A Rare Case of Pancreatic Neuroendocrine Tumor Recurrence and Development of Type 3c Diabetes

Amanda Jacubowsky, DO[1], Tinsa Varughese, DO[1], Lindor Gelin, DO[2], Anne Borja, MD[2]
Lehigh Valley Health Network, Allentown, PA
[1]Department of Medicine, [2]Department of Endocrinology

Introduction

- Yearly, 64,000 people in the United States are diagnosed with pancreatic cancer
  - 90% of cases are exocrine tumors
  - Less than 2% are pancreatic neuroendocrine tumors (PNETs)
- This case highlights a rare case of a patient with recurrent PNET and type 3c diabetes

Case Description

A 60-year-old male with history of type 2 diabetes mellitus on insulin and PNET s/p distal pancreatectomy presented with **abdominal pain** and **watery diarrhea**

Labs: unremarkable CBC, CMP, lipase, insulin, gastrin, glucagon, serotonin, urine 5-HIAA. Elevated Chromogranin A (116 ng/mL), VIP (123 pg/mL), pancreatic polypeptide (526 pg/mL)

Imaging:

- CT Abdomen and Pelvis: negative for acute pathology
- Somatostatin receptor PET Scan: small focus of activity in the pancreatic body (Figure 1)

Outpatient and Hospital Course:

- Trialed on somatostatin analogs → did not help abdominal pain or diarrhea
- Admitted for second distal pancreatectomy with pathology revealing **recurrent PNET** (Figure 2a & 2b)
- Hospital stay complicated by hyperglycemia and labs showing inappropriately low C-peptide (0.2 ng/mL) → diagnosed with **type 3c diabetes.**

Imaging

Figure 1: Positive somatostatin receptor PET scan for activity in pancreatic body (blue arrow)

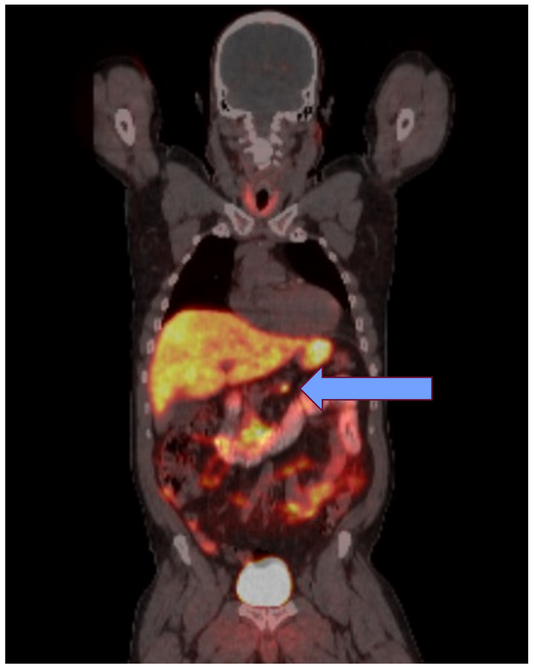
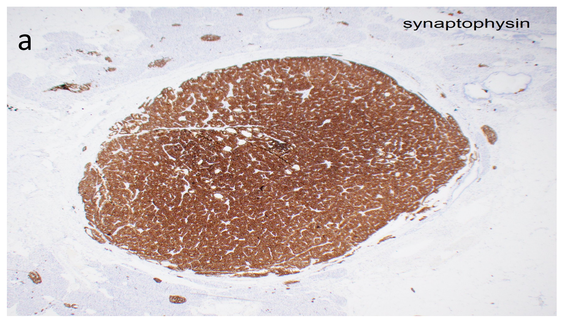


Figure 2: Pathology showing a) Positive synaptophysin staining and b) Positive chromogranin staining



Discussion

- PNETs are a very rare type of cancer that can secrete various functioning and nonfunctioning substances
- Treatment for a functional PNET → surgical resection
- Complications after pancreatectomy: infection, bleeding, pancreatic fistulas, and the development of diabetes requiring insulin
- Type 3c diabetes is seen in patients with chronic pancreatitis, ductal adenocarcinoma, and a history of distal pancreatectomy

References

1) de Ponthaud, C., Menegaux, F., & Gaujoux, S. (2021). Updated Principles of Surgical Management of Pancreatic Neuroendocrine Tumours (pNETs): What Every Surgeon Needs to Know. *Cancers*, *13*(23), 5969. https://doi.org/10.3390/cancers13235969
2) Sun J. (2017). Pancreatic neuroendocrine tumors. *Intractable & rare diseases research*, *6*(1), 21–28. https://doi.org/10.5582/irdr.2017.01007
3) Tan, Q. Q., Wang, X., Yang, L., Chen, Y. H., Tan, C. L., Zhu, X. M., Ke, N. W., & Liu, X. B. (2020). Analysis of recurrence after resection of well-differentiated non-functioning pancreatic neuroendocrine tumors. *Medicine*, *99*(24), e20324. https://doi.org/10.1097/MD.0000000000020324
4) Hart, P. A., Bellin, M. D., Andersen, D. K., Bradley, D., Cruz-Monserrate, Z., Forsmark, C. E., Goodarzi, M. O., Habtezion, A., Korc, M., Kudva, Y. C., Pandol, S. J., Yadav, D., Chari, S. T., & Consortium for the Study of Chronic Pancreatitis, Diabetes, and Pancreatic Cancer(CPDPC) (2016). Type 3c (pancreatogenic) diabetes mellitus secondary to chronic pancreatitis and pancreatic cancer. *The lancet. Gastroenterology & hepatology*, *1*(3), 226–237. https://doi.org/10.1016/S2468-1253(16)30106-6
5) Kim, K. J., Jeong, C. Y., Jeong, S. H., Ju, Y. T., Jung, E. J., Lee, Y. J., Choi, S. K., Ha, W. S., Park, S. T., & Hong, S. C. (2011). Pancreatic Diabetes after Distal Pancreatectomy: Incidence Rate and Risk Factors. *Korean journal of hepato-biliary-pancreatic surgery*, *15*(2), 123–127. https://doi.org/10.14701/kjhbps.2011.15.2.123