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Dyspnea and Chest Pain in a Young Woman Caused by a Giant Pericardial Lymphohemangioma: Diagnosis and Treatment

Sean P. Heffron, MD, MS, MSc¹, Carlos L. Alviar, MD¹, Christopher Towe, MD², Benjamin P. Geisler, MD, MPH¹, Leon Axel, MD, PhD^{1,3}, Aubrey C. Galloway, MD², and Adam H. Skolnick, MD¹

¹Department of Medicine, Leon H. Charney Division of Cardiology, New York University Langone Medical Center, New York University School of Medicine, New York, NY

²Department of Surgery, Division of Cardiothoracic Surgery, New York University Langone Medical Center, New York University School of Medicine, New York, NY

³Department of Radiology, New York University Langone Medical Center, New York University School of Medicine, New York, NY

Abstract

We describe a 21-year old woman who presented with chest pain and dyspnea on exertion, who was found to have a large pericardial mass. Multimodality imaging was instrumental in narrowing the differential diagnosis and planning surgical treatment, which included coronary artery bypass and right heart reconstruction. The final pathologic diagnosis was lymphohemangioma – the largest ever to be reported in the literature.

Case Presentation

A 21-year old woman presented with complaints of reduced exercise tolerance and nonexertional left-sided chest pain. An ECG was unremarkable, but chest x-ray revealed an enlarged cardiac silhouette (Supplemental Figure S1). Echocardiography revealed a moderate-sized circumferential pericardial effusion as well as a large heterogenous mass adjacent to the right heart (Supplemental Movie S2). The mass was noted to compress the right atrium, but did not result in hemodynamic obstruction (Supplemental Movie S3).

To further characterize the mass, the patient underwent cardiac magnetic resonance imaging. The mass extended from the base of the heart, wrapping anteriorly to abut the right ventricle, and extended in front of the heart from the diaphragmatic surface to the aortic arch, with dimensions of 11.2 × 6.2 × 14.9 cm (Figure 1, Supplemental Movie S4). The mass partially encased the ascending aorta and completely encased the right coronary artery (Figure 2).

Address for Correspondence: Adam H. Skolnick MD, Associate Professor of Medicine, New York University School of Medicine, 530 First Avenue, HCC Suite 3D, New York, NY 10016, USA, Tel. 1-212-263-2674; Fax. 1-212-263-6470, adam.skolnick@nyumc.org.

Conflicts of interest: none

Consent

Written informed consent was obtained from the patient for publication of this Case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Fluid-filled spaces, seen first on echocardiography, were further characterized with heterogeneous T1 and T2 hyperintensity (Figure 2, Supplemental Figure S5). There was also heterogeneous late gadolinium enhancement. The mass did not appear adherent to the parietal pericardium (Supplemental Movie S6). Computed tomographic imaging of the chest, abdomen and pelvis showed no evidence of metastatic disease.

The clinical presentation, in combination with imaging findings, suggested a benign tumor, most likely a hemangioma, although sarcoma and rhabdomyoma remained in the differential. The risk of sampling error and bleeding, given the encasement of the right coronary artery, made radiographically-guided biopsy an unpalatable option. The mass was felt to be provoking significant symptoms, so a complete en bloc resection was deemed warranted. Coronary angiography was performed preoperatively, which demonstrated branches of the right coronary and left circumflex coronary arteries to be supplying the mass (Supplemental Movies S7 and S8).

At surgery, the mass was found to be adherent to most of the right atrium, the anterior annulus of the tricuspid valve and a small portion of the right ventricle. The tricuspid valve leaflets were not involved. The right coronary artery was found to be encased by the mass, as suggested by imaging. The mass was able to be resected en bloc; however, this required nearly complete resection of the right atrium, the tricuspid annulus and a portion of the right ventricle. A porcine tricuspid valve prosthesis was placed and porcine small intestine submucosa extracellular matrix was used to reconstruct the right atrium (Supplemental Movie S9). A right internal mammary to distal right coronary artery anastomosis was also constructed.

The final pathologic diagnosis was cavernous lymphohemangioma (Supplemental Figures S10 – S13).

Discussion

An abnormal collection of lymphatics, as well as blood vessels, lymphohemangiomas generally occur in the head and neck. While cardiac hemangiomas make up approximately 5% of benign primary cardiac tumors¹, lymphangiomas and lymphohemangiomas are exceedingly rare. There are fewer than two dozen cases of cardiac lymphangiomas in the literature, the majority occurring in patients under ten years of age.² Given the low likelihood of spontaneous resolution and high recurrence rates, complete surgical resection is recommended in symptomatic patients.³ A single case of a cardiac lymphohemangioma has been reported.⁴ To our knowledge, this is the largest pericardial or cardiac vascular tumor (hemangioma/lymphangioma/lymphohemangioma) to be reported.

The patient was discharged home on post-operative day #12. Six months postoperatively, right ventricular function returned to normal, electrical conduction was preserved and repeat MRI demonstrated no evidence of recurrence.

Conclusions

This case demonstrates the utility of multi-modality imaging in narrowing the diagnosis and planning treatment of cardiac masses.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgments

Disclosures

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Brief Summary

We describe a 21-year old woman who presented with chest pain and dyspnea on exertion, who was found to have a large pericardial mass. Multimodality imaging was instrumental in narrowing the differential diagnosis and planning surgical treatment, which included coronary artery bypass and right heart reconstruction. The final pathologic diagnosis was lymphohemangioma – the largest to be reported in the literature.

Figure 1.
Sagittal black blood sequence view of the pericardial mass demonstrates its extent from the diaphragm to above the aortic arch.

Figure 2.
T2 weighted sagittal image demonstrating heterogeneous hyperintensity of the pericardial mass as well as encasement of the right coronary artery (arrows) by the mass.