

Ischemic Mitral Prolapse Unveiled by Ventricular Tachycardia

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Abstract

While mitral valve prolapse (MVP) is generally regarded as a benign valvular heart condition, it can be associated with significant adverse outcomes, including sudden cardiac death (SCD) due to ventricular arrhythmias (VA). This article presents a clinical case illustrating a 65-year-old patient with MVP associated with characteristic echocardiographic signs, including mitral annular disjunction and the Pickelhaube sign. Magnetic resonance imaging (MRI) confirmed the presence of myocardial fibrosis, a potential arrhythmic substrate. The objective of this article is to emphasize the importance of echocardiographic assessment in identifying SCD risk factors in MVP patients and highlighting the complexity of this condition. Successful mitral valve replacement surgery was performed, demonstrating the effectiveness of early intervention.

Subject Areas

Cardiology, Echocardiography, Cardiac Arrhythmia, Cardiac Imaging

Keywords

Mitral Valve Prolapse, Ventricular Arrhythmias, Mitral Annular Disjunction, Pickelhaube Sign, Myocardial Fibrosis, Surgical Intervention

1. Introduction

Mitral valve prolapse (MVP), once regarded as a relatively benign valvular heart condition, has emerged as a condition potentially carrying significant and adverse outcomes, notably sudden cardiac death (SCD) resulting from ventricular arr-hythmias (VA). While initially considered benign, MVP is now recognized for its potential gravity, especially when linked to life-threatening arrhythmic events. [1] [2] [3]

Understanding MVP's complex clinical spectrum is critical in identifying those individuals at heightened risk of fatal arrhythmias, constituting a challenging task within the realm of cardiovascular medicine. This study presents a compelling case of arrhythmic MVP, shedding light on the diverse clinical expressions that this condition can manifest.

In addition to clinical manifestations, the identification of specific electrocardiographic and imaging findings associated with SCD in MVP patients is crucial for risk stratification. However, discerning the small subset of individuals at elevated risk for fatal arrhythmias remains intricate. This study delves into an intriguing case, providing insights into the complex clinical landscape of MVP, ultimately aiming to contribute to enhanced understanding and improved risk assessment in this subset of patients. [4] [5] [6]

By exploring this unique case, we underscore the importance of a comprehensive approach to MVP, emphasizing the need for meticulous evaluation, precise risk stratification, and timely intervention to prevent potentially devastating outcomes. Through this exploration, we strive to bridge the knowledge gap surrounding MVP, ultimately guiding improved patient care and outcomes.

The objectives of this research are:

- Illustrate a Clinical Case: Present a clinical case of a 65-year-old patient with MVP and suffering from ventricular tachycardia (VT), highlighting the diverse clinical manifestations of this condition.
- Emphasize the Importance of Echocardiographic Assessment: Highlight the crucial role of echocardiographic evaluation, including transthoracic and transesophageal approaches, in identifying sudden cardiac death (SCD) risk factors in MVP patients.
- Demonstrate the Complexity of the Condition: Showcase the complexity of mitral valve prolapse (MVP) and its potential to evolve into complex and potentially life-threatening clinical scenarios, challenging the previous notion of it being a benign condition.
- Confirm the Link with Myocardial Fibrosis: Confirm, through magnetic resonance imaging (MRI), the presence of myocardial fibrosis, suggesting a potential arrhythmogenic substrate.
- Demonstrate the Effectiveness of Early Intervention: Highlight the effectiveness of promptly performed mitral valve replacement surgery in the presented case.
- Stress the Importance of Prevention: Emphasize the need for future research to better understand the mechanisms of sudden cardiac death associated with MVP and to develop more effective prevention strategies.

2. Case Presentation

The patient, R. H, a 65-year-old individual, was hypertensive and diabetic. He was admitted due to ventricular tachycardia at 180 bpm, which was terminated by external electrical cardioversion (ECV). The post-crisis electrocardiogram

(ECG) showed a regular sinus rhythm with negative T waves in the inferior leads. Echocardiographic examinations, including transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE), revealed the presence of fibroelastic mitral valves with ballooning of the posterior leaflet (Figure 1), bivalvular prolapse with rupture of an accessory chord in the A2 segment, resulting in massive eccentric mitral regurgitation consisting of two distinct jets (Figure 2). Additionally, cardiac abnormalities such as non-dilated left ventricular hypertrophy with a 70% ejection fraction (EF), left atrial dilation, and suspected pulmonary hypertension were noted. Global longitudinal strain (GLS) of the left ventricle (LV) showed more pronounced impairment in its lateral segment, corresponding to the location of the papillary muscle (Figure 3). Magnetic resonance imaging (MRI) confirmed the presence of fibrosis in the LV papillary muscles (Figure 4). Coronary angiography did not reveal significant lesions. Following these investigations, an urgent mitral valve repair was indicated. Surgical exploration revealed severe anatomical lesions, including retraction of the mitral leaflets, rendering repair unfeasible, leading to valve replacement. The patient recovered well post-surgery.



Figure 1. TTE image displaying the appearance of fibroelastic mitral valve with chordal elongation (the arrow).



Figure 2. Color Doppler Echocardiography (TTE) demonstrating severe Mitral Regurgitation (MR) (the arrow).



Figure 3. Global Longitudinal Strain (GLS) of the Left Ventricle (LV) showed more pronounced impairment in its lateral segment, corresponding to the location of the papillary muscle (arrow localization).



Figure 4. Cardiac MRI image showing the location of fibrosis in the LV papillary muscles (the arrows).

3. Discussion

The diagnosis of mitral valve prolapse (MVP) can be suspected in patients presenting with a wide range of symptoms, including chest pain, exertional dyspnea, fatigue, dizziness, anxiety, palpitations, and syncope related to hypotension. Resting electrocardiography (ECG) may sometimes reveal abnormalities such as flattening or inversion of T waves in the inferior leads. However, it is essential to note that these signs are not specific to MVP and can be observed in other cardiac conditions. [2] [4] [7]

Transthoracic echocardiography (TTE) is the gold standard for evaluating patients with MVP. It allows for the detection of important signs related to the risk of ventricular arrhythmias, including mitral annular disjunction. This phenomenon is characterized by detachment of the annular roots from the ventricular myocardium, causing a displacement of 2 to 10 mm during systole. Another significant sign is the Pickelhaube sign, which manifests as a high-velocity mid-systolic spike in the tissue Doppler velocity profile of the lateral annulus of the mitral valve. This phenomenon occurs when the prolapsed posterior leaflet stretches the postero-medial papillary muscle, creating strong traction on the adjacent postero-basal left ventricular wall. [1] [2] [6]

These phenomena, particularly mitral annular disjunction and the Pickelhaube sign, have recently been associated with an increased risk of sudden cardiac death (SCD) in MVP patients. This underscores the crucial importance of echocardiographic assessment to identify these potential risk factors and guide patient management. [6] [7]

Transesophageal echocardiography (TEE) is an even more advanced imaging modality for evaluating MVP patients. It provides detailed visualization of the mitral valve and its surrounding structures, allowing for better characterization of anatomical abnormalities and disease severity [1] [2] [8]. Additional studies, including magnetic resonance imaging (MRI) and histology, have contributed to the understanding of MVP pathophysiology and its complications. They have revealed the presence of myocardial fibrosis, particularly in the papillary muscles and the inferior-basal left ventricular wall. This fibrosis is likely caused by myocardial stretching resulting from mitral leaflet prolapse. These findings suggest that MVP may contribute to the creation of an arrhythmic substrate in the heart, potentially increasing the risk of ventricular arrhythmias. [7] [8] [9]

Arrhythmic mitral valve prolapse is a rare manifestation of a condition generally considered benign. Echocardiographic signs such as mitral annular disjunction and the Pickelhaube sign are crucial elements in assessing the risk of ventricular arrhythmias and SCD in MVP patients. Understanding the underlying mechanisms of this condition remains an essential area of research to improve patient management and prevent severe complications. [4] [6] [7]

4. Conclusions

The study of this intriguing clinical case sheds light on the often underestimated aspect of mitral valve prolapse (MVP), a condition traditionally considered benign. The careful observation of this 65-year-old patient reveals a compelling association between MVP, its distinctive echocardiographic features, and potentially fatal complications, including ventricular arrhythmias and sudden cardiac death.

The echocardiographic data unveiled significant anomalies, notably mitral annular disjunction and the Pickelhaube sign, underscoring the crucial role of imaging in assessing MVP patients. Furthermore, MRI confirmed the presence of myocardial fibrosis, suggesting a potential arrhythmogenic substrate.

The clinical implications of this case go beyond the individual patient, emphasizing the urgent need for comprehensive evaluation of patients with MVP to detect potential signs of sudden death risk. Ventricular arrhythmias, particularly ventricular tachycardia, are formidable complications that require heightened vigilance and regular monitoring to prevent fatal cardiac events. This clinical case reminds us that mitral valve prolapse, despite its apparent benignity, can emerge as a major player in complex clinical scenarios. It highlights the imperative of thorough and regular assessment of MVP patients, emphasizing the importance of imaging, such as echocardiography, to identify markers of sudden death risk. The multidisciplinary approach, combining clinical assessments, imaging data, and in-depth research, is crucial to unravel the complexity of this condition and guide adequate preventive management to enhance the quality of life and save lives.

Conflicts of Interest

The authors declare no conflicts of interest.

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