Abstract citation ID: bvac150.189

Adrenal

PSAT013

Elevated Plasma Free Metanephrines with No Evidence of Pheochromocytoma in a Patient with Multiple Endocrine Neoplasia Type 2A

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Background: Adrenal medullary hyperplasia (AMH) is arbitrarily defined by adrenal medullary proliferation measuring less than 1 cm while larger lesions are considered pheochromocytoma. Some patients with MEN2 present with adrenal medullary hyperplasia (AMH), which is a precursor of pheochromocytomas.

Clinical Case: A 22-year-old female with a past medical history of MEN2A. She is positive for germline mutation in RET proto-oncogene at 10q11.2 exon 11, position 634 from cysteine to arginine.

She was evaluated for a 1cm left thyroid nodule, which required total thyroidectomy for Medullary Thyroid Cancer. Pathology revealed a 0.5cm deposit on the largest dimension, clear margins, and no angiovascular, or lymphovascular invasion, surgery was complicated by hypoparathyroidism. She complained of intermittent anxiety and fatigue but was otherwise asymptomatic. Physical examination is unremarkable except for thyroidectomy scar and multiple exostoses.

On 8/5/2011 plasma-free metanephrines were elevated at 83 pg/ml (<57pg/ml) and Free normetanephrine was 45 pg/ml (<145pg/ml).

Over the next years, her plasma-free metanephrine levels have been checked approximately every 6 months ranging between 63-106pg/ml with free plasma normetanephrine in normal ranges (30-81pg/ml). However, on 2/2021, her plasma normetanephrine level increased to 304pg/ml. More recently, on 11/30/2021 her free normetanephrine levels reached 671pg/ml and free Metanephrine 131pg/ml. She had two MRIs of the abdomen and pelvis over the years and both showed unremarkable adrenal glands. NM I-123 MIBG scintigraphy reported physiologic symmetric activity in the adrenal glands. Finally, she had GA-68 DOTATATE PET/CT with no suspicious radiotracer avid lesion in the abdominopelvic organs. Given the persistent elevation on plasma metanephrines, we decided to start the patient on Doxazosin prophylactically while we continue biochemical and imaging surveillance.

Conclusion: Our patient has a persistent elevation in plasma-free metanephrines with no evidence of pheochromocytoma in abdominal imaging. The earliest abnormality in the adrenal medulla in patients with MEN 2 is diffuse hyperplasia with a reduction in the cortical/medullary ratio (1). This hyperplasia is considered to be a precursor of pheochromocytoma in patients with MEN 2 and in our case the most likely explanation for the progressive elevation in plasma-free metanephrines (1). There is a strong molecular relationship between AMH and Pheochromocytoma (2), close monitoring for progression to pheochromocytoma is necessary for all patients with AMH. The presence of

markedly elevated plasma metanephrines in an asymptomatic patient with no evidence of pheochromocytoma poses a therapeutic dilemma for preemptive use of alpha blockade and the potential need for a medical alert bracelet.

References: (1) Cho, K et al "Adrenal Medullary disease in Multiple Endocrine Neoplasia II". AJR 134: 23-29. (2) Korpershoek, E et al "Adrenal Medullary Hyperplasia Is a Precursor Lesion for Pheochromocytoma in MEN2 Syndrome". Neoplasia. 2014 Oct; 16(10): 868–873

Presentation: Saturday, June 11, 2022 1:00 p.m. - 3:00 p.m.