Case of Mitral Valve Prolapse – Associated Sudden Cardiac Death in Pregnancy

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Patient: Female, 34-year-old
Final Diagnosis: Ventricular arrhythmia
Symptoms: Sudden cardiac death
Medication: —
Clinical Procedure: Echocardiography • electrocardiogram • magnetic resonance imaging
Specialty: Cardiology

Objective: Unusual clinical course
Background: Mitral valve prolapse (MVP) is a frequent echocardiographic finding that can be accompanied by symptoms ranging from a benign course to occasionally catastrophic complications, such as heart failure, and rarely, sudden cardiac death. Female sex, younger age, physiological or psychological stress, electrical instability, and changes in the structure of the mitral apparatus all seem to be risk factors for fatal ventricular arrhythmias in patients with MVP. We report a case of MVP-related cardiac arrest in a pregnant woman, which is rarely reported.

Case Report: A 34-year-old woman who had collapsed at home from cardiac arrest was transported to the hospital. She had no history of cardiac diseases and was 8 weeks pregnant. Premature ventricular complexes and sinus tachycardia were observed on the 12-lead electrocardiogram as she arrived at the Emergency Department. The second cardiac arrest she experienced while in the hospital was observed to be from torsades de pointes. Further investigations revealed severe mitral valve regurgitation due to posterior leaflet prolapse and regional hypokinesis of the inferior wall and interventricular septum.

Conclusions: Ventricular arrhythmia is a frequent finding of mitral valve regurgitation. However, it rarely results in serious consequences. Malignant arrhythmic mitral valve regurgitation can result in sudden cardiac death; therefore, physicians need to be aware of patients with MVP who exhibit characteristics of a potential high-risk profile in order to avoid tragic outcomes.

Keywords: Death, Sudden, Cardiac • Mitral Valve Prolapse • Pregnancy • Tachycardia, Ventricular

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Background

Mitral valve prolapse (MVP) represents 2% to 3% of echocardiographic findings in the general population [1]. MVP often has a benign course; nevertheless, complications such as arrhythmias, endocarditis, and cerebral ischemia can be extremely troubling or even fatal [2]. The risk of sudden cardiac death due to MVP is underestimated. Indeed, our knowledge of the underlying pathomechanisms is limited. As a result, several studies have been carried out to identify the high-risk profile of individuals with MVP. However, most of these studies were retrospective and therefore unable to establish a causal relationship between MVP and sudden cardiac death [1-4]. We present the case of a pregnant woman who, after experiencing an out-of-hospital cardiac arrest, was hospitalized with a substantial MVP, confirmed by echocardiography.

Case Report

A 34-year-old woman was preparing herself before work and suddenly collapsed, without any preceding suspected symptoms. She was in her eighth week of pregnancy, G2P1001, and had no notable history of cardiac diseases. Her first pregnancy was uneventful, and she was not on any medications except for multivitamins at that period. Additionally, to the extent that her family members were aware, there were no abnormal cardiac findings detected during her prenatal visits. Her husband, who was a pharmacist, administered urgent basic life support for her, and successfully transported her to a local hospital. At the time of admission, a 12-lead electrocardiogram (ECG) revealed sinus tachycardia with premature ventricular complexes (Figure 1). Arterial blood gas showed no acidosis or electrolyte disorders. Computed tomography ruled out hemorrhagic stroke and high-risk pulmonary embolism as causes of sudden cardiac death. Unfortunately, a second episode of cardiac arrest attributed to torsades de pointes was recorded on the monitor. She was sent to our hospital for additional investigations upon resuscitation.

The patient was admitted to the Intensive Cardiac Care Unit of the Interventional Cardiology Department at the University Medical Center of Ho Chi Minh City for a comprehensive examination. Upon arrival, her patient’s Glasgow coma scale was 4. There was a 4/6 mid-systolic murmur, heard loudest at the cardiac apex. The patient’s renal and thyroid functions were normal. After the cardiac arrest, cardiac troponin T and liver enzymes rose and then returned to the normal range. A 24-h Holter ECG demonstrated sinus rhythm, with short PR interval, prolonged QTc, multifocal premature ventricular complexes, bigeminy, couplets, and nonsustained ventricular tachycardia (Figure 2). On echocardiography, the chordae tendineae of the mitral valve were intact. Posterior leaflet P1 and P2 prolapse caused severe eccentric mitral valve regurgitation with vena contracta of 7.29 to 9.2 mm (Figure 3). A transthoracic echocardiogram did not reveal any mitral annular displacement.

![Figure 1. Electrocardiogram of the patient at the time she was admitted to the Emergency Department after the first cardiac arrest. The 12-lead electrocardiogram revealed sinus tachycardia with premature ventricular complexes (arrow).](image-url)
Figure 2. Holter electrocardiogram of the patient demonstrated sinus rhythm with short PR interval, prolonged QTc, multifocal premature ventricular complexes, bigeminy (star), couplets (asterisk), and non-sustained ventricular tachycardia (arrow).

Figure 3. Mitral posterior leaflet prolapse and severe mitral valve regurgitation on echocardiography.
left atrium was moderately dilated. The left ventricular systolic function was preserved and hyperkinetic. However, the mid inferior wall, mid inferoseptum, and apical septum exhibited decreased contractility. Cardiac magnetic resonance imaging confirmed severe mitral regurgitation (Figure 4). On T2-weighted sequences, there was no evidence of cardiac edema. Late gadolinium enhancement increased in the subendocardial layer of the posterior wall suggested the existence of fibrotic tissue (Figure 5). A myocarditis viral panel was negative. Due to the patient’s condition, we were legally required to terminate the pregnancy, with her husband’s approval. Following 2 cardiac arrests, further management required a more intrusive treatment. The patient was unconscious, on mechanical ventilation, and at risk for hospital-acquired pneumonia at the time. The prognosis for her neurological recovery was extremely poor. Following the termination of the pregnancy, a left heart catheterization was conducted for a diagnostic assessment of coronary artery diseases. A coronary angiogram revealed no stenosis or obstruction (Figure 6). The severity of the mitral valve regurgitation and the patient’s symptoms warranted a surgical intervention for the mitral valve [5]. Beside surgical repair, targeted catheter ablation and implantable cardioverter defibrillation (ICD) are generally indicated for arrhythmias mitral regurgitation [5]. However, owing to her unfavorable neurological prognosis, her family refused further operations and invasive procedures but gave consent only for an ICD implantation, since it was lifesaving. MVP can cause heart failure symptoms and myocardial damage. The mainstay of guideline-directed medical therapy for heart failure should include angiotensin-converting enzyme inhibitors and angiotensin receptor blockers, beta blockers, aldosterone antagonism, and sodium-glucose co-transporter 2 inhibitors [6]. Beta blockers, in particular, are effective for the treatment of ventricular arrhythmias, as well as for reverse left ventricular function in experimental mitral regurgitation [6]. The patient was given metoprolol (25 mg) and lisinopril (10 mg) daily, in addition to having an ICD implanted. She stayed in the hospital for 3 weeks, during which time hospital-acquired pneumonia developed and was resolved with a course of meropenem. In her regular monthly check-up with us in the outpatient clinic, we discovered that she had another episode of ventricular tachycardia, which was terminated successfully by her ICD 3 weeks after she was discharged from the hospital. At the latest examination, 2 years after her admission, the patient was well. Her transthoracic

**Figure 4.** Severe mitral regurgitation on cardiac magnetic resonance imaging. Mitral regurgitation jet (arrow).
Figure 5. Late gadolinium enhancement increased in the subendocardial layer of the posterior wall suggested the existence of fibrotic tissue (red arrow).

Figure 6. Angiogram shows non-obstructive coronary arteries.

echocardiogram results were almost unchanged and indicated no processing of left ventricular dysfunction.

Discussion

This young female patient was highly suspected of having malignant arrhythmic MVP. We were unable to explore the cardiovascular history of her family and the patient herself. This female patient’s mitral valve disorder might have presented but was not sufficiently addressed during prenatal visits. A thorough physical examination followed by echocardiography and ECG would qualify this patient for further assessment by a cardiologist, which would improve her prognosis.

MVP is a common valvular heart disease and constitutes the most frequent cardiac abnormality in pregnant women [7]. This patient was unlikely to have infiltrative myocardopathies, since she did not have hypertension, thyroid disorders, or myeloproliferative diseases. In addition, her echocardiographic abnormalities were primarily thought to be the results of mitral regurgitation rather than of infiltrative myocardopathies. Signs of pericardial thickening or bilateral atrial enlargement were not identified.
The course of MVP is benign, except when it involves severe mitral regurgitation that leads to heart failure; nonetheless, certain types of MVP are also associated with fatal malignant ventricular arrhythmias [7]. Essayagh et al found in their cohort analysis of solitary MVP patients that ventricular arrhythmias occurred often but rarely resulted in tragic outcomes [8]. A prospective cohort study of Italian patients under the age of 35 found that MVP was the third most prevalent cause of cardiac arrest, after arrhythmogenic right ventricular dysplasia and coronary artery disease [4]. The incidence of sudden cardiac death in individuals with MVP is estimated at 0.2% to 0.4% per year [1]. Han et al suggested that a high-risk profile included female patients with a median age of 30, probable physiological or psychological stress, frequent premature ventricular complexes or ventricular arrhythmias on Holter ECG, and bileaflet MVP and moderate mitral regurgitation on echocardiography [2]. Evidence of electrical instability and altering structure of the mitral apparatus, such as left ventricular fibrosis in the papillary muscles and inferobasal wall, mitral annulus disjunction, and systolic curling, has been proposed by studies concerning the causal relationship between MVP and sudden cardiac death [1,3]. The majority of cases with severe mitral regurgitation are managed with surgery [5]. Correction of the flail leaflet mitral regurgitation has been associated with a decreased incidence of sudden cardiac death [5]. However, after surgical mitral regurgitation repair or replacement, ICD implantation is still indicated as a class I recommendation for individuals who experience sudden cardiac death due to ventricular tachycardia [5]. More research into a risk stratification model is needed to determine who should get primary prevention measures and more in-depth diagnostic procedures to reduce cardiac events.

Conclusions

The primary causes of maternal mortality are cardiac diseases [9]. Myocardial infarction, peripartum myocardiopathy, and arrhythmias are potential consequences of the profound changes in hemodynamic and hormonal state that occur during pregnancy [10]. It is possible that our patient had MVP since her first pregnancy, and that it had been unnoticed, as she remained asymptomatic. In addition to the importance of echocardiography in diagnosis, increasing experience with MVP-complicated arrhythmias is noteworthy. There are currently no recommendations for the risk assessment or treatment of MVP-related ventricular arrhythmias. Electrophysiological and cardiac magnetic resonance investigations are limited to certain cases. Prophylactic ICD implantation and surgical repair or replacement of the mitral valve are not commonly indicated because of the lack of evidence from prospective and randomized control studies [1]. To manage ventricular arrhythmias, beta-blockers remain the criterion standard, and ICD implantation for secondary prevention is also recommended [11].

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Department and Institution Where Work Was Performed

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Declaration of Figures’ Authenticity

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