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

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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.