

Negative Urinary Fractionated Metanephrines and Elevated Urinary Vanillylmandelic Acid in a Patient with a Sympathetic Paravesical Paraganglioma

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Received date: Jan 06, 2015, Accepted date: Jan 07, 2015, Published date: Jan 9, 2015

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Clinical Image

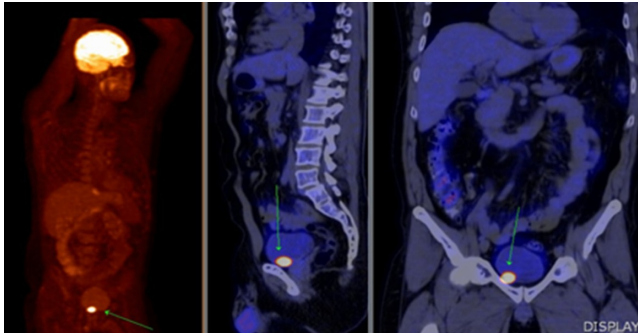


Figure 1: Negative Urinary Fractionated Metanephrines and Elevated Urinary Vanillylmandelic Acid in a Patient with a Sympathetic Paravesical Paraganglioma.

Keywords: Paraganglioma; Positron-emission tomography; Neuroendocrine tumors; Urinary bladder neoplasms; Vanillylmandelic acid

A 50-year-old man with diffuse abdominal pain had a pelvic CT scan that revealed a bladder tumor. Biopsy was performed by urology and the patient suffered a hypertensive crisis during the procedure. He hadn't had a personal or family history of hypertension, flushing, palpitations or syncope. With a conclusive pathology of paraganglioma (immunohisto-chemistry reported intense and diffuse positive for chromogranin and synaptophysin, focal reactivity for the S100 protein and a proliferation index Ki67% of 1%), biochemical tests were carried out. Fractionated urinary metanephrines were normal: total metanephrines: 614 ug/24 hrs (RV <900 ug/24 hrs), normetanephrine: 423 ug/24 hrs (RV <600 ug/24 hrs), metanephrine: 141 ug/24 hrs (RV <300 ug/24 hrs); chromogranin A: 6.40 ng/ml (RV: 1.9-15 ng/ml) but vanillylmandelic acid was elevated: 28.2 mg/24hrs (RV <13.6 mg/24

hrs). An 18 fluorodeoxyglucose PET/CT study (18 FDG PET/CT) showed an abnormal glucose uptake in the bladder with 16.9 SUVs. No distant metastases were reported. Surgical resection was performed successfully and antihypertensive medication was discontinued. The patient remains asymptomatic and normotensive (unmedicated). Results of genetic testing are pending [1-3]. Bladder paragangliomas (bPGL) are sympathetic tumors that represent 0.06% of all bladder tumors. Approximately 50-60% of these tumors present with adrenergic symptoms; catecholamine crisis during urination might occur in 30% and hematuria is the main manifestation in 60%. Up to 40% of bPGL are malignant. 18 FDG PET/CT are a sensitive test to evaluate malignancy. It has shown a good usefulness in evaluating the extension of this sickness due to the fact that it may identify lesions undetected by tomography or magnetic resonance imaging. Its sensitivity is about 74% in patients with metastatic PGL, and it can reach 100% in SDHB (succinate dehydrogenase complex, subunit B) gene mutation carriers.

Acknowledgements

We thank José Luis Torres Grajales and Clínica Las Americas for helping us to get the image.

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