

Different clinical presentations of arrhythmogenic right ventricular cardiomyopathy in two brothers

Aritmojenik sağ ventrikül kardiomyopatinin iki kardeşte farklı klinik görümlerle ortaya çıkışı

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Introduction

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is characterized by progressive fatty or fibrofatty infiltration of the right ventricular myocardium, which constitutes a substrate for electrical instability and a focus of ventricular arrhythmias (1). Patients usually present with ventricular tachycardias (VT) with left bundle branch morphology that may lead to sudden death, especially in healthy young adults (2). In vast majority of the patients, ARVC is a familial disease frequently transmitted as an autosomal dominant trait (3). Echocardiography, computer tomography and magnetic resonance imaging (MRI) are the most frequently used methods for the assessment of the right ventricle in the diagnosis of ARVC. The prognosis is poor in patients with progressive heart muscle disease when arrhythmias and heart failure appear (4).

We investigated a family consisted of 8 members. The father died suddenly at the age of 45, the mother, two sisters and one brother were healthy and two brothers had ARVC.

Case 1

A 25-year-old man was admitted to the emergency room with chest pain and syncope. He was unconscious and had a marked hypotension. A rapid sustained ventricular tachycardia (300 beats/min) was diagnosed (Fig. 1). The arrhythmia was immediately converted to sinus

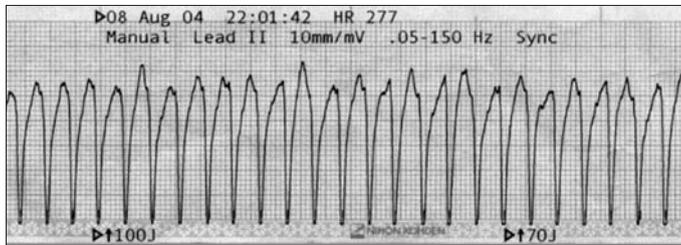


Figure 1. The monitor recording of rapid ventricular tachycardia

rhythm with electrical cardioversion. During sinus rhythm inverted T waves in V1-V4 and frequent ventricular ectopic beats were observed. Transthoracic echocardiography (TTE) revealed a dilated right atrium and ventricle with diffuse wall hypokinesia in right ventricle, but the left ventricle appeared normal (Fig. 2). Color Doppler showed a mild

tricuspid regurgitation. The diagnosis was ARVC and an implantable cardioverter/defibrillator was placed for life-threatening sustained ventricular tachycardia.

Case 2

This patient was the brother of the patient in case 1. He had right heart failure (NYHA class III), cyanosis and clubbing. The patient was 20

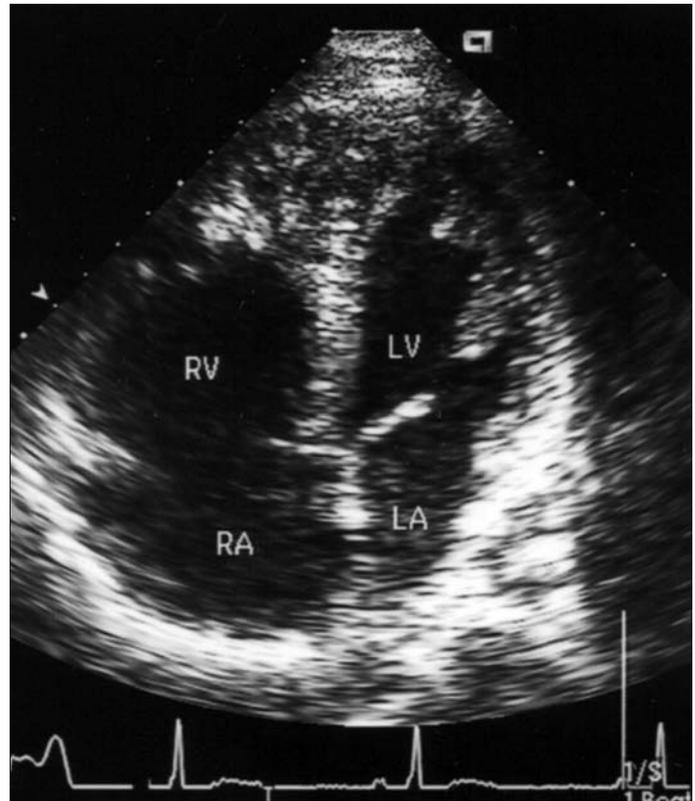


Figure 2. Apical four-chamber echocardiographic view of the heart shows dilated right atrium and ventricle, myocardial trabeculation at right ventricular apex and reduced thickness of right ventricular free wall in Case 1

years old when heart failure occurred as NYHA class II. Twelve-lead electrocardiogram (ECG) showed inverted T waves and epsilon wave in V1-V4 (Fig. 3). Frequent ventricular ectopic beats were recorded during 24-hour ECG monitoring. Chest X-ray and respiratory function tests were normal. Hemoglobin value (22 gr/dl) was significantly greater than normal (12-14 gr/dl). Arterial O₂ saturation was low (80%). The TTE findings were similar with case 1. Transesophageal echocardiographic (TEE) examinations revealed a right-to-left interatrial shunt through a patent foramen ovale (PFO). Contrast TEE also confirmed the right to left shunting. In addition to TEE, cardiac catheterization revealed right to left shunt through the interatrial septum. Magnetic resonance imaging showed that there was fatty replacement of the myocardium at the apical and free wall of the right ventricle (Fig. 4).

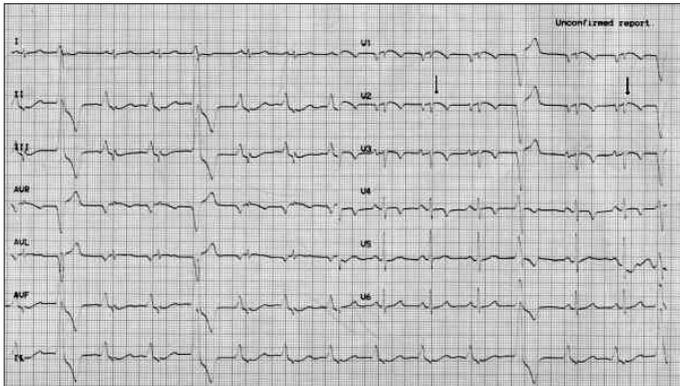


Figure 3. The 12-lead ECG shows inverted T waves (V1-4) and epsilon waves (V1-2) in case 2. Reduced QRS complex amplitude and pulmonary P waves are seen

Discussion

Arrhythmogenic right ventricular cardiomyopathy is a familial disease characterized by dilatation and regional wall hypokinesia of the right ventricle and left bundle branch block VT on ECG. The main clinical courses are ventricular arrhythmias, sudden death and especially right heart failure (4). The diagnosis of ARVC is based on the presence of familial inheritance, the structural and functional pathologies of right ventricle, arrhythmias, depolarization and repolarization abnormalities on ECG.

The ECG abnormalities are shown in 70% of ARVC patients (5). T wave inversion in the precordial leads is the most common repolarization abnormality. Complete or incomplete right bundle branch block, prolongation of right precordial QRS duration and a small, discrete wave just beyond the QRS in particularly V₁, designated as epsilon wave, are the depolarization abnormalities. Epsilon wave is detected in 5-30% of the cases and if present, it is a pathognomonic for ARVC (6). Our both cases had repolarization abnormalities and only Case 2 had epsilon wave.

Ventricular arrhythmia as in case 1 is the most classical finding in ARVC. However, severe right heart failure as in Case 2 is an unusual clinical presentation. The different clinical presentations of two brothers were the most striking clinical aspect. Cyanosis and clubbing were the remarkable physical examination finding in case 2. Marked polycythemia and slight hypoxia were also found in this case. The potential etiology of cyanosis and clubbing were investigated and cardiac right to left shunt through the PFO was detected in case 2. Cubero et al. (7) had also previously reported a similar case with ARVC. The mechanisms underlying right to left shunting in these pathologies are associated with marked elevations in right ventricle afterload and/or increased right atrial pressure. We think that increased right atrial pressure due to right ventricular failure may be the reason of right to left shunting in our patient.

Echocardiography, computer tomography and MRI are the most frequently used methods for the assessment of the right ventricle. Tissue Doppler and strain echocardiography and multidetector computer tomography have a diagnostic role of ARVC as different new imaging techniques (8). The echocardiographic evaluation is a sufficient method if the right ventricular structural and functional abnormalities are prominent. The MRI is also noninvasive method that is reliable in distinguishing the fat from the muscle. Increased signal intensity in regions of replacement of its myocardium with fat can be clearly visualized by MRI. Reduced right ventricular ejection fraction can also be shown by MRI (9). We found a significant trabeculation in right ventricular apex on TTE in both patients. The MRI also confirmed the structural alteration of the right ventricle.

Arrhythmogenic right ventricular cardiomyopathy is a progressive heart muscle disease. The patients with VT, right ventricular or biventricular pump failure have poor prognosis (4). Therapeutic options include antiarrhythmic drugs, catheter ablation and implantable cardioverter defibrillator (ICD), which is the most effective therapy against arrhythmic sudden death for ventricular arrhythmias. We considered the ICD implantation for case 1 because of life-threatening rapid VT. A diuretic, angiotensin converting enzyme inhibitors and warfarin were prescribed for case 2. His functional capacity was still 3 at the 6th month of treatment. Heart transplantation was planned for case 2 because of refractory heart failure.

Conclusion

In conclusion, VT and inverted T waves in precordial leads in sinus rhythm are the classical findings of arrhythmogenic right ventricular cardiomyopathy. However, the different clinical features of the same heart disease should be kept in mind.

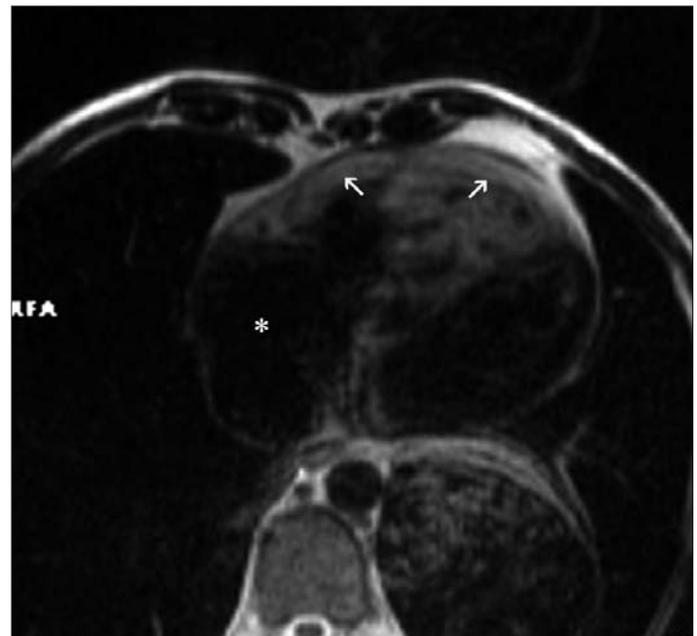


Figure 4. T2 weighted MR image of heart showed that diffuse hyperintensity of right ventricular free wall and apex due to fatty infiltration (arrows). Note markedly dilated right atrium (asterisk)

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Re-operation for the mechanical valve obstruction with a beating heart technique in an elderly patient with compromised ventricular function

Bozulmuş ventrikül fonksiyonlu yaşlı hastada çarpan kalp tekniği ile mekanik kapak obstrüksiyonu için reoperasyon

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Introduction

Conventional mitral valve replacement (MVR) consists of cardiopulmonary bypass (CPB), cross-clamping, and cardioplegia. Reperfusion injury is a phenomenon that can occur in classic technique (1). Beating heart valve surgery by perfusing the heart continuously with blood eliminates the ischemia and avoids reperfusion injury. In addition, this technique may have other advantages, as the heart is under more physiologic condition than the cardioplegic arrested state especially in patients who have compromised ventricular functions (2-4).

Case report

An 85 years old male patient admitted with symptoms of dyspnea and palpitation. He underwent MVR operation 2 years ago.

No mechanical valve sounds were detected during cardiac auscultation. Patient was in NYHA functional class III. Electrocardiography revealed atrial fibrillation and bilaterally lung edema observed on chest X-ray. Echocardiography showed 13/6 mmHg peak/mean mitral diastolic gradient, and no leaflet motion on the prosthetic valve. Also, ejection fraction (EF) was calculated as 30% and pulmonary artery pressure (PAP) was measured as 60 mmHg. During X-ray scope examination no leaflet motion was observed.

Emergency re-operation was performed. Before the re-median sternotomy, femoral artery and vein were explored. Aortic and bi-caval

cannulation were performed. Coronary sinus (CS) and right upper pulmonary vein were also cannulated. Operation was started with the use of standard CPB without cross-clamping the aorta. Pulmonary vein was continuously vented. When the patient was put in the Trendelenburg position, the left atrium was opened. We observed that the leaflets of prosthesis were stuck. During excision of the valve and the left atrial thrombectomy aorta were clamped and continuous retrograde CS perfusion with oxygenated warm blood was started. Retrograde CS perfusion rate was kept between 400-500 ml/min and perfusion pressure kept between 50-60 mmHg. Possible myocardial ischemia was monitored electrocardiographically and with measuring of blood gas changes of the returned blood from the aortic venting in every 10 minutes during CS perfusion. Prosthetic valve was excised and bileaflet mechanical valve was replaced. After closing the left atrium and de-airing of the heart, aortic cross-clamp was released and retrograde perfusion was stopped. Weaning from the CPB occurred smoothly with 5 mcg/kg/min dopamine support. Operation was performed without any complication and there was no postoperative cerebrovascular event.

Discussion

Although major technological advances have been made in myocardial protection, perioperative adverse affects caused by myocardial ischemia and reperfusion injury have not been completely eliminated. Therefore, great effort is made to prevent reperfusion injury during such procedures