Unusual Case of a Hormone-Secreting SDHC Head and Neck Paraganglioma

Hans K. Ghayee1; Brandon Isaacson2; Megan Farley3; Arielle Click4; Karel Pacak5; and Jerry W. Shay4

1Department of Internal Medicine, University of Texas Southwestern Medical Center, Dallas, TX
2Department of Otolaryngology, University of Texas Southwestern Medical Center, Dallas, TX
3Department of Cancer Genetics, Simmons Comprehensive Cancer Center, University of Texas Southwestern Medical Center, Dallas, TX
4Department of Cell Biology, University of Texas Southwestern Medical Center, Dallas, TX
5National Institutes of Health, Bethesda, MD

Case: A 66 year-old man with history of primary hyperparathyroidism was being evaluated for a parathyroidectomy. His labs at the time showed PTH= 214pg/ml (12-88), calcium= 10.9mg/dL (8.4-10.3), creatinine= 1.48mg/dL (0.6-1.4), albumin= 3.7g/dL(3-5). A DXA scan of the left femoral neck showed a T-score -1.6 and distal forearm T-score -2.2. Sestamibi scan surprisingly revealed: destructive lytic processes with increased radiotracer uptake in the left skull base as well as uptake in a lymph node in the superior mediastinum. Increased uptake was also seen in the posterior and inferior to the right thyroid lobe, which represented a parathyroid adenoma. Magnetic resonance imaging (MRI) revealed a 4.8cm x 5.3cm x 6.2 cm left jugular foramen mass representative of a glomus jugulare paraganglioma. A 24 hr urine study revealed: dopamine= 909pg/ml (<60), epinephrine= 177pg/ml (<84), norepinephrine= 5655pg/ml (<420), total metanephrines = 2939mcg/24hrs (<832mcg/24hrs), normetanephrines = 2665mcg/24hrs (<676mcg/24 hrs). Genetic testing was consistent with a SDHC mutation c.397C>T. Unlike most cases of SDHC mutations, this patient’s tumor was secreting elevated amounts of catecholamines and the tumor size was quite large. The differences between this specific patient’s hormone-secreting SDHC tumor versus the more common, silent SDHC tumors were investigated by expression studies. Conclusion: It should not be assumed that head and neck paragangliomas are non-secretory unless measurements of catecholamines and metanephrines have been evaluated. Distinction between hormone secreting and non-hormone secreting paragangliomas requires further research.