

CLINICAL CARDIOLOGY

IMAGING VIGNETTE: CLINICAL VIGNETTE

Exertional Angina in a Young Woman Caused by Large Cardiac Paraganglioma



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ABSTRACT

Coronary ischemia is uncommon in patients in their third decade of life. We present a 21-year-old woman with classic exertional angina secondary to a large cardiac paraganglioma. Cardiac paragangliomas are rare extra-adrenal neuroendocrine tumors that arise from chromaffin cells. Cardiac symptoms can be related to catecholamine excess or anatomical compression. (J Am Coll Cardiol Case Rep 2024;29:102209) © 2024 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/>).

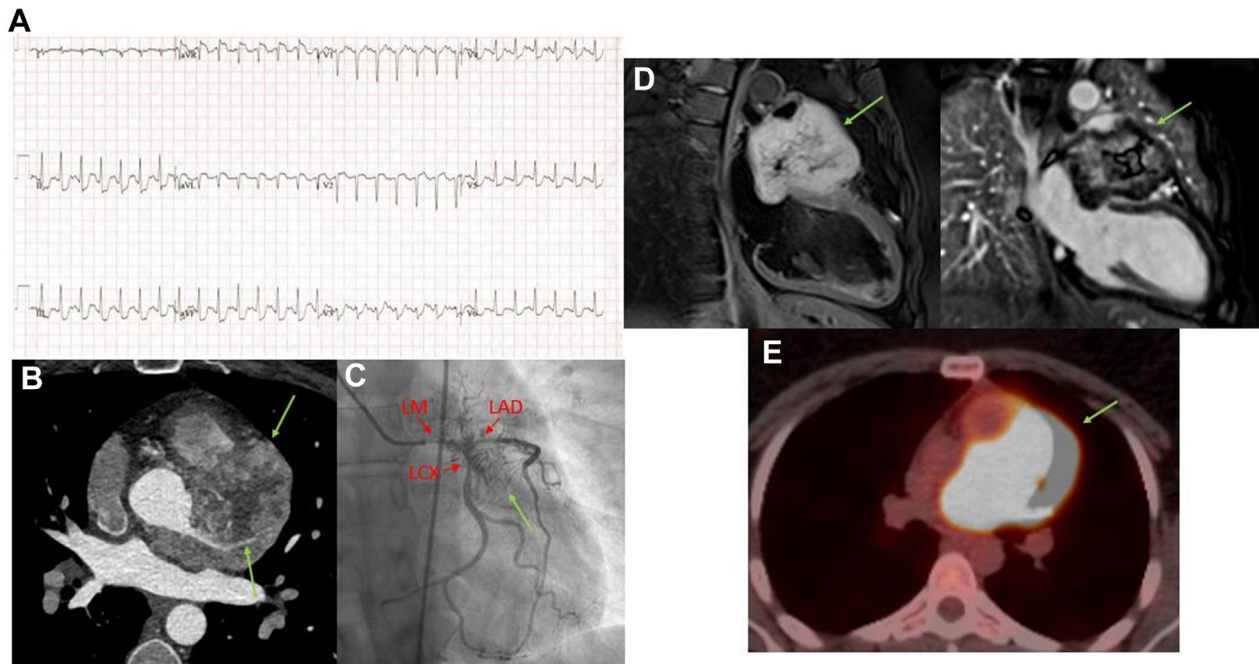
A 21-year-old woman with no significant medical history presented to her primary care provider because of a 6-month history of exertional chest pain and shortness of breath. She was initially treated for exercise-induced asthma and anxiety without improvement in her symptoms. For further evaluation, exercise treadmill echocardiography was performed. The patient exercised for 5 minutes and 16 seconds on a standard Bruce protocol and experienced chest pain. Both electrocardiographic evidence and echocardiographic evidence of coronary ischemia were observed (**Figure 1A**, **Video 1**)

Considering the patient's demographics and absence of a family history of premature coronary artery disease, traditional atherosclerotic heart disease was deemed unlikely. Computed tomographic coronary angiography was performed to assess coronary anomaly and spontaneous coronary dissection. A large, 75 × 41 mm, heterogeneous, highly vascular mediastinal mass was detected encasing the left coronary arteries with anatomically significant compression (**Figure 1B**). As part of the preoperative evaluation, coronary angiography was performed and confirmed severe stenosis of the left main, anterior descending, and circumflex arteries. Extensive coronary blood flow to the tumor was visualized (**Figure 1C**, **Videos 2 and 3**). The differential diagnosis of these findings included paraganglioma, angiosarcoma, or hemangioendothelioma. Cardiac magnetic resonance with cine and first-pass perfusion imaging confirmed a large, well-circumscribed, hypervascular mass with areas of central necrosis (**Videos 4 and 5**). T2-weighted imaging with fat saturation demonstrated hyperintense T2 signal (classic "light bulb appearance"), and post-contrast evaluation revealed prominent heterogeneous gadolinium uptake most consistent with a paraganglioma (**Figure 1D**). Positron emission tomography-computed tomography showed avid fluorodeoxyglucose uptake of the mediastinal mass without hypermetabolic adenopathy or any evidence of metastasis (**Figure 1E**). With further work-up the serum normetanephrine was shown to be elevated.

Our patient underwent surgical tumor resection and pulmonary homograft placement (because of the pulmonary artery compression), left internal mammary artery to left anterior descending and saphenous vein graft to obtuse marginal bypass, surgical ligation of left coronaries, and placement of central venous arterial

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FIGURE 1 Diagnostic Imaging of Cardiac Paraganglioma

(A) Ischemic stress electrocardiogram shows deep horizontal ST-segment depression in leads II/III/AVF and V₄-V₆. (B) Computed tomography coronary angiography reveals tumor encasement of the left coronary arteries and heterogeneous contrast uptake by the tumor (green arrow). (C) Coronary angiography confirms severe compression of the left main (LM), anterior descending (LAD), and circumflex (LCX) arteries. Abundant coronary blood flow to the paraganglioma is also demonstrated (green arrow). (D) T2-weighted fat subtraction cardiac magnetic resonance image shows intense T2 activity and "light bulb" appearance of a well-circumscribed mass. Image after use of gadolinium contrast material shows extensive gadolinium uptake with a central core of necrosis (green arrow). (E) Fluorodeoxyglucose-positron emission tomography shows avid fluorodeoxyglucose uptake by the tumor and no evidence of lymphadenopathy or metastasis (green arrow).

extracorporeal membrane oxygenation. Pathologic analysis from that procedure confirmed the diagnosis of cardiac paraganglioma. Sadly, the patient unfortunately died of postoperative adverse effects.

Cardiac paragangliomas are extraordinarily rare extra-adrenal neuroendocrine tumors that arise from chromaffin cells.¹ There is a wide variability in this tumor's hormonal functional activity, as demonstrated in 17% to 79% of patients in the available literature.² Our young patient presented with classic exertional angina but without any associated risk factors. Her symptoms were attributed to severe compression of the coronary vasculature. Although coronary ischemia is rare in patients with this demographic, classic exertional angina dictates additional diagnostic evaluation with multimodality imaging, as illustrated in this case.

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KEY WORDS angina, coronary compression, paraganglioma

 **APPENDIX** For supplemental videos, please see the online version of this paper.