Arrhythmogenic right ventricular cardiomyopathy mimicking right ventricular outflow tract tachycardia

Sağ ventrikül çıkış yolu taşikardisini taklit eden aritmojenik sağ ventrikül kardiyomiyopatisi

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Ventricular tachycardia may be mistaken for right ventricular outflow tract tachycardia in patients with arrhythmogenic right ventricular cardiomyopathy. A 27-year-old man had complaints of palpitations and syncope. The admission electrocardiogram (ECG) showed sustained monomorphic ventricular tachycardia with left bundle branch block and inferior axis morphology. The ECG obtained during sinus rhythm was normal. Transthoracic echocardiography showed both ventricles in normal function and size. During electrophysiologic study, ventricular tachycardia was induced consistent with the clinical tachycardia. It was thought to originate from the left ventricular outflow tract and was terminated by radiofrequency ablation. However, the patient presented again, after a year, complaining of palpitations. The admission ECG was similar to that obtained before with sustained ventricular tachycardia, whereas the ECG during sinus rhythm showed negative T waves in leads V1-3. During electrophysiologic study, another ventricular tachycardia was induced with left bundle branch block and horizontal axis morphology as well as that consistent with the clinical tachycardia. The former was terminated spontaneously. The presence of a different morphology and negative T waves on the ECG suggested arrhythmogenic right ventricular cardiomyopathy. On angiography, the right ventricle was dilated and hypocontractile. Cardiac magnetic resonance imaging confirmed the diagnosis by showing decreased wall thickness and wall motion abnormality in the right ventricle.

Key words: Arrhythmogenic right ventricular dysplasia/diagnosis; cardiac electrophysiology; cardiomyopathies; diagnosis, differential; electrocardiography; tachycardia, ventricular/etiology.

Aritmojenik sağ ventrikül kardiyomiyopatisi olan hastalarda ventrikül taşikardilerinin farklı formları bazen sağ ventrikül çıkış yolu taşikardisi ile karıştırılabilir. Çarpıntı ve bayılma yakınmaları ile başvuran 27 yaşındaki erkek hastanın elektrokardiyogramında (EKG), sol dal bloku ve inferiyor eksen morfolojisinde, uzamış monomorfik ventrikül taşikardisi izlendi. Sinüs ritminde çekilen EKG ise normal idi. Transtorasik ekokardiyografide sol ve sağ ventrikül fonksiyonları ve boyutları normal bulundu. Elektrofizyolojik çalışmada, klinik ventrikül taşikardisi ile uyumlu ve sağ ventrikül çıkış yolundan kaynaklandığı düşünülen ventrikül taşikardisi oluşturuldu ve aritmi radyofrekans ablasyon ile sonlandırıldı. Hasta bir yıl sonra çarpıntı yakınmasıyla tekrar başvurdu. Başvuru EKG'sinde bir yıl öncekine benzer uzamış ventrikül taşikardisi izlenirken, sinüs ritminde çekilen EKG'de ise, V1-3 derivasyonlarında T dalgası negatifliği izlendi. Elektrofizyolojik çalışmada, klinik ventrikül taşikardisi ile uyumlu ventrikül taşikardisi yanı sıra sol dal bloku ve yatay eksen morfolojisinde ikinci bir ventrikül taşikardisi oluştu. Bu aritmi kendiliğinden sonlandı. Farklı morfolojide ikinci bir ventrikül taşikardisi oluşması ve EKG'de T dalgası negatifliği olması üzerine hastada aritmojenik sağ ventrikül kardiyomiyopatisi olabileceği düşünüldü. Sağ ventrikül anjiyografisinde, sağ ventrikül genişlemiş ve kasılması azalmış olarak izlendi. Bu tanı, manyetik rezonans görüntülemede sağ ventrikül duvarında incelme ve duvar hareket bozukluğu görülmesi ile doğrulandı.

Anahtar sözcükler: Aritmojenik sağ ventrikül displazisi/tanı; kalp elektrofizyolojisi; kardiyomiyopati; ayırıcı elektrofizyolojisi; kardiyomiyopati; ayırıcı tanı; elektrokardiyografi; taşikardi, ventrikül/etyoloji.

Idiopathic ventricular tachycardia is observed in patients with non-structural diseases and responds to medical as a well as ablation therapy.[1] The most common type of idiopathic ventricular tachycardia is right ventricular outflow tract (RVOT) tachycardia. Ventricular tachycardia associated with arrhythmogenic right ventricular cardiomyopathy (ARVC) may sometimes mimic RVOT tachycardia. Arrhythmogenic right ventricular cardiomyopathy originates from the right ventricle, and occurs together with re-entrant

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ventricular tachycardia of left bundle branch block configuration. These arrhythmias are frequently exacerbated by the secretion of catecholamine due to exercise.[2] It is very important to distinguish between ventricular tachycardia associated with ARVC, and RVOT tachycardia. This is due to the fact that ARVC is responsible for approximately 20% of sudden death incidents among patients below the age of 35, and may require aggressive treatment modalities such as implantation of cardioverter defibrillator.[3]

CASE REPORT

Sustained monomorphic ventricular tachycardia was observed on the electrocardiogram (ECG) of a 27-year-old man who presented to our hospital with complaints of palpitations and syncope. The patient's tachycardia was of left bundle branch block and inferior axis morphology (Figure 1a). The ECG obtained during normal sinus rhythm was normal (Figure 1b). There was no history of sudden death or any known cardiac disease in the patient's family medical history. Normal left and right ventricular functions and dimensions were observed on the transthoracic echocardiography. There was no valvular heart disease or wall motion abnormality.

During electrophysiologic study, ventricular tachycardia was induced by programmed stimulation from the right ventricle, which was consistent with left bundle branch block and clinical ventricular tachycardia of inferior axis morphology. Records were obtained from the right ventricular apex, left ventricular apex and the RVOT, and it was concluded that the tachycardia was of RVOT origin. The arrhythmia was terminated by radiofrequency ablation on the right ventricular anterior free wall.

The patient presented again one year later with complaints of palpitation. A left bundle branch block and sustained ventricular tachycardia of inferior axis morphology was observed on the ECG at presentation. The ECG performed with the patient in sinus rhythm demonstrated a negative T-wave in leads VI-3, which was not observed in his previous ECG (Figure 1c). The repeat transthoracic echocardiography revealed similar findings as in the previous analysis. The electrophysiologic study by programmed stimulation demonstrated a ventricular tachycardia consistent with clinical tachycardia, as well as left bundle branch block and a second ventricular tachycardia of horizontal axis morphology (Figure 1d). This arrhythmia terminated spontaneously. Presence of another ventricular tachycardia of a different morphology and the negative T-wave observed on the ECG suggested a possible ARVC. Right ventricular angiography was thus performed. The right ventricle was observed to be dilated and hypocontractile, consistent with ARVC. This diagnosis was confirmed by decreased wall thickness and wall motion abnormality in the right ventricle, as observed on magnetic resonance imaging.

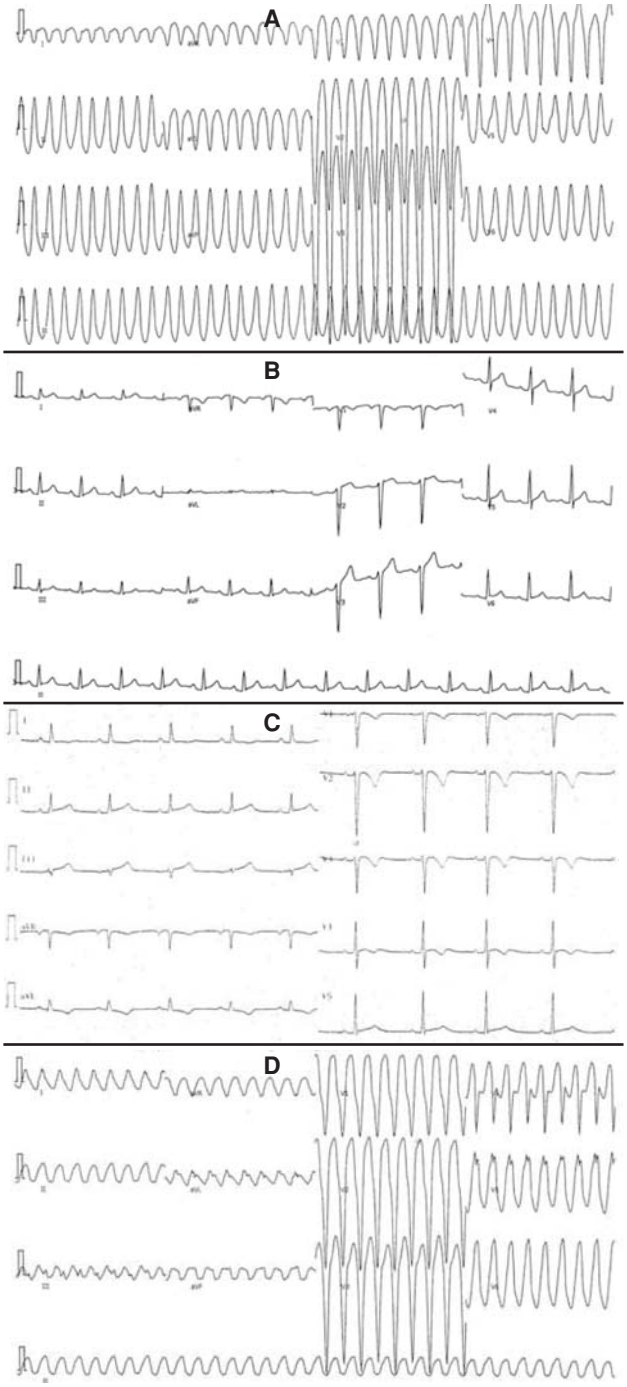

Figure 1. Electrocardiograms of the patients obtained **(A)** during palpitation, **(B)** during sinus rhythm, and **(C)** one year later during sinus rhythm. **(D)** Induced left bundle branch block and ventricular tachycardia of horizontal axis morphology during the electrophysiologic study.

DISCUSSION

Induction of ventricular tachycardia during the first electrophysiologic study should suggest a re-entrant tachycardia. This is due to the fact that RVOT tachycardias do not occur by programmed stimulation, but by triggered activity. RVOT tachycardias may be induced during electrophysiologic studies by administration of drugs such as isoproterenol. The single morphologic nature of arrhythmias which occur during electrophysiologic studies should suggest RVOT tachycardia, whereas more than one morphology suggests ventricular tachycardia associated with ARVC. Presence of late potentials during electrophysiologic studies is another indication of ARVC. Ventricular tachycardia associated with ARVC was not considered in our patient because at presentation there was no family medical history related to the condition, ventricular tachycardia induced during the first electrophysiologic study was of a single morphology, and also because the ECG and transthoracic echocardiography obtained during sinus rhythm were normal. Consequently we did not use isoproterenol during electrophysiologic study and did not explore late potentials. We performed radiofrequency ablation for ventricular tachycardia. However, we considered the diagnosis of ventricular tachycardia associated with ARVC following observation of a negative T-wave on the ECG obtain during sinus rhythm and the occurrence of ventricular tachycardias of different morphologies during the electrophysiologic study, when the patient presented again one year later with similar complaints. The right ventricular angiography and magnetic resonance imaging performed were also consistent with ARVC.

ARVC should also be considered in the differential diagnosis when different morphologies of ventricular tachycardia are observed during electrophysiologic studies. Transthoracic echocardiography findings are not always adequate in the diagnosis of ARVC. Right ventricular angiography and magnetic resonance imaging should also be performed in addition to transthoracic echocardiography in case of any suspicion of ARVC.

It is imperative to remember that ARVC should also be considered in the differential diagnosis of patients who present with RVOT tachycardia, since this would affect determination of the various treatment strategies.

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