

A Three-Leaflets Mitral Valve Associated to Three Papillary Muscles in a Patient with Hypertrophic Cardiomyopathy

Válvula Mitral com Três Folhetos Associada a Três Músculos Papilares em Paciente Portador de Cardiomiopatia Hipertrofica

Antonio Tito Paladino^{1,2}, Luiz Antônio Vasconcelos¹, Leonardo Mello Guimarães de Toledo¹, Natalia Massoni¹, Mariana Oliveira Rezende¹, Natasha Soares Simões dos Santos¹, David Costa de Souza Le Bihan¹, Rodrigo Bellio de Mattos Barretto¹, Jorge Eduardo Assef¹

¹Departamento de Ecocardiografia, Instituto Dante Pazzanese de Cardiologia, São Paulo, SP, Brazil. ²Departamento de Emergência Cardiovascular, Instituto Dante Pazzanese de Cardiologia, São Paulo, SP, Brazil.

Abstract

Mitral valve malformations are considered rare even in their association with other anomalies, except for mitral valve prolapse, which is found in approximately 10% of the population. Mitral valve abnormalities may affect any portion of the valve or subvalvular apparatus (annulus, cusp, tendinous cords or papillary muscles).^{1,2} Mitral valve anatomy and function can be assessed by echocardiography, resonance or cardiac tomography, and is essential for potential therapeutic programming.²

Introduction

Malformations of the mitral valve are rare, even considering the association with other anomalies, exception made to mitral valve prolapse, that can be observed in almost 10% of the population.¹ Any part of the valvular or subvalvular apparatus can be affected (annulus, leaflets, tendinous cords and papillary muscles), causing stenosis or insufficiency.

Echocardiography, cardiac resonance and cardiac tomography can evaluate mitral anatomy and function precisely, being a cornerstone to diagnosis and treatment of such anomalies.³

Case Report

A 57-year-old woman with hypertrophic cardiomyopathy was referred for routine evaluation in our echo lab. Her transthoracic echocardiogram revealed asymmetric ventricular hypertrophy, with septal predominance and peak rest gradient in outflow tract of 152 mmHg. Interestingly, the mitral valve had an unexpected appearance of a Shamrock, with three leaflets and their corresponding papillary muscles (figure 1).

Keywords

Heart Defects, Congenital; Echocardiography; Magnetic Resonance Imaging; Tomography.

Mailing Address: Antonio Tito Paladino •

Instituto Dante Pazzanese de Cardiologia, Avenida Dr. Dante Pazzanese, 500, Vila Mariana. CEP 04012-909, São Paulo, SP, Brazil
E-mail: atpf40@gmail.com

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Doppler evaluation revealed moderate regurgitation without significant stenosis. There was also a systolic anterior movement of valve apparatus. Biventricular function was preserved and no other cardiac structural malformations were found. Mild pericardial effusion was seen, without any hemodynamic compromise.

For a better diagnostic evaluation, cardiac magnetic resonance imaging was performed. In cine images (2, 3, 4 chambers), three papillary muscles were well individualized, with tendinous cords attached to them. Two muscles were located in their usual position (mid part of inferolateral and anterolateral walls). Additionally, an accessory papillary muscle could be seen emerging from the basal segment of the inferolateral wall, which had been observed in two-dimensional transthoracic echocardiogram (Figure 2).

Three-dimensional transesophageal echocardiography confirmed the three papillary muscles, attached to three functional leaflets through tendinous cords.

Discussion

Tricuspid mitral valve has been described, associated or not with hypertrophic cardiomyopathy. Supernumerary papillary muscles and leaflets are conceivable, since both have the same embryology origin.^{4,5}

In previous studies, three-leaflet mitral valves have been associated with left ventricle outflow obstruction, associated or not with hypertrophic cardiomyopathy.³ This association, also described by other authors, is noteworthy and could suggest a relationship between the genetic disorders observed in hypertrophic cardiomyopathy and the embryology of papillary muscles and mitral leaflets.^{3,6}

In this case, we need to emphasize that 3D transesophageal echocardiography was the imaging technique that allowed a better view of the mitral valve, with its “three-leaflet” morphology.

Authors' contributions:

Research creation and design: Paladino Filho AT. Data acquisition: Paladino Filho AT, Rezende M, Soares N, Massoni N. Data analysis and interpretation:

Vasconcelos LA, Toledo L. Manuscript writing: LeBihan

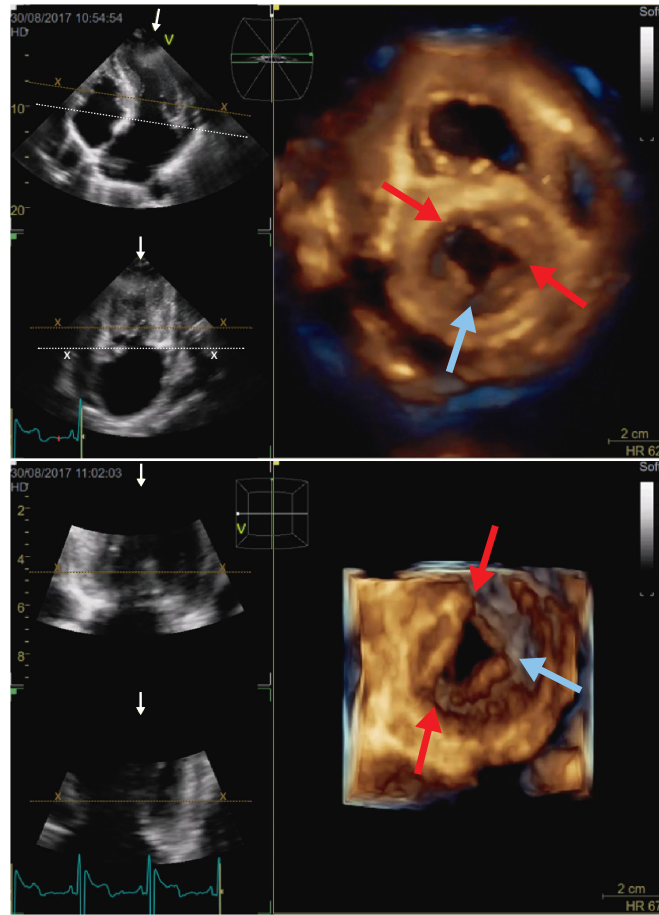


Figure 1 – Red arrows: Anterolateral and posteromedial commissure. Blue arrow: “Extra commissure” dividing the posterior leaflet.

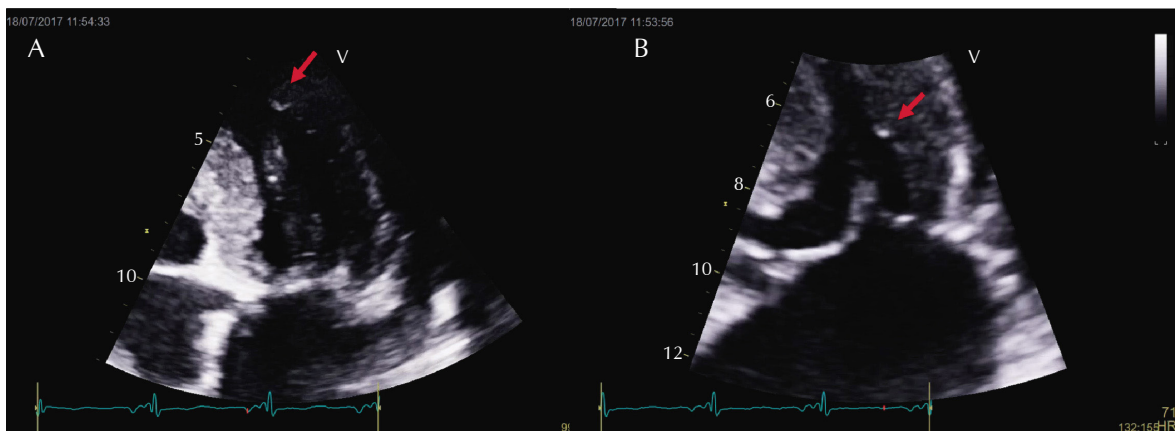


Figure 2 – A) Apical implant of antero-lateral papillary muscle. B) Extra papillary muscle (basal position).

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Conflict of interest

The authors declare that there is no conflict of interest regarding this manuscript.

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