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STUDIUM PRZYPADKU / *CLINICAL VIGNETTE*

Asymptomatic course of arrhythmogenic right ventricular cardiomyopathy

Bezobjawowy przebieg arytmogennej kardiomiopatii prawej komory

Agnieszka Sioma[1], Łukasz Mazurkiewicz[1, 2], Magdalena Marczak[2], Ewa Kucharczyk-Petryka[1], Jacek Grzybowski[1]

[1]Department of Cardiomyopathy, Institute of Cardiology, Warsaw, Poland
[2]Magnetic Resonance Unit, Institute of Cardiology, Warsaw, Poland

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a primary myocardial disorder characterised by progressive fibro-fatty transformation of right ventricle (RV) or both ventricles, which constitutes a substrate for electrical instability. Typical manifestation comprises electrocardiographic repolarisation abnormalities, RV dilatation with systolic dysfunction, and potentially malignant ventricular arrhythmias. Contemporary diagnostic criteria heavily depend on qualitative analysis of ventricular dilatation and systolic dysfunction, which need to be accompanied by the visual identification of regional wall motion abnormalities in order to qualify as minor or major disease-defining criteria. We present a case of a 52-year-old male who was referred to our institution with suspicion of ARVC, for further evaluation. He had had abnormal electrocardiogram (ECG) (suggesting ARVC) for 21 years of his life and dilated RV in echocardiography preformed 15 years ago. He denied syncope or any chest pain, and his exercise tolerance was good. He had no family history of ARVC or sudden death. Physical examination was normal as well as blood tests including N-terminal pro B-type natriuretic peptide and troponin T. ECG showed sinus bradycardia 50/min with the epsilon waves superimposed over the right bundle branch block and negative T-waves in leads V1–V6 (Fig. 1A, B). 24-h Holter ECG registered sinus rhythm at the average of 67/min with no complex ventricular arrhythmias. Cardiac magnetic resonance revealed normal size and function of left ventricle and severely dilated RV (RV end-diastolic volume index was 161 mL/m^2) with markedly compromised systolic function (RVEF 23%), vast aneurysmatic regions, and diffuse thinning of free RV wall (Fig. 1C, D, E). No severe myocardial fibrosis was found, despite a tiny focus in the inferior junction point. Computed tomography scan of coronary arteries was normal. The clinical manifestations of ARVC are varied and age-dependent. In this case we demonstrate that even severe RV malfunction, which fulfilled all ARVC diagnostic criteria, may be asymptomatic and not generate complex ventricular arrhythmias. Due to the progressive nature of the disease asymptomatic ARVC patients should have an annual follow-up with Holter guidance and exercise testing. Currently there is no evidence for medical nor implantable treatment in this group. Our recommendations for this patient, included avoiding endurance physical activity. The diagnostic pattern was limited due to lack of genetic testing, which should be performed in subjects suspected for the disease and in family members of known ARVC patients.

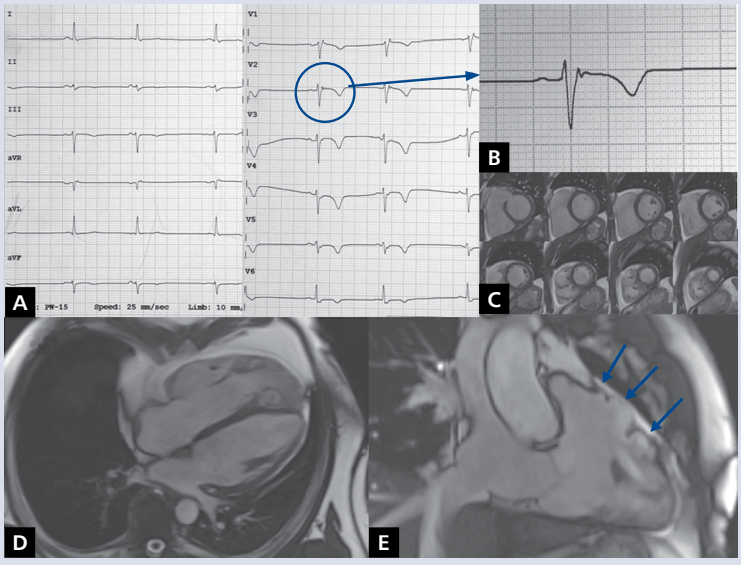

Figure 1. The standard 12-lead electrocardiogram with the epsilon waves superimposed over the right bundle branch block and negative T-waves in leads V1–V6 (**A**) and a magnification of QRS in V2 showing epsilon wave (**B**). A short axis stack showing normal size left ventricle and enlarged right ventricle (**C**). SSFP still, four-chamber frame (**D**). SSFP still, long axis frame showing multiple, vast aneurysmatic areas within the free wall of right ventricle (arrows) (**E**)

Address for correspondence:
Jacek Grzybowski, MD, PhD, Department of Cardiomyopathy, Institute of Cardiology, ul. Alpejska 42, 04–628 Warszawa, Poland, e-mail: jgrzybowski@ikard.pl

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