Cardiac Paraganglioma
Multimodality Imaging of a Rare Tumor

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ABSTRACT
Paragangliomas are rare tumors of chromaffin cells arising from an extra-adrenal location. Unlike pheochromocytomas, they are seldom functional. We present a case of pericardial paraganglioma incidentally encountered on an echocardiographic study, focusing on the characteristic features the tumor demonstrates on different imaging modalities. (Level of Difficulty: Intermediate.) (J Am Coll Cardiol Case Rep 2021;3:273-5) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 38-year-old man with a family history of early-onset coronary artery disease proceeded on a scheduled basis for echocardiographic evaluation. Findings were compatible with normal cardiac structure and function; however, an echogenic, difficult-to-illustrate mass was noted from the suprasternal view beneath the aortic arch and the right pulmonary artery. Color Doppler imaging with low-velocity scale yielded findings suggestive of a mass with a rich blood supply with continuous flow demonstrated on the feeding arteries (Figure 1A, Video 1).

A magnetic resonance imaging scan was subsequently performed, revealing a well-circumscribed mass between the left atrium, the ascending aorta, the right atrium, and the superior vena cava and under the right pulmonary artery. The mass was isointense to myocardium on T1-weighted images and highly hyperintense on T2-weighted, non-fat-suppressed images. These imaging features were considered as potentially consistent with a diagnosis of paraganglioma.

A computed tomographic coronary angiography followed, which confirmed the presence of the aforementioned mass, with its dimensions calculated at 5.2 × 3.7 × 4.6 cm. Abundant blood perfusion was demonstrated, with the feeding arteries arising from both the right coronary artery and branches of the left circumflex artery (Figure 1B).

To confirm or rule out an adrenergic or neuroendocrine tumor and exclude possible other tumor body location, iodine-123 metaiodobenzylguanidine scan and technetium-99m tectrotyd scintigraphy were performed. Planar and hybrid images were obtained by using a hybrid single photon emission computed tomography/computed tomography camera (GE Discovery NM/CT 670, Milwaukee, Wisconsin). Scintigraphic images revealed an increased uptake of both radiotracers in the anatomical region between the left and right atrium of the heart under the right pulmonary artery, confirming a mass with both adrenergic and neuroendocrine characteristics, most probably paraganglioma (Figures 1C and 1D).

The patient was scheduled for surgical resection of the mass, and a pre-operative invasive coronary angiography was performed, confirming the findings of computed tomographic coronary angiography with respect
to coronary anatomy and tumor perfusion (Figures 1E and 1F, Videos 2 and 3). The patient eventually underwent complete surgical excision of the mass, which was intraoperatively found to be partially intrapericardial, at the region of the transverse sinus.

Paragangliomas are rare tumors of chromaffin cells arising from an extra-adrenal location and can be associated with inherited syndromes in some cases. Unlike pheochromocytomas, they are rarely functional (1). Intrapericardial paragangliomas are extremely rare (2) and tend to be located in the aortopulmonary groove at the level of the para-aortic sympathetic ganglia (1). They are frequently clinically silent, with symptoms not occurring until the tumor has enlarged sufficiently to result in compression phenomena, clinically expressed as angina or dyspnea.

In the present case, the paraganglioma constituted an incidental finding of an off-label echocardiographic study in an asymptomatic individual. Although multimodality imaging and a multidisciplinary medical team are cornerstones in establishing a definite diagnosis and ultimately offering a curative treatment, our case highlights the value of the meticulous examination of paracardiac spaces and structures when performing a routine echocardiographic study.

**FIGURE 1** Multimodality Imaging of a Pericardial Paraganglioma

(A) Suprasternal echocardiographic view, where under the aortic arch and the right pulmonary artery, an echogenic mass was found and on color Doppler imaging with low-velocity scale appears as a richly perfused blood mass with continuous flow of the feeding arteries (a). Yellow arrows show the mass. (B) Computed tomography reveals a well-circumscribed mass under the ascending aorta and the right pulmonary artery. Yellow arrows show the mass. (C) Scintigraphic images with technetium-99m tetrotyd revealed an increased uptake of the radiotracer in the anatomical region between the left and right atrium of the heart and under the right pulmonary artery, confirming a mass with neuroendocrine characteristics (increased expression of somatostatin receptors 2, 3, 5). (D) Scintigraphic images with iodine-123 metaiodobenzylguanidine scan confirms that the mass possesses adrenergic (increased uptake of iodine-123 metaiodobenzylguanidine) characteristics. (E, F) Selective angiographies of the (E) left and (F) right coronary arteries, demonstrating the feeding arteries of the mass, arising from both the right and the left circumflex (CMX) coronary artery. 99mTc = technetium-99m; Ao = aorta; LAD = left anterior descending coronary artery; MIBG = metaiodobenzylguanidine; PA = pulmonary artery; RCA = right coronary artery; RPA = right pulmonary artery.
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APPENDIX For supplemental videos, please see the online version of this paper.