Primary Cardiac Angiosarcoma: Diagnostic Utility of Computed Tomography and Cardiac Magnetic Resonance

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Primary cardiac tumors are infrequent, but their clinical presentation is variable and their associated mortality is high. By the time they produce symptoms and the diagnosis is made, they have usually progressed to a large mass causing considerable hemodynamic compromise. Although Doppler echocardiography is the usual technique for the initial diagnosis, magnetic resonance imaging or computed tomography can offer more anatomically useful information. We describe 3 patients with cardiac angiosarcoma, 2 in the right atrium and 1 in the left ventricle, the latter diagnosed by computed tomography-guided biopsy. All 3 patients underwent surgery, but the short-term course was fatal in all cases.

Key words: Cardiac angiosarcomas. Echocardiography. Magnetic resonance. Biopsy. Computed tomography.

INTRODUCTION

Primary cardiac tumors (PCTs) are rare. Around 25% of these tumors are malignant and 90%-95% of malignant tumors are sarcomas.2 They may remain clinically silent for a long time or cause a range of cardiac and systemic symptoms that mimic other diseases.3-6

The preferred initial method for detecting cardiac tumors is echocardiography, but a complete assessment of the heart and surrounding tissue may sometimes be difficult with this technique. Magnetic resonance (MR) imaging and computed tomography (CT) can provide precise information on the extent of involvement of the tumor.4,7-9 Moreover, these techniques allow simultaneous evaluation of cardiac structures and surrounding tissue and, in certain instances, a guided biopsy is possible.

We report the use of different imaging techniques to determine the clinical and morphologic characteristics of the hearts of 3 patients who underwent interventions for cardiac angiosarcomas.

CASE STUDY

Between 1995 and 2002, 3200 patients underwent open-heart surgery. Three of these (0.0009%) were pa-
tients who had cardiac masses diagnosed by imaging techniques. Diagnosis of cardiac angiosarcoma was confirmed by biopsy in all 3 patients. These patients are discussed below.

**Case 1**

The patient was a 19-year-old man with functional grade 3 dyspnea, skin and mucous membrane pallor, and a grade 3/6 systolic murmur audible in the fourth and fifth right intercostal spaces. The echocardiogram showed a solid mass of variable density measuring 6×4.5 cm in the right atrium (RA) that had invaded the atrial septum (Figure 1A). In the enhanced T1-weighted MR images (Figure 1B), a proliferative mass that had invaded almost the entire right ventricle (RV) was evidenced, displacing the atrial septum to the left. Moderate left pleural effusion and retrocardiac atelectasis were also found. No signs of metastasis to abdominal organs were observed with abdominal MR imaging or echography. Surgical intervention confirmed a large RA tumor that had invaded the RV through the tricuspid valve. The base of attachment extended towards the RA free wall and towards the opening of the inferior vena cava. No hepatic metastasis was observed in abdominal CT or echography. The tumor was resected and the atrium reconstructed with a bovine pericardial patch. Biopsy confirmed the diagnosis of angiosarcoma. After 1 year of follow up, the patient presented with progressive dyspnea and refractory congestive heart failure and died.

**Case 2**

A 32-year-old woman with dyspnea at rest and of emaciated appearance was experiencing tachycardia on admission to hospital. A grade 3/6 diastolic murmur—maximal in the tricuspid region—was audible and she presented jugular engorgement and leg edema. The echocardiogram revealed a solid mass measuring 6×4 cm (Figure 2A and B) with regions of low signal intensity. The mass had a base of attachment to the atrial septum and invaded the tricuspid valve causing severe flow obstruction with maximum tricuspid gradient of 15 mm Hg. The patient went into cardiogenic shock and required emergency heart surgery. In the RA, a large tumor measuring 7×4 cm was found that invaded the RV through the tricuspid valve. The base of attachment extended towards the RA free wall and towards the opening of the inferior vena cava. No hepatic metastasis was observed in abdominal CT or echography. The tumor was resected and the atrium reconstructed with a bovine pericardial patch. Biopsy confirmed the diagnosis of angiosarcoma. After 1 year of follow up, the patient presented with progressive dyspnea and refractory congestive heart failure and died.

**Case 3**

A 50-year old man with atypical pain in the left side of the chest, asthenia, and adynamia presented with weight-loss and tachycardia at rest. In the echocardiogram, a solid mass could be seen measuring 8×11 cm with transparent regions in the lower, lateral and apical walls of the LV (Figure 3A). The enhanced T1-weighted MR images (Figure 3B) showed a large tumor of 14×11 cm, with variable T2-weighted signal strength. Lytic lesion of the costal arch and mild pleural effusion were also observed. Chest CT (Figure 3C and D) defined the edges of the tumor and allowed a guided biopsy without complications. The biopsy confirmed diagnosis of primary angiosarcoma. After 6 months, the patient presented with refractory heart failure and died.

**DISCUSSION**

Malignant PCTs may present with any cardiac symptom but heart failure and systemic embolism are the most common ones initially. Two of our
patients showed signs of heart failure and one had atypical chest pain.

In this series of patients, the initial diagnostic technique was echocardiography, although MR imaging provided greatest additional information. Thus, the extent of the tumor could be precisely defined, so facilitating the decision on how to proceed (the approach used and whether to resect the tumor). Computed tomography allowed diagnosis of the third patient by guided biopsy prior to surgery.

Echocardiography has the advantage of being a cheap and readily available technique that can be done at the patient’s bedside with no contraindications. However, it is sometimes difficult to define what structures have been invaded by the tumor. A benign tumor may also be falsely diagnosed, as in the second of our patients, because the echocardiographic images resemble those of atrial myxoma, something that has been reported occasionally in the medical literature.

Magnetic resonance imaging has the advantage of providing better image quality and a more precise characterization of the tumor (density, extent, and invaded cardiac structures). It also allows assessment of involvement of other neighboring structures at the same time. The most significant characteristics of malignancy in the MR images are the size of the base of attachment of the tumor to the myocardial wall, the tumor size, compression of the cardiac chambers or the great vessels, pericardial or extracardial involvement and signs of tumor necrosis. Although there are no pathognomonic signs, MR images show the angiosarcoma as a tumor mass, usually with no spindle structures, and with weak areas of signal intensity in the enhanced T1-weighted scan, attributed to regions of tumor necrosis. In rhabdomyosarcomas, the intermediate signal intensity in the enhanced T1-weighted scan is similar to that found in adjacent myocardium. Fibromas appear heterogeneous with the same signal intensity in the enhanced T1-weighted scan and the malignant nature of the tumor can be discerned with enhanced T1-weighted images.

Computed tomography is better able to define calcification, the study time is shorter, and the technique is useful when MR imaging is contraindicated. Guided transthoracic biopsies are possible with this technique in some patients.

CONCLUSIONS

We present 3 patients with angiosarcoma in whom echocardiography provided preliminary diagnosis. Magnetic resonance best established the extent of the tumor and CT allowed diagnosis by means of biopsy in one patient. Surgery confirmed the definitive diagnosis and the short-term outcome was death in all 3 patients.

REFERENCES

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