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Arrhythmogenic right ventricular cardiomyopathy complicated by thrombus formation (RCD code: III-4)

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Abstract

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is caused by replacement of myocardial cells by lipocytes and fibrocytes. The disease is associated with life-threatening ventricular arrhythmias. Echocardiography is an easily accessible and useful diagnostic tool in these patients We present a case of an ARVC patient with high risk of sudden cardiac death and thrombus formation in the right ventricle. JRCD 2015; 2 (5): 165–168

Key words: sudden cardiac death, vantricular arrhythmia, echocardiography, implantable cardioverter-defibrillator

Background

Depletion of the right ventricle myocardial cells is the primary mechanism of arrhythmogenic right ventricular cardiomyopathy (ARVC) development. Cardiomyocytes, probably in consequence of apoptosis, are gradually replaced by lipocytes and fibrocytes. Majority of affected patients are male and in approximately a third of cases a hereditary, autosomal dominant trait can be identified [1].

Echocardiography is an easily accessible and useful diagnostic tool in ARVC patients [2]. Dilation along with structural and wall motion abnormalities of right ventricle (RV) can all be visualized. Areas of tissue distortion are the sites initiating reentry arrhythmias, most commonly ventricular tachycardia (VT) with ECG features of left bundle branch block (LBBB). During sinus rhythm (SR) ECG typically shows incomplete or complete right bundle branch block (RBBB), T waves inversion in V1–V3 leads and intraventricular conduction abnormalities – an epsilon wave [1,3]. Sudden cardiac death (SCD) risk is elevated with highest risk in young patients (<35 years of age), those with poorly tolerated ventricular arrhythmias or left ventricle (LV) involvement [1,4].

We present a case of an ARVC patient with high risk of SCD and thrombus formation in RV.

Case description

A 42-year old male was urgently admitted to Cardiology Department due to hemodynamically unstable paroxysmal tachycardia. He had a history of recurrent palpitations with three documented incidents of pharmacologically treated VT 12 years before. Consequently, he had been taking a maintenance dose of 160 mg of sotalol daily for the last several years. He reported good exercise capacity with no angina symptoms. There was no family history suggesting hereditary character of the disorder.

ECG on admission showed a wide QRS complex tachycardia of altering morphology with frequency of approximately 200 bpm. After promptly performed electrical cardioversion SR of 70 bpm was restored but intraventricular conduction abnormalities persisted.

Transthoracic echocardiography (TTE) revealed RV dilation (dimension of 37 mm in parasternal long axis view) with areas of hypo- and dyskinesis, fibrous trabeculas and wall thinning with numerous recesses (Figure 1). In the apical 4-chamber view an additional structure within the RV was visualized. It had dimensions of 11 × 17 mm and echogenicity corresponding to an unorganized thrombus (Figure 2). Doppler color flow mapping showed presence of tricuspid regurgitation with an area of 6 cm^2 (and right atrium

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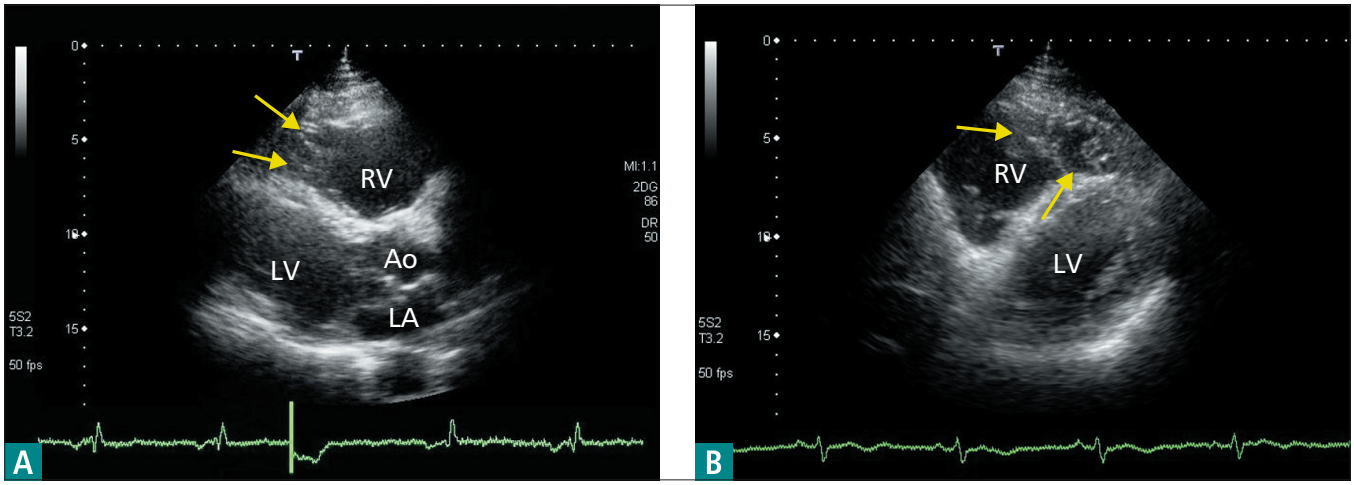

Figure 1. Transthoracic echocardiography. Parasternal long-axis (A) and short-axis views (B). Structural abnormality of the right ventricle (arrows). LV – left ventricle, RV – right ventricle, LA – left atrium, Ao – aorta

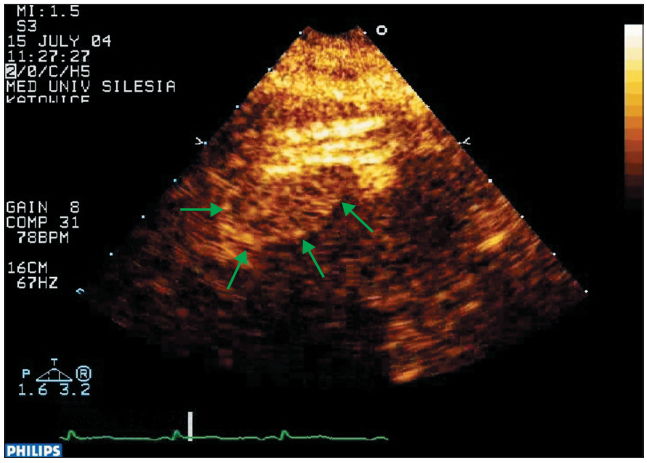

Figure 2. Transthoracic echocardiography. Apical four chamber view. An additional structure in the right ventricle (arrows)

area of 22 cm^2), maximum velocity of 2.3 m/s and RV systolic pressure of 32 mm Hg.

Next, ECG-based studies were continued. 24-hour holter monitoring registered numerous pairs of ventricular premature beats. Late ventricular potentials were detected on a signal-averaged registration with total QRS time of 134 ms, QRS low voltage (< 40 uV) phase duration of 68 ms and the last 40 ms of QRS amplitude of 5.03 uV.

Coronary angiogram, performed to exclude ischemia-induced arrhythmia, showed no stenotic lesions.

Cardiac magnetic resonance imaging confirmed previously described morphology abnormalities within the RV.

Finally, the diagnosis of ARVC complicated by VT and RV thrombus formation was established. Due to recurrent character of the arrhythmia the patient was scheduled for implantation of an implantable cardioverter defibrillator (ICD). The procedure was preceded by temporary increase of sotalol daily dose to 320 mg and 4 weeks of oral anticoagulation therapy after which the resolution of RV thrombus was documented by TTE. After ICD implantation the pharmacotherapy continued with 240 mg of sotalol daily and oral anticoagulation.

Discussion

We present a case of a patient with ARVC in whom anatomical and functional abnormalities resulted in RV thrombus formation.

Literature reports have recently described a number of ARVC patients wwith presence of intracardiac embolic material, either in RV [5, 6] or, less frequently, in RA [7]. Thrombi formation does not seem directly related to rhythm disturbances. Regional anatomical and wall motion abnormalities with local flow impediment are probably more significant. A case of ARVC with coexistent prothrombin gene mutation was also reported in which pharmacotherapy-resistant thrombophilia and recurrent intracardiac thrombi formation eventually necessitated heart transplantation [8].

Hemodynamic instability during VT and relatively young age of our patient pointed at a higher risk of SCD. As long-term antiarrhythmic pharmacotherapy proved inefficient ICD implantation was indicated but required prior treatment of RV thrombus was required.

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