J of Nuclear Medicine Technology, first published online June 13, 2017 as doi:10.2967/jnmt.116.182212

NUCLEAR IMAGING IN A CARDIAC PARAGANGLIOMA

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Conflict of interest:
Nada Almenieir, Shawn Karls, Vilma Derbekyan, and Robert Lisbona declare that they have no conflict of interest.

Compliance with Ethical Standards:
No funding was obtained from any source.
All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Manuscript word count: 728 words.

Running Title:
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Abstract: We report a case of a cardiac paraganglioma in the right atrioventricular groove, where the use of different nuclear medicine studies aided in the diagnosis.

Keywords: Paraganglioma, Cardiac, Succinate dehydrogenase/deficiency.

Introduction:

Nuclear imaging is useful for the evaluation of suspected paragangliomas. ^{123}I/^{131}I-MIBG scintigraphy is the initial recommended study, however it is associated with high false-negative rate, as the sensitivity is 18%–50% (*Taïeb et al*). Additional nuclear imaging with ^{111}In-octreoscan can be used with a sensitivity and specificity of 94 and 75% (*Telischi et al*). Also, studies show that FDG-PET is as specific as MIBG for detection of the primary tumor and metastases and even more sensitive than 123I-MIBG and CT/MRI for detection of metastatic disease (*Timmers et al*). Mutations in the SDHx genes are associated with paragangliomas. All hereditary *SDHx* paraganglioma syndromes have an autosomal dominant inheritance pattern with varying penetrance (*Welander et al*).

Case Report:

A 25-year-old female was referred to cardiology for palpitations. An echocardiogram showed a large mass anterior to the right ventricle, measuring 6x5 cm. The mass appeared to be compressing the RV in the right AV groove. A CT scan of the chest (figure 1) and a cardiac MR were performed for evaluation. The CT scan showed the mass to be heterogeneously hyperenhancing, with a central stellate "scar" centered on the right atrium, with imaging characteristics suggestive of a cardiac hemangioma. Cardiac paraganglioma was in the differential. The cardiac MR showed the lesion to be predominantly solid with several small cystic components, but it did not otherwise contribute to the characterization of the lesion.

Symptomology were suspicious for a neuroendocrine tumor, with an elevated serum Chromogranin A, 24-hour urine, Norepinephrine, Dopamine and Normetanephrine. An Octreoscan was performed and showed intense increased uptake by the mass, figure 2. For the evaluation of treatment options, an ^{131}Iodine-MIBG scan was performed and showed only mild heterogenous radiotracer accumulation in the mass, figure 3. An FDG PET/CT was obtained to rule out metastasis, figure 4. It showed the mass to be intensely hypermetabolic with central necrosis, SUVmax was 31, with no metastasis. A MUGA scan was performed to evaluate cardiac dynamics, which showed no impairment (LVEF 64%, RVEF 52%).

The patient underwent surgical resection of the mass. The right ventricle was repaired with a bovine pericardial patch. As the pre-surgery coronary angiogram showed the proximal RCA to be dilated with a large branch feeding the mass, coronary artery bypass grafting of the RCA was performed. Pathology confirmed the mass to be a paraganglioma, with a Ki-67 labeling index of 16%. Chromogranin A levels dropped from 380.7 to 26 ng/mL, 24-hour urinary Norepinephrine also decreased from 2249 to 101 nmol/d, Dopamine from 26096 to 535 nmol/d, and normetaneprhine from 1266 to 66 nmol/d. Immunohistochemistry showed cytoplasmic loss of Succinate Dehydrogenase Complex Iron Sulfur Subunit B (SDHB) expression. The patient is currently undergoing genetic testing.

Discussion:

In this case, an intracardiac tumor produced a diagnostic dilemma. While radiographic studies provided structural information, an Octreoscan confirmed the clinical suspicion of a neuroendocrine tumor. ^{18}F-FDG PET can also be useful, with the additional value of quantitative SUV. Although the link between the malignant potential of paragangliomas and Ki-67 has not been fully established, in our case the high ki-67 expression was associated with high SUVmax.

Conclusion:

Iodine-MIBG scans are associated with a high false negative in paragangliomas. ^{111}In-octreotide scintigraphy and ^{18}F-FDG PET are useful secondary scans.

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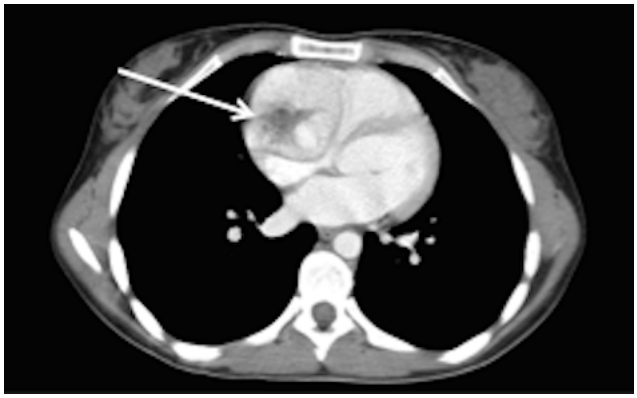

Fig 1. (A) CT scan of the chest showing the hyperenhancing mass with hypoenhancing thin rim and central stellate "scar".

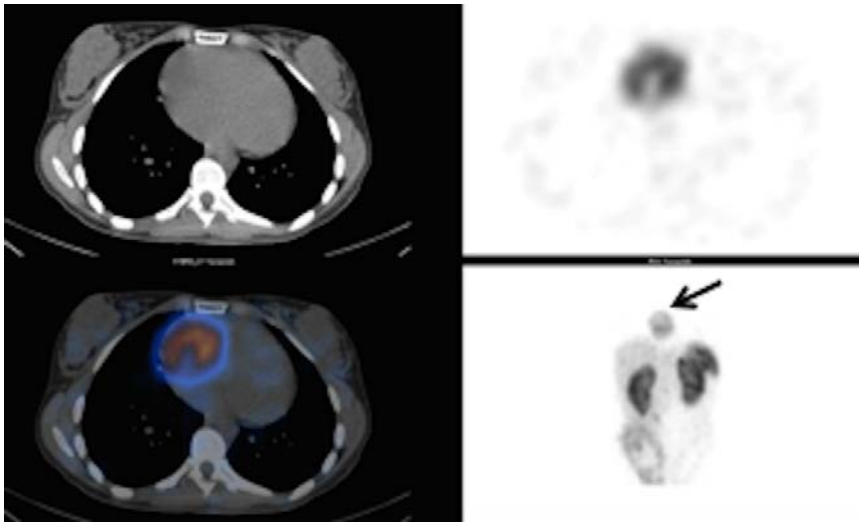

Fig 2. An ^{111}Indium-Octreoscan SPECT/CT showing focal intense increased uptake by the mass.

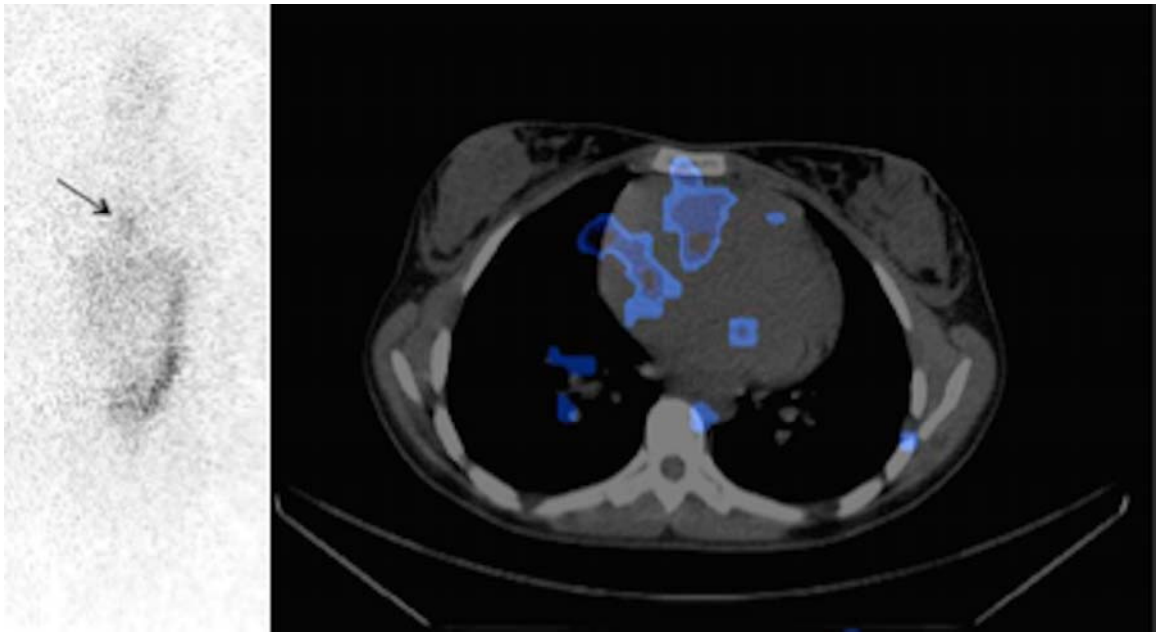

Fig 3. 131 Iodine-MIBG anterior planar image, and SPECT/CT showing only mild heterogeneous radiotracer accumulation in the mass.

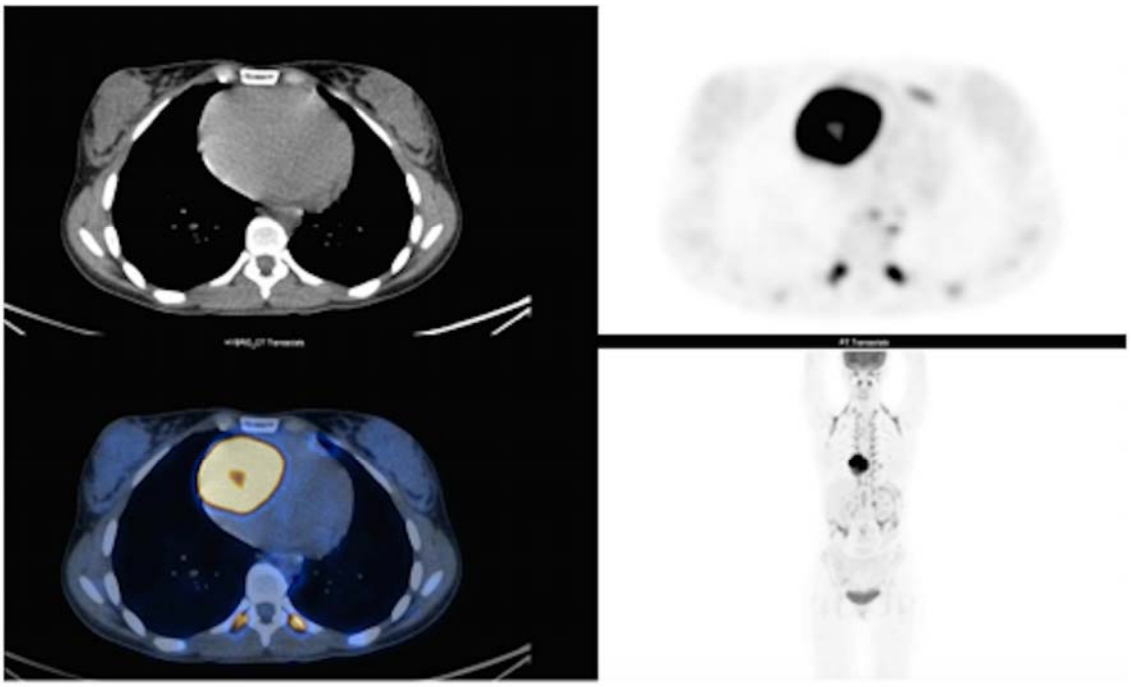

Fig 4. ^{18}F-FDG PET/CT showing the mass to be intensely hypermetabolic with central necrosis.