Malignant Arrhythmia in a 40-Year-Old Man

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Case Report

A 40-year-old white man with a chief concern of a heart valve problem presented to the emergency department with onset of palpitations and diaphoresis, preceded by several days of flulike symptoms. He was afebrile and alert, with a blood pressure of 98/62 mm Hg. The electrocardiogram showed a wide-complex tachycardia at a rate of 218/min with right bundle-branch block (RBBB) morphologic features and northwest frontal axis. The results were positive for Brugada and Josephson signs. There was evidence of fusion beats with intermediate QRS morphologic features and width (1st and 17th beats along lead II).

The emergency services staff promptly administered an intravenous bolus of amiodarone (Cordarone), 300 mg, but the patient soon became hemodynamically unstable. Defibrillation was initiated but aborted because of spontaneous conversion to a narrow complex rhythm. A subsequent electrocardiogram showed atrial fibrillation with inferior biphasic T-wave abnormalities. A physical examination revealed a soft S1 sound and pansystolic murmur at the apex radiating to the left axilla. There was no midsystolic click. Echocardiography revealed anterior mitral valve prolapse (MVP) with severe posteriorly directed mitral regurgitation (MR) and dilated left atrium. The left ventricle (LV) was mildly dilated with an ejection fraction of 53%. Subsequent telemetry monitoring revealed recurrent ventricular ectopic beats of similar RBBB morphologic features. An amiodarone infusion was started on the patient.

Discussion

The patient’s diagnosis was malignant arrhythmic MVP syndrome. Magnetic resonance imaging revealed late gadolinium enhancement in the posterior papillary muscles and midinferior LV wall, suggestive of MVP-induced scarring.

The clinical challenge was formulation of a treatment strategy to control symptoms, increase chance of survival, improve function, and prevent arrhythmia recurrence. The evidence of LV systolic dysfunction (ejection fraction <60%) in the context of severe primary MR is a class I indication (level of evidence B) for a mitral-valve operation in the American Heart Association / American College of Cardiology guidelines for management of valvular heart disease, and atrial fibrillation is a class IIa indication (level of evidence B). Intraoperative arrhythmia ablation can be performed conjunctively. Although percutaneous catheter ablation of ventricular tachycardia (VT) and/or atrial fibrillation is a reasonable option, this does not address the patient's need for mitral-valve operations. Radiofrequency ablation of clinically dominant ventricular ectopic foci was effective in alleviating symptoms and reducing appropriate implanted cardioverter/defibrillator (ICD) shocks in a small group of patients with arrhythmic MVP, but randomized trial data on hard clinical endpoints has yet to emerge. Our patient with structural heart disease and spontaneous sustained VT, regardless of hemodynamic stability, should receive an ICD to prevent death from a sudden cardiac event; the implantation can be performed after a mitral-valve surgical procedure.

The plan was surgical mitral valve repair with intraoperative arrhythmia ablation followed by device therapy. Electrophysiologic study performed with voltage mapping revealed scars and double potentials from the tip to the base of the posterior papillary muscles, extending to 2 cm toward the septum. There was inducible VT with the RBBB morphologic features as described above. Coronary angiography revealed that the patient’s coronary arteries were normal. The patient underwent open minithoracotomy and surgical mitral valve repair. Intraoperative examination revealed scarring over the posterior papillary muscles, around which basal circumferential cryoablation was performed. Atrial fibrillation cryoablation and left atrial appendage plication were completed conjunctively. Three weeks after mitral valve repair, a subcutaneous ICD was implanted. At outpatient visits up to 6 months, the patient remained in sinus rhythm and free of symptoms.

Recent studies have found that MVP, traditionally considered a benign condition, has arrhythmogenic potential. Autopsy and clinical studies report that MVP is an important underdiagnosed cause of arrhythmic sudden death and idiopathic out-of-hospital cardiac arrest. Ventricular arrhythmic risk is independent of LV function or severity of MR.

Malignant MVP syndrome is typically encountered in young women with bileaflet MVP and a midsystolic click, which were absent in this case; however, the combination of MVP, a polymorphic or RBBB-type VT of LV origin with prevailing baseline inferior repolarization abnormalities, and frequent outflow-tract ventricular ectopic activity should prompt the physician to consider this unifying diagnosis.

Malignant MVP syndrome is effected by multifactorial mechanisms of electrical instability. Previous studies report that most patients with arrhythmic MVP have late gadolinium enhancement in the papillary muscles and inferobasal LV wall, suggesting scarring-related reentrant electrical instability. These findings suggest a myocardial stretching insult attributable to abnormal traction, increased systolic velocity, and excursion of papillary muscles. Furthermore, mechanical stretching also induces altered membrane depolarization indicative of ventricular arrhythmias. Thus, there is a theoretical electrophysiologic basis for MVP intervention for arrhythmia control, although results from surgical intervention have been variable.