

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.

Angiosarcomas cardíacos primarios: utilidad de la tomografía computarizada y la resonancia magnética cardíaca en su diagnóstico

Los tumores cardíacos primarios (TCP) malignos son infrecuentes, con presentación clínica variada y elevada mortalidad. Cuando producen síntomas y en el momento del diagnóstico, suelen corresponder a masas tumorales muy evolucionadas y con un importante compromiso hemodinámico. El ecocardiograma-Doppler acostumbra ser la técnica diagnóstica inicial, pero en ocasiones las imágenes de resonancia magnética (RM) o de tomografía computarizada (TC) pueden ofrecer más información anatómica. Presentamos los casos de 3 pacientes con angiosarcoma cardíaco, 2 de aurícula derecha y 1 de ventrículo izquierdo, este último diagnosticado por biopsia dirigida mediante tomografía computarizada; los 3 fueron intervenidos quirúrgicamente, pero con evolución fatal a corto plazo.

Palabras clave: Angiosarcoma cardíaco. Ecocardiografía. Resonancia magnética. Biopsia. Tomografía computarizada.

INTRODUCTION

Primary cardiac tumors (PCTs) are rare. Around 25% of these tumors are malignant¹ 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 assess-

ment 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-

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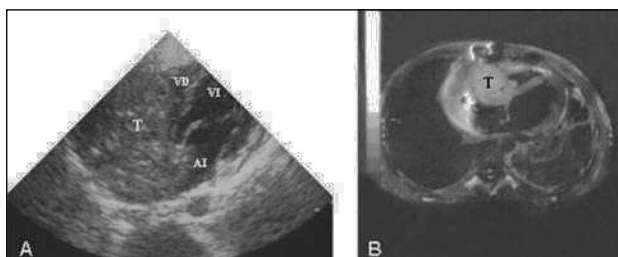


Figure 1. A: transthoracic echocardiogram in subcostal 4-chamber projection showing a tumor (T) that occupies almost the entire right atrium and part of the right ventricle (RV). B: spin-echo enhanced T1 axial plane image of invasion of the right atrium by the tumor. LA indicates left atrium; LV, left ventricle.

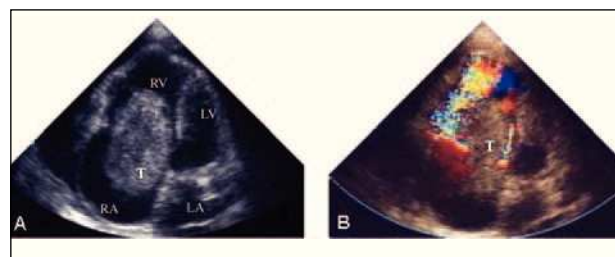


Figure 2. A: Transthoracic echocardiogram in 4-chamber apical projection, showing an oval tumor (T) in the right atrium (RA) that has invaded the right ventricle (RV). A central region of lower signal intensity suggests a region of necrosis. B: Turbulent flow in color Doppler imaging due to obstruction of the tricuspid valve. LA indicates left atrium; LV, left ventricle.

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 and the septal region of the tricuspid valve. The tumor was resected, the tricuspid valve reinserted and a venous bypass to the right coronary artery performed. The biopsy confirmed diagnosis of primary angiosarcoma. After 6 months, the patient presented with refractory 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 large solid RA 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 8.1 × 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 14.1 × 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 angiosarcoma. Abdominal CT and echography revealed no metastasis to abdominal organs. The tumor could not be resected during surgery because the extent and anatomical involvement were too great. The patient died 10 days after the operation.

DISCUSSION

Malignant PCTs may present with any cardiac symptom,^{1,3,4,10} but heart failure and systemic embolism are the most common ones initially.^{5,11} Two of our

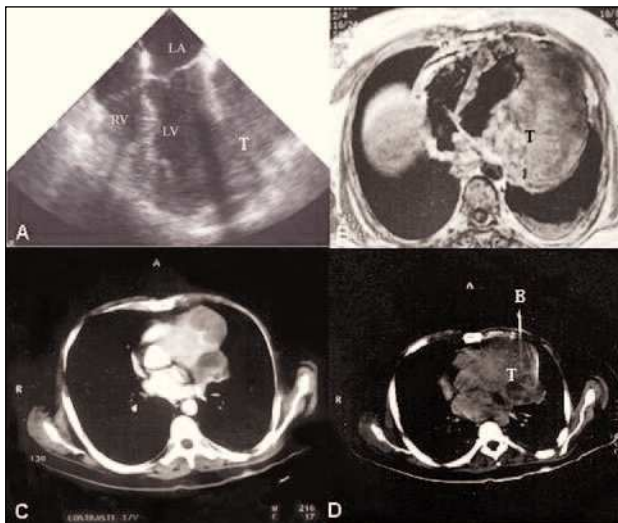


Figure 3. A: transesophageal echocardiogram showing a solid tumor (T) with a heterogeneous structure attached to the lateral wall of the left ventricle (LV). B: spin-echo enhanced T1 axial plane image showing a large solid tumor with involvement of the LV lateral wall, and with variable signal intensity indicative of tumor necrosis. C: helicoidal computed tomography with contrast showing section of the aortic root. The left heart outline is deformed by a multi-lobed mass with regions of variable intensity. D: computed tomography with biopsy needle (B). LA indicates left atrium; RV, right ventricle.

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,^{4,12} so facilitating the decision on how to proceed (the approach used and whether to resect the tumor).^{8,13} 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.¹⁴

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.¹⁵ 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.^{9,16} In rhabdomyosarcomas, the intermediate signal intensity in the enhanced T1-weighted scan is similar to that found in adjacent myocardium.¹⁷ Fibromas appear heterogeneous with the same signal intensity in the enhanced T1-weighted scan¹⁶ 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.¹⁸

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

1. Abad C. Tumores cardíacos (II). Tumores primitivos malignos. Tumores metastásicos. Tumor carcinoide. *Rev Esp Cardiol* 1998; 51:103-14.
2. Roberts WC. Primary and secondary neoplasms of the heart. *Am J Cardiol* 1997;80:671-82.
3. Reynen K. Frequency of primary tumors of the heart. *Am J Cardiol* 1996;77:107.
4. Sarjeant JM, Butany J, Cusimano RJ. Cancer of the heart. Epidemiology and management of primary tumors and metastases. *Am J Cardiovasc Drugs* 2003;3:407-21.
5. Grande AM, Ragni T, Vigano M. Primary cardiac tumors. A clinical experience of 12 years. *Tex Heart Inst J* 1993;20:223-30.
6. Jiménez Mazuecos J, Fuentes Manso R, Segovia Cubero J, Toquero Ramos J, Oteo Domínguez JF, Alonso Púlpon Rivera L. ¿Es útil el trasplante cardíaco como tratamiento del sarcoma cardíaco primario? *Rev Esp Cardiol* 2003;56:408-11.
7. Winkler M, Higgins CB. Suspected intracardiac masses: evaluation with MR imaging. *Radiology* 1987;165:117-22.
8. Lund JT, Ehman RL, Julsrud PR. Cardiac masses: assessment by MR imaging. *AJR Am J Roentgenol* 1989;152:469.
9. Bruna J, Lockwood M. Primary heart angiosarcoma detected by computed tomography and magnetic resonance imaging. *Eur Radiol* 1998;8:66-8.
10. Roldán Torres I, Martí Bonmatí L, Peláez Gonzalez A. Tumor cardíaco que simula miocardiopatía hipertrófica. *Rev Esp Cardiol* 2003;56:734.
11. Perchinsky MJ, Lichtenstein SV, Tyers GF. Primary cardiac tumors: forty years' experience with 71 patients. *Cancer* 1997;79: 1809-15.
12. Schwartzman PR, White R. Imaging of cardiac and paracardiac masses. *J Thorac Imaging* 2000;15:265-73.

13. Freedberg RS, Krozon I, Runnancik WN. The contribution of magnetic resonance imaging to the evaluation of intracardiac tumors, diagnosed by echocardiography. *Circulation* 1988;77:96-103.
14. Hasegawa T, Nakagawa S, Chino M, Kunihiro T, Ui S, Kimura M, et al. Primary cardiac sarcoma mimicking benign myxoma: a case report. *J Cardiol* 2002;39:321-5.
15. Siripornpitak S, Higgins CB. MRI of primary malignant cardiovascular tumors. *J Comput Assist Tomogr* 1997;21:462-6.
16. Araoz PA, Eklund HE, Welch TJ. CT and MR imaging of primary cardiac malignancies. *Radiographics* 1999;19:1421-34.
17. Villacampa VM, Villarreal M. Cardiac rhabdomyosarcoma: diagnosis by MR imaging. *Eur Radiol* 1999;9:634-7.
18. Chiles C, Woodard PK, Gutierrez FR, Link KM. Metastatic involvement of the heart and pericardium: CT and MR imaging. *Radiographics* 2001;2:439-49.