

Right Ventricular Myxoma

Cristina Schneider,¹ Maria Natividade Santos Costa Lopes,¹ Simone Ferreira Leite,¹ Camila Costa Oliveira,¹ Nestor Sabatovicz Jr,² Maria Cristina Resende²

Hospital Anchieta;¹ Hospital Santa Lúcia,² Distrito Federal – Brazil

Introduction

Primary tumors of the heart and pericardium are uncommon, with their incidence ranging from 0.001% to 0.2% in necropsy series, with benign histological characteristics in 75% of the cases.^{1,2}

Cardiac myxomas account for about 50% of benign neoplasms, being unique in most cases. They are preferentially located in the left atrium, with an incidence of 70 to 80%, 18% in the right atrium and, more rarely, from 2 to 5% in the ventricles, or multicentric.³⁻⁶ Myxomas that originate in the right ventricle (RV) and obstruct the right ventricular outflow tract (RVOT) are extremely rare in children.⁵

Patients present at least one of the characteristics of the classical triad described, which includes constitutional, embolic or obstructive symptoms.²

These tumors arise in the endocardium from a mesenchymal or multipotent subendocardial cell, forming a mass, often covered by thrombi, with an average size of 5 cm in diameter, weighing 50 g to 60 g, usually encased in a myxomatous stroma and, histologically, distinguish themselves from organized thrombi because they are covered with endothelium.²

This study reports the case of a young oligosymptomatic patient with massive myxoma in RV, an extremely rare location.

Case Report

A 18-year-old female patient presented malaise and indisposition two months before the consultation. Hypothyroidism was diagnosed. Medicated with levothyroxine and partial improvement, the patient reported, however, chest pain when performing more intense physical activities, tachycardia, asthenia, vertigo and visual disturbances, which improved with rest. On physical examination, HR 111 bpm, BP 101 x 76 mmHg and discrete systolic murmur in the apex, with no other abnormalities.

Electrocardiogram showed sinus tachycardia (102 bpm), right atrial overload and incomplete right bundle branch block.

Keywords

Mixoma/ diagnostic imaging; Mixoma/ surgery; Ventricular Dysfunction Right; Echocardiography/ methods; Magnetic Resonance Spectroscopy/ methods.

Mailing Address: Maria Natividade Santos Costa Lopes • SHIN QI 02, Conj. 04, Casa 16. Postal Code 71.510-040, Lago Norte, Brasília, DF — Brazil

E-mail: naticostalopes@gmail.com

Manuscript received October 25, 2017; revised November 7, 2017; accepted November 7, 2017

DOI: 10.5935/2318-8219.20180007

Transthoracic echocardiogram revealed a large lobulated mobile heterogeneous mass taking up the entire RV cavity (about 5.4 x 2.7 cm), invading both the RVOT and the inflow tract, generating ventricular filling restriction. It was not possible to identify its place of insertion. There was no obstruction to RVOT (Figure 1A).

Chest computed tomography (CT) with intravenous contrast, as well as magnetic resonance imaging (MRI), showed solid expansive bilobed formation in the RV, with a pedicle originating in the interventricular septal subendocardium, filling this chamber and extending to the atrial cavity and RVOT, without infiltrative characteristics (Figure 1B).

The patient underwent surgery to do excision of the mass taking up the entire RV with adhesions and destruction of the tricuspid valve, adhered to the interventricular septum, tricuspid valve repair and closure of a small atrial septal defect, a surgical finding. The procedure was uneventful (Figure 2A).

Anatomopathological study revealed a myxoid-like tumor amidst bleeding and vasocongestion, containing small cells, either isolated or arranged in a row, with eosinophilic cytoplasm, diffusely distributed, not showing atypia or mitotic figures. The study concluded it was a RV myxoma (Figure 2B).

The patient was discharged for outpatient follow-up and the postoperative echocardiogram showed absence of tumor mass, tricuspid repair with moderate degree insufficiency and preserved biventricular systolic function.

Discussion

Cardiac myxoma affects patients in the age range of 15 to 80 years, with an approximate average of 50 years,^{2,4} slight female predominance (5:4) and, despite having a benign histological character, can lead to an adverse outcome, being responsible for incapacitating complications.^{2,3,5}

RV myxomas usually cause symptoms resulting from RVV obstruction, and symptoms of syncope, pulmonary embolism and sudden death may occur.^{6,7} In the literature, there are few reports of isolated cases of RV myxomas, in most cases occurring with RVOT obstruction. Seong et al.⁷ reported the case of a patient with a very similar condition to ours, where a large myxoma originated in the RV, did not obstruct the RVOT and presented dense adhesion to the tricuspid valve leaflet and, as in our case, surgery also included tricuspid valve repair.

Due to the lack of specificity of the symptoms, diagnosis of cardiac tumors is due to clinical suspicion, since they can simulate systemic diseases, valvular diseases, cardiomyopathies and pericardiopathies. Imaging methods, particularly echocardiography, are the tests of choice for diagnosis,⁷ with excellent sensitivity (95%) in detecting the tumor, which may reach 100% when associated with transeosophageal echocardiography.^{3,6}

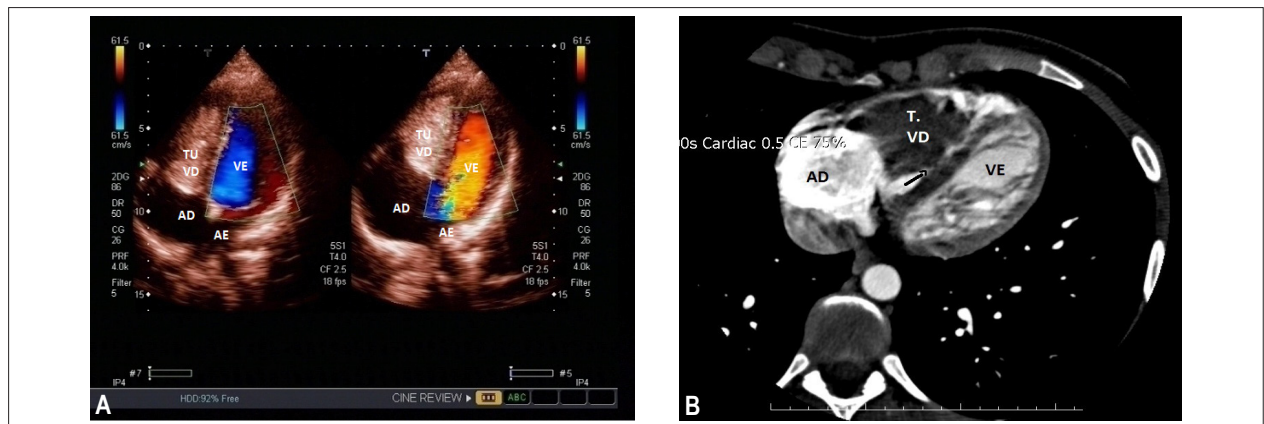


Figure 1 – A: Transthoracic echocardiogram, apical view — Tumor taking up the entire cavity of the right ventricle. B: Computed tomography showing insertion of mass in the interventricular septum.

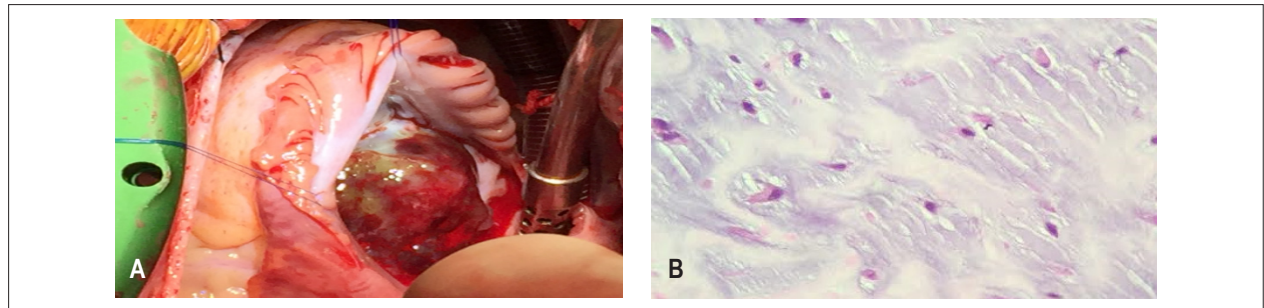


Figure 2 – A: Appearance of the tumor after opening of the right atrium. B: Histopathological aspect, confirming myxoma diagnosis.

CT is useful for determining tumor fixation point, degree of intramural invasion, pericardial involvement and extracardiac structures, while MRI has an excellent correlation with histopathological findings, 3-dimensional view, facilitating the definition of tumor localization and its mobility, being considered the tests of choice to characterize the cardiac tumors.^{8,9}

Definitive diagnosis of myxoma needs confirmation with histopathology tests.

In this case, which illustrates an uncommon spot of tumor appearance, the symptoms presented by the patient were rather nonspecific, but the correlation between the findings in the imaging studies and the anatomopathological study confirmed the diagnosis of myxoma.

Treatment is the surgical removal of the tumor with a good margin of safety, to avoid relapses, although its incidence is very low. As a preventive measure of these recurrences, regular follow-up of the patient is recommended, including regular echocardiographic evaluation.⁸ This has been the procedure adopted for this case.

Authors' contributions

Data acquisition: Schneider C, Lopes MNSC, Leite SF, Oliveira CC, Resende MC; Manuscript drafting: Schneider C, Lopes MNSC; Selection of references: Schneider C, Lopes MNSC, Oliveira CC; Intraoperative documentation: Sabatovicz Jr N, Resende MC.

Potential Conflicts of Interest

There are no relevant conflicts of interest.

Sources of Funding

This study had no external funding sources.

Academic Association

This study is not associated with any graduate program.

References

1. Motta AAR, Colen Filho E, Colen EA, Vieira JAS, Alves MAP, Borges MF, et al. Left atrial myxoma: report of three cases. *Rev Bras Cir Cardiovasc.* 1997;12(4):377-83. doi:http://dx.doi.org/10.1590/S0102-76381997000400011
2. Barbuto C, Sueth DM, Pena FM, Vieira MA, Franklin MM, Teixeira MA. Mixoma atrial esquerdo. *Rev SOCERJ.* 2006;19(2):180-3.
3. Lobo Filho JG, Sales DLS, Borges AEPP, Leitão MC. Mixoma de átrio direito com prolapso para o ventrículo direito. *Rev Bras Cir Cardiovasc.* 2006;21(2):217-20. doi: http://dx.doi.org/10.1590/S0102-76382006000200017
4. Manfroi W, Vieira SR, Saadi EK, Saadi J, Alboim C. Múltipla recorrência de mixomas cardíacos com embolia pulmonar aguda. *Arq Bras Cardiol.* 2001;77(2):161-3. Doi: http://dx.doi.org/10.1590/80066-782x2001000800007
5. Wiegard G, Sieverding L, Kramer U, Haen S, Hofbeck M. Multimodality imaging of a subtotally obstructive right ventricular myxoma in a asymptomatic 10- year-old girl. *Ann Pediatr Cardiol.* 2014;7(2):158-9. doi:10.4103/0974-2069.132519
6. Gribaa R, Slim M, Kortas C, Kacem S, Ben Salem H, Quali S, et al. Right ventricular myxoma obstructing the right ventricular outflow tract: a case report. *J Med Case Rep.* 2014;8:435-7. doi:10.1186/1752-1947-8-435
7. Ho Cho S, Shim MS, Kim W. We the right ventricular myxoma which attached to the tricuspid valve: sliding tricuspid valvuloplasty. *Korean J Thorac Cardiovasc Surg.* 2015; 48(3):228-30. doi:10.5090/kjtcs2015.48.3.228
8. Lacey B W, Lin A. Radiologic evaluation of right ventricular outflow tract myxoma. *Tex Heart Inst J.* 2013;40(1):68-70. PMID:PMC3568267
9. Ponczek MAK, Feitosa FS, Olivetti NQS, Introcaso MC, Ianni B, Mady C. Mixoma em Ventrículo Direito com Embolização Pulmonar. *Rev bras ecocardiogr imagem cardiovasc.* 2010;23(4):108-11.