

Mitral Hemi-Arcade: A Rare Variant of a Rare Disease



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INTRODUCTION

Papillary muscle hypertrophy has been classically reported in the literature as a part of hypertrophic obstructive cardiomyopathy (HOCM).^{1,2} The standard treatment is surgical debulking when septal myectomy is done after failure of medical therapy.³ However, there are few reports in which this anomaly exists in the absence of HOCM, such as in association with Fabry's disease, in which it can constitute up to 20% of the left ventricular mass.⁴ We report a case in which this variant, along with congenitally abnormal papillary muscle insertion into the anterior mitral leaflet, created a dynamic left ventricular outflow tract (LVOT) obstruction.

CASE PRESENTATION

A 31-year-old Caucasian man with a medical history of ulcerative colitis was referred to our facility for echocardiography after an episode of

pleuritic left-sided chest pain that spontaneously resolved. He did not have a history of hypertension, aortic stenosis, or kidney disease. Electrocardiography showed anterolateral early repolarization changes and T-wave inversion in lead III (Figure 1).

Physical examination revealed a heart rate of 68 beats/min. Blood pressure was 130/80 mm Hg. Jugular venous pressure and the carotid vessels were normal. S1 and S2 were physiologic. There was a soft systolic murmur at the lower left sternal border that got softer with Valsalva maneuver and sitting forward. The patient had normal peripheral pulses and no carotid bruits.

Transthoracic echocardiography showed a normal ejection fraction of 60% and mildly increased thickness of the septum at 13 mm. There was also mildly increased left ventricular wall thickness with increased apical anterior and anterolateral wall thickness of 11 mm. LV mass was estimated to be 267 g, and left ventricular mass index was 129 g/m², with relative left ventricular wall thickness of 0.55. Interestingly,

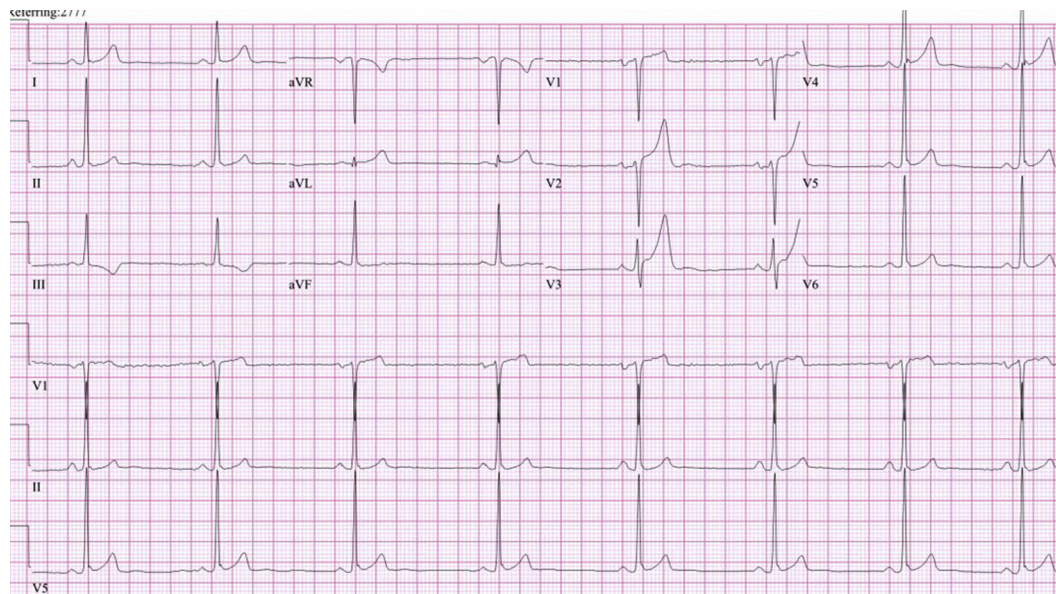


Figure 1 Electrocardiogram showing anterolateral early repolarization changes and T-wave inversion in lead III.

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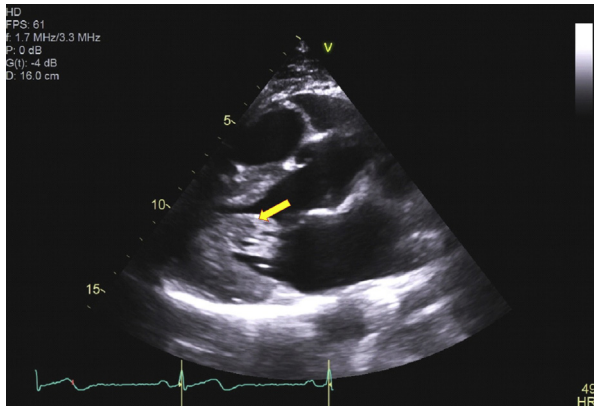


Figure 2 Parasternal long-axis echocardiographic view showing hypertrophied papillary muscle (*yellow arrow*).

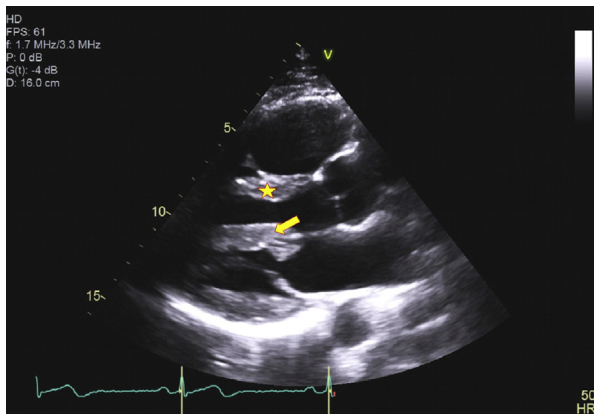


Figure 3 Parasternal long-axis view showing hypertrophied septum (*star*) and papillary muscle (*yellow arrow*).

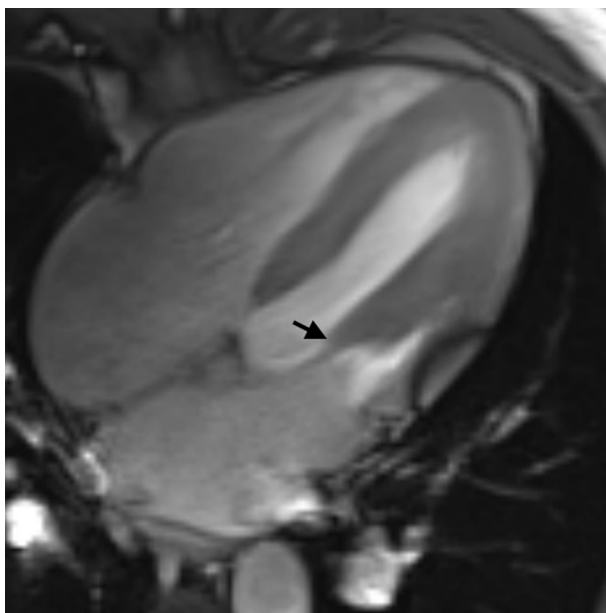


Figure 4 Systolic image from a four-chamber steady-state free-precession cine image demonstrating an enlarged and elongated anterior papillary muscle (*arrow*) inserting directly onto the anterior mitral valve leaflet and blocking a portion of the LVOT.

although the mitral valve leaflets were normal, there was marked hypertrophy of the anterolateral papillary muscle, which directly inserted into the midportion of the anterior mitral leaflet (Figures 2 and 3, Videos 1–4). There was no LVOT obstruction at rest or with Valsalva maneuver. Global average longitudinal strain was -18% . The left atrium size and diastolic function were normal.

To further characterize these findings and investigate the possibility of hypertrophic cardiomyopathy, such as myocardial crypts or delayed myocardial enhancement, the patient underwent cardiac magnetic resonance imaging. Cardiac magnetic resonance imaging confirmed the presence of elongated and enlarged papillary muscles with short chordae tendineae. The anterolateral papillary muscle was abnormal, with a large portion of the muscle inserting directly onto the anterior mitral valve leaflet. The posterior medial papillary muscle was not elongated, and the subvalvular apparatus associated with it appeared normal with normal chordae tendineae (Videos 5 and 6). The anterior papillary muscle encroached upon the LVOT. Extending from this insertion, there was some thickened soft tissue along the free edge of the anterior mitral valve leaflet, which involved roughly two-thirds of the leaflet free edge but did not communicate with the posterior medial papillary muscle, because of the normal subvalvular apparatus in this location disrupting potential mitral arcade formation and resulting in a mild “hemi-arcade” appearance (Figure 4).⁵ There was mild to moderate eccentric mitral regurgitation, which could have been exacerbated by this “hemi-arcade” configuration. There was no delayed late gadolinium enhancement and no myocardial crypts.

During exercise oxygen consumption testing, the patient reached maximal effort, obtaining 96% of maximal predicted heart rate, but the maximal oxygen consumption was 20.8 mL/kg/min, which was 48% of predicted. Pulmonary ventilation/carbon dioxide production slope was 46, and oxygen consumption/heart rate was 51% of predicted, suggesting a cardiac output limitation (Figure 5). To attempt to explain the impaired cardiac output during exercise, the patient underwent stress echocardiography. With stress, the LVOT maximal instantaneous gradient was 73 mm Hg (Figures 6–8, Video 7). The mechanism of obstruction was likely LVOT crowding from the papillary muscle and cavity obliteration.

The patient was started on β -blockers and was doing well at follow-up. There were several discussions regarding exercise restrictions. He was advised against competitive sports and encouraged to exercise as per recommendations for patients with provokable gradients.^{6,7}

DISCUSSION

Congenital abnormalities of the mitral valve apparatus can be valvular, supravulvar, or subvalvular, leading to various degrees of mitral regurgitation and/or stenosis. As first described by Layman and Edwards⁸ in 1967, a classic mitral arcade is a subvalvular abnormality characterized by a fibrous tissue along the free edge of the mitral valve leaflet communicating with the posterior papillary muscle via an elongated posterior papillary muscle that inserted onto the valve or shortened and thickened posterior papillary muscle. In the case described here, the posterior papillary muscle and its subvalvular apparatus (chordae) were normal, and the “arch” that would form the arcade was broken, resulting in a “hemi-arcade.”⁹ This hemi-arcade did not restrict the anterior mitral leaflet, as it would in a complete arcade, thus allowing the development of a dynamic LVOT obstruction.

Solitary hypertrophied papillary muscle, at times silent and discovered only during autopsy, has been reported to cause electrocardiographic abnormalities including T-wave inversions and U waves¹⁰ and atrial fibrillation.^{5,11} The abnormal echocardiographic features

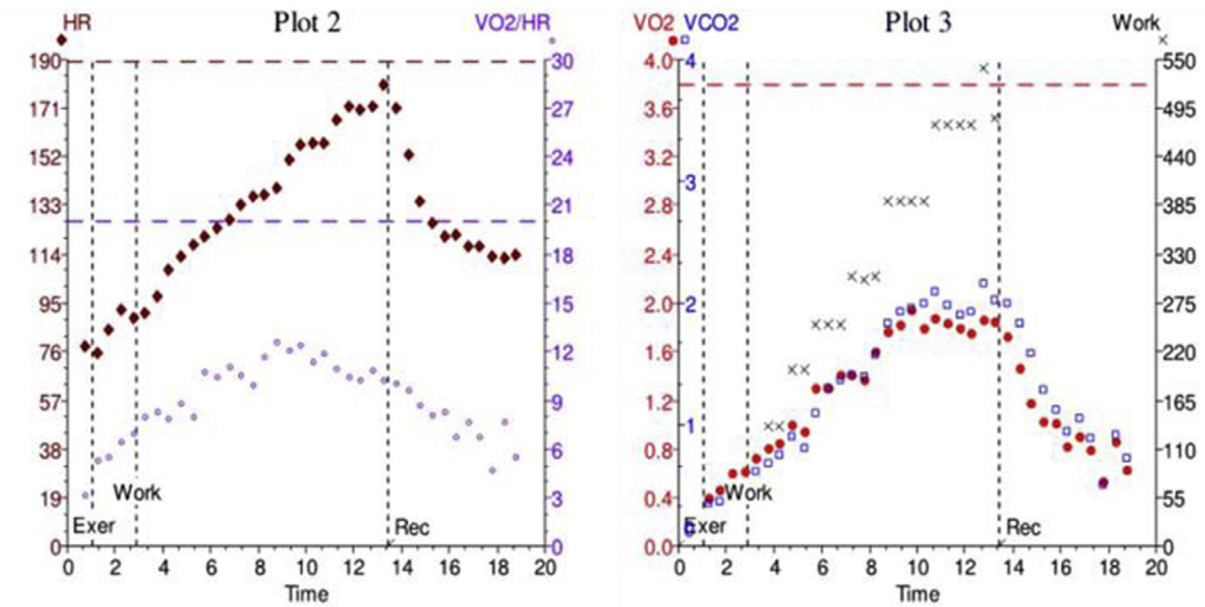


Figure 5 Cardiac output limitations during cardiopulmonary exercise testing. *Exer*, Exertion; *Rec*, recovery.

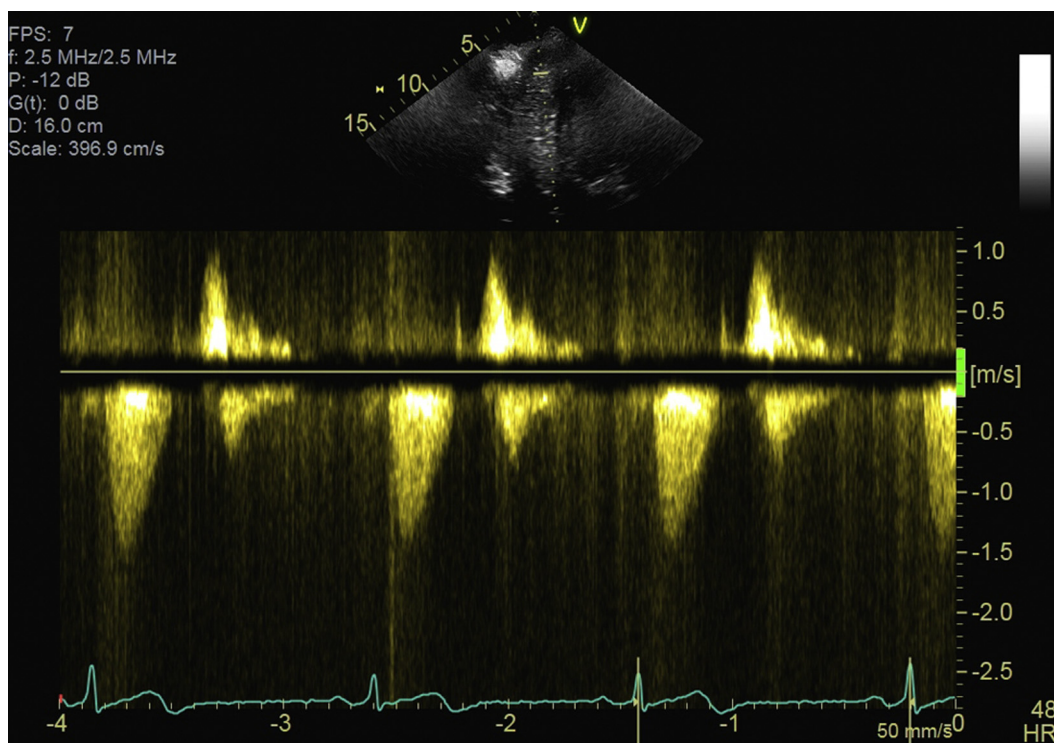


Figure 6 Continuous-wave Doppler of the LVOT at rest.

of the papillary muscles described in our case in the absence of additional features of HOCM should raise suspicion for the presence of congenital papillary muscle anomalies. Similar to HOCM, stress testing provides added value to assess LVOT obstruction with exercise. Previously postulated mechanisms for LVOT obstruction include the drag effect, whereby the mitral valve or subvalvular apparatus gets pushed toward the septum because of high-velocity force of flow and abnormal anatomy and position of the anterior

mitral leaflet and subvalvular apparatus.¹² Del Guzzo and Sherrid¹³ suggested that the bulky papillary muscle creates turbulent flow, leading to subaortic fibrosis, stenosis, and direct systolic traction that may explain the obstruction and septal hypertrophy. Similar to other forms of LVOT obstruction, surgery with debulking of the papillary muscle and likely septal myectomy or myotomy is indicated on the basis of the presence of associated symptoms and the degree of outflow obstruction.¹⁴

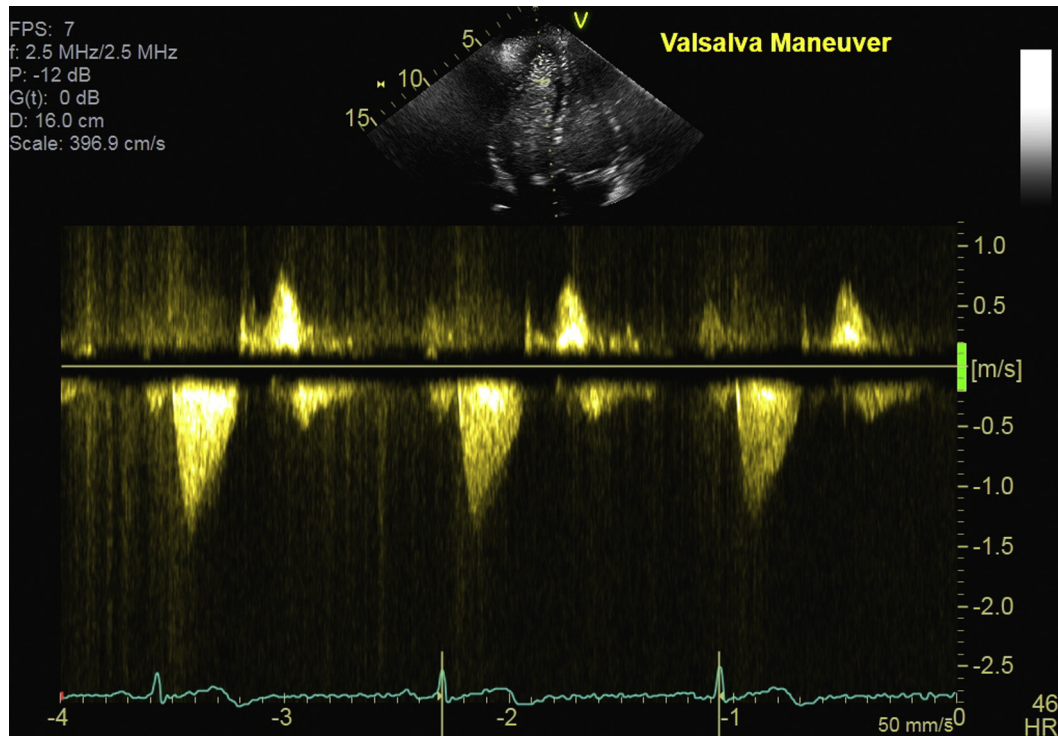


Figure 7 Continuous-wave Doppler of the LVOT with Valsalva maneuver.

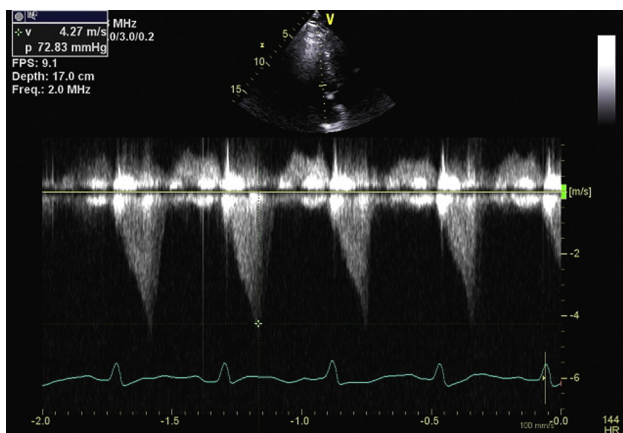


Figure 8 Continuous-wave Doppler of the LVOT during stress.

CONCLUSION

Solitary hypertrophied and abnormally inserted papillary muscle is a very rare isolated entity. We describe a case in which this anomaly caused significant obstruction and exercise impairment in an otherwise healthy and asymptomatic young man. This anomaly should be suspected when an abnormal papillary muscle is noted on echocardiography without other features of hypertrophic cardiomyopathy. Additional testing such as exercise echocardiography and cardiac magnetic resonance could confirm this congenital abnormality.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <http://dx.doi.org/10.1016/j.case.2017.06.007>.

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