

Mitral Valve Prolapse in a Healthy Middle Age Female Subject

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Abstract

This case study illustrates that a perfectly healthy person can develop mitral valve prolapse (MVR) without any warning or heart ailment. A 61 year old healthy looking female with no prior known heart disease and excellent tolerance to exercise complained of developing shortness of breath. On physical exam, she was diagnosed with the new onset of heart murmur. Transthoracic echocardiography (TTE) exam was done which confirmed the prolapsing of the P2 segment of the posterior leaflet of the mitral valve. She underwent mitral valve replacement. Subsequent TTE ultrasound exam showed no mitral insufficiency.

INTRODUCTION

Mitral valve prolapse (MVP) is a common valve abnormality which affects 2-3% of the general population.¹ It is characterized by a systolic displacement of one or both mitral leaflets below the mitral annulus plane into the left atrium (LA).² MVP can be distinguished into primary or 'nonsyndromic' MVP, and secondary or 'syndromic' MVP. In the non-syndrome case, the findings from the Farmingham Heart Study (FHS)¹ suggest that MVP is a progressive disease affecting predominantly middleaged individuals. In the syndromic case, MVP occurs in the presence of connective tissue disorders such as Marfan syndrome, Loeys-Dietz syndrome, Ehlers-Danlos, osteogenesis imperfecta, pseudoxanthoma elasticum, and the recently reported 'aneurysmsosteoarthritis syndrome'.3,4 MVP has also been observed in hypertrophic cardiomyopathy.5 MVP can be associated with significant mitral regurgitation (MR), bacterial endocarditis, congestive heart failure, and even sudden cardiac death, of which the substantial percentage being asymptomatic young individuals.6,7

MVP is a medical entity that is not fully understood although being known for a long time. It was first described by "mid-systolic click" back in 1887 by Cuffer

and Barbillon.8 Later in 1963 Barlow9 demonstrated the presence of MR by angiography in patients with "mid systolic click" murmur. The etiology of mitral valve prolapse can be degenerative (myxomatous), fibroelastic deficiency, acute rheumatic disease, Marfan's syndrome, bacterial endocarditis, papillary muscle rupture after acute myocardial infarction (MI) and acute ischemia. Various symptoms associated with MVP include atypical chest pain, exertional dyspnea on exertion, syncope and clinical findings are low blood pressure, leaner build and electrocardiographic repolarization abnormalities. During physical exam the classic auscultatory finding in MVP is a midto-late systolic click frequently associated with a high-pitched, late systolic murmur; however its specificity is limited. Physical examination and twodimensional transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) are the diagnostic gold standards for MVP.

Here a case is discussed about a 61 year old healthy looking female who is diagnosed with a recent onset of murmur

CASE REPORT

A healthy 61 years old female with no known chronic health problems presents for a transthoracic echocardiography for a recent onset of murmur. She reports that until recently she had excellent exercise

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tolerance and was able to hike steep mountain trail - "White House" trail in 'Canyon de Chelly' locally. This year she did that hike twice with her daughter without any issue. Most recently she had to stop three times coming up to catch her breath. She feels severely dizzy if she walks quickly and her heart "pounds". She is not had any chest pain but has had some recent heartburn. No orthopnea or PND. Apart from this she appears in good health and distress free. A TTE exam was performed on a GE Vivid S6 a dedicated echocardiography ultrasound machine using a M4S sector array transducer. A series of images were obtained from different windows, mainly parasternal long axis (PLAX), parasternal short-axis (PSAX) and apical views. In the parasternal long axis view, Figure 1, the posterior leaflet of the mitral valve show systolic displacement below the mitral annulus and into the left atrium. There is significant anteriorly directed mitral regurgitation, Figure 1b, which is consistent

with posterior leaflet defect. The left atrium is also dilated due to increased pressure. The mitral valve leaflets seen in the parasternal short axis are shown in Figure 2. At the onset of systolic phase the anterior and posterior leaflets should co-apt completely but in Figure 2 the P2 segment of the posterior leaflet is seen to prolapse and shows significant regurgitation at the onset of systolic phase (Figure 2b). Apical 4 chamber view of the mitral leaflet is shown in Figure 3. The P2 segment of the posterior leaflet is prolapsed during the systolic phase. Significant regurgitation due to prolapse of the P2 segment is shown in the CW Doppler spectrum, Figure 4. Patient was sent to University of Arizona Medical Center in Tucson for possible mitral valve repair or replacement. Mitral valve was eventually replaced. Patient was given a prosthetic mitral valve. Repeated TTE after valve replacement showed no regurgitation. This is shown in the apical view, Figure 5.



Figure 1. Parasternal long axis (PLAX) view recorded in patient with mitral valve prolapse (a) left atrium is dilated and the posterior leaflet is displaced into the left atrium (b) color flow shows retrograde jet pointing anteriorly.



Figure 2. Parasternal short axis (PSAX) view (a) note the posterior P2 segment is prolapsed at the onset of systolic phase (b) color flow demonstrate regurgitation at the start of the systolic phase

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Figure 3. Apical four chamber view showing prolapse of P2 segment of the posterior leaflet of the mitral valve. Note mild dilation of the left atrium



Figure 4. Continuous wave spectrum showing significant regurgitation during the systolic cycle

DISCUSSION AND CONCLUSION

The mitral valve is composed of two leaflets, a longer anterior with narrower base and a shorter posterior with broader attachment. Anatomically, the two leaflets may each be divided into three sections, lateral, middle and medial. There are two major classifications used to designate components of each leaflets, the Carpentier classification¹⁰ and the Duran classification. Carpentier nomenclature is based on leaflet indentation, while in the Duran classification scallops are grouped based on chordal attachments. The modified Carpentier classification, which is a combination of Carpentier and Duran nomenclature, is anatomically more precise than the classic Carpentier scheme, but is less widely used

Carpentier's¹⁰ widely recognized nomenclature describes three posterior leaflets scallops, the lateral (P1), middle (P2) and medial (P3) and the three anterior segments – lateral (A1), middle (A2) and



Figure 5. Apical view after mitral valve replacement. Note there is no regurgitation jet.

medial (A3)¹⁰. Most cases of prolapse involve the posterior middle (P2) scallop, which is easily identified on long axis TTE images. The posterior lateral scallop P1 is not well seen on long axis but best visualized on the apical four views. In this study the prolapse of the P2 segment is well seen in the apical four-chamber and long axis view. Since the leaflets are not thickened the prolapse is considered 'non-classic'. Prolapse with thickening greater than 5 mm is termed as 'classic'. Thus TTE can confirm the diagnosis of MVP. However, 2D transesophageal echocardiography (TEE) and 3D TEE are more effective in identifying prolapsing MV segments because of imaging several planes and can simulate the surgeon's view of the mitral valve.

The prevalence of non-syndromic MVP (classic or nonclassic) during the early days of 2D echocardiography was high. It ranged from 5 to 15%. This was due to erroneous assumption that the mitral valve was planar. Later the anatomy of mitral valve was revised to show

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that the mitral annulus is in fact saddle shaped and reported that MVP preferentially afflicted women and older individuals⁷. However, it was disproven by the Framingham Heart study (FHS)¹ which showed that MVP overall prevalence rate is 2.4% and is independent of age and gender. It is evenly distributed amongst middle age men and women. Thus suggesting, MVP is a progressive disease which affects predominantly middle-aged individuals. This seems true as is seen in the present case study where the subject's age is 61 years.

In conclusion, the new onset of heart murmur in a healthy 61 year old female subject with no prior known heart disease was related to the prolapsing of P2 segment of the posterior leaflet of the mitral valve. This was confirmed by transthoracic echocardiography (TTE). The case further illustrates that a perfectly healthy person can develop MVR without any prior warning. The patient underwent mitral valve replacement and TTE confirmed the new replaced valve was free from regurgitation.

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Citation: Amarjit Singh. *Mitral Valve Prolapse in a Healthy Middle Age Female Subject. Archives of Radiology.* 2019; 2(1): 6-9.

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