

Ventricular Tachycardia Triggered by Physical Exercise as the First Manifestation of Arrhythmogenic Right Ventricular Dysplasia: A Case Report and Review of the Literature

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Abstract

Arrhythmogenic right ventricular dysplasia/cardiomyopathy is an inherited cardiac disorder that predisposes to sudden cardiac death, particularly in young patients and athletes. Physical exercise promotes the phenotypic expression of the disease and accelerates the onset of life-threatening ventricular arrhythmias. We report the case of a 42-year-old male who was diagnosed with arrhythmogenic right ventricular dysplasia after his first episode of ventricular tachycardia that occurred during a football match.

Keywords: Arrhythmogenic right ventricular dysplasia • Syncope • Ventricular tachycardia • Exercise

Introduction

Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy (ARVC/D) is an inherited condition characterized histologically by the rarefaction of cardiomyocytes and their substitution with fibro-fatty tissue [1]. It is an autosomal dominant disease with incomplete penetrance and variable phenotypic expression that results from a genetic defect of the cardiac desmosome and predisposes patients to ventricular arrhythmias, right ventricular failure and, sudden cardiac death [2]. The diagnosis is usually achieved using the task force criteria [1]. Non-invasive imaging play an essential role in the diagnosis and typically assess segmental motion abnormalities of the right ventricular wall in addition to right ventricular dilation or dysfunction. Besides, Magnetic Resonance Imaging (MRI) may show the intramyocardial lipofibromatous composition of the Right Ventricle (RV) [3]. The therapeutic strategy is based on risk stratification. It includes lifestyle changes, antiarrhythmic drugs, catheter ablation, Implantable Cardioverter-Defibrillator (ICD), and heart transplantation [4]. Exercise is one of the main triggers of a life-threatening arrhythmia and sudden death in addition to the symptomatic progression of right ventricular dysfunction in ARVC/D [5].

Case Report

We present the case of 42-year-old man with no personal or family history of cardiovascular disease, unexplained syncope or sudden cardiac death whose clinical symptoms started with the onset of syncope during a football game. On physical examination, apart from a regular tachycardia, vital signs were stable and physical examination revealed no cardiac or other systemic abnormalities. We performed twelve-lead Electrocardiogram (ECG) that showed sustained Ventricular Tachycardia (VT) with left bundle branch configuration (Figure 1). Ventricular tachycardia was reduced by defibrillation after brief general anesthesia. Biological parameters were within normal

ranges. A coronary angiography was performed and revealed normal arteries. Transthoracic Echocardiography (TEE) revealed apical trabecula-like stacked plates aspect and dilation of the RV (Figure 2). There were no abnormalities in the left ventricular ejection fraction and segmental wall motion. MRI showed foci of fat in RV (Figure 3), dilated RV (Figure 4), and dyskinesia of the right ventricular anterior wall. The diagnosis of ARVC/D was made according to the new task force criteria, and the decision was made to implant a mono-chamber cardioverter defibrillator.

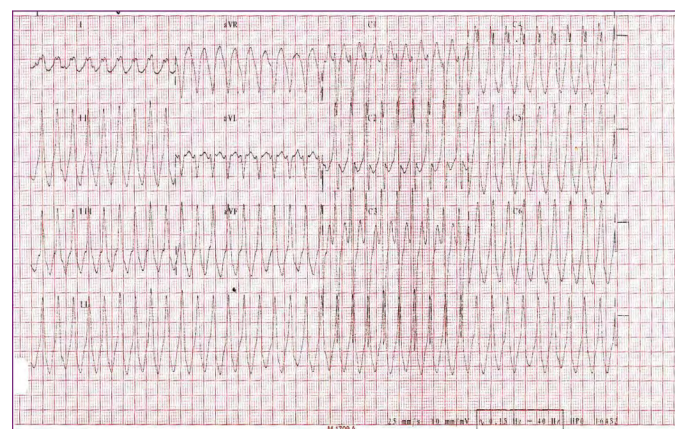


Figure 1. Twelve-lead ECG showing sustained ventricular tachycardia with left bundle branch configuration.

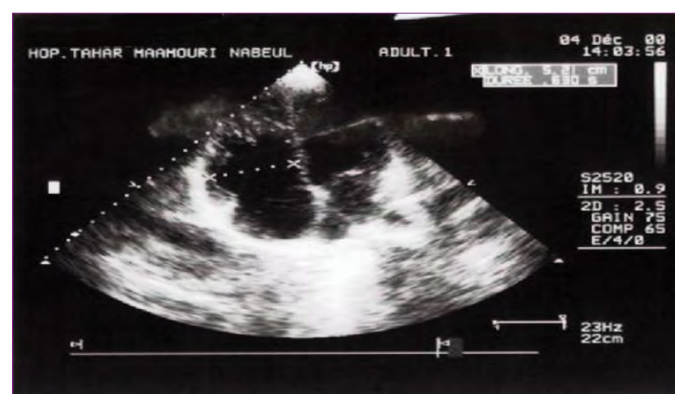


Figure 2. Transthoracic echocardiography, apical four chambers view showing right ventricular dilation with structural abnormalities (apical trabecula-like stacked plates aspect).

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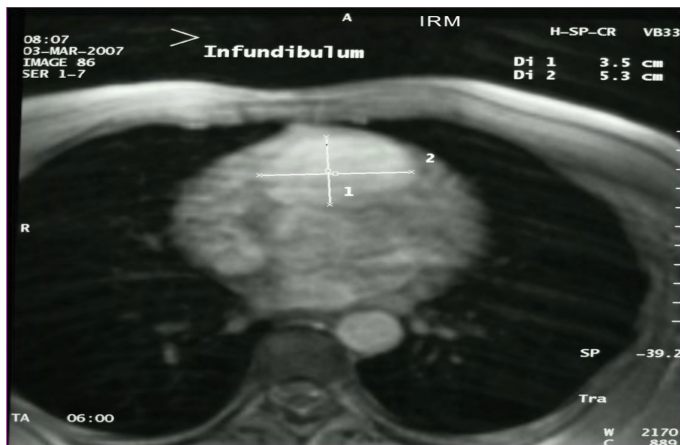


Figure 3. MRI showing the infiltration of the infundibular region of the right ventricle with fat.



Figure 4. MRI showing right ventricular dilation.

Discussion

ARVC/D is a genetically determined myocardial disorder typically transmitted with an autosomal dominant pattern of inheritance with low penetrance and variable phenotypic expression [1]. Its prevalence ranges from 1:2500 to 1:5000 [2]. Most studies reported a higher incidence among men with a male to female ratio 3:1 [2]. There is a large spectrum of clinical manifestations that occur generally between the second and fourth decades of life [1]. They range from asymptomatic subjects to life-threatening arrhythmias, refractory heart failure, and sudden cardiac death [1]. Histologically, ARVC/D is characterized by segmental myocyte's depletion and progressive fibrofatty replacement of the myocardium associated with a variable degree of inflammatory infiltrates [2]. These degenerative changes progress from the epicardium to the endocardium resulting therefore to aneurysmal dilation of the right ventricular wall that predominates in the so-called « triangle of dysplasia » involving the infundibular, apical and sub-tricuspid regions [1]. Left ventricular involvement, although infrequent, is usually associated with an unfavorable outcome [2-5]. Genotyping had identified several genes mutations linked to ARVC/D. Indeed, genes encoding components of the desmosome, nuclear envelope, intermediate filament, area composita, sarcomere, and sodium channel have been incriminated [2-5]. Defective desmosomes represent almost 60% of these mutations [6]. These proteins play a key role in tissue integrity by mediating cell-to-cell adhesion and cytoskeletal linkages. Besides, they are important mediators in the regulation of different signaling pathways [3]. Therefore, mutations in different desmosomal proteins may lead to electrical instability with loss of conduction and arrhythmias [7]. Physical exercise has an obvious deleterious role in the natural history of ARVC/D. It may facilitate the

mechanical uncoupling of myocytes which leads to extensive cell death and the substitution of the myocardial tissue with inflammatory infiltrates, fibrosis, and adipocytosis [8]. On one hand, the desmosomal dysfunction accelerated by exercise may lead to electric instability and is associated with more adverse events manifested by VT, ventricular fibrillation, and sudden cardiac death [5]. On the other hand, the increase of afterload and wall stress may also cause rapid deterioration of the right ventricular function leading thus to refractory heart failure [5]. Several combined clinical, electrical, morpho-functional, and histopathologic criteria were proposed by the task force to reach a clinical diagnosis [1]. ARVC/D is certain in the presence of 2 major criteria, 1 major plus two minor or four minor criteria [6].

Twelve-lead electrocardiogram is a valuable diagnostic test that typically shows T wave inversion in the anterior precordial leads, incomplete right bundle branch block, QRS fragmentation, post-excitation epsilon waves and, terminal QRS activation delay [9]. Non-invasive imaging modalities represent an integral part of a diagnostic evaluation of ARVC/D too. TEE assesses structural and functional anomalies including RV dilation and/or global dysfunction and segmental wall motion abnormalities [1]. CMR is the gold standard imaging technique. It can visualize segmental RV wall motion abnormalities, RV dilation and dysfunction, the intramyocardial lipo-fibromatous infiltration, thinning and aneurysms of the RV wall, hypertrabeculation and delayed enhancement images [10]. Once the diagnosis is made, the next step comprises risk stratification. Several parameters were considered as independent predictors of adverse events such as the onset of unexplained syncope, non-sustained VT, RV and/or left ventricular dysfunction, RV/right atrial dilation, heart failure, young age at diagnosis, male gender, complex genotypes, inferior T wave inversion, fragmented QRS and precordial QRS amplitude ratio <0.48 [4]. Current therapeutic strategies are essentially palliative. The main objectives consist in reducing the morbidity and the mortality of ARVC/D and slowing the progression of the disease. Restriction of intense physical activity is always recommended. Therapeutic management tools include antiarrhythmics drugs, catheter ablation, ICD and heart transplantation. Among these, ICD is the only effective preventive measure. Unquestionably indicated in secondary prevention of sudden cardiac death, it is also recommended in intermediate and high-risk patients [4].

Conclusion

ARVC/D is an inherited cardiomyopathy that predisposes to fatal arrhythmias and sudden cardiac death. Physical exercise plays a pivotal role in the promotion and progression of this disease, which may partly explain the variability of its phenotypic expression. The diagnosis is based on a multi-parametric approach and treatment is essentially symptomatic.

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