

Rare Cause of Chest Pain in Young Patient: Left Ventricular Myxoma

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Introduction

Cardiac masses represent 0.2% of all tumors; three quarters of these tumors are benign and 50% are myxomas, with an incidence of 0.0017% in the general population. Of all myxomas, 75% occur in the left atrium, and less than 5% occur in the left ventricle (LV).^{1,2}

Despite being a benign tumor, a myxoma can cause significant morbidity and mortality by affecting the intracardiac blood flow, triggering embolic events and arrhythmia by infiltrating the electrical conduction system of the heart.^{3,4} Since myxoma is often found in the left atrium, current estimates are that only 5% of all myxomas affect the right or left ventricle.^{1,4}

The symptoms are usually associated with mitral valve obstruction.⁵ Therefore, it is commonly accompanied by dyspnea, atypical chest pain, syncope, heart failure and thromboembolic phenomena.^{2,4,5}

Case Report

We report the case of a 24-year-old patient that has had, for 6 months, atypical chest pain, palpitations and shortness of breath. Patient with negative history for type 2 diabetes mellitus, systemic arterial hypertension, nephropathy, thyroid disease, smoking and alcoholism. At the examination, she was in good general condition, conscious and oriented, with peripheral pulses present and symmetrical, blood pressure of 115/80 mmHg, quiet precordium, no fremitus or murmurs, as well as positive vesicular murmurs in both hemithoraxes and absence of adventitious sounds. No lower limb edema and normal abdominal examination. The investigation was initiated with an electrocardiogram, which showed sinus and regular rhythm, with no abnormalities. Chest x-ray and exercise test were also performed, with normal results. Transthoracic echocardiogram (TTE) (Figure 1) showed a rounded mass adhered to the postero-medial LV wall, not quite mobile, with regular borders, homogeneous contents, and no internal Doppler flow. The mass did not compromise

LV flow, the mitral valve, the subvalvular apparatus or the inflow tract of this chamber, and no pedicle was found in the examination. Using planimetry, the mass was estimated at 1.72 × 1.73 cm, area of 1.73 cm² and circumference of 4.65 cm (Figures 1A and 1B). In this case, the mass did not affect the blood flow, as color Doppler revealed LV diastolic filling flow with normal velocities and no evidence of a significant pressure gradient in the ventricular inflow tract (Figure 1C), confirmed by transesophageal echocardiography (Figure 1D, Videos 1 and 2). All of these data were essential for a proper surgical planning and to demonstrate a potential mass influence on the blood flow.

To complement the investigation, cardiac magnetic resonance imaging was performed, whose findings were compatible with left ventricular papillary fibroelastoma. Priority surgical treatment was chosen for a complete mass excision. Longitudinal median sternotomy was performed using cardiopulmonary bypass, followed by cardioplegic cardiac arrest and left atriotomy in the longitudinal direction. The tumor was approached through the mitral valve and was found in the posterior LV wall just below the posterior leaflet of the mitral valve at the junction between P2 and P3, which did not compromise either the muscle structure or the subvalvular apparatus. The tumor was single and located, with an estimated diameter of 1.5 to 2 cm, with a calcified appearance. It was completely excised along the margin of muscle at the base, to avoid relapses (Figure 2). The mitral valve was then tested and proved to be competent. Left atrial suture, and withdrawal of the patient from extracorporeal circulation were performed, with the heart under sinus rhythm. The piece was sent for histopathological study, which showed findings compatible with cardiac myxoma (Figure 3). The patient progressed clinically well and uneventfully. Control TTE showed normal mitral valve and normal LV blood flow. On the 10th postoperative day, the patient was discharged from the hospital in good clinical condition and was followed for 4 years in an outpatient setting, with no further episodes of chest pain, dyspnea, palpitations or other complaints over these four years.

Discussion

The clinical presentation of the myxoma may be nonspecific, with chest pain, dyspnea, fatigue, palpitations⁵ and, often, diagnosis is delivered by incidental finding of the mass. Weight loss may also occur, although the patient did not present any. Diagnosis can be challenging, especially in young patients, as in this report.

Depending on the size, location and characteristics of the mass, symptoms secondary to the obstructive process (such as functional mitral stenosis), abnormal intracardiac blood flow (stroke) and infiltration of the electrical conduction system of the heart (causing arrhythmia) may occur. Tumor fragments

Keywords

Chest Pain; Dyspnea; Tachycardia; Syncope; Echocardiography/diagnostic Imaging; Myxoma/surgery; Heart Neoplasms.

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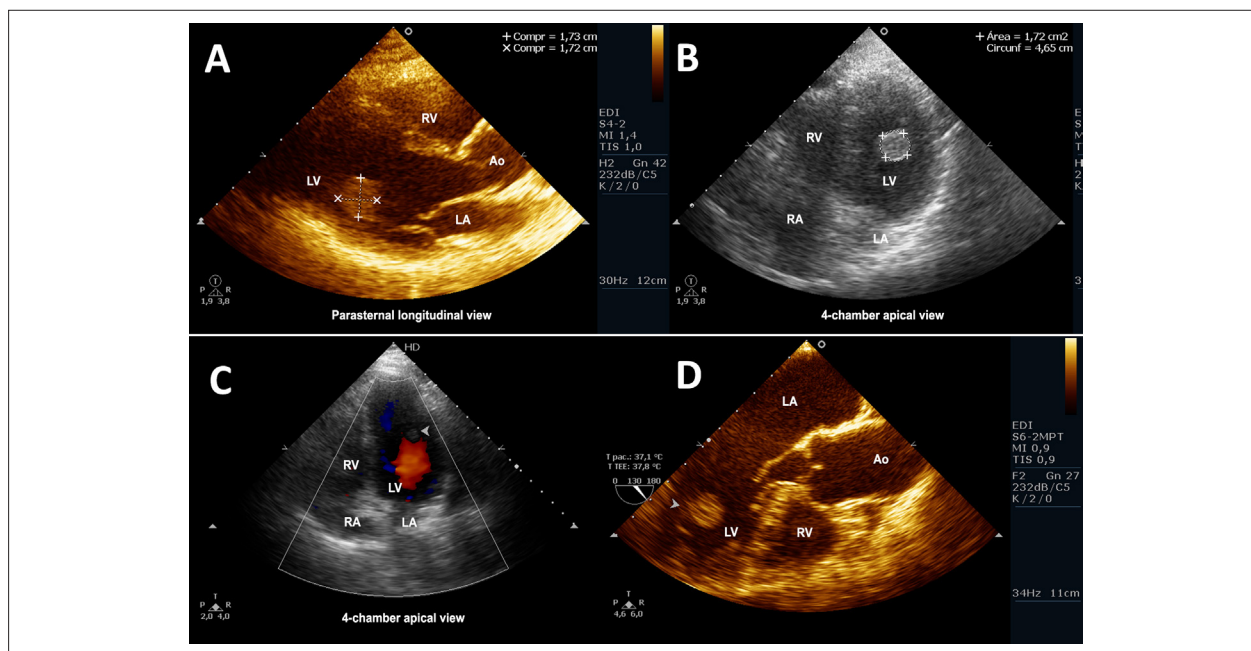


Figure 1 – (A) Transthoracic echocardiography. Longitudinal parasternal view showing a circular mass measuring 1.7 × 1.7 cm, located in the postero-medial left ventricular (LV) wall not compromising the mitral subvalvular apparatus. (B) Transthoracic echocardiography. 4-chamber apical view showing, by planimetry, a mass with an approximate area of 1.73 cm² and circumference of 4.65 cm. (C) Transthoracic echocardiography. 4-chamber apical view showing, on color Doppler, LV diastolic filling flow with normal velocities and no evidence of any significant pressure gradient in the ventricular inflow tract. (D) Transesophageal echocardiogram at 130° confirming the presence of mass adhered to the postero-medial LV wall. RV: right ventricle; LA: left atrium; Ao: aorta; RA: right atrium.

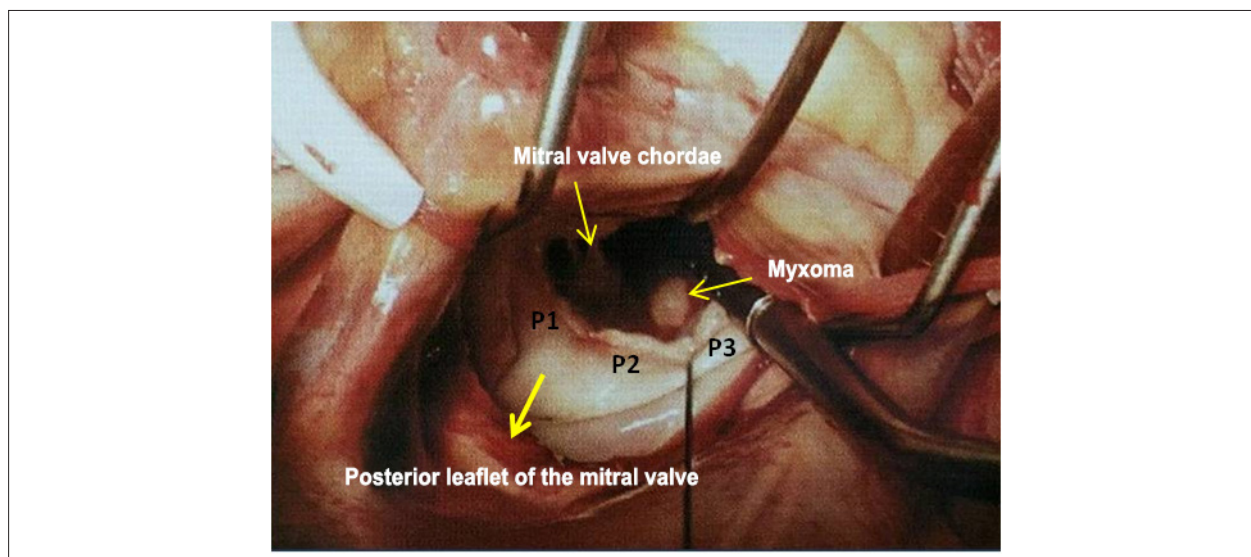


Figure 2 – Left atriotomy for transvalvular approach of left ventricular myxoma, showing calcified mass at the base of the postero-medial papillary muscle (at the P2/P3 level of the posterior leaflet of the mitral valve) not compromising the subvalvular or muscular structure. The tumor was single and located, with an estimated diameter of 1.5 to 2 cm.

may promote pulmonary or systemic embolism, depending on the location of the myxoma.⁴

Therefore, the initial presentation may be in the presence of a stroke, as found by Long and Gao,² who described 15 cases of cerebral embolization secondary to myxoma. Some tumors (about 4%) are completely asymptomatic and diagnosed incidentally.

Another form of presentation occurs when the cardiac myxoma is one of the components of Carney's syndrome. It is an autosomal dominant inherited disorder (typically occurring in several family members) in which schwannoma, cardiac and/or extracardiac myxomas, endocrine tumors and skin pigmentation disorders may occur.^{6,7} It is estimated that this syndrome is responsible for 5 to 7% of cardiac myxomas,

Case Report

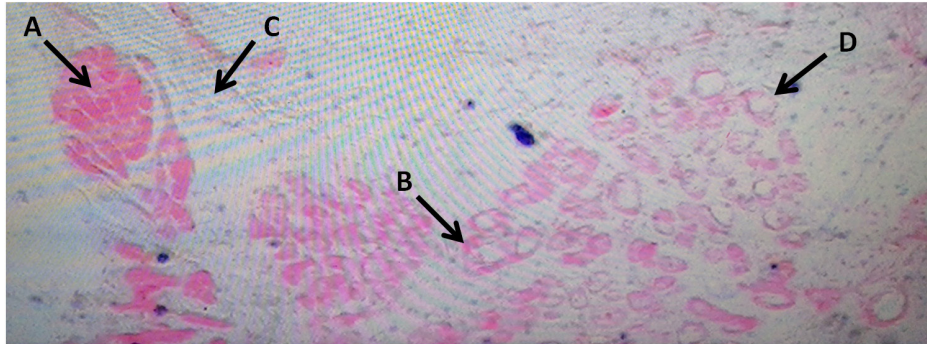
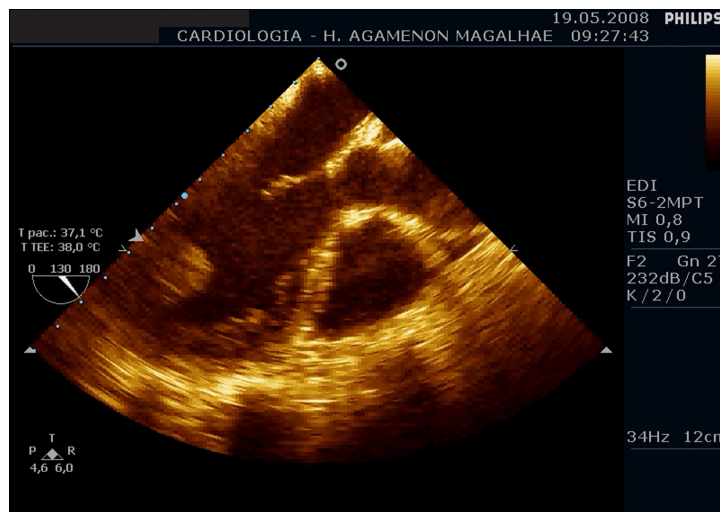


Figure 3 – Pathology study: benign neoplasia consisting of star-shaped or globular cells (A), endothelial cells (B) and mature smooth muscle cells inserted in a loose connective tissue (C), eosinophilic cells, in which blood vessels of different diameters (D), often congestive, are observed. These findings are consistent with a cardiac myxoma.



Video 1 – Transesophageal echocardiogram confirming the presence of mass adhered to the postero-medial left ventricular wall. Access the video here: http://departamentos.cardiol.br/dic/publicacoes/revistadic/2019/v32_1/video_v32_1_260_ingles.asp

occurring in younger patients with greater chances of relapse compared to sporadic myxomas.⁶ Although the case discussed in this manuscript is a 24-year-old patient, the possibility of Carney's syndrome was ruled out because none of the other characteristics were present. Besides, outpatient follow-up was performed for more than 4 years with no signs of relapse.

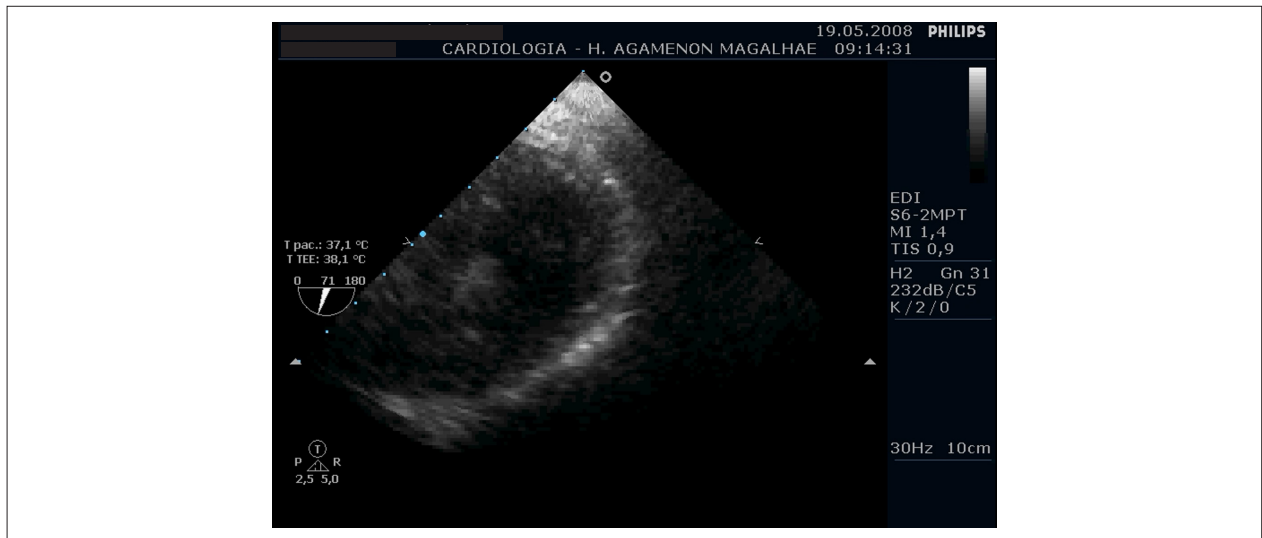
For the investigation, TTE is currently the first-line test to be performed because of its immediate availability, and its non-invasive and inexpensive nature. In addition, it allows the simultaneous analysis of valve function and cardiac chamber. When associated with Doppler, TTE allows the detection of stenosis and/or valve defects and small valve masses (< 1.0 cm).³

In some situations, the differential diagnosis of myxoma and thrombi can be challenging, especially when the left atrial thrombus presents as a large organized mass, making them indistinguishable on TTE. In these cases, clinical and echocardiographic findings alone are not sufficient to safely differentiate one another, so a more detailed examination is required.³

Computed axial tomography differs from the TTE by showing the detailed mass morphology, location and extent. In addition, it can analyze adjacent structures (pericardium and large vessels) with risk of metastatic infiltration.³ A T1 magnetic resonance imaging scan shows a myxoma and allows delineating ovoid or rounded masses and lobed masses with no signs of heterogeneity. T2 scans show masses with hyperintensity, depending on the content of the lesion. In addition to a myxoid substance, magnetic resonance imaging may reveal thrombi, calcification, cystic or necrotic substance and bleeding areas.⁸

The preferred treatment is surgical resection, which has an excellent prognosis. As found in other studies, Barreiro et al.⁹ reported a 0% mortality rate at 30 days to 1 year postoperatively. Considering late mortality (3 months to 22 years) after surgery, the figures range from 0.4% to 5% of the cases, with most deaths related to embolic events.

In this case, the patient presented nonspecific symptoms, but the echocardiography findings associated to cardiac magnetic resonance imaging enabled the diagnosis and recommendation



Video 2 – Transesophageal echocardiogram on color Doppler confirming the presence of mass adhered to the postero-medial left ventricular wall not compromising the mitral valve, the subvalvular apparatus or the left ventricular inflow tract. Access the video here: http://departamentos.cardiol.br/dic/publicacoes/revistadic/2019/v32_1/video_v32_1_260_ingles.asp

of the surgery. The pathology study of the resected mass confirmed it to be LV cardiac myxoma. As reported in this case, postoperative follow-up is recommended for the early identification of relapses, although they are infrequent.¹⁰

Authors' contributions

Data acquisition: Góes GHB, Calado Filho IP, Sobral Filho DC; Data analysis and interpretation: Góes GHB, Ferraz Neto JCS, Calado Filho IP, Lima RC, Sobral Filho DC; Manuscript writing: Góes GHB, Ferraz Neto JCS; Critical revision of the manuscript as for important intellectual content: Góes GHB, Calado Filho IP, Lima RC, Sobral Filho DC.

Potential Conflicts of Interest

There are no relevant conflicts of interest.

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Academic Association

This study is not associated with any graduate programs.

References

- Fakhari S, Bilehjani E. A large left ventricle myxoma presenting with epigastric pain and weight loss. *Case Rep Cardiol*. 2016;2016:9018249.
- Long Y, Gao C. Brain embolism secondary to cardiac myxoma in fifteen chinese patients. *ScientificWorldJournal*. 2014 Mar 9;2014:718246.
- Plana JC. Three-dimensional echocardiography in the assessment of cardiac tumors: the added value of the extra dimension. *Methodist Debakey Cardiovasc J*. 2010;6(3):12-9.
- Alvarado-Castro C, Vega-Brizneda MP, Matijasevic-Arcila E, Maldonado-Escalante JD, Buitrago-Sandoval A. Left ventricular myxoma. *Rev Colomb Cardiol*. 2017;24(3):300.e1-300.e5.
- Nehaj F, Sokol J, Mokan M, Jankovicova V, Kovar F, Kubaskova M, et al. Outcomes of patients with newly diagnosed cardiac myxoma: a retrospective multicentric study. *Biomed Res Int*. 2018 Feb 6;2018:8320793.
- Bandettini WP, Karageorgiadis AS, Sinaii N, Rosing DR, Sachdev V, Scherthaner-Reiter MH, et al. Growth hormone and risk for cardiac tumors in Carney complex. *Endocr Relat Cancer*. 2016;23(9):739-46.
- Carney JA. Carney complex: the complex of myxomas, spotty pigmentation, endocrine overactivity and schwannomas. *Semin Dermatol*. 1995;14(2):90-8.
- Braggion-Santos MF, Koenigkam-Santos M, Teixeira SR, Volpe GJ, Trad HS, Schmidt A. Magnetic resonance imaging evaluation of cardiac masses. *Arq Bras Cardiol*. 2013;101(3):263-72.
- Barreiro M, Renilla A, Jimenez JM, Martin M, Al Musa T, Garcia L, et al. Primary cardiac tumors: 32 years of experience from a Spanish tertiary surgical center. *Cardiovasc Pathol*. 2013;22(6):424-7.
- Wang Z, Chen S, Zhu M, Zhang W, Zhang H, Li H, et al. Risk prediction for emboli and recurrence of primary cardiac myxomas after resection. *J Cardiothorac Surg*. 2016 Feb 2;11:22.