Metastatic Cardiac Paraganglioma: A Rare Subset of Mediastinal Paragangliomas

Running Title: "Metastatic Cardiac Paraganglioma"

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Case Presentation: A 55-year-old man with a history of hypertension, hyperlipidemia, type 2 diabetes and unexplained tachycardia was transported from his primary care clinic to the emergency department for severe hypotension, chest tightness, and dizziness. An electrocardiogram showed ST elevations in the inferior leads prompting admission and emergent cardiac catheterization. The coronary angiogram (Figure 1) revealed complete obstruction of the right coronary artery with extensive collateral vasculature and a possible aneurysm or epicardial mass. He underwent an aortic CT (Figure 2) and cardiac MRI which demonstrated a 7.3 cm hypervascular mass arising from the visceral pericardium, insinuated between the proximal aorta and pulmonary trunk with parasitized blood supply from the right coronary artery. Additionally, an indeterminate hepatic lesion was discovered. Further evaluation with Gallium-68 Dotatate PET-CT (Figure 3) revealed an intensely avid mediastinal mass and slightly avid liver lesion. During subsequent endocrinology evaluation, the patient reported palpitations elicited with 3+ cups of coffee daily but denied headache, diaphoresis, pallor, flushing, nausea, abdominal pain, diarrhea, rash, tremor, agitation, or family history of tumor. Physical exam revealed no abnormal skin findings. 24 hour urine studies revealed normal metanephrines and epinephrine but with elevated norepinephrine 159mcg/24hr [15-80], dopamine (undetectably high), and 5-Hydroxyindoleacetate 15mg/24hr [0-6]. Plasma fractionated metanephrines were normal. What is the diagnosis?

Answer

Liver biopsy confirmed the diagnosis of metastatic paraganglioma, specifically cardiac paraganglioma due to its primary location. Palpitations and tachycardia resolved with alpha and beta-adrenergic blockade. Generally, first line treatment is surgery. Due to the presence of metastases at diagnosis, the patient is currently undergoing four cycles of neoadjuvant chemotherapy to improve the surgical outcome. Genetic testing, including SDHx evaluation, is pending. There are less than 200 reported cases of mediastinal paraganglioma in the literature, even less for cardiac paraganglioma as present in our patient. Dopamine secreting paragangliomas are rare and usually silent, leading to large tumor size and metastases prior to detection. Lack of clinical symptoms may be due to the vasodilatory effects of dopamine. There are only a handful of dopamine secreting mediastinal paragangliomas reported in the literature, but dopamine evaluation was useful in this case due to the minimal change in catecholamines and metanephrines and the negative plasma fractionated metanephrines. Gallium-68 Dotatate is the most sensitive and specific imaging test for detecting paragangliomas. In this case an indeterminate liver lesion was identified and provided a target for biopsy, ultimately altering initial management.

Figure 1
The proximal RCA fills a vascular structure with an abrupt cutoff and blush of contrast representing the parasitized blood supply of the patient's cardiac paraganglioma (arrows).

Figure 2
Sagittal image revealing a large hyper-enhancing mass along the anterior margin of the right heart (line), insinuating between the right ventricular outflow tract and aortic root. Also note a large pericardial effusion (arrow).

Figure 3
Scout view demonstrating intense abnormal dotatate uptake within the mediastinum (right arrow) and uptake above background within the right liver lobe (left arrow). There is physiologic uptake within the salivary glands, liver, spleen, adrenals, bowel, and urinary collecting system.

References

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