Case Reports

Benefits and Limitations of Multimodality Imaging

in the Diagnosis of a Primary Cardiac Lymphoma

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Primary cardiac tumors are far rarer than tumors metastatic to the heart. Angiosarcoma is the primary cardiac neoplasm most frequently detected; lymphomas constitute only 1% of primary cardiac tumors.

We present the case of a 55-year-old woman with a recently diagnosed intracardiac mass who was referred to our institution for consideration of urgent orthotopic heart transplantation. Initial images suggested an angiosarcoma; however, a biopsy specimen of the mass was diagnostic for diffuse large B-cell lymphoma. The patient underwent chemotherapy rather than surgery, and she was asymptomatic 34 months later.

We use our patient's case to discuss the benefits and limitations of multiple imaging methods in the evaluation of cardiac masses. Certain features revealed by computed tomography, cardiac magnetic resonance, and positron emission tomography can suggest a diagnosis of angiosarcoma rather than lymphoma. Cardiac magnetic resonance and positron emission tomography enable reliable distinction between benign and malignant tumors; however, the characteristics of different malignant tumors can overlap. Despite the great usefulness of multiple imaging methods for timely diagnosis, defining the extent of spread and the hemodynamic impact, and monitoring responses to treatment, we think that biopsy analysis is still warranted in order to obtain a correct histologic diagnosis in cases of suspected malignant cardiac tumors. ***(Tex Heart Inst J 2014;41(6):657-9)***

Primary cardiac tumors are rare, compared with tumors metastatic to the heart.[1] A multimodality imaging approach is highly useful for timely diagnosis, defining the extent of spread and the hemodynamic impact, and monitoring response to treatment. We present the case of a woman in whom images suggested one type of cardiac tumor, whereas analysis of a biopsy specimen identified another. We discuss the benefits and limitations of various imaging methods and describe how we reached a final diagnosis.

Key words: *Diagnostic imaging/methods; heart neoplasms/diagnosis/pathology/radiography; lymphoma/diagnosis/radiography; treatment outcome*

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Case Report

In December 2011, a 55-year-old white woman presented at a regional hospital with a 6-month history of gradually progressive edema and dyspnea on exertion. Her medical history included Hashimoto thyroiditis. Physical examination revealed lower-extremity edema. Computed tomograms (CT) of the chest showed an intracardiac mass (Fig. 1). A transthoracic echocardiogram (TTE) revealed a large right atrial mass that extended into the right ventricular (RV) inflow and prevented coaptation of the tricuspid valve leaflets (Fig. 2). The mass almost completely obliterated the right atrial cavity and resulted in functional tricuspid stenosis (mean pressure gradient, 14 mmHg). A large pericardial effusion raised concern about impending tamponade. Cardiac magnetic resonance (CMR) images showed that the tumor infiltrated the right atrial and RV anterior and septal walls, thus impairing RV function, and extended across the interatrial septum into the left atrium and the retroaortic space at the level of the aortic root (Figs. 3 and 4). The mass was heterogeneous on T2-weighted imaging and was mostly hyperintense because of edema; a few low-intensity areas implied tumor necrosis. Post-contrast T1-weighted images showed patchy enhancement: focal areas of hypoenhancement suggested necrosis, and surrounding areas of hyperenhancement suggested fibrosis. The presumed diagnosis was angiosarcoma, and the patient was referred to our institution for consideration of urgent orthotopic heart transplantation.

Results of whole-body positron emission tomography (PET) with use of ^{18}fluorodeoxyglucose (^{18}FDG) revealed a hypermetabolic intracardiac mass and no evidence of

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distant metastatic disease (Fig. 5). We used CMR imaging to decide on a road map, then performed a TTE-guided biopsy of the mass from an inferior vena caval approach. Pathologic evaluation of the biopsy specimen revealed diffuse infiltration by large, round anaplastic cells, and results of immunophenotyping were diagnostic for CD20+ diffuse large B-cell lymphoma (Fig. 6).

The patient underwent chemotherapy with rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone. Over time, serial TTEs showed nearly complete resolution of the mass with no mechanical complications, improved RV inflow and function, and a reduction in size of the pericardial effusion (Fig. 7). In October 2014, the patient reported that she was asymptomatic and doing well.

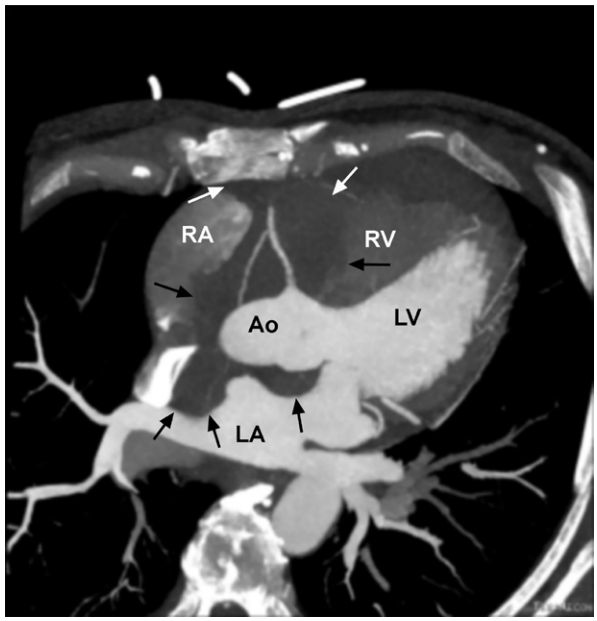

Fig. 1 *Computed tomogram shows a hypoattenuating intracardiac mass (arrows) extending from the right atrium (RA) into the right ventricle (RV) and across the interatrial septum into the left atrium (LA).*

Ao = aortic root; LV = left ventricle

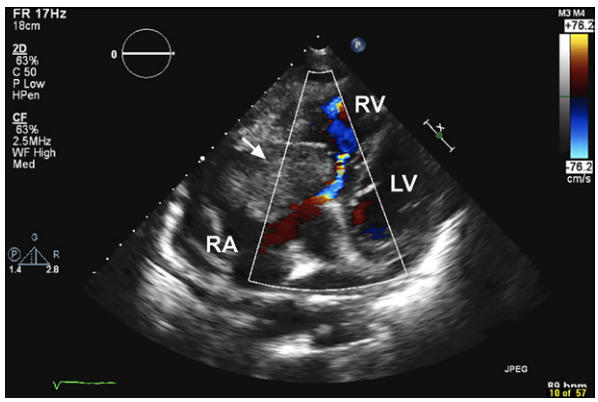

Fig. 2 *Echocardiogram (modified parasternal short-axis view) shows the mass (arrow) obliterating the cavity of the right atrium (RA) and causing functional tricuspid stenosis.*

LV = left ventricle; RV = right ventricle

Supplemental motion image is available for Figure 2.

Discussion

Only 25% of primary cardiac tumors are malignant, and approximately three quarters of these are sarcomas.[2] Lymphomas constitute only 1% of primary cardiac tumors. In immunocompetent adults, approximately

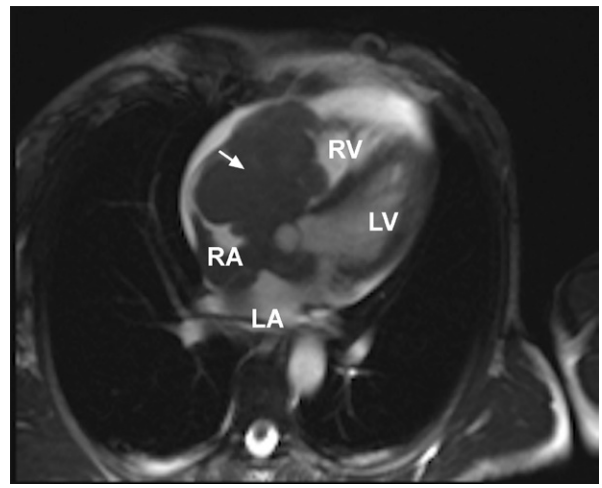

Fig. 3 *Cardiac magnetic resonance image shows the mass (arrow) in the right atrium (RA) and right ventricle (RV). The tumor invades the RV free wall and extends across the interatrial septum into the left atrium (LA).*

LV = left ventricle

Supplemental motion image is available for Figure 3.

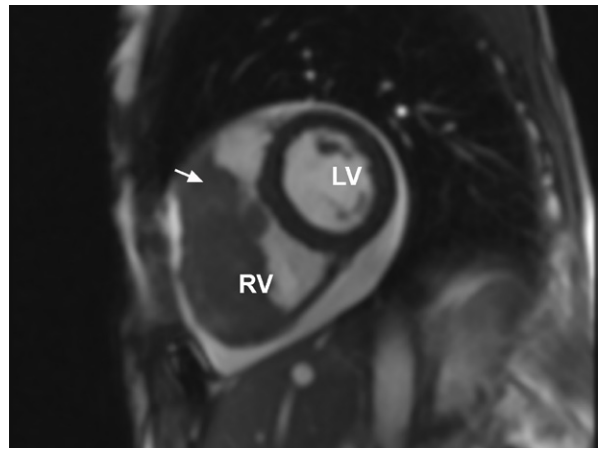

Fig. 4 *Cardiac magnetic resonance image (parasternal short-axis view) shows the mass (arrow) in the right ventricle (RV).*

LV = left ventricle

Supplemental motion image is available for Figure 4.

80% of primary cardiac lymphomas are diffuse B-cell tumors that most often involve the right atrium and RV.[3] Most patients present late, after large growths have caused right-sided heart failure and pericardial effusion. A multimodality imaging approach to cardiac masses greatly aids their diagnosis and characterization, and is essential for monitoring response to treatment.

In our patient, CT was the first method to diagnose the intracardiac mass. Although CT is limited in its ability to define intracardiac spread, it is helpful in defining extracardiac spread. In comparison, CMR enables better soft-tissue and tumor characterization, and it is superior in revealing tumorous infiltration of the myocardium and pericardium. Transthoracic echocardiography, which enables real-time imaging, is valuable in the evaluation of physiologic and hemodynamic effects of a mass and in serial monitoring of treatment response. ^{18}Fluorodeoxyglucose PET/CT is crucial in determining whether a mass is benign or malignant, and in confirming cardiac isolation. Certain features, such as the presence of necrosis or valvular involvement on CT or CMR, or lower ^{18}FDG avidity on PET, suggest an angiosarcoma rather than a lymphoma.[4] The use of CMR and PET can reliably enable a distinction between benign and malignant tumors; however, there is a considerable overlap of characteristics among different malignant tumors. Sarcomas have a poor prognosis and necessitate surgery when feasible, whereas lymphomas often respond to chemotherapy.

Despite the great usefulness of multiple imaging methods in characterizing cardiac tumors of a suspected malignant nature, biopsy is still warranted in order to reach a correct histologic diagnosis.[5] In our patient's case, fragmented care did not enable the streamlined use of different imaging methods—certain studies were performed at a regional hospital before the patient's transfer to our facility. When possible, in view of healthcare costs, it is prudent to use imaging judiciously, to minimize redundancy.

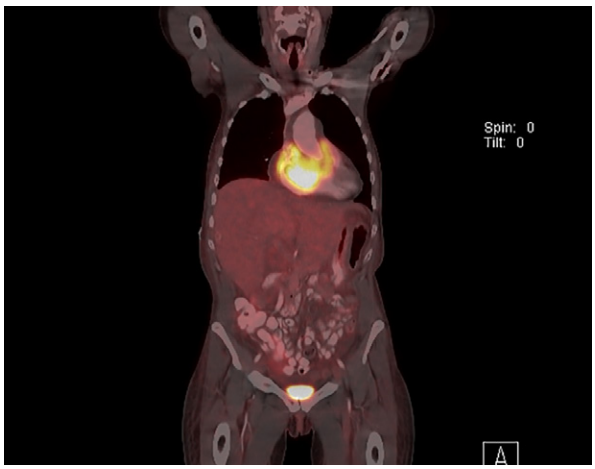

Fig. 5 *Positron emission tomogram (coronal section) shows cardiac isolation. The bright area in the chest is the hypermetabolic intracardiac mass.*

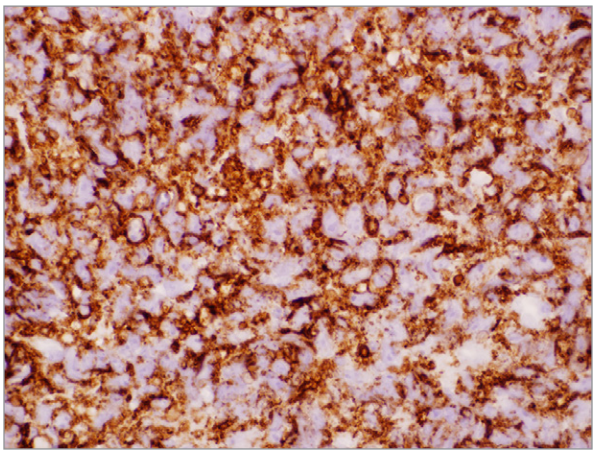

Fig. 6 *Photomicrograph shows diffuse reactivity of all cells for CD20 antibody, confirming B-cell lymphoma (immunoperoxidase stain, orig. ×400).*

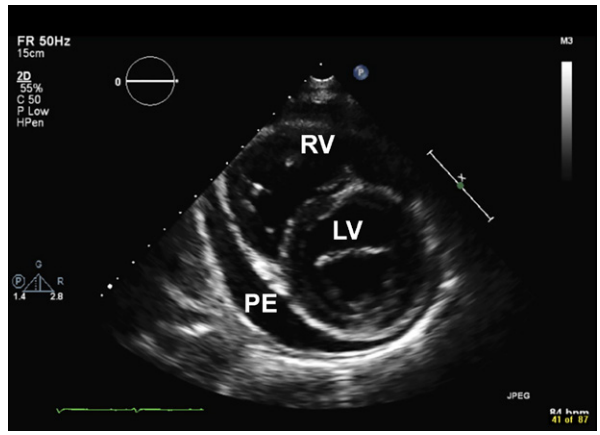

Fig. 7 *Echocardiogram (parasternal short-axis view) shows nearly complete resolution of the mass in the right ventricle (RV) after the patient's chemotherapy.*

LV = left ventricle; PE = pericardial effusion

Supplemental motion image is available for Figure 7.

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