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CASE REPORT

INTERMEDIATE

CLINICAL CASE

Cardiac Arrest in a Patient With Arrhythmic Mitral Valve Prolapse Syndrome

Multiple Possible Etiologies

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ABSTRACT

Arrhythmic mitral valve prolapse syndrome is associated with a high risk of death. A 60-year-old man with arrhythmic mitral valve prolapse syndrome was monitored with an implantable loop recorder. Nine months later dyspnea developed, followed by cardiac arrest. Echocardiography showed mitral valve chordal rupture. He underwent successful surgical mitral valve repair. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2021;3:1769–1773) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 60-year-old man with mitral valve prolapse, mitral annulus disjunction (**Figure 1A** [red arrowheads], Video 1), palpitations, and frequent premature ventricular contractions (PVCs) on Holter monitoring was included in a prospective study with continuous heart rhythm monitoring by implantable loop recorder (ILR). Nine months after ILR insertion, he presented to the emergency department for chest discomfort and progressive shortness of breath over the last 2 days. On admission, he was orthopneic, hypoxemic, apyrexial, and hemodynamically stable. Physical examination revealed a regular pulse, diminished breath sounds on the right side, and bilateral coarse crackles on pulmonary auscultation. Cardiac auscultation revealed a precordial systolic murmur. On admission, the electrocardiogram (ECG) showed sinus tachycardia, right bundle branch block, and frequent PVCs with right bundle branch block and a superior axis configuration (**Figure 2**). He had no arrhythmic symptoms at the time of admission. Shortly after admission, he had a sudden cardiac arrest.

LEARNING OBJECTIVES

- To evaluate the multiple factors that can trigger cardiac arrest in the setting of arrhythmic mitral valve prolapse syndrome.
- To consider the possible role of chordal rupture by continuous chordal stretching in arrhythmic mitral valve prolapse syndrome.
- To evaluate the remaining risk of ventricular arrhythmias in patients operated for mitral valve prolapse.

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ABBREVIATIONS AND ACRONYMS

CMR = cardiac magnetic resonance

ECG = electrocardiogram

ILR = implantable loop recorder

PVC = premature ventricular contraction

PAST MEDICAL HISTORY

The patient was followed regularly because of his mitral valve prolapse with moderate mitral regurgitation. He had palpitations and a high PVC burden of 9% on 24-hour Holter monitoring. He had no other comorbidities and did not use any medication. He was recruited into a prospective study investigating the incidence of ventricular arrhythmias in arrhythmic mitral valve prolapse syndrome and underwent insertion of an ILR (Medtronic Reveal LINQ11). ILR interrogation revealed episodes of nonsustained ventricular tachycardia, thus supporting the diagnosis of arrhythmic mitral valve prolapse syndrome (1). Study protocol cardiac magnetic resonance (CMR) showed mildly impaired left ventricular systolic function (ejection fraction, 50%) and posterior mitral valve prolapse and mitral annulus disjunction of a maximal 14-mm distance (**Figure 1A** [red arrowheads], Video 1). Contrast-enhanced CMR showed inferolateral left ventricular wall fibrosis by late gadolinium enhancement and no papillary muscle involvement (**Figures 3A and 3B**).

DIFFERENTIAL DIAGNOSIS

The patient had no other known cardiac disease susceptible to ventricular fibrillation. There were no symptoms, clinical signs, or ECG changes suggestive of ischemic cardiac disease. The QT interval was normal. There was no suspicion of illicit drug abuse.

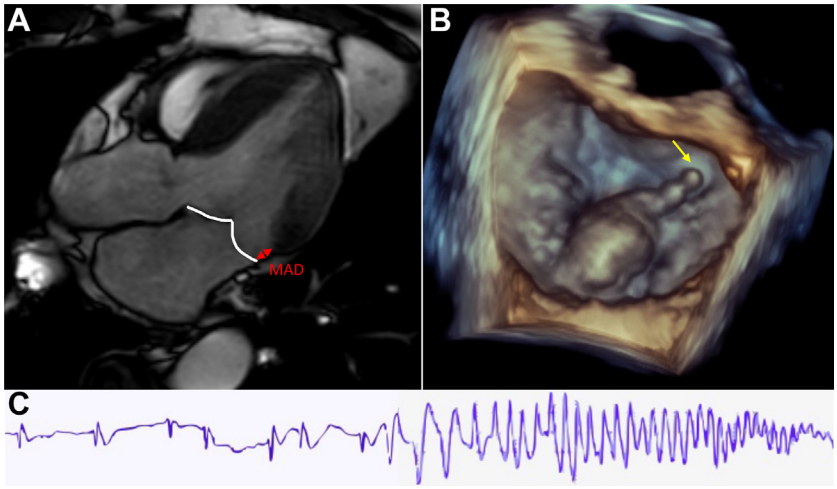
INVESTIGATIONS

After the cardiac arrest, ILR interrogation confirmed polymorphic ventricular tachycardia followed by ventricular fibrillation (**Figure 1C**). Blood sampling showed inflammatory markers, a positive D-dimer result, slightly elevated troponin T, high N-terminal pro-B-type natriuretic peptide, and slight hypokalemia. Chest radiography and computed tomography showed bilateral signs of pulmonary edema and bilateral pleural effusion. Pulmonary embolism was excluded. Nasopharyngeal secretion and blood samples were negative for infection. Echocardiography demonstrated severe mitral regurgitation secondary to ruptured chordae tendineae and mitral middle posterior scallop prolapse with flail (**Figure 1B** [yellow arrow], Video 2) 9 months after the diagnosis of moderate mitral regurgitation. Coronary angiography showed no significant stenosis of the left anterior coronary artery.

MANAGEMENT

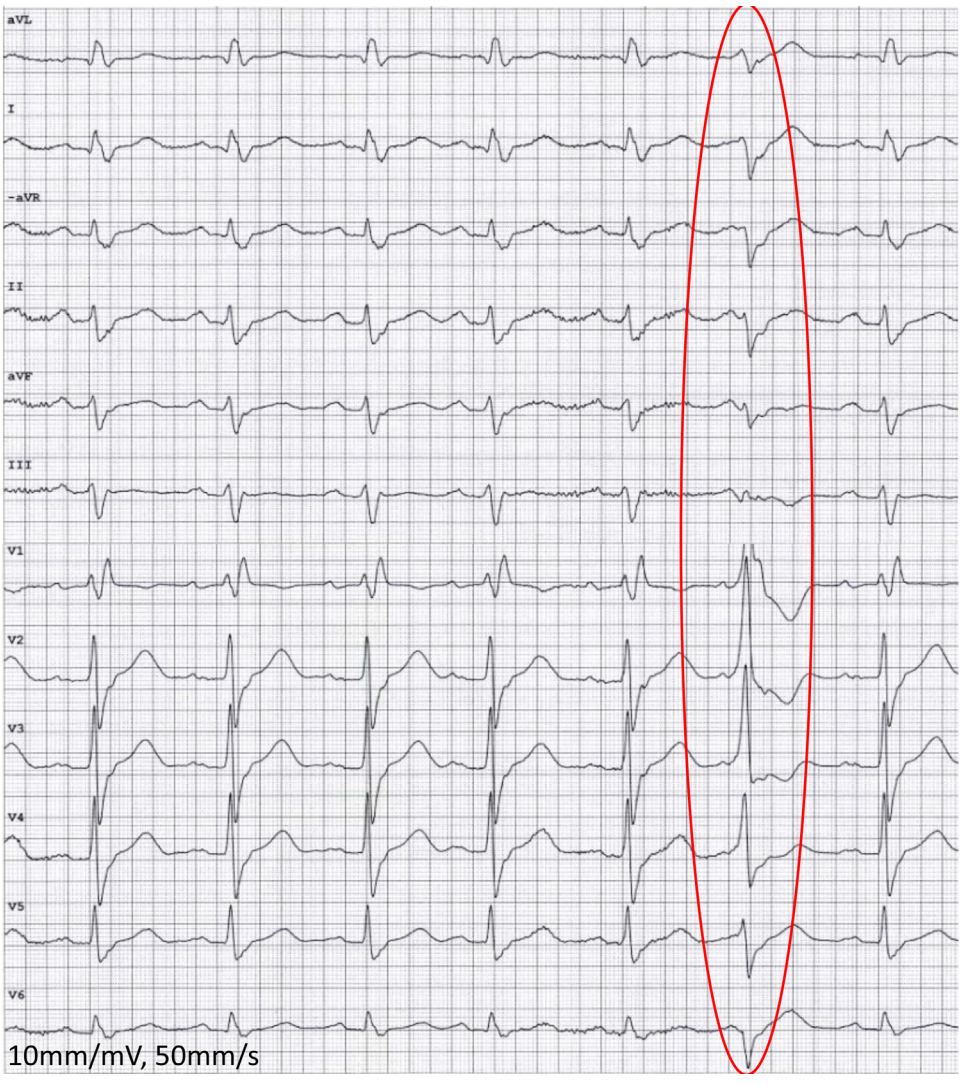
The patient needed advanced critical care after the cardiac arrest, including circulatory support. He was treated with mitral valve repair and additional tricuspid annuloplasty indicated by preoperatively diagnosed secondary tricuspid regurgitation with a

FIGURE 1 Schematic Summary of the Clinical Case



(A) Cardiac magnetic resonance 3-chamber view at end-systole showing posterior mitral valve prolapse and disjunction of the mitral annulus **(red arrowheads). (B)** A 3-dimensional transesophageal echocardiographic surgeon perspective of the mitral valve showing a prolapsed middle scallop of the posterior mitral valve leaflet and chordal rupture with flail **(yellow arrow). (C)** Polymorphic ventricular tachycardia degenerating into ventricular fibrillation registered by the implantable loop recorder. MAD = mitral annulus disjunction.

FIGURE 2 Electrocardiogram



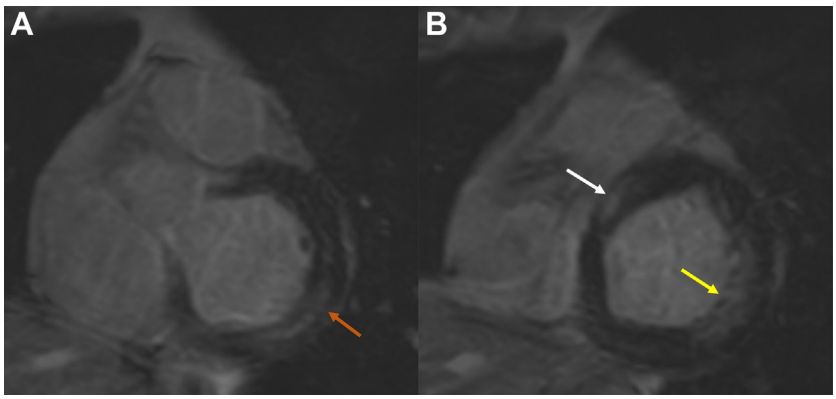
Electrocardiogram on admission showing sinus tachycardia (100/min), right bundle brunch block, and a premature ventricular contraction of left ventricular origin **(red oval frame).**

dilated tricuspid annulus. Postoperative echocardiography showed satisfactory valvular repair with moderately impaired left ventricular function. He underwent implantation of cardiac resynchronization therapy defibrillator as a result of a total atrioventricular block and an expected high percentage of ventricular pacing.

DISCUSSION

Arrhythmic mitral valve prolapse syndrome is a recently defined condition including frequent ventricular arrhythmias in the setting of mitral valve prolapse and/or mitral annulus disjunction. Ventricular arrhythmias range from frequent PVCs and/or nonsustained ventricular tachycardias to ventricular fibrillation and cardiac arrest (1). The arrhythmic mechanisms are presumed to include increased systolic stretching of the papillary muscles, inducing ventricular ectopy by depolarization-induced triggered activity and myocardial fibrosis forming the substrate for micro-re-entry arrhythmias (2). Our patient had focal myocardial fibrosis on contrast-enhanced CMR imaging, moderate mitral

FIGURE 3 Cardiac Magnetic Resonance Late Gadolinium Enhancement Sequences



(A and B) Arrows show myocardial fibrosis with midwall **(orange arrow, A)** and patchy subendocardial **(yellow arrow, B)** distribution located at the basal lateral left ventricular wall and septal fibrosis with midwall distribution **(white arrow, B).**

regurgitation, and a high PVC burden on Holter monitoring; all are recognized markers of severe arrhythmias in patients with arrhythmic mitral valve prolapse syndrome (2,3). Our patient had no previous syncope or documented sustained ventricular arrhythmias. He was considered at high risk for ventricular arrhythmias but was not fulfilling current guidelines for primary prevention implantable cardioverter-defibrillator implantation. Therefore, he was included in our prospective study of ILR monitoring of patients with arrhythmic mitral valve prolapse syndrome.

However, our patient had a cardiac arrest caused by a mechanism different from the mechanisms involved in sudden cardiac death in arrhythmic mitral valve prolapse syndrome. One may, however, speculate that the chordal rupture induced the ventricular arrhythmia through activation of the underlying arrhythmogenic substrates associated with this syndrome. Our patient represents a typical case of mitral valve prolapse secondary to fibroelastic deficiency with an increased risk of chordal rupture. We may speculate that mitral valve prolapse of a single scallop and stretching of tissue by the mitral annulus disjunction progressively leads to chordal thinning, followed eventually by chordal rupture (4). Whether myocardial and papillary muscle fibrosis further contributes to the chordal rupture is not known. Therefore, in the era of increased attention paid to arrhythmic mitral valve prolapse syndrome, it is important not to forget the risk of chordal rupture as a possible complication of mitral valve prolapse and mitral annulus disjunction. Chordal rupture leads to sudden aggravation of mitral regurgitation, pulmonary edema, and ultimately life-threatening arrhythmia. Neurohormonal activation and sympathetic nervous system activation have proarrhythmic effects in pulmonary edema (5), thereby potentially precipitating the arrhythmic event in our patient.

Ventricular arrhythmias diminish after mitral valve surgery according to a previous report (6), and lower mortality from ventricular arrhythmias was reported in patients with mitral valve prolapse after mitral valve surgery (3). However, more studies are needed to assess the risk of arrhythmias after valvular surgery both in patients with arrhythmic mitral valve prolapse syndrome and in patients with mitral valve prolapse secondary to fibroelastic deficiency.

FOLLOW-UP

Echocardiographic reassessment 3 months postoperatively showed improved left ventricular function. Cardiac resynchronization therapy defibrillator interrogation demonstrated 90% biventricular pacing and PVCs.

CONCLUSIONS

Life-threatening arrhythmias are presumably the result of traction on the papillary muscles in patients with arrhythmic mitral valve prolapse syndrome. However, mitral valve prolapse and mitral annulus disjunction also predispose to chordal rupture with secondary acute mitral regurgitation, as a precipitating factor for cardiac arrest.

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KEY WORDS chordae, mitral valve, ventricular fibrillation

APPENDIX For supplemental videos, please see the online version of this paper.