An Incidentaloma in the Cardiology Clinic

To the Editor:

Paragangliomas are rare neuroendocrine tumors of neural crest origin. In this report, we describe the exhaustive diagnostic workup involved in the diagnosis of a para-
ganglioma, which was first noted as an incidental mass in the proximity of left atrium on a routine transthoracic echocardiogram (TTE).

CASE REPORT

A 55-year-old man with a history of valvular cardiomyopathy, hypertension, diabetes mellitus, and nonobstructive coronary artery disease presented to the cardiology clinic for follow-up after undergoing bioprosthetic aortic valve replacement for severe aortic regurgitation 2 years ago. He denied any dys-
pnea, palpitations, chest pain, or syncope. Physical exami-
nation was remarkable for blood pressure of 127/85 mm Hg and an early-peaking systolic murmur at the right upper sternal border. He was taking aspirin, simvastatin, carvedilol, losartan, and furosemide. A baseline TTE was ordered for follow-up on the bioprosthetic valve function.

On the TTE, the left ventricular ejection fraction and the bioprosthetic valve function were normal, but an incidental well-circumscribed mass measuring 35 × 22 mm was noted in the proximity of the left atrium (Figure, A). On the basis of the TTE images, it could not be ascertained whether the mass was intracardiac or extracardiac. Therefore, a transesophageal echocardiogram was performed, which revealed similar characteristics of the mass as on the TTE, but it was also limited in localizing the lesion (Figure, B, Video, A, available online). Color Doppler (Video, B, available online) suggested a highly vascular mass, concerning for malignancy. Thus, the patient was sent for a cardiac magnetic resonance imaging for further evaluation, which demonstrated a 37 × 24 × 47-mm extracardiac oval mass adjacent to the posterior wall of the left atrium with homogeneously increased T2 signal intensity (Figure, C). On postcontrast T1 fat-saturated image, the mass had significant homogeneous enhancement (Figure, D). The differential diagnosis for this mass included lymphangioma, teratoma, hemangioma, or paraganglioma, or, less likely, lymphoma, pericardial mesothelioma, or metastatic tumor. The patient did not have any systemic signs or symptoms to suggest a malignant process. To further characterize the mass, a positron emission tomography computed tomography scan was completed. The mass was noted to have a maximum standardized uptake value of 18.7, suggestive of a malignant process (Figure, E).

Video-assisted thoracic surgery was performed to biopsy the mass. The gross architecture of the obtained specimen showed a neoplasm forming nests (Figure, F). The cells were mildly pleomorphic with smudgy nuclei. Otherwise, the nuclei had a relatively homogenous chromatin pattern with barely visible nucleoli. The specimen was stained with antibodies directed against cytokeratin, chromogranin, synaptophysin, OCT-4, C-KIT, CD20, and S-100. The neoplastic cells were positive only for chromogranin (Figure, G) and synaptophysin (Figure, H), suggesting a diagnosis of paraganglioma. Elevated urine and plasma metanephrines with normal parathyroid hormone and calcitonin levels were consistent with functioning pheochromocytoma. A 123I-metaiodobenzylguanidine scan was completed, which revealed localized disease as described before without metastasis.

DISCUSSION

Intracardiac or extracardiac paraganglioma is a rare tumor of neural crest origin. The presentation varies on the basis of the functional status and location of the tumor. Most of the mediastinal paragangliomas are nonfunctional, and approximately 90% of them are benign. The usual presentation is due to the mass-like effect of the tumor on the surrounding structures causing dyspnea, dysphagia, or ischemic symp-
toms. Surgical resection of functioning paraganglioma can be complicated because of the location and vascular nature of the tumor.

CONCLUSIONS

Because of the location of the tumor and the patient’s history of a sternotomy, surgical resection was technically difficult. The patient was given alpha-blockers and referred for 131I-metaiodobenzylguanidine treatment. This case underscores
the challenges associated with diagnosis and management of intracardiac or extracardiac paragangliomas.

Danesh K. Kella, MD
Rupak Desai, MBBS
Leon Rubinsztein, MD
Jeranfel Hernandez, MD
Andro Kacharava, MD, PhD

aDepartment of Medicine
Emory University School of Medicine
Atlanta, Ga

bDepartment of Cardiology
Atlanta Veterans Affairs Medical Center
Ga

cDepartment of Radiology
Atlanta Veterans Affairs Medical Center
Ga

http://dx.doi.org/10.1016/j.amjmed.2016.10.033

References

SUPPLEMENTARY DATA
Supplementary Videos accompanying this article can be found in the online version at http://dx.doi.org/10.1016/j.amjmed.2016.10.033.