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Effects of mitral valve repair on ventricular arrhythmia in patients with mitral valve prolapse syndrome: A report of two cases

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The incidence of sudden cardiac death due to ventricular tachyarrhythmias is two-fold in patients with mitral valve prolapse (MVP) compared to the general population, with the risk enhanced by mitral regurgitation (MR) [1]. According to current guidelines, severe MR is considered an indication for surgical treatment as class I level of recommendation when the patient is symptomatic and has atrial fibrillation, left ventricular (LV) enlargement, decreased LV ejection fraction, or a high pulmonary arterial pressure. Nevertheless, current guideline does not list the occurrence of ventricular tachyarrhythmias alone as an indication for surgical management of MR [2]. Herein, we report 2 cases of MVP with associated refractory ventricular tachyarrhythmias which disappeared after successful MV repair.

The first patient was a 35-year-old Caucasian woman with a history of hypertension who complained of syncope starting with chest pain, palpitation, and dyspnea. On admission, her blood pressure was 130/70 mm Hg with an irregular pulse rate (80 bpm). At physical examination, a grade 2/6 systolic murmur and mid-systolic click were detected. Electrocardiography (ECG) showed sinus rhythm and frequent premature ventricular contractions (PVCs) associated with a right bundle branch block, inferior axis morphology and wide QRS complexes (Fig. 1). In addition, cardiac monitoring revealed short runs of nonsustained ventricular tachycardia (max of 280 bpm; Fig. 2), and ECG Holter monitoring revealed nonsustained ventricular tachycardia. Cardiac enzymes were within normal limits. Despite the fact that she had a normal coronary angiography 3 years before current hospitalization, she underwent computed tomographic angiography of coronary arteries, which showed normal coronary arteries. Echocardiographic examination revealed severe MR because of bileaflet MVP with a LV ejection fraction of 55%. The patient refused radiofrequency ablation of PVCs. Consequently, she was discharged with sinus rhythm on regimens of losartan, metoprolol, and amiodarone. The patient was subsequently readmitted because of frequent episodes of palpitation and syncope. Since the patient had severe MR along with a reduced LV function, she underwent MV repair performed by implementing a P2 triangular resection and sliding plasty of the 2 other posterior scallops. In addition, an AnnuloFlex® annuloplasty ring #34 (Sorin-Carbomedics, Austin, TX, USA) was used to stabilize the mitral ring. Surgery was uneventful and she was discharged on metoprolol (47.5 mg, daily). Eighteen months after surgery, Holter monitoring showed sinus rhythm without ventricular tachyarrhythmia. Echocardiographic examination revealed a LV ejection fraction of 50% and mild MR.

The second patient was a 67-year-old Caucasian woman with a history of moderate-to-severe degenerative MR who was admitted with a history of frequent palpitations, an episode of aborted sudden cardiac death, and near-syncope episodes while on metoprolol 200 mg per day. The baseline ECG showed frequent PVCs with a right bundle branch block and superior axis and a QRS width > 160 ms. Previous ECG recording showed some episodes of nonsustained ventricular tachycardia (the longest was 300 bpm). Echocardiography revealed a flail posterior MV leaflet and severe MR with an LV ejection fraction of 60%. The diameters of left atrium and end-systolic left ventricle were 37 and 30 mm, respectively. Her angiography showed normal coronary arteries. Accordingly, she was scheduled to undergo MV repair. The MV was repaired with posterior leaflet triangular resection and implantation of a 32-mm Memo 3D annuloplasty ring (Sorin Biomedica Cardio S. R. L., Saluggia, Italy). At discharge, ECG showed sinus rhythm with no ventricular ectopy. Three and half years after surgery, the patient is treated with atorvastatin (20 mg, once daily) and metoprolol (50 mg, twice daily). She is asymptomatic with normal sinus rhythm at ECG and no

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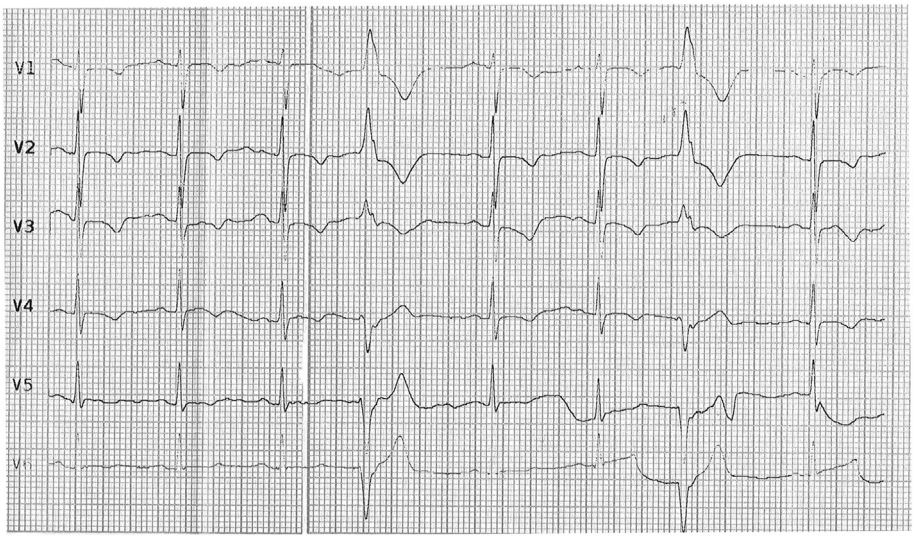

Fig. 1. Electrocardiogram shows a wide QRS complex and an R morphology in lead V_1.

ventricular tachyarrhythmias during Holter monitoring. Echocardiographic examination showed no significant MR and LV ejection fraction was 60%.

The MVP syndrome is a clinical entity with such various symptoms as atypical chest pain, palpitations, syncope, and anxiety. The incidence of life-threatening ventricular arrhythmias leading to death has been found to occur occasionally [1]. In a series of MVP patients surviving cardiac arrest, the main predictors of life-threatening ventricular arrhythmias included bileaflet MVP, female sex, biphasic or inverted T waves in the inferior leads, and PVCs of the outflow tract alternating with a papillary muscle or fascicular origin [3]. However, the association between MVP and ventricular arrhythmias remains a matter of debate, with some etiologies having been postulated such as traction on the papillary muscles, endocardial friction lesions, coronary microemboli from platelet-fibrin aggregates on the MV, transient ischemia due to mechanical changes in the coronary blood flow, improper autonomic tone [3], and fibrosis of the papillary muscles and inferobasal left ventricular wall [4]. Some echocardiographic parameters such as LV dilatation and dysfunction, MV anterior leaflet length [5], isovolumetric relaxation time, and moderate-to-severe MR [6] have also been found to be the predictors of ventricular arrhythmias. Cardiac magnetic resonance imaging and multislice computed tomography can also be used to identify the focus of arrhythmias in the papillary muscles or myocardial tissues [7].

ECG studies have been demonstrated to be valuable for detecting the origin of ventricular arrhythmias in the setting of MVP. There are some ECG features that help us distinguish the origin of ventricular arrhythmias. In case of MVP, ventricular arrhythmias originating from the papillary muscles should be differentiated from those originating from the mitral annulus adjacent to the papillary muscles or LV fascicles, since some arrhythmias are idiopathic and others may result from structural heart disease [8]. In addition, the catheter ablation of ventricular arrhythmias originating from the papillary muscles can be challenging and carries a lower success rate due to the complex structure of the papillary muscles [9]. Arrhythmias of the papillary muscles have an R or qR morphology in lead V_1, QRS complex of >160 ms, and R/S ratio ≤ 1 in lead V_6 in the LV anterolateral region [10], while fascicular arrhythmias exhibit a right bundle branch block pattern and higher prevalence of an $r < R'$ pattern in V_1 [8]. In addition, ventricular arrhythmias originating from the mitral annulus have longer QRS durations than fascicular ones; and among ventricular arrhythmias with a superior axis, the presence of $R \geq S$ in lead V_5 is associated with 100% sensitivity and 100% specificity for differentiating the mitral annulus from the papillary muscle and fascicular origins [8]. Both of our patients had a QRS complex of >160 ms and an R morphology in lead V_1, which indicated PVCs with an origin at the papillary muscles.

Based on the latest guidelines, only symptomatic patients or patients with LV enlargement or dysfunction, atrial fibrillation, or pulmonary hypertension in the presence of severe MR should undergo MV repair. Furthermore, prophylactic MV repair has been considered without the above-mentioned criteria, provided that the valve is repairable in advanced repair centers with experienced teams [1]. On the other hand, ventricular arrhythmias have yet to be considered an indication for MV repair in MVP cases [2]. In addition to these notions, Enriquez-Sarano et al. [11] reported that early MV repair should be taken into account in MR cases during the evaluation of patients based on the following criteria: 1) age; 2) comorbidities; 3) concomitant non-MV diseases; 4) valve reparability; 5) MR severity; 6) severity of MR consequences, including heart failure, reduced LV ejection fraction, atrial fibrillation, elevated pulmonary pressure, hormonal activation with B-type natriuretic peptide elevation, and reduced functional capacity; and 7) associated coronary disease [11].

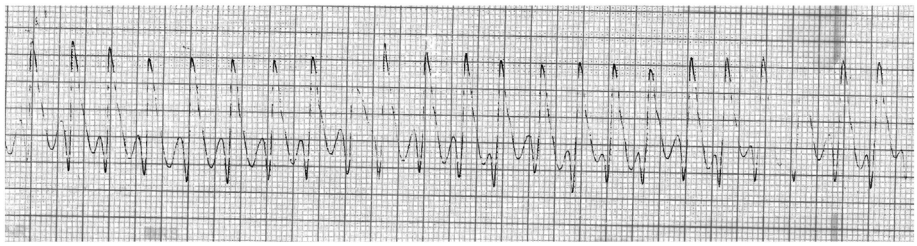

Fig. 2. Electrocardiogram shows runs of nonsustained ventricular tachycardia.

The only case report published thus far demonstrated that in a patient with the MVP syndrome and concomitant refractory ventricular tachyarrhythmias, early MV repair lessened the incidence of ventricular arrhythmias [12]. These 3 cases, including the present cases, have shown promising outcomes in a setting of a very little-known-about phenomenon. Nonetheless, MVP patients with ventricular tachyarrhythmias require due attention with a particular focus on 2 main aspects: 1) proper imaging modalities and electrophysiologic studies, and 2) long-term outcomes of early MV repair compared with those of medical management in asymptomatic MVP patients presenting with ventricular tachyarrhythmias.

Conflict of interest

The authors report no relationships that could be construed as a conflict of interest.

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