A Tale of Two NETs: A Pheochromocytoma Masquerading as a Pancreatic NET

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Neuroendocrine tumors (NETs) are rare endocrine neoplasms with myriad of clinical manifestations. We present a case of two different NETs in a patient to increase physician awareness and highlight the importance of prompt multidisciplinary approach to avoid catastrophic complications.

Case Presentation

A 58-year-old female with history of difficult to control hypertension, diabetes mellitus and gastroesophageal reflux was referred to an oncology office for suspected metastatic pancreatic neuroendocrine tumor.

She had undergone abdominal imaging for known hepatitis B, and a cystic pancreatic lesion was discovered in addition to a hypervascular right adrenal nodule which was thought to represent metastatic disease. She endorsed flushing, headache, gastroesophageal reflux and cough in addition to recent worsening in glycemic control, but denied any diarrhea or rashes. Otherwise complete review of systems was unremarkable.

Physical exam:
BP 188/90 mm of Hg, HR 83. Physical exam revealed an obese female, otherwise no significant findings.

Notable Laboratory and Imaging studies:
- Chromogranin A 205 ng/ml, post clonidine: 413 pg/ml
- Plasma free metanephrines 552 pg/ml, post clonidine: 413 pg/ml
- a.m. cortisol 1 ug/dl following dexamethasone suppression test.
- EUS with biopsy confirmed WHO grade 1 neuroendocrine tumor.
- Ga-68 Dotatate scan: radiotracer uptake in the pancreatic tail corresponding to the known pancreatic neuroendocrine tumor. Focal radiotracer uptake in the right adrenal gland corresponds to the previously seen adrenal gland nodule.

Discussion

- Pancreatic NETs arise sporadically, however they can be associated with genetic syndromes including multiple endocrine neoplasia type 1 (MEN1), von Hippel-Lindau syndrome, neurofibromatosis type I or tuberous sclerosis. Surgery is the only known cure for sporadic pancreatic NETs, whether functional or nonfunctional. Tumor functionality, grade, and stage are important factors in choosing patients for surgical treatment and determining the operative approach.

- Pheochromocytomas are rare catecholamine secreting tumor with malignant potential that arises from chromaffin cells of the adrenal medulla.

- Biochemical confirmation of the diagnosis using total fractionated urine metadrenalines or plasma-free metadrenalines should be followed by radiological evaluation to locate the tumor (4).

- Treatment of pheochromocytomas involves resection of the pheochromocytoma following appropriate medical preparation with alpha-adrenoreceptor blockade, calcium channel antagonists or alphametyrosine (5).

- Our patient was diagnosed with 2 separate NETs: a pancreatic neuroendocrine tumor and a pheochromocytoma. She was placed on Doxazosin with excellent control of her blood pressure with plans for staged resection of her two NETs.

- Appropriate biochemical testing completely changed her medical management strategy and provided hope for curative treatment.

Teaching Points

- All patients with adrenal incidentalomas should be evaluated for the possibility of malignancy and subclinical hormonal hyperfunction.
- Prompt surgical intervention is crucial because untreated pheochromocytoma may result in significant cardiovascular complications.
- Multidisciplinary approach is necessary for appropriate diagnosis, management and follow up of patients.

Bibliography