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A Rare Case of a Non- Functioning Pancreatic Neuroendocrine Tumor

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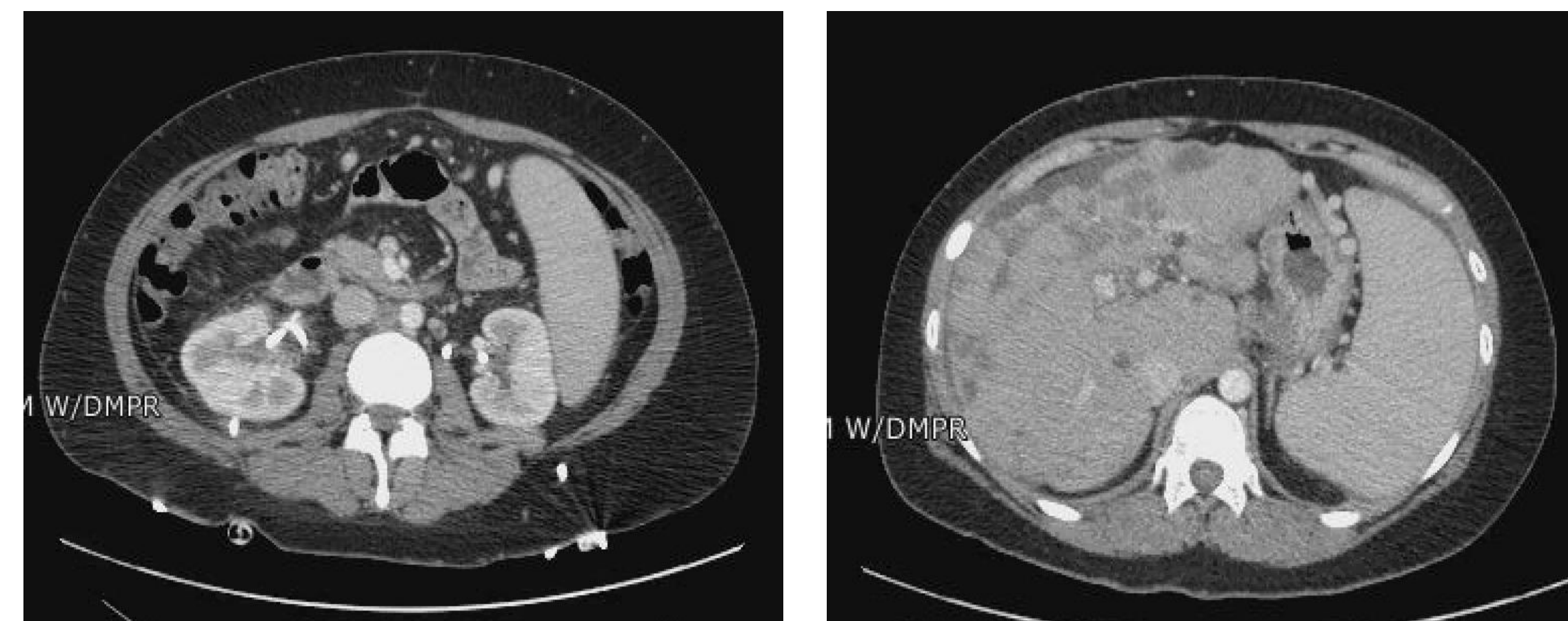
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INTRODUCTION

- Pancreatic neuroendocrine tumors (PNETs) are very rare and represent about 1–2% of all pancreatic tumors.
- Non-functional PNETs (NF-PNETs) represent approximately 85% of all PNETs. They are mostly found incidentally at an advanced stage because patients are asymptomatic and the tumors do not secrete hormones that would manifest with symptoms.
- More than 95 percent of the pancreatic neoplasms arise from the exocrine component of the pancreas. Neoplasms arising from the endocrine pancreas are neuroendocrine tumors (NETs).
- PNETs, also known as islet cell tumors, originate in the islets of Langerhans. PNETs are rare neoplasms of the pancreas, with an incidence of 1/100,000/year and account for about 1-2% of all pancreatic neoplasms [2].
- Computed tomography (CT) is the imaging modality of choice, but other diagnostic imaging tools need to be investigated for earlier detection of this insidious and fatal disease.
- Surgical treatment is currently preferred, but only palliative chemotherapy and radiation is currently available for advanced disease.
- We report a case of 32-yr-old female who initially presented with increasing abdominal girth, decreased mobility, and lower extremity swelling up to the hip and she was incidentally found to have a widely metastatic intermediate grade NF-PNET stage IV T2N0M1 to the liver, peritoneum, bones, and gallbladder.

CASE REPORT

- This is a case of a young female who underwent a computed tomography of the abdomen and pelvis (CT) in 2016 that showed a massively enlarged liver with diffuse metastases and her largest lesion measured 5.8x4.9cm.
- The patient also had a 3.5x3.6 cm pancreatic tail mass as well as a 10.3x11 cm mass in her pelvic areas extending to the vaginal wall:



- The patient underwent a core biopsy of her liver on 01/28/2016 and her pathology showed a metastatic neuroendocrine tumor with Ki 67 of 40% and a mitotic rate of 10 per 10 HPF.
- Treatment with capecitabine and temozolomide began shortly after.
- A repeat CT of the abdomen and pelvis on 06/24/2016 showed significant decrease in her pancreatic mass and liver metastases.
- Chromogranin A level had decreased from 5800 to 35 ng/dL. She completed 6 cycles of chemotherapy and then had an exploratory laparotomy, lysis of adhesions, and bilateral oophorectomy with the gynecology service.
- The patient was found to have metastatic disease with peritoneal deposits to her sigmoid colon and her peritoneum. Her treatment was changed to octreotide and everolimus postoperatively until

the MRI of her abdomen in mid 2017 showed progression of liver metastases.

- The patient was then changed to doxorubicin, 5-fluorouracil (5-FU), and streptozocin.
- Repeat imaging showed improvement of her metastases to the liver.
- The patient received four cycles of treatment and she then received Y90 to her liver that completed in early 2018.
- A repeat MRI of the abdomen showed stable disease in her liver.
- A bone scan shortly after showed multiple bone lesions and she was having significant bone pain. She received one cycle of platinum and etoposide and she felt that her bone pain was improving.
- A gallium 68 PET CT was recommended to clarify if the patient had an overexpression of somatostatin receptors and this would be more consistent with a slower growing NET. The patient would then receive treatment with Peptide Receptor Radionuclide Therapy (PRRT).
- The patient was diagnosed with widely metastatic intermediate grade NF-PNET stage IV T2N0M1 to the liver, peritoneum, bones, and gallbladder.

DISCUSSION

- According to the World Health Organization (WHO) classification, three classes of NETs can be identified based on histology and pathology: well-differentiated NETs can be classified as G1 tumors, when they express <2 mitoses/10 HPF and ≤2% Ki-67 index;

as G2 tumors, when they express 2-20 mitoses/10 HPF and 3-20% Ki-67, whereas neuroendocrine carcinomas (NECs) usually belong to G3 category, with >20 mitoses/10 HPF and >20% Ki-67 index [1].

- The patient had Ki 67 of 40%.
- Potential risk factors for PNETs include diabetes, smoking, a previous history of chronic pancreatitis, and genetic factors (hereditary endocrinopathies, including multiple endocrine neoplasia types I) [2], However the patient had no risk factors for this disease.
- Clinical management involves a multidisciplinary approach, but surgery remains the only curative therapy for early-stage disease.
- Radical surgery for pancreatic NET offers a 5-year survival rate of 45-92%, highlighting the importance of accurate diagnosis and appropriate referral of patients for surgery [3].

CONCLUSION

- Because of its rarity, randomized controlled studies have not been done and current treatment recommendations are based primarily on case series and individual treatment approaches [4].
- Further research is under way on newer, investigational drugs.

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