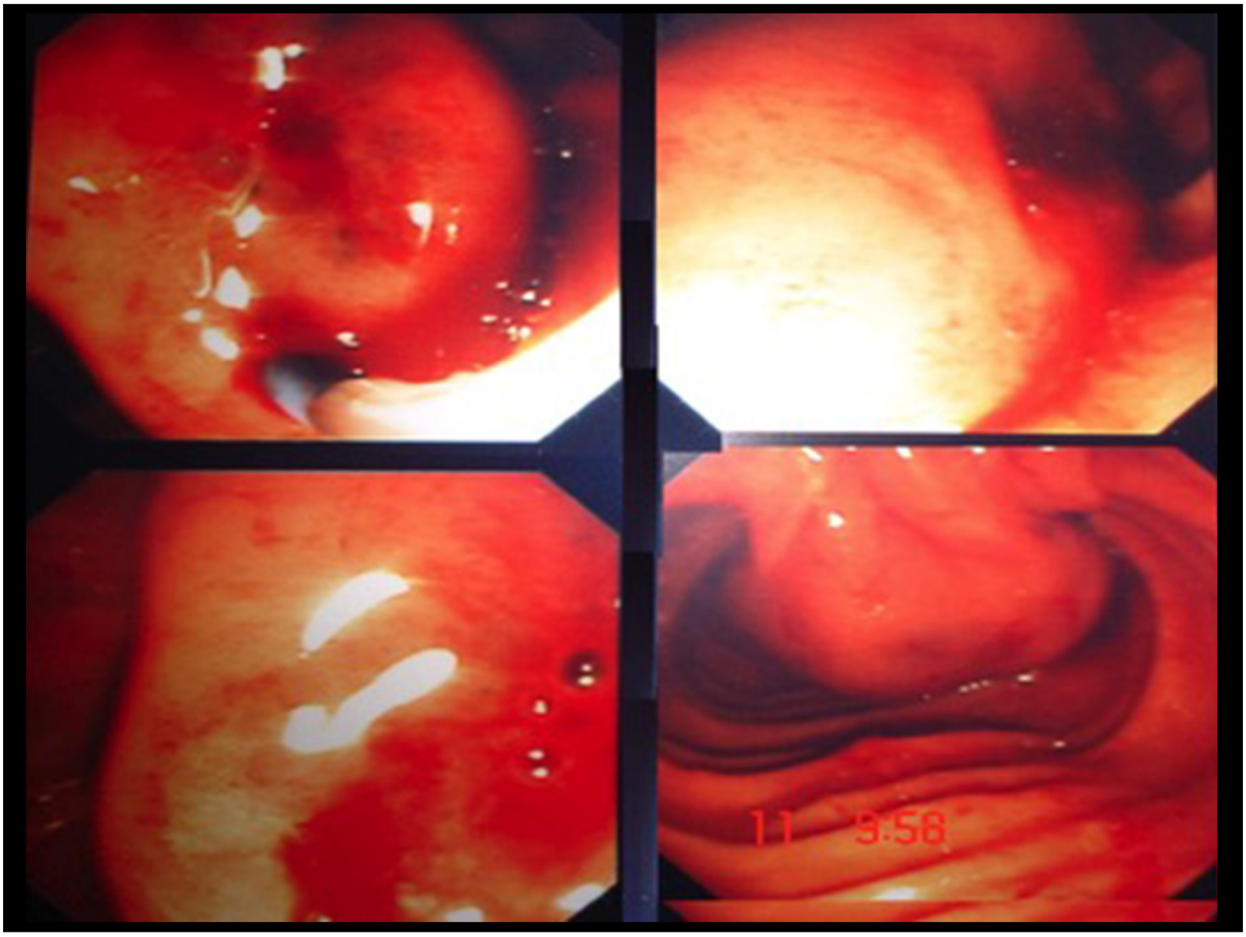

Figure 2. Upper gastrointestinal endoscopy showing a pedunculated ulcerated 5 cm duodenal mass near the papilla, with bleeding upon contact.

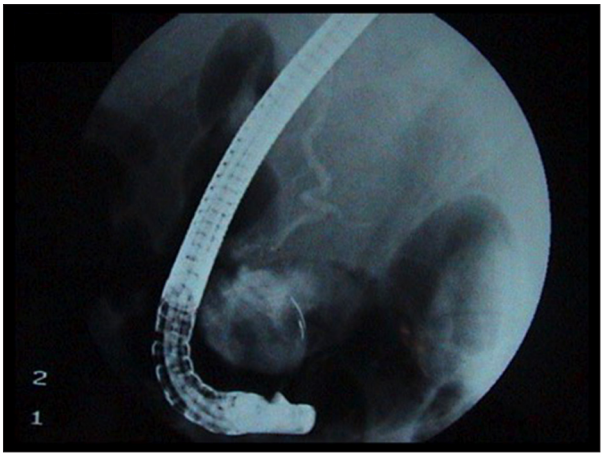

Figure 3. An endoscopic retrograde cholangiopancreatography showing an irregular mass with contrast pooling around it.

with contrast pooling around it, but there was no evidence of extension into the common bile duct or the pancreatic duct (Figure 3). Finally, an endoscopic ultrasound revealed a 5 cm mass restricted to the mucosal layer and absence of nodal metastasis. Giving the aforementioned results, a transduodenal excisional biopsy was performed; starting with a median laparotomy followed by full mobilization of the duodenum. A longitudinal incision over the second duodenal portion revealed the mass that was very close to the major papilla. An en passant cholecystectomy was performed, and methylene blue was injected through the cystic remnant in order to identify the papilla, after which the mass was locally excised and a specimen sent for pathology.

Macroscopically, the mass was lobulated, measuring 4.2×2.7×1.5 cm, involving the submucosal and muscularis layers with ulceration of the overlying mucosa. Microscopically, it consisted of fusiform cells, with no evidence of nuclear atypia or mitotic activity. On immunohistochemistry studies, the tumor cells were diffusely positive for cytokeratin antibodies, and focally positive for synaptophysin and chromogranin. Based on the aforementioned histological and immunohistochemistry findings, the diagnosis of carcinoid tumor was established.

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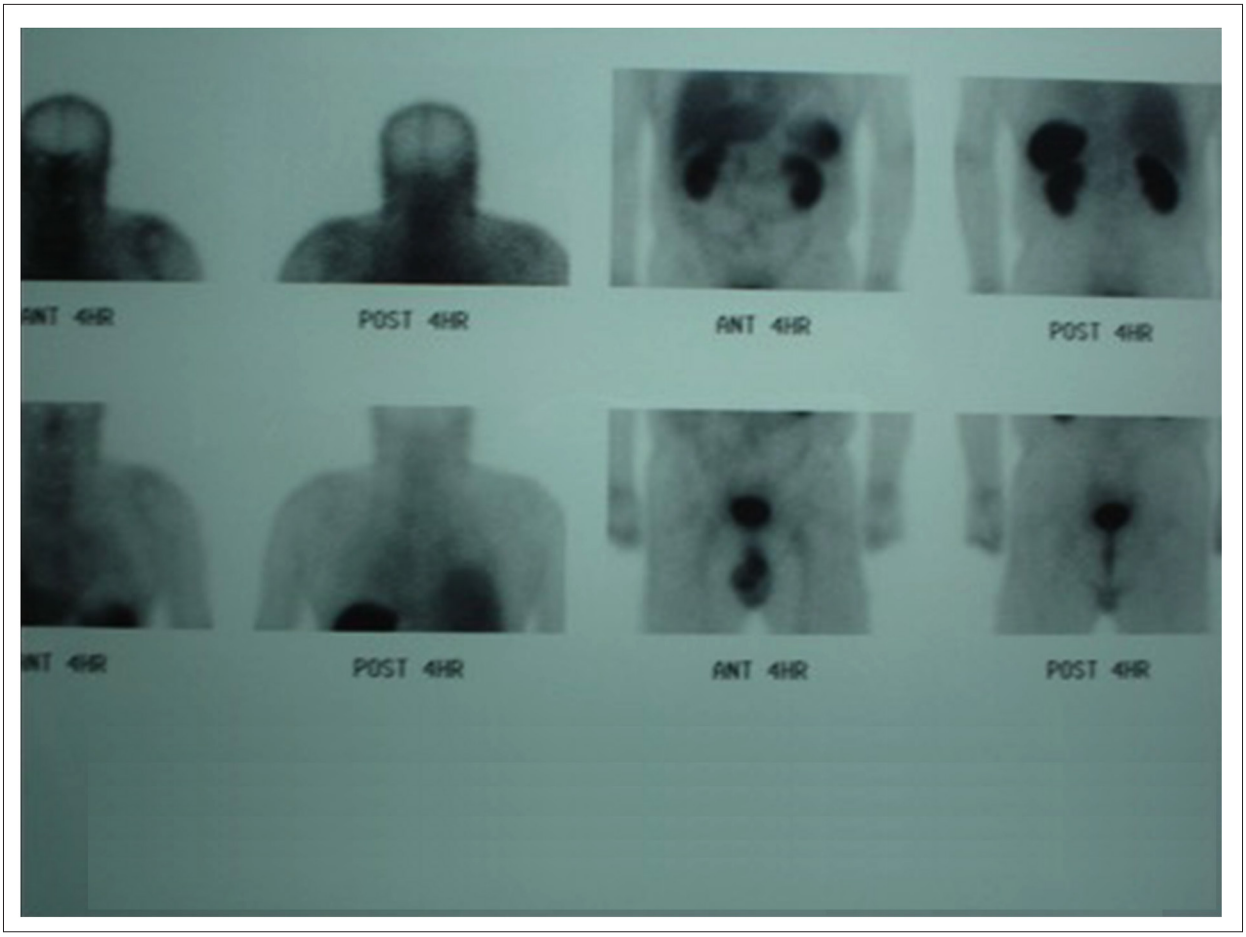

Figure 4. Follow-up octreotide scintigraphy performed 3 months after the operation with no uptake of radioactive material.

A follow-up octreotide scintigraphy 3-months post-operatively showed no evidence of radioisotope uptake (Figure 4). Serum insulin, insulin, C-peptide, gastrin, vasoactive intestinal peptide, pancreatic polypeptide, glucagon, somatostatin, chromogranin A and B, and calcitonin, as well as urinary 5-hydroxy-indole acetic acid (5-HIAA) levels, were within the normal range.

The patient recovered smoothly and had no major complaints in during the 3-month follow-up period.

Discussion

Small bowel tumors account for about 2% of all gastrointestinal neoplasms and only 0.3% of all neoplasms [1]. The most common types of gastrointestinal cancers are carcinoids and adenocarcinomas [7]. The data are conflicting concerning which one of these 2 types is the most common. Even though carcinoid tumors are rare neoplasms, they are the most common gastrointestinal neuroendocrine tumors. Their incidence has increased in the last several decades; most likely due to increasing recognition [5]. The duodenum and the ampulla are considered to be rare locations for carcinoid neoplasms, accounting only for 2% and 0.05% of all carcinoids, respectively [8].

Tumors of the ampulla of Vater are defined as arising in the last centimeter of the common bile duct where it passes through the wall of the duodenum and the ampullary papilla [9]. Distinguishing between true ampullary cancers and periampullary tumors is critical, the former accounting for about 7% of all periampullary cancers [10]. The distinction between a duodenal carcinoid and an ampullary carcinoid should be made as they have different clinical behaviors and long-term outcomes. In one study, Makhlouf et al. addressed the differences in clinical, histological, and immunohistochemical characteristics between ampullary carcinoid and non-ampullary duodenal carcinoid. Jaundice and association with von Recklinghausen disease are the clinical features distinguishing ampullary carcinoids from duodenal carcinoids. As for histological and immunohistochemical features, psammoma bodies, secretion of somatostatin, and an insular growth pattern characterize ampullary carcinoids; whereas the cribriform pattern of growth and secretion of gastrin characterize duodenal carcinoids. The Makhlouf et al. study concluded that the tumor size and the

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mitotic index had no correlation with the risk of metastasis in ampullary carcinoids, but significantly affected the metastatic potential of duodenal carcinoids [11]. Ricci et al. found no correlation between nodal positivity and the ampullary carcinoid tumor size [12].

No definitive statement can be made regarding the treatment options of ampullary carcinoids as there are only a few cases reported in the literature. However, authors have concluded treatment options based on their experience with duodenal carcinoids. Although long-term survival has been achieved by local excision, Ricci concluded that radical excision may be the treatment of choice for carcinoids of the ampulla of Vater [12]. Most experts still recommend either a Whipple or a pylorus preserving pancreaticoduodenectomy (PPPD) for tumors over 2 cm in size [13]. A successful endoscopic approach has also been reported for ampullary carcinoids [14].

Ampullary carcinoids are more advanced at presentation and have a worse overall survival than duodenal carcinoids (98 *vs.* 143 month respectively). However, ampullary carcinoids that are managed by local resection have a similar overall survival with locally resected duodenal carcinoids [15].

Conclusions

Our patient had a 5 cm peripapillary carcinoid tumor that was managed by local excision, and the patient had long-term survival and no clinical evidence of recurrence.

This was a case of a periampullary "nonampullary" tumor, a term that we cannot find in review articles, raising the following question: Should periampullary duodenal carcinoids be considered as ampullary ones?

Department and Institution where work was done

Mount Lebanon Hospital, Beirut, Lebanon.

Conflicts of interest

None.

References:

1. Hatzaras I, Palesty JA, Abir F et al: Small-bowel tumors: epidemiologic and clinical characteristics of 1260 cases from the Connecticut Tumor Registry. Arch Surg, 2007; 142(3): 229–35
2. Waisberg J, Joppert-Netto G, Vasconcellos C et al: Carcinoid tumor of the duodenum: A rare tumor at an unusual site. Case series from a single institution. Arq Gastroenterol, 2013; 50(1): 3–9
3. Helland S, Prøsch A, Viste A: Carcinoid tumours in the gastrointestinal tract – A population-based study from Western Norway. Scand J Surg, 2006; 95(3): 158–61
4. Chandrasegaram MD, Gill AJ, Samra J et al: Ampullary cancer of intestinal origin and duodenal cancer – A logical clinical and therapeutic subgroup in periampullary cancer. World J Gastrointest Oncol, 2017; 9(10): 407–15
5. Pinchot SN, Holen K, Sippel RS, Chen H: Carcinoid tumors. Oncologist, 2008; 13(12): 1255–69
6. Vanoli A, La Rosa S, Klersy C et al: Four Neuroendocrine tumor types and neuroendocrine carcinoma of the duodenum: Analysis of 203 cases. Neuroendocrinology, 2017; 104: 112–25
7. Terada T: Malignant tumors of the small intestine: A histopathologic study of 41 cases among 1,312 consecutive pecimens of small intestine. Int J Clin Exp Pathol, 2012; 5(3): 203–9
8. Ozsoy M, Ozsoy Y, Canda A et al: The rare malignancy of the hepatobiliary system: Ampullary carcinoid tumor. Case Rep Med, 2011; 2011: 173036
9. Jean M, Dua K: Tumors of the ampulla of vater. Curr Gastroenterol Rep, 2003; 5(2): 171–75
10. Jemal A, Siegel R, Ward E et al: Cancer Statistics, 2008. Cancer J Clin, 2008; 58(2): 71–96
11. Makhlouf H, Burke A, Sobin L: Carcinoid tumors of the ampulla of Vater: A comparison with duodenal carcinoid tumors. Cancer, 1999; 85(6): 1241–49
12. Ricci J: Carcinoid of the ampulla of vater. Local resection or pancreaticoduodenectomy. Cancer, 1993; 71(3): 686–90
13. Gilani N, Ramirez FC: Endoscopic resection of an ampullary carcinoid presenting with upper gastrointestinal bleeding: A case report and review of the literature. World J Gastroenterol, 2007; 13(8): 1268–70
14. Panzeri F, Crippa S, Castelli P et al: Management of ampullary neoplasms: A tailored approach between endoscopy and surgery. World J Gastroenterol, 2015; 21(26): 7970–87
15. Randle RW, Ahmed S, Newman NA, Clark CJ: Clinical outcomes for neuroendocrine tumors of the duodenum and ampulla of Vater: A population based study. J Gastrointest Surg 2014; 18: 354–62

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