

When the Skin Speaks and the Gut Follows: How diarrhea and a rash lead to a diagnosis of pancreatic neuroendocrine malignancy

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Introduction: Neuroendocrine tumors (NETs) are a heterogeneous group of neoplasms arising from neuroendocrine cells. Pancreatic neuroendocrine tumors (PanNETs) represent a subset that are either functional tumors manifesting hormone-specific syndromes or nonfunctional tumors with nonspecific presentations. Nonfunctional PanNETs are often found incidentally at a later stage, typically due to mass effect or metastases to the liver. Early recognition is crucial for timely, targeted treatment of functional PanNETs.

Case: An 85-year-old male with hypertension, peripheral arterial disease, COPD, benign prostatic hypertrophy, recently diagnosed eczematous and psoriasiform dermatitis, glaucoma, and venous insufficiency presented with one week of persistent diarrhea consisting of four loose stools daily. The patient denied abdominal pain, fever, chills, appetite loss, or sick contacts. Lab studies revealed severe hypokalemia and elevated liver enzymes. A right upper quadrant ultrasound showed hepatomegaly with complex cystic lesions. Computed tomography imaging revealed a large peripherally enhancing lesion in the right lobe of the liver with central fluid attenuation suspicious for cystic degeneration versus necrosis, multiple solid and cystic hepatic lesions consistent with metastases, and a 3.4 cm enhancing pancreatic body mass concerning for a neuroendocrine tumor. A liver biopsy confirmed metastatic, low-grade neuroendocrine tumor, consistent with a pancreatic primary. A transthoracic echocardiogram was unrevealing. Niacin levels were low, while AFP, CEA, and CA19-9 were unremarkable. A glucagon level was elevated to 535 pg/mL. A diagnosis of pancreatic neuroendocrine tumor with hepatic metastases, complicated by carcinoid syndrome and pellagra was made. The patient continued multivitamin, prednisone taper, and discharged with oncology follow up and was found to have a glucagonoma.

Discussion: This case demonstrates the delayed finding of glucagonoma, a functional PanNET. As PanNETs metastasize to the liver, patients may experience carcinoid syndrome due to the vasodilatory effects of active amines, peptides, and prostaglandins. Common symptoms are flushing and diarrhea. Patients may become niacin deficient, leading to pellagra or heart failure. Prior to the onset of diarrhea, our patient was diagnosed with recurring eczematous and psoriasiform dermatitis on skin biopsy 6 months prior. This can be attributed to early necrolytic migratory erythema often associated with glucagonoma or due to his niacin deficiency. Dermatitis and diarrhea together are two of the three typical findings of pellagra. On further questioning, both the patient and his family endorsed a cognitive decline and irritability over the prior 4 months, thus completing the 3rd finding of dementia or neuropsychiatric findings classic for pellagra. The findings of a recurring skin rash and persistent diarrhea should raise the possibility of a PanNET on the differential diagnosis list.

Citations

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