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Arrhythmogenic biventricular cardiomyopathy, ascending aortic aneurysm, left main coronary stenosis and complex symptomatic arrhythmias in a middle-aged marathoner: An exceptional association in the same subject

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Abstract

A 65-year-old man, marathoner since he was twenty, with no cardiovascular risk factors or comorbidity, presented at the Emergency Department with a persistent interscapular pain and palpitation. The electrocardiogram showed a rapid ventricular tachycardia, urgently treated by electrical cardioversion. The echocardiogram showed a huge supravalvular aortic aneurysm, 67 mm, with bicuspid aortic valve (treated after few weeks by open heart surgery). Coronary angiography revealed an isolated moderate left main coronary stenosis. Subsequently, the patient presented several episodes of supraventricular arrhythmias. After the first ventricular tachycardia episode, he was hospitalized twice more, owing to ventricular tachycardia recurrences. A cardiac magnetic resonance was then performed. Arrhythmogenic biventricular cardiomyopathy diagnosis was hypothesized owing to the presence of five major 2010 International Task Force Criteria. The main point of this case report is the coexistence in the same subject of all described cardiovascular complications caused or favored by excessive endurance exercise, never described before in the literature, to the best of our knowledge.

Keywords

exercise; cardiovascular disease; athletes

Abbreviations

ARVD: Arrhythmogenic right ventricle cardiomyopathy; EPS: Electrophysiological study; ICD: Implantable cardioverter defibrillator; LBBB: Left bundle branch block; RBBB: Right bundle branch block; RV: Right ventricle; VT: Ventricular tachycardia

Introduction

Regular physical exercise is effective in the prevention and treatment of conditions such as hypertension, coronary artery disease, and diabetes [1]. Although there are certainly benefits to exercise, recent research focusing on the high end of the exercise dose-response range, suggests that chronic endurance training can cause myocardial changes. It is well known that vigorous exercise in unfit, sedentary individuals can trigger an adverse cardiac event, but this now appears to be true in fit

individuals as well [2-3]. In this case report, we describe the strange coexistence of all the described cardiovascular complications in the same subject caused or favored by excessive endurance exercise. A genetic predisposition can be suspected but excessive endurance training has certainly contributed to the onset and progression of the majority of these complications. Furthermore, traits of sport addiction can be recognized in this subject [4].

Case Presentation

A 65-year-old man, marathoner since he was twenty, presented at the Emergency Department on 27 March 2016 with a persistent interscapular pain and palpitation. He had no significant medical history other than occasional palpitation for a few months. The Electrocardiogram (ECG) showed a wide QRS complex tachycardia, 190 bpm, Right Bundle Branch Block (RBBB) morphology and superior axis. Urgent electrical cardioversion was performed with a positive result. The echocardiogram showed a huge supravalvular aortic aneurysm and bicuspid aortic valve with calcified cusps and mild regurgitation. Subsequent computed tomography angiography confirmed the aneurysm, reaching the aortic arch, maximum diameter 67 mm, with no signs of dissection. Furthermore, the echocardiogram showed a moderate enlargement of the Right Ventricle (RV), with hypertrophy and hypokinesis. Coronary angiography revealed an isolated moderate left main coronary stenosis. During hospitalization, the patient presented several episodes of paroxysmal supraventricular arrhythmias (atrial flutter, fibrillation and tachycardia). On 13 April 2016 he underwent open heart surgery: Aortic valve replacement with a bioprosthesis Carpentier-Edwards Perimount Pericardial Magna n° 27 and supracoronary ascending aortic replacement with a 30 mm Hemashield graft, without post-operative complications. On 27 May 2016 the patient underwent an Electrophysiological Study (EPS), on therapy with amiodarone and metoprolol: No arrhythmic events were induced. In spite of our recommendations, the patient started endurance training again. On 18 June and 23 July 2016, he was hospitalized again owing to Ventricular Tachycardia (VT) recurrences with Left Bundle Branch block (LBBB) morphology and superior axis. On the second occasion palpitation started soon after intense physical activity. Metoprolol and amiodarone were stopped owing to severe bradycardia and QT prolongation. On 14 September 2016 a cardiac magnetic resonance was performed in a tertiary referral hospital. Arrhythmogenic biventricular cardiomyopathy diagnosis was hypothesized owing to the presence of five major 2010 International Task Force Criteria [5]: 1) ECG: Epsilon waves in right precordial leads (Figure 1); 2) ECG: inverted T waves in right precordial leads in the absence of RBBB; 3) non sustained and sustained VT, LBBB morphology with superior axis; 4) Echocardiogram: Presence of a RV apical aneurism (Figure 2) and parasternal long axis RV out-flow tract > 32 mm; 5) Cardiac magnetic resonance: presence of RV akinesia in the basal segment of the free wall and ratio of RV end-diastolic volume to body surface area > 110 ml/m2; in T1-weighted black blood turbo-spin-echo sequences, obtained with fat suppression, evidence of suspected mid-myocardial fatty infiltrations in the anterolateral right ventricle free wall (Figure 3); furthermore, the left ventricle was dilated, globally hypokinetic, with akinesia in the middle-basal segment of the posterior wall; late gadolinium enhancement imaging showed the presence of gadolinium in the akinetic areas of both ventricles. On 15 September 2016 the patient underwent a second EPS without antiarrhythmic drugs; two types of VT (LBBB with superior axis and RBBB with superior axis) and a common atrial flutter were induced. Radiofrequency ablation procedure was successfully performed to treat both ventricular and supraventricular arrhythmias. After that, the

patient refused Implantable Cardioverter Defibrillator (ICD) insertion procedure, even if strictly indicated. In addition, correctly informed about the risks, he started endurance training again. On 27 December 2016 he experienced a new VT recurrence (180 bpm, LBBB morphology with superior axis), urgently treated by electrical cardioversion. In January 2017 the patient underwent a new EPS. Two types of VT were induced but radiofrequency ablation procedure failed, probably owing to the epicardial origin of arrhythmias. At that moment the patient accepted the ICD implantation procedure. He was discharged on therapy with sotalol, aspirin, atorvastatin and ramipril. One year later no symptoms or significant arrhythmic events occurred.

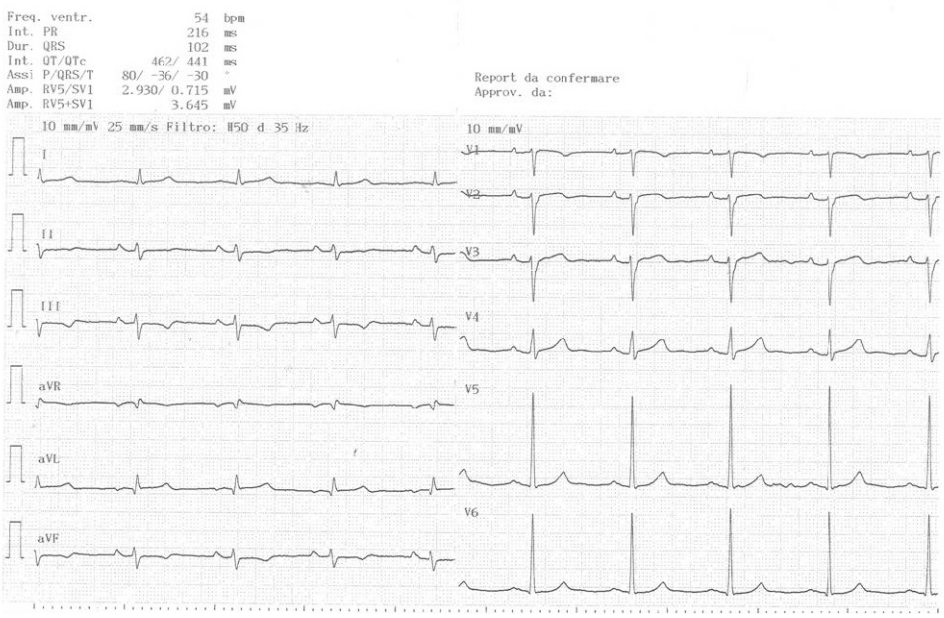

Figure 1: Electrocardiogram: Epsilon waves are evident between the end of the QRS complexes and the beginning of the ST segments in leads V2.

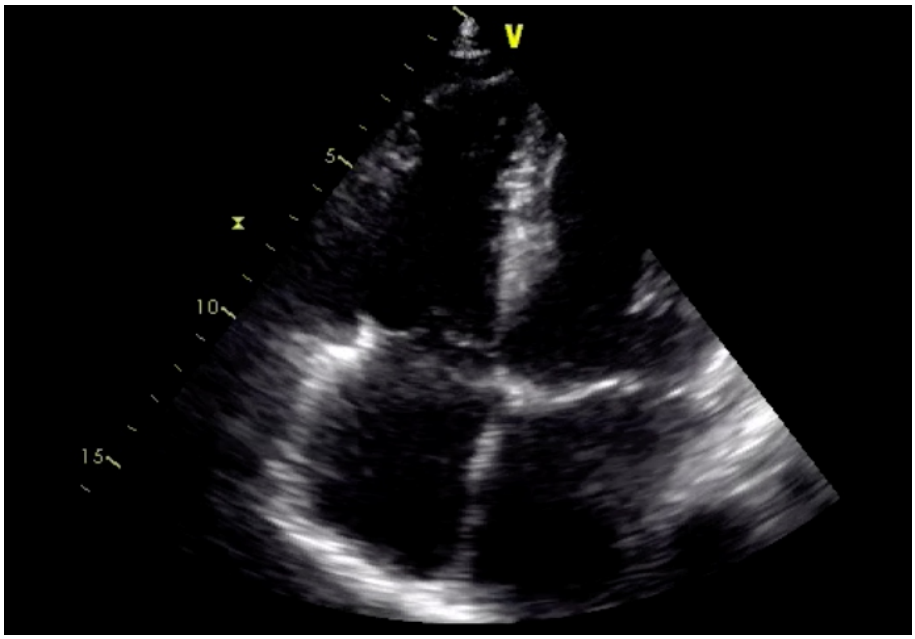

Figure 2: Apical Four Chamber Echocardiogram View: A focal dyskinetic bulging in the apical segment of the right ventricle is evident.

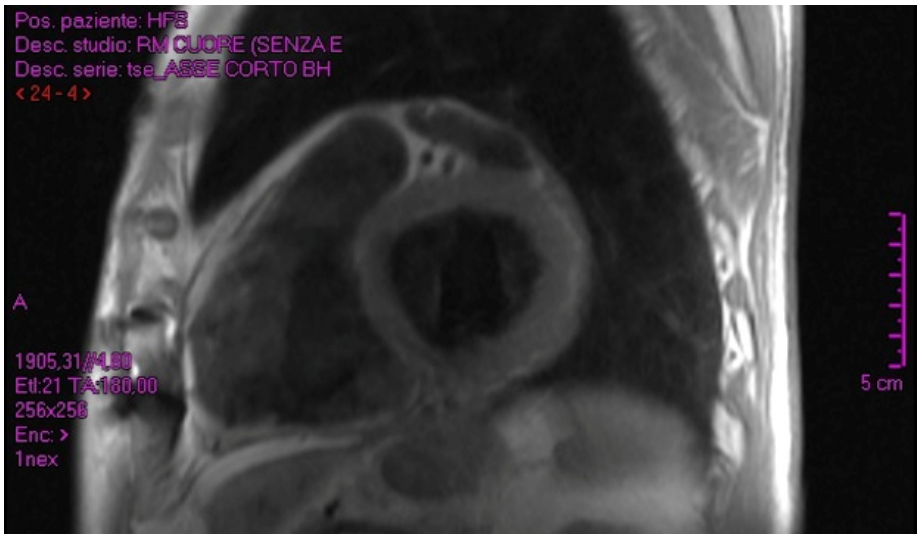

Figure 3: Cardiac Magnetic Resonance Imaging (T1-weighted black blood turbo-spin-echo sequence obtained with fat suppression): two areas of suspected mid-myocardial fatty infiltrations in the anterolateral right ventricle free wall are evident.

Discussion

While obesity has been on the rise in developed countries, there has also been a simultaneous increase in the number of individuals participating in ultra-endurance events. Years of endurance training could lead to long term consequences, such as myocardial fibrosis [6], atrial fibrillation [7-8], ventricular arrhythmias [9], and coronary artery atherosclerosis [10]. Ascending aortic aneurysm and acute aortic dissection have also been described in athletes [11]. In our patient a genetic predisposition for these last diseases can be suspected owing to the presence of bicuspid aortic valve. However, excessive exercise has certainly contributed to the onset and progression of the aortic aneurysm. Considering Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) more and more studies show that endurance exercise can clearly contribute to the early onset, faster progression and worst prognosis in a subject genetically predisposed in some way. The effects of long-term endurance exercise are still unclear. It is believed that the intrinsically thinner walls of the RV and atria could make them more susceptible than the LV to remodeling caused by volume and pressure overload from sustained high-output states [12-13]. The hypothesis of an exercise-induced acquired form of ARVC has also been advanced in recent years [12]. In 2010 La Gerche et al [14]. reported a lower than expected rates of desmosomal gene mutations in some endurance athletes with complex ventricular arrhythmias, advancing the hypothesis that an ARVC-like phenotype may be acquired through intense exercise, without an identifiable genetic predisposition. However, these authors searched mutations in just five candidate desmosomal genes. Since 2000, when the first mutation in a candidate desmosome gene had been identified, sixteen genetic loci had been identified and mutations have been reported in fourteen different genes [15]. It's clear that our knowledge about the genetic predisposition for ARVC is incomplete and continually evolving. Our patient didn't perform a genetic analysis. However, he has no family history of the disease and clinical manifestations appeared in late adulthood (frequently, in patients affected by a familiar form, clinical manifestations appear in adolescence and earliest youth [16]). Despite severe physical injuries and medical recommendations, the patient continued exercising for almost a year, highlighting traits of exercise addiction.

Conclusion

This case report shows the strange coexistence in the same subject of all described cardiovascular complications caused or favored by excessive endurance exercise, never described before in the literature, to the best of our knowledge.

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