JACC: CASE REPORTS VOL. 4, NO. 14, 2022

CASE REPORT

ADVANCED

CLINICAL CASE SERIES: SURGERY AND INTERVENTIONS

Role of Multimodality Imaging and Preoperative Management in Intrapericardial Paragangliomas

Experience From a Case Series

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ABSTRACT

Intrapericardial paragangliomas are rare, highly vascular tumors that frequently adhere to adjacent structures and blood vessels, making surgical resection challenging. In this case series, we discuss the role of multimodality imaging and preoperative embolization in the management of 3 patients presenting with intrapericardial paragangliomas. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2022;4:871–877) © 2022 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/).

Paragangliomas (PGLs) are neuroendocrine tumors outside the adrenal glands. Intrapericardial PGLs are extremely rare.[1] These tumors are highly vascular and frequently adhere to adjacent structures, rendering surgical resection challenging.[2] Therefore, intrapericardial PGLs often necessitate cardiopulmonary bypass (CPB) for surgical resection.[2] In this case series, we discuss the role of multimodality imaging in the diagnostic evaluation of intrapericardial PGLs and the potential of preoperative vascular embolization in decreasing bleeding risk.

LEARNING OBJECTIVES

- To appreciate the role of multimodality imaging in the diagnosis of intrapericardial paragangliomas.
- To highlight the importance of preoperative embolization of feeding arteries of the paraganglioma to aid surgical resection in certain cases.

CASE 1

A 44-year-old man presented with pleuritic chest pain, palpitations, and insomnia. Relevant history included pericardial tamponade needing emergent pericardiocentesis 2 weeks earlier for which he was taking colchicine and ibuprofen. Blood pressure was 114/71 mm Hg, pulse was 88 beats/min, respiratory rate was 17 breaths/min, and temperature was 36.5 °C. A physical exam including cardiopulmonary exam

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received February 4, 2022; revised manuscript received April 26, 2022, accepted April 28, 2022.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2022.04.014

ABBREVIATIONS AND ACRONYMS

CMR = cardiac magnetic resonance

CPB = cardiopulmonary bypass

CT = computed tomography

FDG = fluorodeoxyglucose

LCX = left circumflex artery

PET = positron emission tomography

PGL = paraganglioma

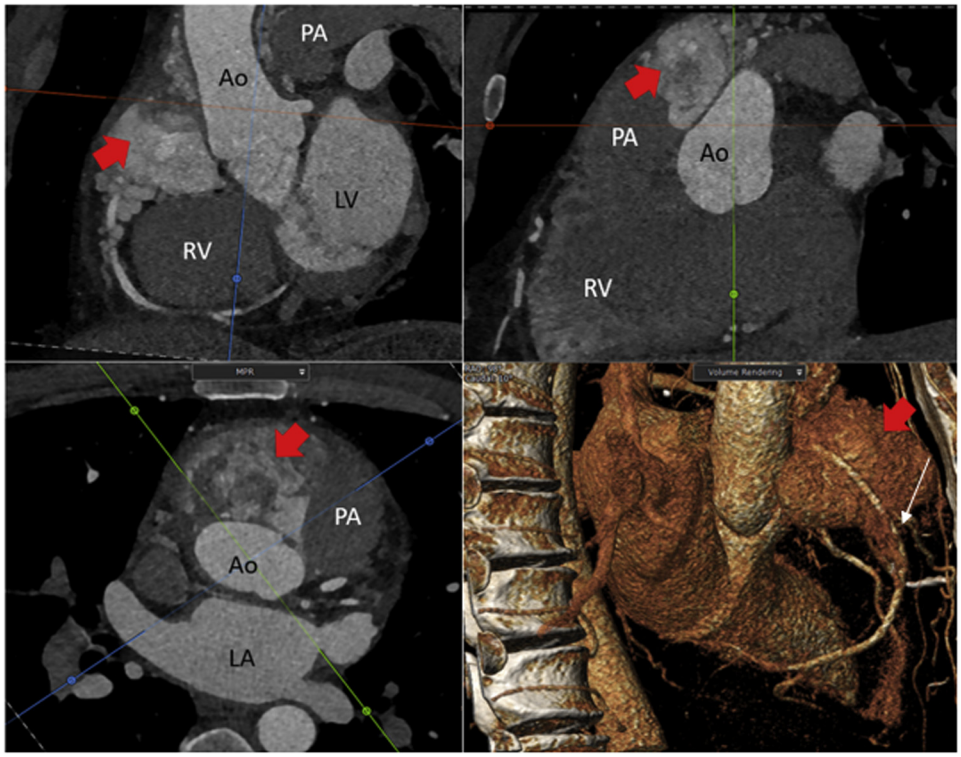
RCA = right coronary artery

TTE = transthoracic echocardiography

was unremarkable. Computed tomography (CT) angiography of the chest showed a markedly heterogeneous, partially hyperdense structure abutting the right cardiac wall (**Figure 1,** Video 1). Unenhanced CT attenuation ranged between 41 and 63 HU. Cardiac magnetic resonance (CMR) showed a large, well-circumscribed hypervascular mass centered in the right atrioventricular groove partially encasing the right coronary artery (RCA) (**Figure 2,** Video 2). Subsequently, positron emission tomography (PET)-CT using F-18 fluorodeoxyglucose (FDG) showed a markedly FDG-avid mass with no evidence of metastasis. Given the characteristics on imaging, the mass was suspicious for PGL versus nonseminomatous germ cell tumor. Transthoracic echocardiography (TTE) demonstrated normal right ventricular function and a large, well-circumscribed anterior mass adjacent to the right heart chambers and RCA without significant extrinsic compression (**Figure 3**). The mass was highly vascular on Doppler TTE (Video 3). Lactate dehydrogenase, alpha-fetoprotein, human chronic gonadotropin, C-reactive protein, and 24-hour urine fractionated catecholamines and metanephrines were normal. A gallium-68 DOTATATE (DOTA-octreotate) PET-CT showed that the mass was markedly DOTATATE-avid—highly suggestive of PGL (**Figure 4**).

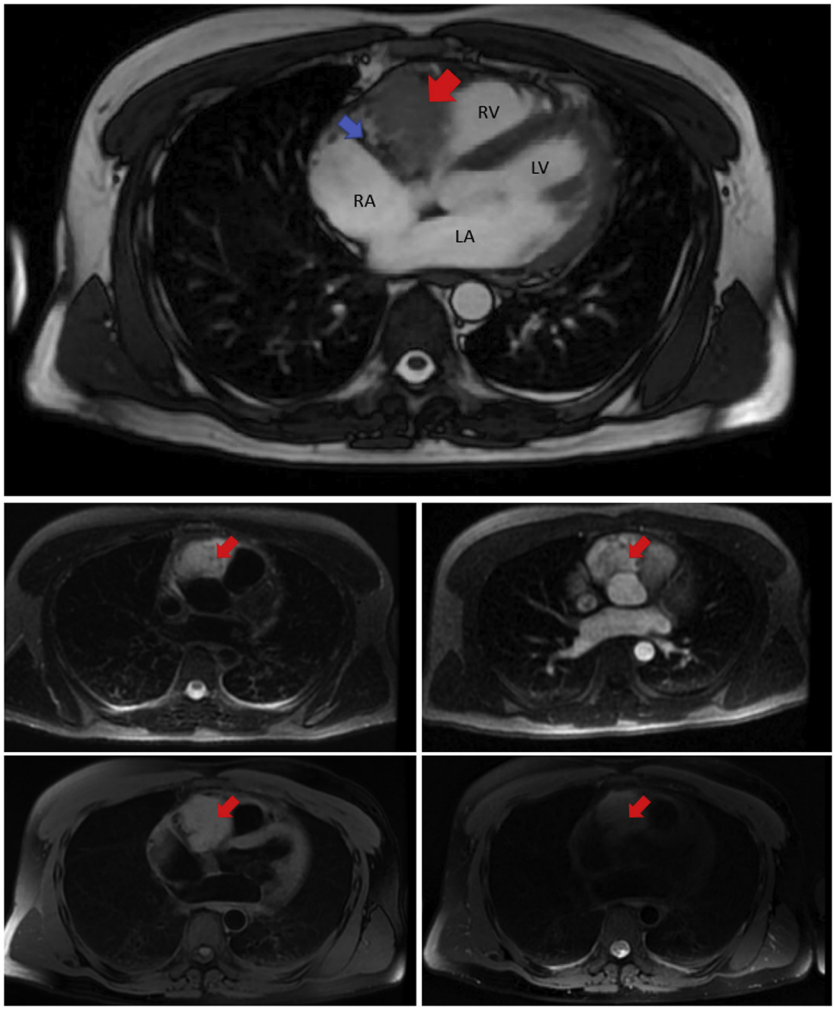
Surgical resection was planned. Appropriate alpha- and beta-adrenergic blockade was started. A gated cardiac CT angiogram with 3-dimensional printing of heart, performed for surgical planning, showed the PGL encased and narrowed the proximal RCA and partially encased the aortic root and ascending aorta. Coronary angiography showed that the proximal RCA was 100% obstructed by the PGL but completely collateralized through left-to-right collaterals (**Figure 5**). It also showed the PGL received feeder vessels from the RCA, left main artery, and left circumflex artery (LCX) (**Figure 5**). Embolization of the feeder vessels was considered but was not feasible.

FIGURE 1 Computed Tomography Angiography of the Chest Showing the Paraganglioma



The mass **(red arrows)** is shown in the rotated coronal view **(left upper)** (Video 1), in the axial view **(left bottom)**, in the rotated sagittal view **(right upper)**, and with 3-dimensional rendering **(right bottom)**. Note that the right coronary artery **(open arrow)** is encased by the mass. Ao = aorta; LA = left atrium; LV = left ventricle; PA = pulmonary artery; RV = right ventricle.

FIGURE 2 Cardiac Magnetic Resonance Imaging of the Paraganglioma



(Top) Axial view shows the mass **(red arrow)** is centrally positioned in atrioventricular groove and partially encases the right coronary artery **(blue arrow)**. **(Middle panels)** The lesion is hyperintense on T2-weighted images **(left)** and shows intense hyperenhancement on perfusion imaging **(right)** suggesting paraganglioma. **(Bottom)** The lesion is nonsuppressed on fat saturation T1 **(left)** and T2 **(right)** sequences distinguishing this from lipoma. RA = right atrium; other abbreviations as in Figure 1.

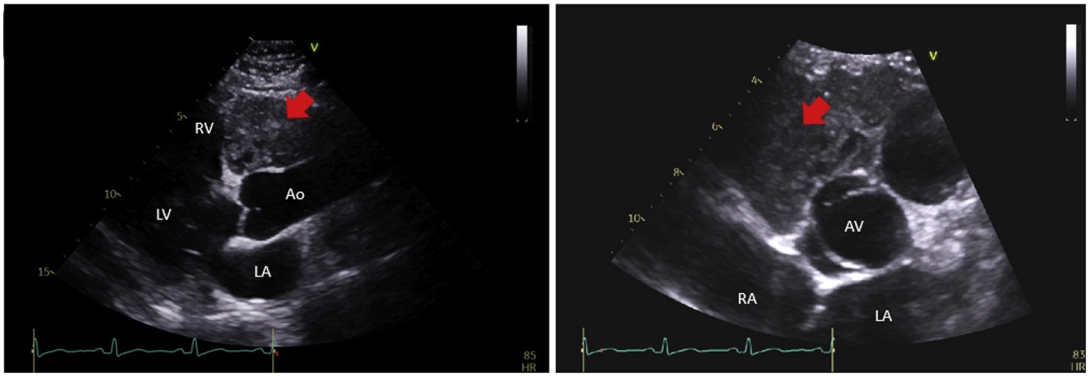
Surgical resection was subsequently performed successfully on CPB.

CASE 2

A 58-year-old woman presented with palpitations, tachycardia, fatigue, and chest pressure worse in the recumbent position for 4 months. She had a history of a presumed mediastinal PGL first diagnosed 14 years prior, which then enlarged from 3 to 5 cm in diameter 6 years later. Blood pressure was 130/70 mm Hg, pulse was 126 beats/min (regular), and respiratory rate was 15 breaths/min. A physical exam was otherwise unremarkable. Holter monitor was unremarkable. CMR showed a large hypervascular mass in the mediastinum posteriorly (7.8 × 6.2 × 6.4 cm) lying immediately superior to the left atrium with imaging features consistent with intrapericardial PGL. Its vascular supply was shown to arise from the proximal descending thoracic aorta, distal RCA, and 2 collateral vessels from the LCX. Her 24-hour urine showed normal fractionated catecholamines and metanephrines.

The patient was initially planned for trans-sternal resection of the intrapericardial PGL, and a

FIGURE 3 Transthoracic Echocardiography of the Paraganglioma



Parasternal long-axis **(left)** and parasternal short-axis **(right)** views showing the PGL **(red arrow)**. The mass clearly abuts the RV and the ascending Ao. Abbreviations as in Figures 1 and 2.

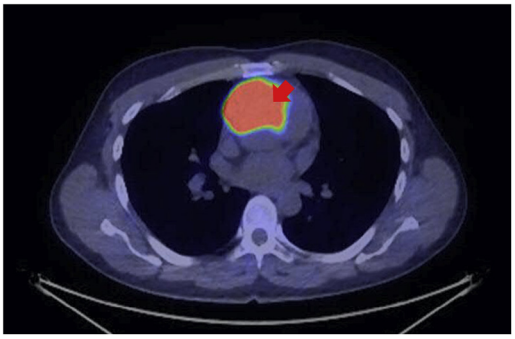
3-dimensional model of the patient's heart was printed (Figure 6, left and middle panels). However, owing to extensive bleeding despite CPB, the surgery was aborted. The patient underwent coiling of the bronchial artery, the 2 feeder vessels of the LCX, and 1 feeder vessel off the distal RCA to devascularize the tumor prior to a second attempt of surgical resection (Figure 7, Video 4). The patient then underwent successful surgical resection of the PGL (Figure 6, right).

CASE 3

A 67-year-old man underwent catheter ablation for atrial fibrillation. Relevant history included resolved tachycardia-induced cardiomyopathy, multifocal thromboembolic strokes, and remote right-sided nephrectomy for an oncocytoma. His post-ablation cardiac CT scan incidentally showed a 1-cm circumscribed mass inferior to the proximal left anterior descending artery with avid enhancement and delayed washout on imaging, which was unchanged from the pre-ablation cardiac CT. CMR demonstrated avid perfusion and T2 hyperintensity of the mass and normal left ventricular function. Given imaging characteristics and location, the mass was suspicious for a PGL. The 24-hour urine fractionated catecholamines and metanephrines were normal. Symptom-wise, the patient described fatigue despite maintaining sinus rhythm. His vital signs included a blood pressure of 142/74 mm Hg and pulse of 58 beats/min. He was started on alpha- and beta-adrenergic blockade in preparation for further testing.

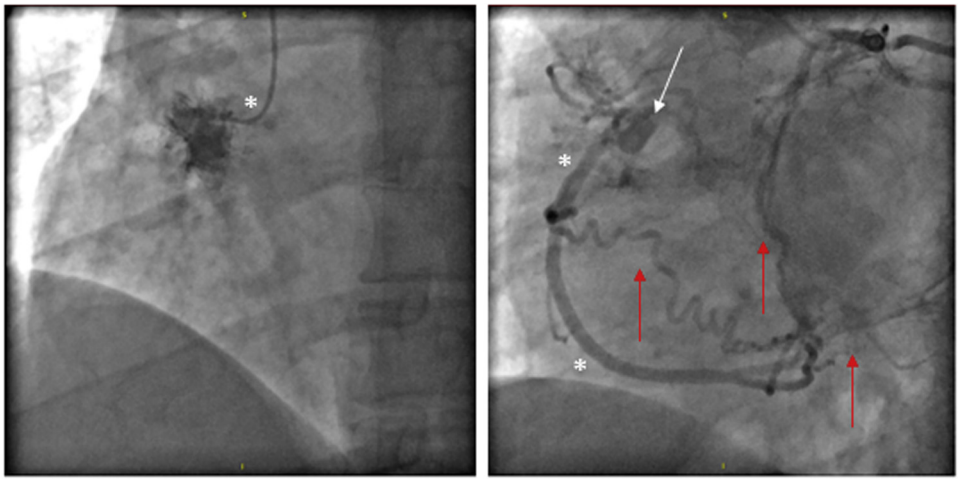
Coronary angiography showed a feeder vessel from the distal left main artery to the mass with an incidental moderate-severe discrete atherosclerotic lesion of the mid left anterior descending artery and no evidence of tumor compression (Figure 8). An exercise nuclear stress test was negative for ischemia. Multidisciplinary discussion including cardiothoracic surgery, cardiology, and endocrinology concluded that given the small size of the tumor and nesting within the great vessels and left main coronary artery (Figure 9), the risks of surgical resection outweighed the benefits. Furthermore, there were concerns that biochemical testing may be falsely negative in the setting of a small PGL size with a possibility of mechanical disruption and potentially fatal

FIGURE 4 Positron Emission Tomography–Computed Tomography Imaging With Gallium-68–DOTATATE of the Paraganglioma



The mass is DOTATATE (DOTA-octreotate)-avid, confirming the diagnosis of paraganglioma.

FIGURE 5 Coronary Angiography Displaying the Relationship of the Paraganglioma to the Coronary Arteries



(Left) complete occlusion of the right coronary artery (RCA) **(asterisk)**. **(Right)** RCA **(asterisk)** was completely collateralized through left-to-right collaterals **(red arrows)**. Moreover, the paraganglioma **(open arrow)** received feeder vessels from the RCA, left main artery, and left circumflex artery.

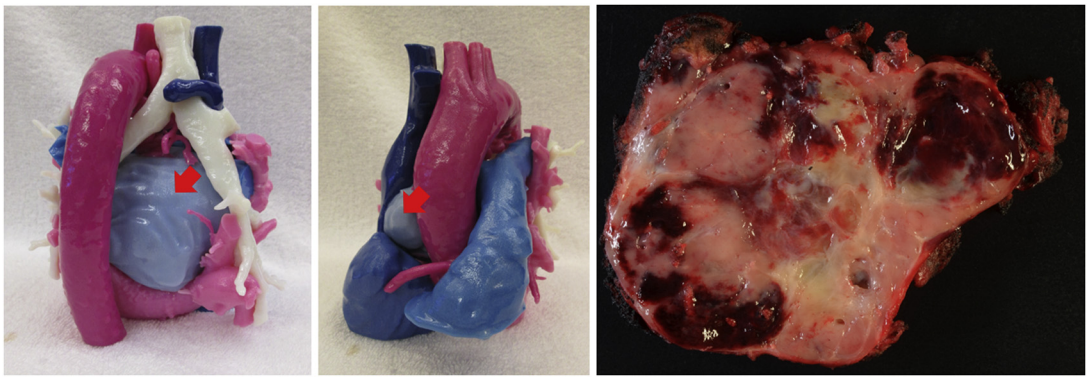
catecholamine secretion. Therefore, the risk of percutaneous embolization was thought to outweigh the benefits. Serial cross-sectional imaging and biochemical monitoring was thought to be the best management course for the patient, who was medically managed for coronary artery disease. The patient was doing well at 6-month follow-up after diagnosis.

DISCUSSION

Intrapericardial PGLs are extremely rare tumors and are most often located adjacent to the left atrium.[1] They can be functional and secrete catecholamines in up to 80% of cases.[1]

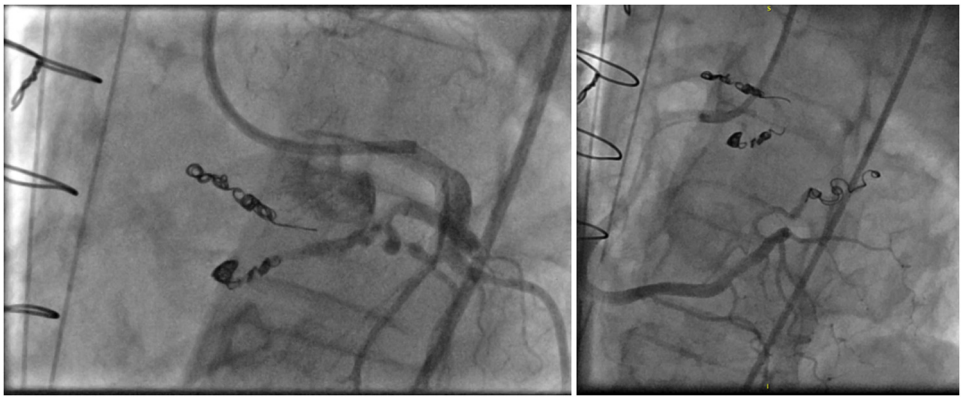
The diagnosis of functional PGLs presenting with sympathetic symptoms usually starts with 24-hour

FIGURE 6 3-Dimensional Modeling and Gross Appearance of the Tissue-Confirmed Paraganglioma



(Left and middle) Three-dimensional–printed model of the heart with the mass in **light blue color** overlying the left atrium **(red arrows)**. **(Right)** Lobulated, well-circumscribed mass with **tan color** and areas of hemorrhage, highlighting the vascular nature of the surgically excised paraganglioma.

FIGURE 7 Coiling of Feeder Branches From the Coronary Arteries to the Paraganglioma



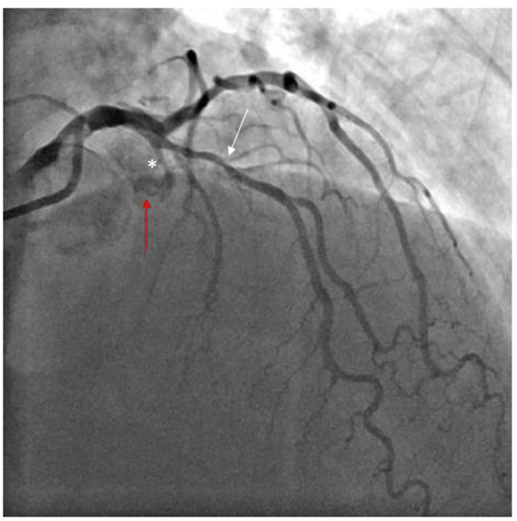
(Left) The left anterior oblique 35° cranial 3° view shows the coiled 2 feeder vessels arising from the left circumflex artery. **(Right)** The left anterior oblique 35° cranial 17° view shows coiling of the feeder artery arising from a distal branch of the right coronary artery (Video 4).

urine collection for fractioned catecholamines and metanephrines.[3] When positive, imaging evaluation starts with CT or magnetic resonance imaging of the abdomen and pelvis to look for pheochromocytoma and extra-adrenal PGL, which accounts for 95% of cases.[3] If abdominal imaging is negative, imaging of the thorax and head and neck or molecular imaging is done next.[4]

On the other hand, around 20% of intrapericardial PGLs are nonfunctioning and diagnosed due to mass effect (eg, chest/back pain, cough, hoarseness) or metastatic disease, or incidentally on imaging.[1] Owing to the distinctive features of PGLs including vascularity and location, imaging characteristics are often enough for presumptive diagnosis before proceeding with surgical resection.[3] On CT scan, findings include intense enhancement with contrast and delayed washout.[4] There could be cystic, calcific, or necrotic components.[5] PGLs typically have unenhanced CT attenuation in the 40- to 50-HU range.[3] On CMR, PGLs are hyperintense on the T2 sequence, usually are hypointense on the T1 sequence, and show dynamic contrast-enhanced features on perfusion sequence.[5] Echocardiography should be performed to assess cardiac function and the relationship between the tumor and cardiac structures.[6] Biochemical testing should be performed in all patients even if asymptomatic. Moreover, it is recommended that genetic testing be performed in all patients with PGL.[3]

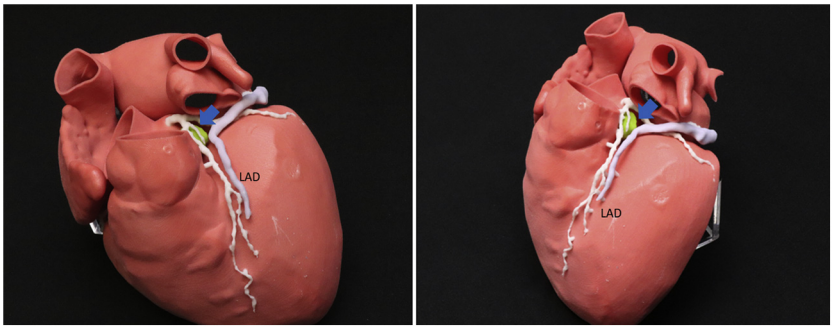
Molecular imaging is helpful in the diagnosis of PGLs and is important to evaluate for metastasis or synchronous tumors. Currently, F-18 FDG or Ga-68-DOTA-coupled peptides (DOTATATE, DOTANOC, and DOTATOC) in conjunction with PET-CT are the preferred modalities with excellent sensitivity and imaging characteristics for staging.[7] Coronary angiography can be helpful in determining coronary

FIGURE 8 Coronary Angiography Showing Feeder Vessels to the Presumed Paraganglioma as Well as Incidental Coronary Artery Disease



Coronary angiogram showing feeder vessel **(asterisk)** arising from the distal left main coronary artery and tumor blush within the paraganglioma **(red arrow)** beneath the left main and proximal left anterior descending arteries. A discrete atherosclerotic lesion **(open arrow)** is seen in the mid left anterior descending artery without evidence of tumor compression.

FIGURE 9 3-Dimensional Modeling of the Heart With the Paraganglioma



Three-dimensional–printed model with the mass in green color **(blue arrow).** Note the small size of the mass and its proximity to the left main artery and left anterior descending artery (LAD).

artery compression due to mass effect, concurrent blockage, and feeder arteries to the PGL and should be considered for surgical planning.

Surgical resection is the definitive treatment for thoracic PGLs. Alpha- and beta-adrenergic blockade should be initiated 7 to 14 days before surgical manipulation, even when biochemical testing is normal.[5] Given the highly vascular nature of PGLs and their frequent proximity to blood vessels, multiple measures can be helpful to decrease intraoperative bleeding, including use of CPB and preoperative embolization of the feeding arteries.[2] The latter was essential in one of our patients in whom surgical resection was not initially possible. This procedure could be considered during surgical planning when high risk of bleeding is anticipated (eg, aorticopulmonary PGLs). Finally, in patients in whom surgical risk is considered to be prohibitive or in those with small noncompressive, nonsecreting PGLs, surveillance with serial imaging and biochemical testing may be appropriate given the slow-growing nature of most PGLs.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS cardiac magnetic resonance imaging, computed tomography, echocardiography, embolization, paraganglioma, surgical resection

APPENDIX For a review of different management strategies of PGLs as well as supplemental videos, please see the online version of this paper.