A Case of Synchronous Malignancies

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Introduction

Pancreatic neuroendocrine tumors (NETs) are rare malignancies, accounting for approximately 1-2% of all pancreatic cancers. Less commonly, pancreatic NETs can be a cause of acute pancreatitis, depending on the size and location of the mass. There have been instances of synchronous carcinoid tumors or various other gastrointestinal NETs with concurrent secondary primary malignancies, however, there have not been previously documented cases of a pancreatic NET with concomitant colon adenocarcinoma presenting as acute pancreatitis.

Case Presentation

73-year-old Caucasian female with no medical history, presented for 1 day of worsening right-sided abdominal pain. The pain began suddenly with no inciting event and she also reported an approximately 20-pound weight loss over the preceding months. Initial lab work showed the patient had an elevated lipase, and computerized tomography (CT) of the abdomen/pelvis demonstrated extensive liver lesions consistent with metastatic disease. Additionally on the CT abdomen/pelvis, the tail of the pancreas had a dilated pancreatic duct with a normal duct size in the proximal duct, indicating a pancreatic duct cut-off sign. CT of the chest was unremarkable. Gastroenterology was consulted for further evaluation and an EGD with EUS was performed the following day revealing a 2cm x 3cm ill-defined mass in the head of the pancreas that was hypoechoic and heterogeneous in appearance. Multiple hypodense lesions were noted throughout the liver. No pancreatic ductal dilatation was noted, and surrounding vasculature appeared normal. Pathology revealed findings consistent with well differentiated grade 2 neuroendocrine tumor and was positive for CK-CAM5.2, CK7, CD56, chromogranin and synaptophysin, along with a KI-67 close to 20%. Hematology/Oncology and Surgical Oncology were consulted. Tumor markers were notable for a slightly elevated CA19-9. Colonoscopy was performed and notable for a 3cm descending colon polyp, which was later revealed to be well-differentiated adenocarcinoma with invasion into the submucosa. The patient was discharged, with instructions to follow up with Hematology/Oncology, Surgical Oncology, and Gastroenterology in the outpatient setting. The patient followed up and obtained a Gallium Dotatate Positron Emission Tomography scan, and is currently in the process of undergoing treatment.

Figure

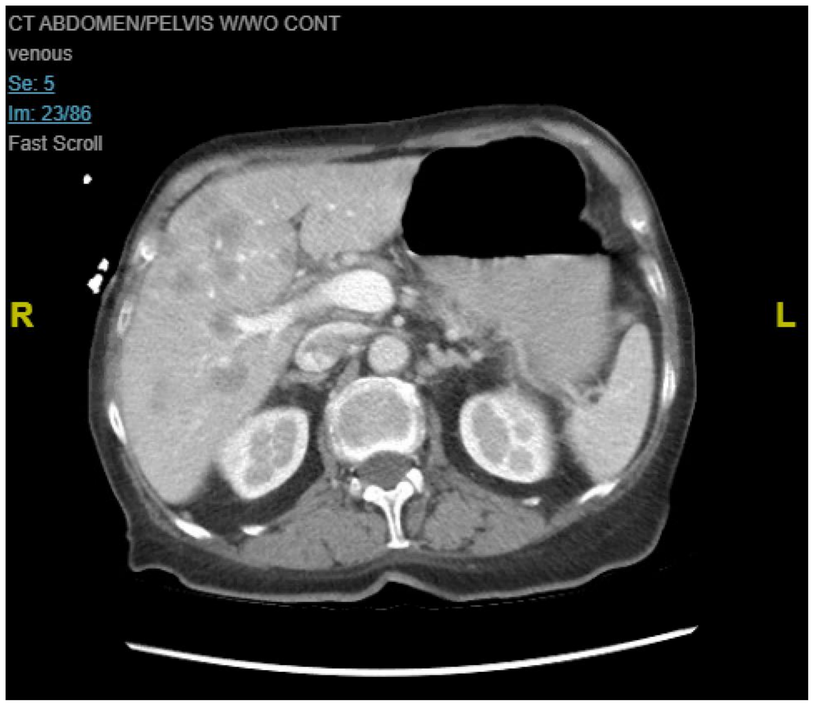

Figure 1: multiple heterogeneic lesions noted in the liver

Discussion

Acute pancreatitis is a common phenomenon that is seen regularly in the inpatient setting. Two out of three criteria are required for diagnosis: abdominal pain, serum lipase or amylase greater than three times the upper limit of normal, or imaging findings [11]. There are a multitude of causes, with the most common being gallstones, alcohol, and medication-induced [2,11]. A less frequently observed etiology is malignancy, as in our case. Amidst the different types of pancreatic neoplasms, NETs are among the rarest [1].

Per Bravo et al., there were 30 reported cases of acute pancreatitis believed to have been caused by pancreatic neuroendocrine tumors as of 2010 [4]. The literature shows that there has been an increase in cases since that time, which can, in part, be attributable to increased diagnosis of asymptomatic, early-stage disease [12,13]. Notwithstanding, pancreatic NETs continue to be a relatively rare malignancy. These can be further subdivided into functioning and nonfunctioning, as in our case. Functional tumors oftentimes are associated with inherited syndromes such as multiple endocrine neoplasia 1, von Hippel-Lindau, tuberous sclerosis or neurofibromatosis type 1 [14]. Per Hallet et al., metastasis was noted in one out of five subjects that presented with NETs of any kind [13]. Due to the relatively asymptomatic nature of nonfunctioning pancreatic NETs, they generally are difficult to diagnose leading to more advanced disease upon presentation [9,14]. Metastases to the liver and the lymphatic system have been noted multiple times in similar cases, and our patient was found to have hepatic involvement as well [4,9,10].

Interestingly, there have been documented occurrences of synchronous NETs and secondary primary malignancies [6,7,9,10]. Yumoto et al. documented a case of concurrent rectal NET and sigmoid colon cancer, while Cokmert et al. presented a case of a NET involving the ampulla of Vater and sigmoid colon cancer [6,9]. Our patient appears to have the only reported case of a pancreatic NET with simultaneous colon adenocarcinoma, presenting as acute pancreatitis. Although the cause of the high incidence of dual tumors is unknown, it is speculated that it could be due to the tumorigenic properties of peptides released by neuroendocrine cells, also termed the field-effect theory [7].

Our patient underwent CT imaging upon admission, which prompted further workup after the discovery of both the pancreatic head mass and hepatic lesions. She then underwent an EGD with EUS for biopsy of the pancreatic mass, with a subsequent colonoscopy to rule out concurrent malignancy. An octreotide scan was set up outpatient to further evaluate neuroendocrine involvement, and the patient has followed up with specialists for further management.

Conclusion

This is the first known case in the literature of a patient presenting with acute pancreatitis, who was found to have both a pancreatic NET and colon adenocarcinoma. As a result of the position of the NET, our patient presented with pancreatitis, ultimately leading to diagnosis of concurrent malignancies for which she can now undergo treatment. Our case hopes to bring awareness to the importance of assessing for synchronous malignancies.

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