Arch Cardiovasc Imaging. 2014 August; 2(3): e20252. DOI: 10.5812/acvi.20252

Published online 2014 May 25. **Case Report**

Multimodality Imaging of a Cardiac Angiosarcoma

Roy Beigel, MD [1]; Joao Carlos Tress, MD [2]; Louise Elizabeth Jane Thomson, MD [1]; Daniel James Luthringer, MD [3]; Alexander Shturman, MD [4]; Alfredo Trento, MD [1]; Robert James Siegel, MD [1,*]

[1]The Heart Institute, Cedars Sinai Medical Center, David Geffen School of Medicine, UCLA
[2]Hospital Universitario Pedro Ernesto
[3]Department of Pathology, Cedars Sinai Medical Center, David Geffen School of Medicine
[4]Department of Cardiovascular Medicine, Western Galilee Hospital, Bar Ilan University

*Corresponding author: Robert James Siegel, The Heart Institute, Cedars Sinai Medical Center, 127 S. San Vicente Boulevard, Suite A3600, Los Angeles, CA 90048. Tel: +1-3104232726, Fax: +1-3104238571, E-mail: Robert.siegel@cshs.org

Received: May 15, 2014; **Accepted:** May 19, 2014

Introduction: While primary malignant tumors of the heart are rare, angiosarcomas are the most common cardiac malignant tumors.
Case Presentation: We describe a 23-year-old woman who presented with a right atrial mass, which was discovered to be a cardiac angiosarcoma. We demonstrate the use of several noninvasive imaging modalities along with pathology confirmation for the definitive and comprehensive diagnosis of a cardiac angiosarcoma, a rare entity by itself.
Conclusions: With the increasing availability of noninvasive imaging techniques, the diagnosis of angiosarcomas can be made at earlier stages. If angiosarcomas are left untreated, their prognosis is very poor. Therapeutic options include surgical excision, chemotherapy, radiation therapy, and heart transplantation or a combination of these.

Keywords: Echocardiography; Magnetic Resonance Imaging; Positron-Emission Tomography

1. Introduction

Primary tumors of the heart, the majority of which are benign, are rare with an incidence at autopsy ranging from 0.0017% to 0.02% (1, 2). The most common cardiac malignancies are sarcomas, and angiosarcomas account for the majority of these malignancies in the adult population, as in the present case. These are malignant neoplasms that more commonly affect the right side of the heart, are highly invasive to surrounding tissue, and have a high potential to metastasize.

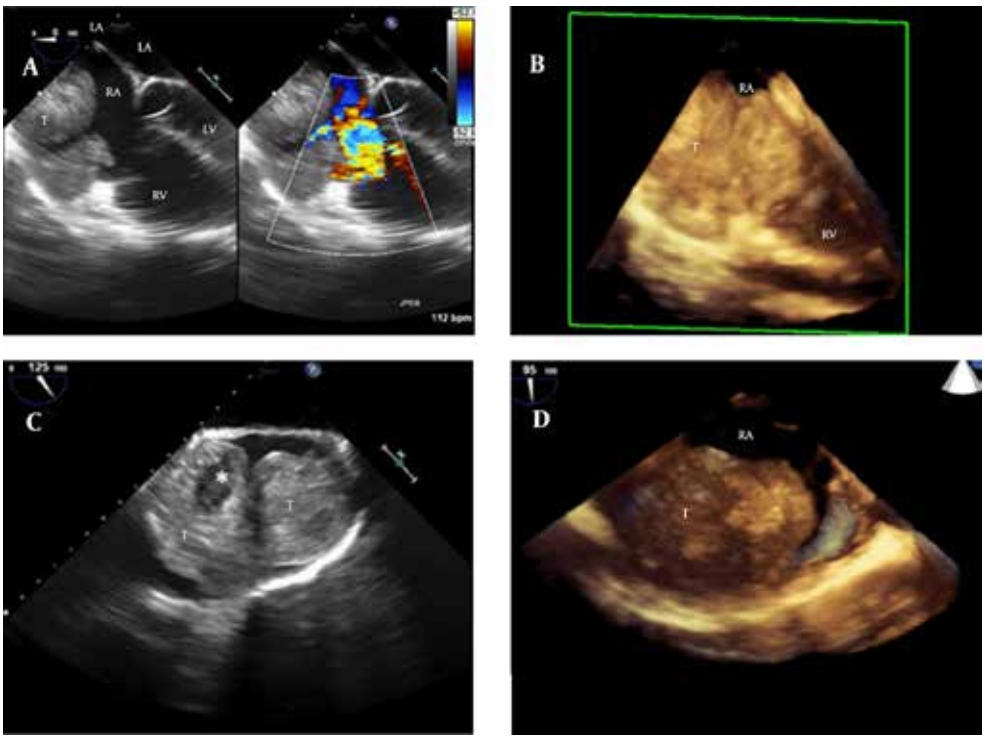
2. Case Presentation

A 23-year-old asymptomatic woman had a screening transthoracic echocardiogram because of a family history of cardiomyopathy and was found to have a mass in the right atrium (RA) and moderate pericardial effusion. Further evaluation by transesophageal echocardiography showed that the mass was attached to the lateral wall of the RA, with partial obstruction of the tricuspid valve, producing a mean diastolic pressure gradient of 4 mmHg (Figure 1). As is shown in Figure 2, cardiac magnetic resonance imaging demonstrated attachment of the mass to the lateral, superior, anterior, inferior, and posterior walls of the RA. There was partial obstruction of the coronary sinus and invasion into the pericardial space, with a moderately large pericardial effusion. Following gadolinium contrast (Gadodamide, Omniscan, Amersham, Piscataway, NJ), there was slow heterogeneous enhancement of the mass (Figure 2D, 2E, and 2F). Fluorodeoxyglucose-positive emission tomography (FDG-PET, Figure 3) did not show metastatic disease and displaced heterogeneous intense hypermetabolism, consistent with a tumor tissue. The patient underwent successful surgical excision of the RA mass (Figure 4A) along with RA and coronary sinus reconstruction with an autologous pericardial patch, creation of right pulmonary window, and drainage of the pericardial effusion. Histologic examination (Figure 4B) confirmed that the mass was a high-grade angiosarcoma, with positive margins along the right atrial free wall.

3. Discussion

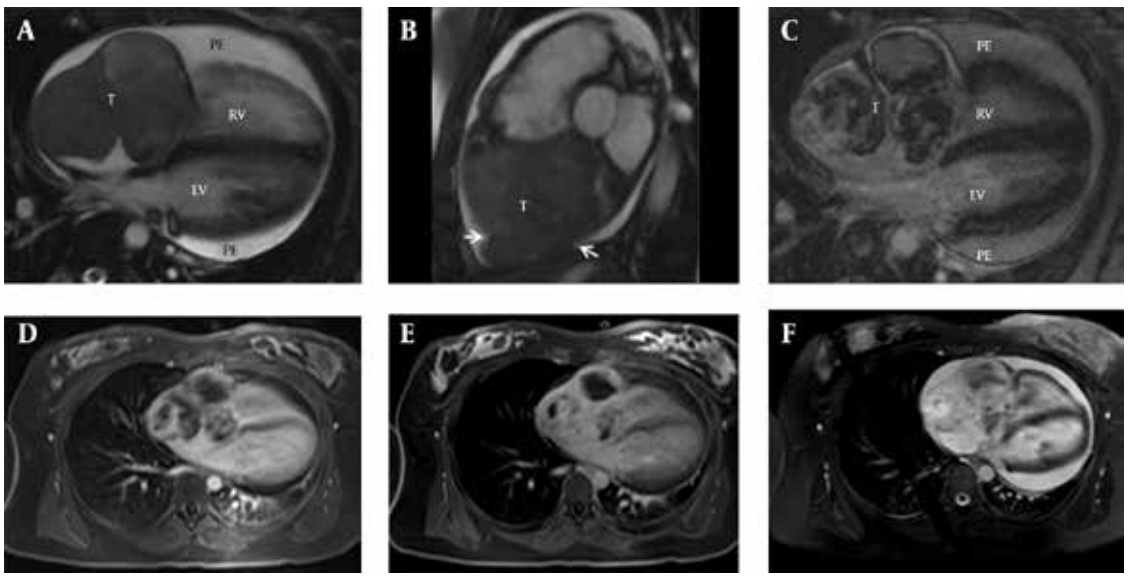
Patients with angiosarcomas usually present with non-specific symptoms and as such these are most often found incidentally or at autopsy. Pericardial effusion is a common finding, although fluid cytology has a low diagnostic yield (2, 3). With the increasing availability of noninvasive imaging techniques, diagnosis can be made at earlier stages. If left untreated, angiosarcomas have a poor prognosis. Therapeutic options include surgical

Figure 1. Transesophageal Echocardiographic Images of the Suspected Mass



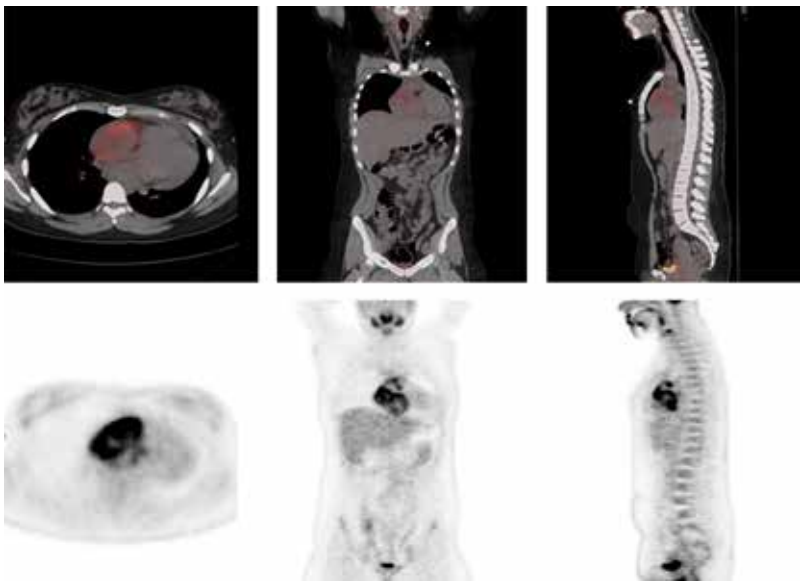
Transesophageal echocardiography, showing an echo-dense finding within the right atrium (RA), consistent with a massive tumor (T): (A) Five-chamber view, showing the mass along with an accelerated flow across the tricuspid valve. (B) Three-dimensional reconstruction of the mass in the RA. (C) Bicaval view, showing that most of the right atrial cavity is compressed by the mass and demonstrating hypodense areas, suggesting of necrosis (asterisk). (D) Three-dimensional imaging from the same view as in C. RV = Right ventricle; LV = Left ventricle.

Figure 2. Magnetic Resonance Imaging of the Suspected Mass



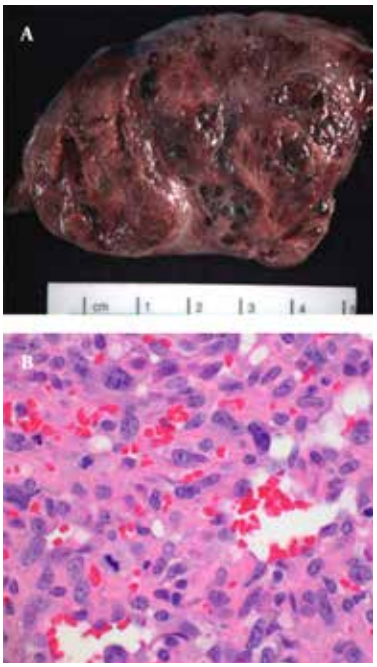
Magnetic resonance imaging: (A) Tumor (T) is shown to be attached to the lateral, superior, anterior, inferior, and posterior walls of the right atrium (RA) along with the presence of moderate to large pericardial effusion (PE). (B) Short-axis image, demonstrating invasion of the tumor to the pericardial space (arrows). Late gadolinium enhancement imaging at 5 minutes after contrast, showing lobulated appearance of peripheral enhancement (C). Serial T1 weighted images, demonstrating progressive enhancement of the central regions of the tumor at 5, 10, and 25 minutes after the administration of gadolinium contrast (D-F). RV = Right ventricle; LV = Left ventricle.

Figure 3. Positive Emission Tomography Imaging of the Suspected Mass



Axial, coronal, and sagittal positive emission tomography (PET) and fused PET/CT images. The tumor in the right atrium is intensely hypermetabolic. There is a physiologic distribution of F18 Fluorodeoxyglucose elsewhere.

Figure 4. Pathological Images of the Explanted Tumor



(A) Gross photograph of the cut surface of the tumor mass, showing a neoplasm with areas of cystic degeneration, hemorrhage, and necrosis, indicative of high-grade vascular malignancy. The tumor measures up to 8.5 cm in greatest dimension and was grossly infiltrative to the right atrial margins of resection. (B) Photomicrograph, demonstrating proliferation of poorly differentiated tumor cells characterized by variation in nuclear size and shape, elevated nucleus to cytoplasm ratio, irregular chromatin, and high mitotic activity. The cells coalesce to form primitive vascular spaces lined by the malignant cells and filled with red blood cells, features indicative of angiosarcoma (600x original magnification).

excision, chemotherapy, radiation therapy, and heart transplantation or a combination of these (2). The reported response to therapy is variable: some authors have reported only a modest response to therapy and a survival of only 6-9 months (2), and the longest survival was 53 months reported in a single case (4).

Authors' Contributions

Roy Beigel and Joao Tress performed literature review and wrote the initial draft of the manuscript. Louise Thomson interpreted MRI and PET imaging studies. Dan Luthringer provided pathology details and analysis of the microscopic specimens of the tumor and wrote the pathology part of paper. Alexander Shturman contributed to literature review, editing, and final revision of the manuscript. Alfredo Trento contributed to the surgical aspects and the pathological examination and reviewed the manuscript. Robert Siegel obtained echocardiographic images and contributed to the editing and final revision of the manuscript. All the authors read, provided feedback, and approved the final version of the paper.

References

1. Reynen K. Frequency of primary tumors of the heart. *Am J Cardiol.* 1996;**77**(1):107.
2. Butany J, Nair V, Naseemudin A, Nair GM, Catton C, Yau T. Cardiac tumors: diagnosis and management. *Lancet Oncol.* 2005;**6**:219–228.
3. Newman D, Klarich K, Kumar G. Echocardiographic Characteristics of Cardiac Angiosarcomas: Mayo Clinic Experience. *J Am Coll Cardiol.* 2013;**61**(10):E1085.
4. Nakamichi T, Fukuda T, Suzuki T, Kaneko T, Morikawa Y. Primary Cardiac Angiosarcoma: 53 Months' Survival After Multidisciplinary Therapy. *Ann Thorac Surg.* 1997;**63**(4):1160–1.