CASE REPORT

The disappearance of nonsustained ventricular tachycardia after surgical repair in a patient with mitral annular disjunction and mitral valve prolapse

Mitral anular disjunction ve mitral kapak prolapsusu olan bir hastada cerrahi tamir sonrası sürekli olmayan ventriküler taşikardinin kaybolması

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Summary– Mitral annular disjunction (MAD) is a structural abnormality defined as the separation of the ventricular myocardium between the mitral valve annulus and the left atrial wall. It is present in some patients with mitral valve prolapse (MVP) and is associated with papillary muscle fibrosis and ventricular arrhythmia. Although it is easy to diagnose, it can be overlooked in daily practice. This study presents the case of a 42-year-old patient who was admitted to the cardiology clinic with complaints of palpitation and syncope. The patient was diagnosed with bileaflet MVP, MAD, and severe mitral regurgitation using transthoracic echocardiography and cardiac magnetic resonance imaging, in which ventricular tachycardia disappeared following subsequent surgical repair.

Özet– Mitral anular disjunction (MAD), mitral kapak anulusu ile sol atriyal duvar arasında ventrikülün miyokarddan ayrılması olarak tanımlanan yapısal bir anormalliktir. MAD, mitral valv prolapsusu (MVP) olan hastaların bir kısmında bulunmakta olup MVP'den bağımsız olarak papiller kas fibrozu ve ventriküler aritmi ile ilişkilidir. Tanısı kolay olmasına rağmen günlük pratikte büyük oranda akla gelmemesi nedeniyle gözden kaçabilmektedir. Bu olguda, çarpıntı ve senkop şikayeti ile kardiyoloji kliniğine başvuran, transtorasik ekokardiyografi ve kardiyak manyetik rezonans görüntüleme ile biliflet MVP, MAD ve ileri mitral yetersizliği tespit edilen ve cerrahi tamir sonrasında ventriküler taşikardinin kaybolduğu 42 yaşındaki olguyu sunmayı amaçladık.

Mitral annular disjunction (MAD) is a structural abnormality defined as the separation of the ventricular myocardium between the mitral valve annulus and the left atrial wall. It is present in some patients with mitral valve prolapse (MVP) and is associated with papillary muscle fibrosis and ventricular arrhythmia. Although it is easy to diagnose, it can be overlooked in daily practice. This study presents the case of a 42-year-old patient who was admitted to the cardiology clinic with complaints of palpitation and syncope. The patient was diagnosed with bileaflet MVP, MAD, and severe mitral regurgitation (MR) using transthoracic echocardiography (TTE) and cardiac magnetic resonance imaging (MRI), for which she underwent subsequent surgical repair.

CASE REPORT

A 42-year-old female patient was followed up for approximately 9 years with a diagnosis of MVP at a cardiology clinic. She was admitted to our cardiology clinic with complaints of intermittent palpitation that caused presyncope-like symptoms and 2 episodes of syncope, independent of exertion. There was no family history of sudden cardiac death. Her physical examination revealed a blood pressure of 110/75 mmHg and a heart rate of 79 bpm. Physical examination also revealed 2/6 late systolic murmur in the mitral area. Biochemical laboratory tests such as electrolytes, liver, and thyroid function tests were within normal limits. Her baseline electrocardiogram (ECG) showed T negativity in the inferolateral leads (D2,

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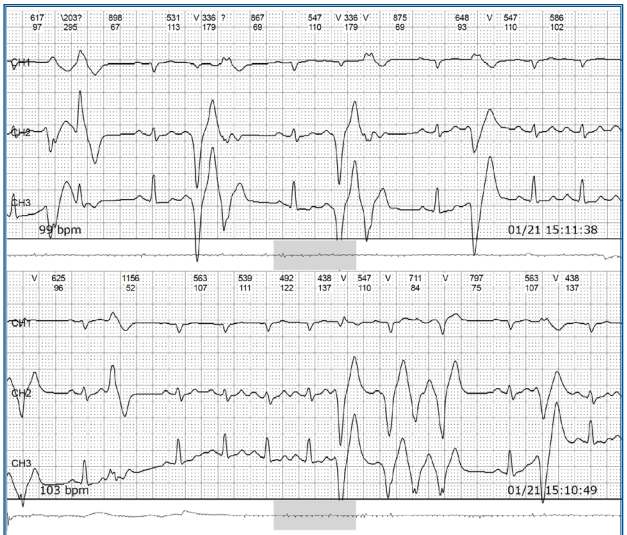

Figure 1. ECG Holter monitoring showing multiple premature ventricular contractions (PVCs) and nonsustained ventricular tachycardia (NSVT).
MVP: mitral valve prolapse.

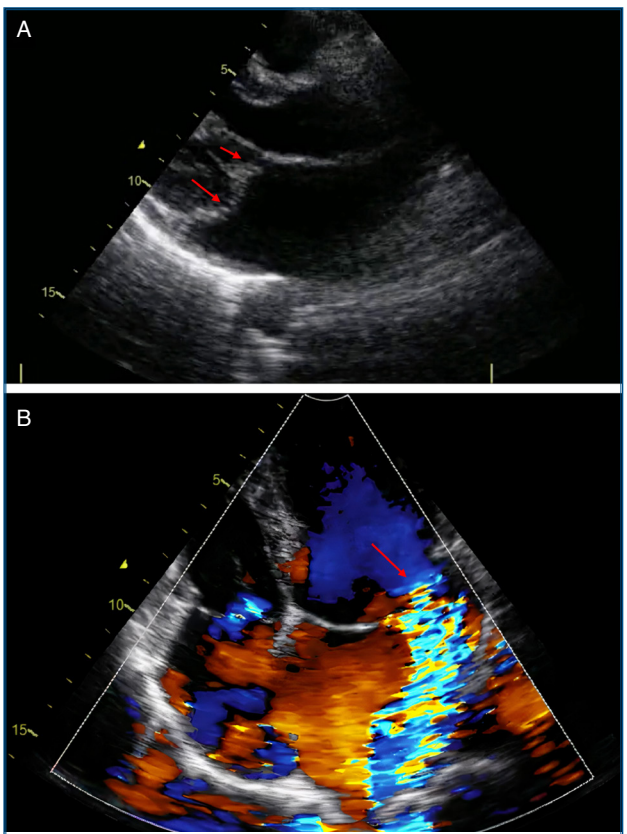

Figure 2. (A) The echocardiographic examination shows bileaflet MVP in the parasternal long-axis view (labeled); **(B)** Color Doppler image showing eccentric severe mitral regurgitation in the apical four-chamber image (labeled).

D3, AVF, V4-6). ECG Holter monitoring revealed nonsustained ventricular tachycardia. Ventricular premature beats were also observed with multifocal right bundle branch block morphology, suggesting that they originated from the left ventricle (Figure 1). Echocardiographic examination revealed enlargement of the left heart chambers (left ventricular end diastolic diameter: 59 mm; left atrial diameter: 45 mm), bileaflet MVP, and severe eccentric MR (Figure 2, Video 1-3*). With TTE, MAD distance was measured to be 10 mm in the apical four-chamber view during late systole. Additionally, left ventricular ejection fraction was 50-55%. Cardiac MRI revealed bileaflet MVP, MAD during systole, severe MR caused by prolapse of A1-P1 and A3-P3 scallops, and late gadolinium enhancement in the left ventricular inferolateral wall (Figure 3). The patient was consulted with cardiovascular surgery, and a decision was made to proceed with valve repair. Coronary arteries were found to be normal in the coronary angiography performed before surgery. Posterior leaflet quadrangular resection was performed. Two neochordae were implanted in the posterior leaflet (P2 and P3), and 1 neochordae in the scallops of the anterior leaflet (A1, A2 and A3). Mitral annuloplasty was applied with a 38-mm Medtronic 3D ring. In the postoperative TTE, mild MR was observed (Figure 4). Nonsustained ventricular tachycardia was not observed in the postoperative ECG Holter monitoring. The patient was discharged as fully recovered. In the postoperative period, she was followed up for 6 months. No syncope attack occurred in this period.

Abbreviations:

3D	*Three-dimensional*
ECG	*Electrocardiogram*
MAD	*Mitral annular disjunction*
MR	*Mitral regurgitation*
MRI	*Magnetic resonance imaging*
MVP	*Mitral valve prolapse*
TEE	*Transesophageal echocardiography*
TTE	*Transthoracic echocardiography*

DISCUSSION

Mitral valve prolapse is a mitral valve disease seen in 2-3% of the general population.[1] Prolapse can involve only the mitral valve or can be seen in more than 1 valve.[2] MVP is associated with an increased risk of arrhythmic complications, including ventricular arrhythmias and sudden cardiac death.[3] Palpitations, especially those due to ventricular arrhythmias, may result in syncope and/or sudden cardiac death.[4]

A diagnosis of MAD means that the atrium-mitral annulus junction is separated from the ventricular muscle.[5] The disjunction distance (the distance from the mitral valve-atrium junction to the ventricular

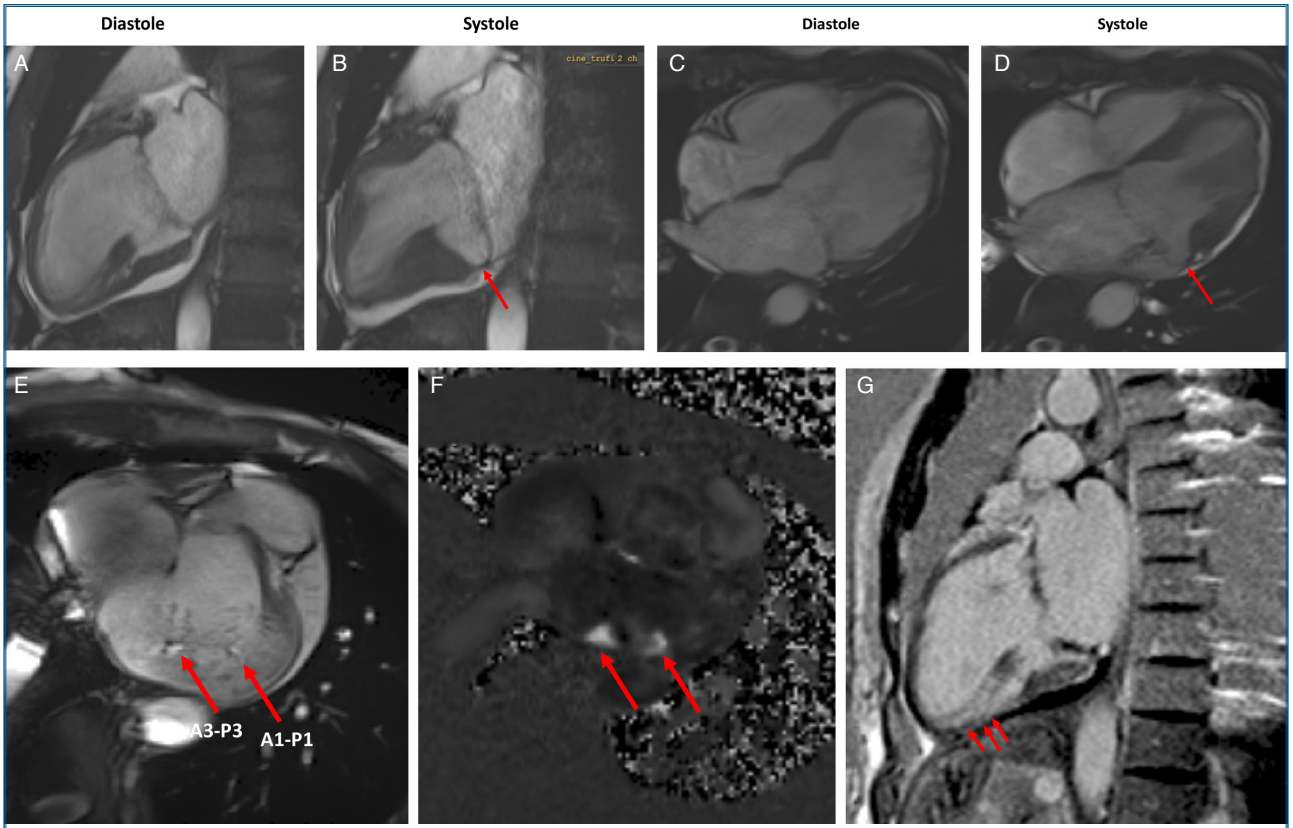

Figure 3. (A-D) Cardiac magnetic resonance imaging shows the MAD in the four-chamber image (labeled); **(E)** Cine image showing MR jets from A1-P1 and A3-P3 scallops; **(F)** Flow mapping showing MR jets from A1-P1 and A3-P3 scallops; **(G)** LGE images showing mild fibrosis at the LV inferolateral wall.
MAD: mitral annular disjunction; MR: mitral regurgitation.

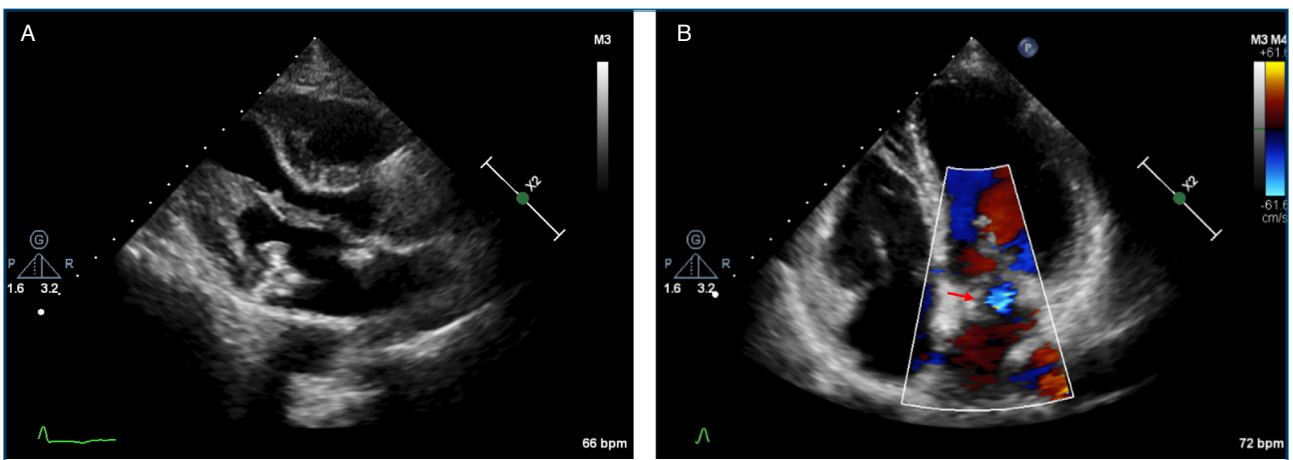

Figure 4. 2D echocardiographic view in the first month after the operation. **(A)** Parasternal long-axis view; **(B)** Apical four-chamber view showed mild mitral regurgitation (labeled).

myocardium) varies, and it can be up to 30 mm.[6] MAD has been reported in 28.3-55% of patients with MVP in echocardiographic studies.[7,8] This is an important issue because, although MAD is common, it is largely overlooked in daily practice. This problem can be explained by the fact that MAD is detected only during ventricular systole. Although it is more common in patients with MVP, it is also seen in people without MVP. In a study of 116 patients with MAD, 22% of the patients had MAD without MVP.[9]

The most common symptom in patients with MAD is palpitation. However, patients with MAD may also have presyncope/syncope and chest pain.[9] Chest pain has been reported more frequently in MVP patients with MAD than in MVP patients without MAD.[6]

Noninvasive imaging methods such as TTE, three-dimensional (3D) echocardiography, transesophageal echocardiography (TEE), and cardiac MRI can often be used for the diagnosis of MAD.[10,11] MAD is seen during the systolic phase in the parasternal long-axis view in echocardiographic evaluation (Figures 2 and 3). Evaluation of MAD with 3D echocardiography can provide a comprehensive picture of its geometry and the structural basis for this functional abnormality by demonstrating the decoupling of annular and ventricular function in relation to anatomic annular disjunction.[10] A curling movement of the basal posterolateral wall of the left ventricle may be demonstrated by TEE.[9] The longitudinal MAD distance can be measured with cardiac MRI. Unlike echocardiography, cardiac MRI can also provide information about myocardial and papillary muscle fibrosis and its localization.[12] Thus, it can contribute to risk assessment. In this case, MAD was detected with both TTE and cardiac MRI. In addition, late gadolinium enhancement was observed in the inferolateral region consistent with fibrosis.

MAD is a structural pathology characterized by left ventricular fibrosis, especially at the posteromedial papillary muscle and inferobasal wall level.[13] Although the mechanism of arrhythmia occurring in patients with MAD is not fully explained, it is thought that fibrosis develops in the papillary muscle and left ventricular inferobasal wall because of the prolapsed valve stretching the chordae, which predisposes the patient to ventricular arrhythmias.[3,14,15] The longitudinal MAD distance has been associated with premature ventricular beats and ventricular tachycardia, and ventricular tachycardia was more common in those with MAD distances greater than 8.5 mm.[6,7,9] In this case, the MAD distance was measured to be 10 mm with TTE.

According to most studies, MAD appears to be associated with ventricular arrhythmias ranging from frequent premature ventricular beats to cardiac arrest.[6,9,16] Most patients with MAD have premature ventricular contractions. In some cases with MAD, after surgical repair of the mitral valve, ventricular arrhythmia has been shown to decrease.[15] In our case, a small number of premature ventricular beats were observed after the operation. However, we do not have the data about the recurrence of arrhythmia in the long term. Because recovery from fibrosis is not expected after surgery, it is thought that alteration of MAD distance may be an effective mechanism in reducing arrhythmia.

In conclusion, MAD is a common structural abnormality in patients with myxomatous mitral valve disease and MVP. Diagnosis can be made using echocardiography and cardiac MRI. Because MAD is associated with ventricular arrhythmias and sudden cardiac death, it should be kept in mind as an important component of diagnostic approach. Future studies need to evaluate short and long term outcome of surgical intervention in patients with MVP and MAD with arrhythmias.

*Supplementary video files associated with this article can be found in the online version of the journal.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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Anahtar Kelimeler: Mitral anular disjunction; mitral valve prolapse; ventriküler taşikardi