Arrhythmogenic cardiomyopathy with primary left ventricular involvement

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Introduction: Arrhythmogenic cardiomyopathy (ACM) is a hereditary heart muscle disease that can affect the isolated right ventricle (RV), both ventricles, or with isolated affection of the left ventricle (LV), in which the healthy myocardium is replaced by fatty and fibrous tissue.[1] It is inherited in an autosomal dominant manner. Genes coding for desmosomal and non-desmosomal proteins are most often mutated. Structural abnormality of the myocardium causes a disturbance in the electrical system of the heart, which manifests as ventricular tachycardia and subsequent sudden cardiac death (SCD).[2] Diagnosis is based on magnetic resonance imaging, echocardiography, and genetic testing. The main goal of treatment is the prevention of sudden cardiac death, with antiarrhythmic drugs to prevent the occurrence of malignant arrhythmias, and primary prevention of SCD by implanting an ICD.[3]

Case report: 22-year-old patient was hospitalized due to recurrent syncope during exertion. Echocardiography showed a normal-sized left ventricle (EDD 48mm), normal-wall thickness (IVSd 9 mm), no segmental contractility loss, EFLV 60%, GLS: -18.2%, slightly decreased values for the basal part of the inferoseptal wall and the medial part of the lateral wall. Diastolic function was normal. E/e 7.6. The right ventricle was normal-sized and without trabeculation, with preserved systolic function. A 12-minute treadmill test was performed, with no discomfort during exercise, and no signs of ischemia on the electrocardiogram, but during recovery, persistent hemodynamically stable ventricular tachycardia with a frequency of 250/min was recorded (**Figure 1**), spontaneously converted to sinus rhythm, and short-term NSVTs were subsequently recorded (**Figure 2**). An MRI of the heart verifies the subepicardial zone of late post-contrast inhibition of the inferoseptal, inferior, and inferolateral walls in the basal and middle third, fulfilling the large structural MR criteria for left ventricular affection with ACM. He was medically treated with the maximum tolerated dose of propranolol. The patient was referred to a tertiary center for further treatment, and an extravascular cardioverter-defibrillator (Medtronic Aurora EV-ICD) was implanted.

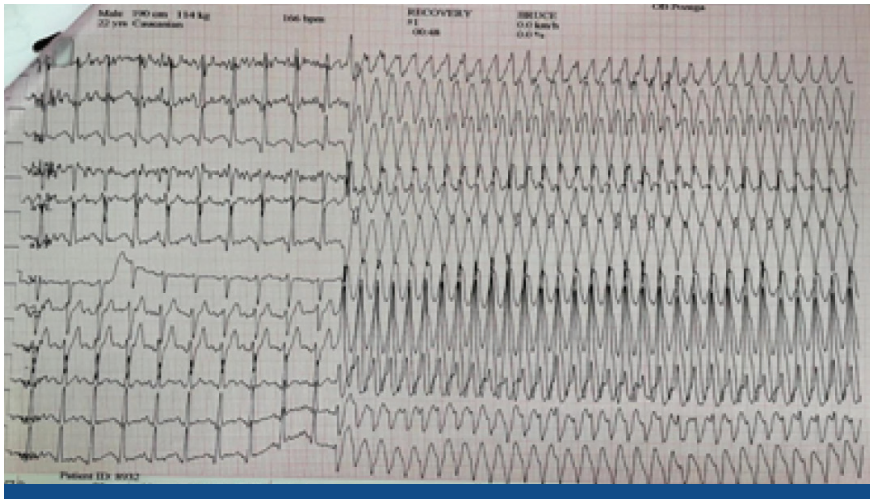

FIGURE 1. Ventricular tachycardia (ECG) on stress test (in recovery phase).

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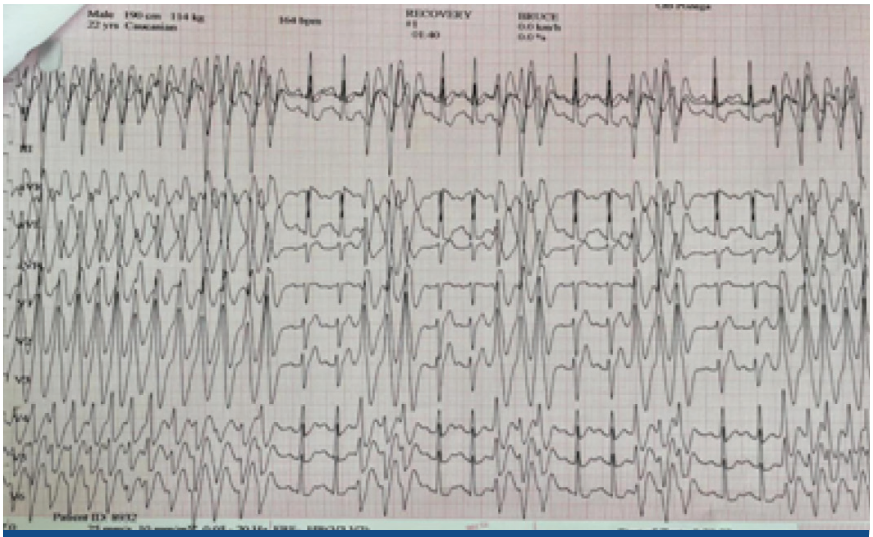

FIGURE 2. Non sustained ventricular tachycardia on stress test (in late recovery phase).

Conclusion: Special caution when examining patients with unexplained syncope during exertion is necessary in order not to overlook a possible malignant arrhythmia in undiagnosed ACM.

LITERATURE

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