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UNUSUAL CASE OF THE LEFT VENTRICULAR ANEURYSMATIC INVOLVEMENT IN ARRHYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY

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Abstract

Arrhythmogenic right ventricular cardiomyopathy is an inherited myocardial disease characterized by fibro-fatty replacement of the right ventricle and is considered the second most prevalent cause of sudden cardiac death in young and athletes. The disease is usually manifested by paroxysms of life-threatening ventricular arrhythmias. In this report, we present a case of a 46 years old male who referred to our clinic with complains of palpitations and irregular heartbeats. The patient presented with a history of palpitations for many years for which he received amiodarone and bisoprolol. During hospital treatment, he developed a paroxysm of hemodynamically unstable wide complex ventricular tachycardia which was treated by electrical cardioversion. The coronary angiography of the patient did not detect any haemodynamically significant stenoses of coronary arteries. The echocardiographic examination revealed concentric hypertrophy of the left ventricle, enlargement of right ventricle and right atrium. The aneurysm was present in the subvalvular region and along the lateral wall of right ventricle right ventricle. The patient was preliminary diagnosed with stable ventricular tachycardia, syncope, arrhythmogenic shock, status after electrical cardioversion and arrhythmogenic right ventricular cardiomyopathy. The cardiac magnetic resonance study was performed for confirmation of clinical diagnosis which revealed the presence of left ventricle aneurysm, right ventricle free wall aneurysm and dilatation. The late gadolinium enhancement revealed the presence of right ventricle fibrosis. The presented case demonstrates that biventricular involvement may be undetected by echocardiography, and cardiac magnetic resonance study with increased experience to interpret myocardial structural and tissue characteristics is considered the most accurate diagnostic technique in arrhythmogenic right ventricular cardiomyopathy.

Keywords: *arrhythmogenic right ventricular cardiomyopathy, left ventricular aneurysm, cardiac magnetic resonance, sudden cardiac death*

Introduction

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an inherited cardiomyopathy characterized by fibro-fatty myocardial replacement leading to the development of ventricular arrhythmias and impairment of ventricular systolic function [*Thiene G et al., 1988; Corrado D et al., 2017; Miles C et al., 2019*]. The disease is inherited by an autosomal dominant manner and clinical manifestations of ARVC are three times more frequent in males than females [*Bauce B et al., 2008*]. The prevalence of ARVC is estimated to be 1:1000-1:5000 [*Basso C et al., 2009; Sen-Chowdhry S et al., 2010*], and it is considered one of the leading causes of arrhythmic cardiac arrest in young people and athletes [*Corrado D et al.,* 2006; *Corrado D et al., 2017a*]. The typical histo-

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pathological feature of ARVC is the loss of right ventricular (RV) myocardium with the substitution of fibrous and fatty tissue [*Basso C et al., 1996*], which is thought to contribute to the development of ventricular arrhythmias by slowing intraventricular conduction and acting as a substrate for arrhythmias through a scar-related macro-reentry mechanism [*Fontaine G et al., 1984*]. The diagnosis of ARVC is based on the Task Force Criteria which include structural, histopathologic, electrocardiographic, familial, arrhythmic, and genetic parameters [*Marcus F et al., 2010*]. The clinical presentation of the disease in adolescents or young individuals consists of palpitations, premature ventricular contractions (PVCs), ventricular arrhythmias, episodes of syncopes, occurring during physical exercise, and cardiac arrest [*Haugaa K et al., 2016; Corrado D et al., 2017b*]. Ventricular tachycardia (VT) with left bundle branch block morphology and superior axis is the typical VT indicating ARVC [*Fontaine G et al., 1984*]. Sudden cardiac death (SCD) may occur in asymptomatic individuals, mostly young people and competitive athletes with a previously undiagnosed arrhythmogenic cardiomyopathy [*Thiene G et al., 1988; Corrado D et al., 1990; 2003; 2006*].

In the typical form of ARVC, the left ventricle (LV) is affected to a lesser extent than the RV; however, there are disease variants characterized by equivalent or even predominant involvement of LV [*Corrado D et al., 1997; Jain A et al., 2010*]. The diagnosis of biventricular or predominant left ARVC may be missed at the onset of symptoms in some patients who present years later with heart failure and are incorrectly diagnosed as having idiopathic dilated cardiomyopathy [*Corrado D et al., 1997; Hulot J et al., 2004*]. Currently, a number of non-invasive cardiac diagnostic tests, including 24-hour Holter ECG, echocardiography and cardiac magnetic resonance (CMR) imaging are used to diagnose ARVC. This report describes a clinical case of a man affected by this rare cardiomyopathy which was manifested by LV aneurysmatic involvement preliminary undetected by transthoracic echocardiography.

To overcome it is possible, due to the uniting the knowledge and will of all doctors in the world

Case description

A 46 years old male was referred for CMR examination to the Clinic of General and Invasive Cardiology of the University Hospital No 1 presenting with complains of palpitations and irregular heartbeats. The patient presented with a history of palpitations for many years for which he has been prescribed a combined treatment with amiodarone and bisoprolol. During the hospital treatment, the patient developed a paroxysm of haemodynamically unstable wide complex VT which was terminated by electrical cardioversion. He underwent coronary angiography, which did not detect any haemodynamically significant stenotic lesions of coronary arteries. The family history was unremarkable. The 24-hour Holter ECG monitoring revealed sinus rhythm with an episode of monomorphic VT consisting of 5 ventricular complexes with a ventricular rate of 120 beats per minute (*bpm*) and without clinical manifestation (Figure 1 A, B).

The transthoracic echocardiography revealed concentric hypertrophy of LV, enlargement of RV and RA. The left ventricular ejection fraction (LVEF) was estimated 45% with no regional LV asynergy. The RV diameter at the basal level was 60 *mm*. The RV aneurysm was present in the subvalvular region and along the lateral wall of RV. The Doppler examination revealed 1st degree mitral regurgitation and 3rd degree tricuspid regurgitation. The patient was preliminary diagnosed with stable VT, syncope, arrhythmogenic shock, status after electrical cardioversion and arrhythmogenic right ventricular dysplasia/cardiomyopathy. The CMR examination was performed in our hospital, which revealed the presence of LV aneurysm, RV free wall aneurysm and dilatation (Figure 2A). The late gadolinium enhancement

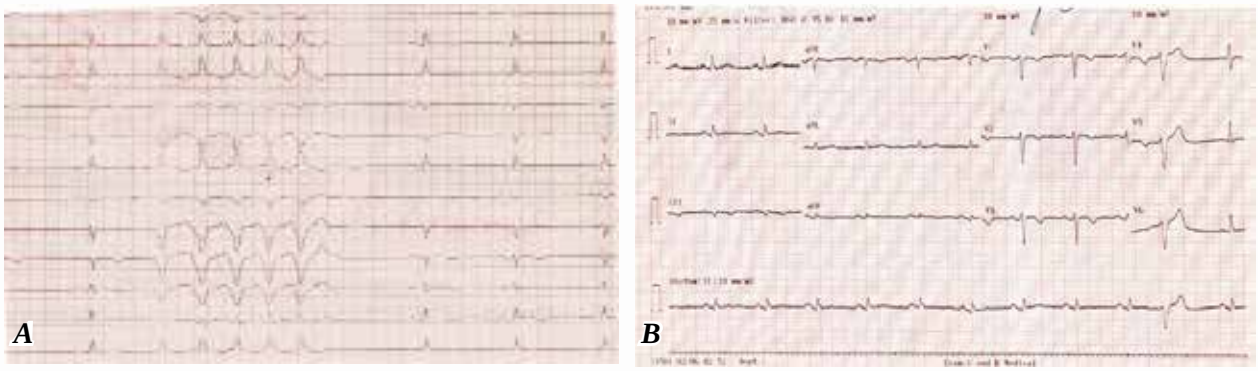

***Figure1. A**-The presence of VT recorded at the patient's admission to CICU; **B**-an episode of monomorphic VT revealed during a 24-hour Holter monitoring. CICU=cardiac intensive care unit; VT=ventricular tachycardia.*

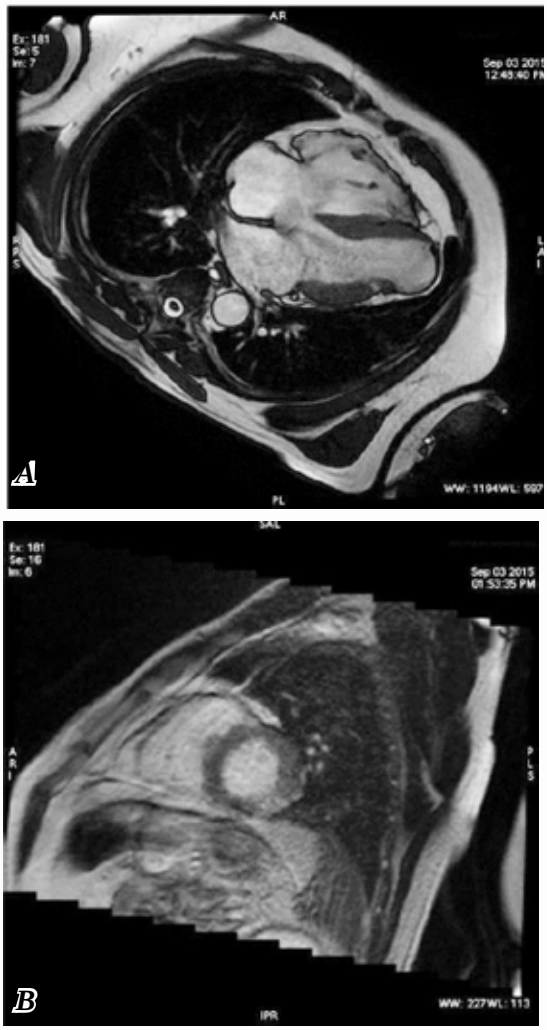

***Figure 2.** Cardiac magnetic resonance imaging: A - Cine study. Long axis view shows RV dilatation with dyskinesia and left ventricle aneurysm (arrows); B - short axis view after late gadolinium enhancement reveals right ventricle wall fibrosis (arrow).*

(LGE) showed the presence of RV fibrosis (Figure 2B). The patient underwent the implantation of a dual-chamber cardioverter defibrillator (ICD) for the secondary prevention of SCD.

Discussion

This report describes a case of LV involvement in a patient with ARVC. The literature data shows that biventricular form of ARVC is common. In fact, previous studies have reported LV involvement ranging between 16% and 76% of cases [*Corrado D et al., 1997; Te Riele A et al., 2013; Berte B et al., 2015*]. The previous study by El Ghannudi and coauthors has shown thatamong 21 patients with ARVC 52% had LV involvement as assessed by CMR [*El Ghannundi S et al., 2015*]. The extent of RV impairment in this study was similar between those with isolated RV involvement and biventricular form of the disease. In another study, Rastegar N. and co-authors studied 78 ARVC mutation carriers of whom 38 had structural abnormalities revealed by CMR, including the impairment of LV in 55% of cases [*Rastegar N et al., 2015*]. In most patients with ARVC, biventricular involvement has been associated with SCD independent of age at death, normal macroscopic appearance of the heart, and participation in competitive sport [*Miles C et al., 2019*]. This difference in findings of LV involvement in this cardiomyopathy is probably conditioned by different imaging techniques used in diagnostic clarifications. In our study, the biventricular involvement was confirmed by the presence of LV aneurysm, RV dilatation and fibrosis by CMR, whereas echocardiographic evaluation failed to reveal the presence of LV aneurysm. Presently, CMR is considered the most accurate non-invasive imaging technique to diagnose this form of ARVC, which may show late gadolinium enhancement (LGE), indicative of myocardial fibrosis, usually within the LV inferior or inferolateral walls [*Sen-Chowdhry S et al., 2008*].

To our knowledge, our clinical case the original report describing the presence of large LV aneurysm in ARVC, and CMR imaging study plays a key role in the diagnostic work-up to assess anatomical, functional and tissue-specific characteristics in this form of cardiomyopathy.

Conclusion

This case demonstrates an uncommon manifestation of ARVC with LV large aneurysmatic involvement. Although LV involvement in ARVC is common, the aneurysm of LV without the presence of coronary pathology may be a rare clinical finding which may lead to the initial is diagnostic approach.

The presented case demonstrates that biventricular involvement may be undetected by echocardiography, and CMR study with increased experience to interpret myocardial structural and tissue characteristics is considered the most accurate diagnostic technique in ARVC.

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