

Case Report

# High-Grade Left Atrial Pleomorphic Sarcoma: Case Report and Review

Martin Ignacio Zapata Laguado<sup>a</sup> Jonathan Orlando Palacios Rojas<sup>b</sup>  
Efraín Alonso Gómez Lopez<sup>c</sup> Martha Lucía Velasco Morales<sup>c</sup>  
Carlos Eduardo Orozco de la Hoz<sup>c</sup>

<sup>a</sup>Department of Internal Medicine, Universidad El Bosque, Bogotá, Colombia;

<sup>b</sup>Department of Critical Care, Universidad la Sabana, Bogotá, Colombia; <sup>c</sup>Department of Coronary Care Unit, Fundación Clínica Shaio, Bogotá, Colombia

## Keywords

Sarcoma · Neoplasms · Heart failure · Heart atria

## Abstract

Primary pleomorphic sarcoma of the left atrium is a rare tumor. There is no actual evidence of the management of this pathological entity, so the main treatment is individualized, surgical management being the cornerstone of the treatment. We present a 78-year-old female who had a clinical picture of heart failure, documenting an atrial mass of the left atrium, with high-grade pleomorphic sarcoma revealed in histopathology. The tumor was surgical removed, with no clinical evidence of residual mass. The tumor recurred again within 3 years, to which the patient succumbed.

© 2017 The Author(s)  
Published by S. Karger AG, Basel

## Case Presentation

A female patient aged 78 years with a clinical record of arterial hypertension consulted the emergency department for appearance of 15 days of worsening in her functional class before the event (NYHA II/IV, now III/IV, with orthopnea, paroxysmal nocturnal dyspnea, and bendopnea). At the first evaluation the patient had normal vital signs according to her age, with positive findings in the physical exam of bilateral rales, abdominojugular test, and

edema of lower limbs. Because of the above it was considered that the patient had a new onset heart failure, and treatment with neurohumoral blockage was started. At the same time a transthoracic echocardiography was done.

The transthoracic echocardiography finding (Fig. 1) was consistent with an atrial mass inside the left atrium, with an area of  $6.4 \times 3.2 \times 6.9$  cm, occupying 60% of the size, without involvement of the mitral valve. The cardiovascular surgery department indicated magnetic resonance imaging of the heart that revealed a mass in the left atrium previously described, dependent on the posterolateral wall, in relation with the pulmonary veins, compressing them without complete obstruction. The mass was of lobulated contours, lightly hyperintense, with respect to the myocardium in T1 and T2 in direct contact with the valvular plane obstructing the output tract in 60–70% and protruding discreetly into the left ventricular cavity (Fig. 2).

The cardiovascular surgeon decides to completely resect this mass, which was done without complications. The sample was sent to pathology (Fig. 3), which reported a malignant, poorly differentiated tumor with epithelioid cells with atypical mitotic figures distributed in sheets accompanied by branched vascular spaces, with areas of hyalinization and necrosis, without recognition of healthy organ. Immunohistochemistry found focal reactivity to CD68 and actin of the smooth muscle, without reactivity to CD34, FLI-1, or S100. The measured proliferation index with Ki-67 was 40%, conclusive of high-grade pleomorphic sarcoma.

The patient was proposed adjuvant therapy with chemotherapy, which she refused. She showed symptomatic complete improvement, and ambulatory follow-up was decided.

The patient came back 3 years later with a clinical picture of decompensated heart failure. Echocardiographic follow-up (Fig. 4) finding tumor recurrence, with pathological fractures and progression of the disease to the woven bone (right humerus, left hip, and lumbosacral region), so it was decided to do conservative and palliative management. A week after hospitalization the patient died.

## Discussion

Cardiac tumors are rare, with an estimated frequency of 0.0017 to 0.33% [1]. Of these tumors, the benign ones correspond to 75% and the malignant ones to 25% [2]. The frequency is similar in males and females, being higher in the right atrium, later left atrium, and both ventricles [3]. The difference with benign tumors, because of the similarity in the figures, lies in the clinical course and the histology [4]. Metastatic tumors are 20–40 times more common than primary malignant cardiac neoplasms [5]; 20% of malignant tumors of the heart present with metastasis. The order of frequency is lung, liver, bones, thoracic lymph nodes, adrenal, and brain [4, 6].

The locoregional recurrence or the appearance of metastasis happens during the first year [7]. Patients with malignant heart tumors of the left side have more frequent distant metastases, while the malignant heart tumors of the right side have more locally advanced disease [7].

In the largest study carried out on cardiac masses, it was found that the average age at appearance of the disease was  $47.1 \pm 16.1$  years [8]. The median follow-up was 51.2 months; until the time to analysis, 69.7% had died, 43% had metastasis, 44.9% of the patients with metastasis had pulmonary metastasis, and 20.9% metastasis to the brain [8].

The clinical presentation of these tumors is very heterogeneous, and the symptoms are due to compromise in ventricular afterload, arrhythmias, embolic events, and metastasis [9,

10]. The patients also can present dyspnea (48%), chest pain (22%), heart failure (13%), and pericarditis (5%). In some cases there have been reports of neurological symptoms, neurovascular syndrome [8, 11], and in some cases cardiogenic shock [12].

Early and immediate detection to start a therapeutic strategy are important to improve the survival of patients [13]. As an initial diagnostic approach, echocardiography could be a useful tool. Magnetic resonance imaging of heart could be utilized in the evaluation of cardiac masses, as it provides contrast resolution to soft tissues and can evaluate extension to the myocardium [14]. Transesophageal echocardiography has proven to be a noninvasive and relatively effective tool [15]. The combination of echocardiography with magnetic resonance imaging of the heart could be the used to differentiate tumor or thrombus [15].

The forecasting characteristics are unspecific; nevertheless, a worse prognosis in patients with high mitotic index, extensive tumor necrosis, infiltration to myocardium, and high grade has been documented [2]. The survival in those patients is less than 1 year with a diagnosis of rhabdomyosarcoma [2]. The main factor related to the survival in patients with these types of malignancies is related to the total resection of the mass, reaching a survival twice as high when the mass is resected totally. Despite of advances in treatment, the prognosis of those patients is bad [16]. The common cause of death after microscopic resection is local recurrence of the tumor, which appears in half of the patients, with survival until 16.5 months [17, 18]. Another factors related to a better prognosis is the anatomical localization, more than the histological type [19].

The use of chemotherapy and radiotherapy is limited [20] to adjuvant therapy in cases where resections are not totally achieved, or in cases where the pathology demonstrates aggressive behavior. The limit of radiotherapy is also well known in cases where sarcoma is shown in pathology, as the radiobiology of these tumors tends to have resistance to it [20].

The use of chemotherapy its related to better survival in patients without metastasis [19]. Recent data suggest that taxanes tend to provide a better survival than standard therapy (ifosfamide + doxorubicin) [8].

### Statement of Ethics

The authors have no ethical conflicts to declare.

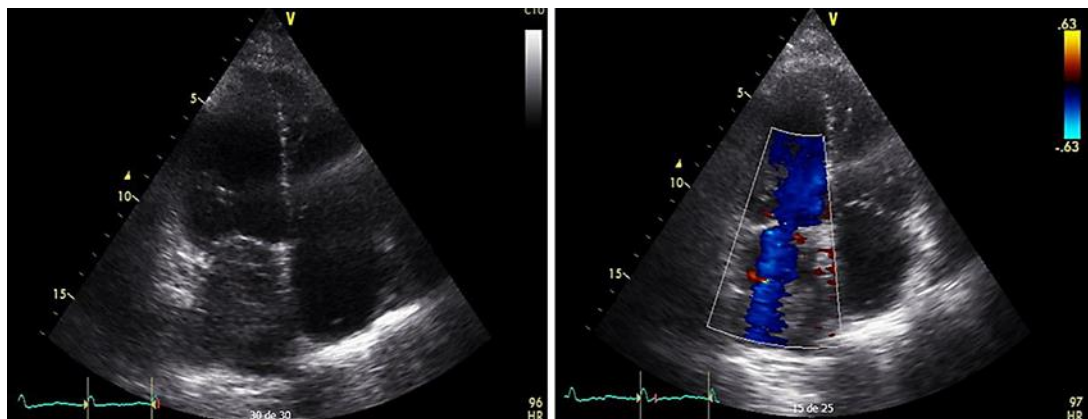
### Disclosure Statement

The authors have no conflicts of interest to disclose.

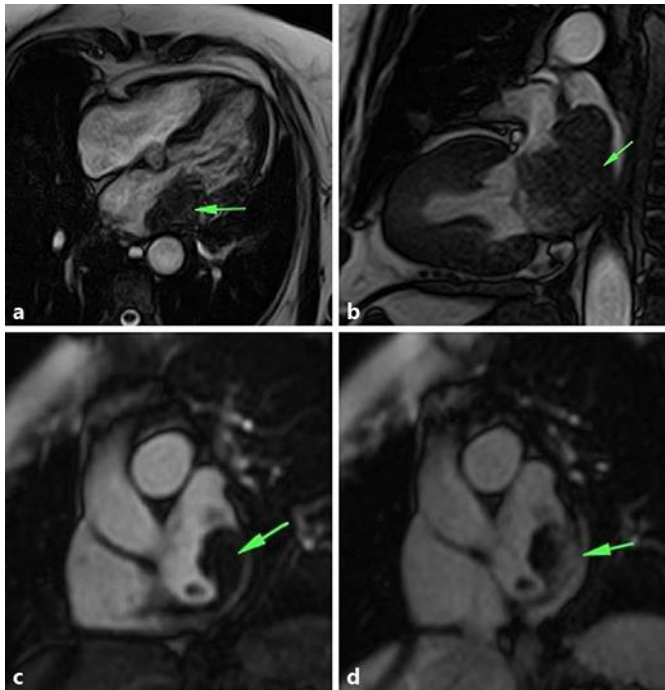
### References

- 1 Corradi D, Contini GA, Gherli T, Nicolini F: A left atrial myxomalike rhabdomyosarcoma. *J Thorac Cardiovasc Surg* 2012;144:e7–e10.
- 2 Burke A, Tazelaar H, Butany J, El-Demellawy D, Loire R, Geva T, et al: Cardiac sarcomas; in Travis W, Brambilla E, Muller-Hermelink H, Harris C (eds): *Pathology and Genetics of Tumours of the Lung, Pleura, Thymus and Heart*. Lyon, IARC Press, 2004, pp 273–281.
- 3 Ma J, Sun JP, Chen M, Zhang L, Xu N, Wang J: Left atrial rhabdomyosarcoma. *Circulation* 2014;129:e503–e505.
- 4 McAllister HA Jr: Primary tumors and cysts of the heart and pericardium. *Curr Probl Cardiol* 1979;4:350–352.

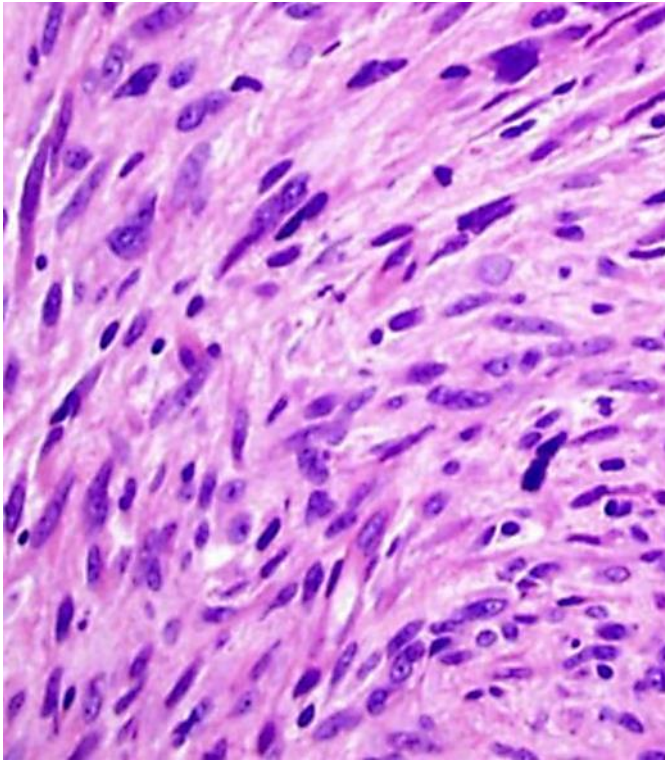
- 5 Fernandes F, Soufen HN, Ianni BM, Arteaga E, Ramires FJ, Mady C: Primary neoplasms of the heart. Clinical and histological presentation of 50 cases. *Arq Bras Cardiol* 2001;76:231–237.
- 6 McManus B, Lee CH: Chapter 69: Primary tumors of the heart; in Libby P, Bonnow RO, Mann DL, et al (eds): *Braunwald's Heart Disease*, ed 8. Philadelphia, Elsevier Science, 2008, pp 1815–1828.
- 7 Ibrahim A, Luk A, Singhal P, Wan B, Zavodni A, et al: Primary intimal (spindle cell) sarcoma of the heart: a case report and review of the literature. *Case Rep Med* 2013;2013:461815.
- 8 Isambert N, Ray-Coquard I, Italiano A, Rios M, Kerbrat P, et al: Primary cardiac sarcomas: a retrospective study of the French Sarcoma Group. *Eur J Cancer* 2014;50:128–136.
- 9 Modi A, Lipnevicius A, Moorjani N, Haw M: Prolonged survival with left atrial spindle cell sarcoma. *Interact Cardiovasc Thorac Surg* 2009;8:703–704.
- 10 Shanmugam G: Primary cardiac sarcoma. *Eur J Cardiothorac Surg* 2006;29:925–932.
- 11 Caballero PE: Left atrial sarcoma presenting as cerebral infarction. *Neurologist* 2008;14:131–133.
- 12 Hsieh CH, Seak CJ, Chiu TF, Chen JC, Li CH: An uncommon cause of heart failure: cardiac sarcomas in the left atrium. *J Emerg Med* 2011;40:e123–e124.
- 13 Canadyova J, Setina M, Smetanova S, Mokravec A: Leiomyosarcoma of the left atrium. *Asian Cardiovasc Thorac Ann* 2008;16:e7–e9.
- 14 Eswaran P, Devadoss P, Narasimhan LS, Kannan K: Synovial sarcoma of the heart: a case report and literature review. *J Cancer Res Ther* 2015;11:659.
- 15 Patil HR, Singh D, Hajdu M: Cardiac sarcoma presenting as heart failure and diagnosed as recurrent myxoma by echocardiogram. *Eur J Echocardiogr* 2010;11:E12.
- 16 Mehta N, Desai A, Shivdasani B, Suryawanshi S, Mehta AB, et al: Left atrial spindle cell sarcoma – case report. *Indian Heart J* 2012;64:416–419.
- 17 Burke AP, Cowan D, Virmani R: Primary sarcomas of the heart. *Cancer* 1992;69:387–395.
- 18 Donsbeck AV, Ranchere D, Coindre JM, Le Gall F, Cordier JF, Loire R: Primary cardiac sarcomas: an immunohistochemical and grading study with long-term follow-up of 24 cases. *Histopathology* 1999;34:295–304.
- 19 Kumar N, Agarwal S, Ahuja A, Das P, Airon B, Ray R: Spectrum of cardiac tumors excluding myxoma: experience in a tertiary center with review of the literature. *Pathol Res Pract* 2011;207:769–774.
- 20 Gupta A: Primary cardiac sarcomas. *Expert Rev Cardiovasc Ther* 2008;6:1295–1297.



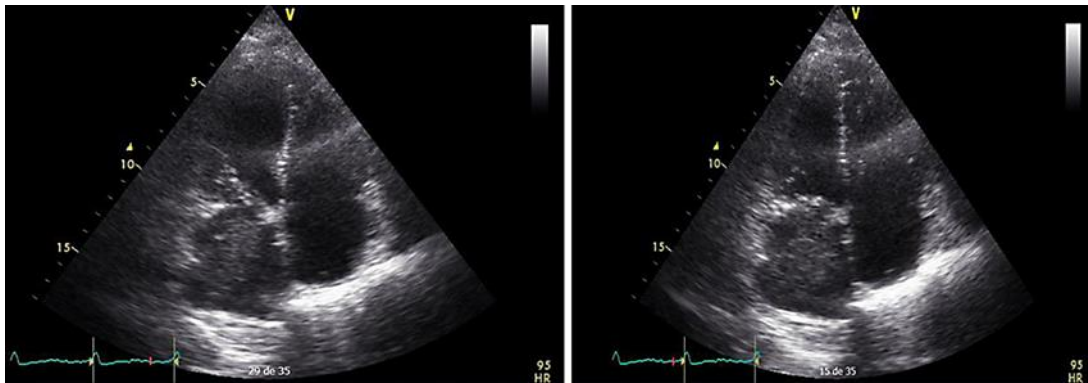
**Fig. 1.** Transthoracic echocardiography, apical projection four chambers, 2D mode, and Doppler color, showing a mass in the left atrium that occupies 60% without mitral regurgitation.



**Fig. 2.** Magnetic resonance imaging of the heart showing a mass dependent on the wall of the left atrium, which extended to the mitral valve plane. **a** SSFP sequence in a four-camera plane. **b** SSFP sequence in a long two-chamber plane showing extension of the mass to the mitral valve plane and to the pulmonary veins. **c, d** Planes of short height at the height of the left atrium during the first step of contrast medium, which shows the rapid capture of contrast in the mass.



**Fig. 3.** Fusiform cells with atypia, pleomorphism, and frequent mitosis. Hematoxylin and eosin stain,  $\times 40$ .



**Fig. 4.** Transthoracic echocardiography, apical projection four chambers, 2D mode, and Doppler color in different phases of the cardiac cycle showing relapse of the initial lesion in the left atrium.