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Left dominant arrhythmogenic cardiomyopathy: A morbid association of ventricular arrhythmias and unexplained infero-lateral T-wave inversion ☆

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Abstract Left-dominant arrhythmogenic cardiomyopathy is a subtype of arrhythmogenic right ventricular cardiomyopathy characterized by early predominant left ventricular involvement. A 34-year-old man presented with palpitations and a history of frequent ventricular extrasystoles of both LBBB and RBBB configuration. Cardiac workup revealed repolarization abnormalities at infero-lateral leads in the absence of diagnostic structural/functional alterations or obstructive coronary artery disease. Six months later he died suddenly. Histopathology was diagnostic for arrhythmogenic right ventricular cardiomyopathy affecting predominantly the left ventricle at subepicardial/midwall myocardial layers. Thus, ventricular arrhythmias accompanied by unexplained infero-lateral T-wave inversion should warn of a possible morbid association underlying left-dominant arrhythmogenic cardiomyopathy.

Keywords: Arrhythmogenic cardiomyopathy; Arrhythmogenic right ventricular cardiomyopathy/dysplasia; Arrhythmias; Sudden death

Introduction

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is characterized by progressive loss of right ventricular myocardium with fibrous or fibrofatty replacement. [1] The pathologic process originates from subepicardial layers of myocardium, particularly at the right ventricular outflow tract, posterior wall and apex. [2] Usually involvement of the left ventricle characterizes advanced disease stages. However, early left ventricular predilection has been increasingly reported. [3,4] The left-dominant subtype is easily under-recognized due to misattribution to other disorders and absence of specific diagnostic criteria for ARVC. [5] Non-specific clinical features include inverted T waves in the infero-lateral leads and arrhythmias originating from the left or both ventricles. [3,4] Histopathology reveals myocyte loss with fibrous or fibrofatty replacement localized at subepicardial and midwall layers of the left ventricular myocardium. Delayed enhancement imaging using cardiac magnetic resonance (CMR) might be informative. [4] Since this condition with major lethal potential is still highly under-recognized in clinical practice, continued enforcement of left-dominant subtype of ARVC as a differential for unexplained ventricular arrhythmias and infero-lateral T-wave inversion is an important lesson to teach.

Case presentation

A 34-year-old man presented with palpitations and frequent ventricular extrasystoles during the last 7 years. Syncope or presyncope were not reported. Family history of cardiomyopathy or sudden death was absent. Resting 12-lead ECG revealed T-wave inversion in leads V5 and V6, and flattening T waves in leads II, III and aVF (Fig. 1). Low QRS voltage was observed in limb leads and left precordial leads. In lead V1 the QRS complex was fragmented with terminal activation duration of 56 ms (normal value < 55 ms). More than 1000 ventricular extrasystoles were recorded in 24-hour Holter monitoring. Ventricular extrasystoles captured on 12-lead resting ECG, showed LBBB and RBBB configuration. Two-dimensional echocardiography revealed right and left ventricular dimensions at the upper normal limits with a normal overall systolic function. End-diastolic diameter was 59 mm for the left ventricle and 30 mm for the

☆ Conflicts of interest: None declared.

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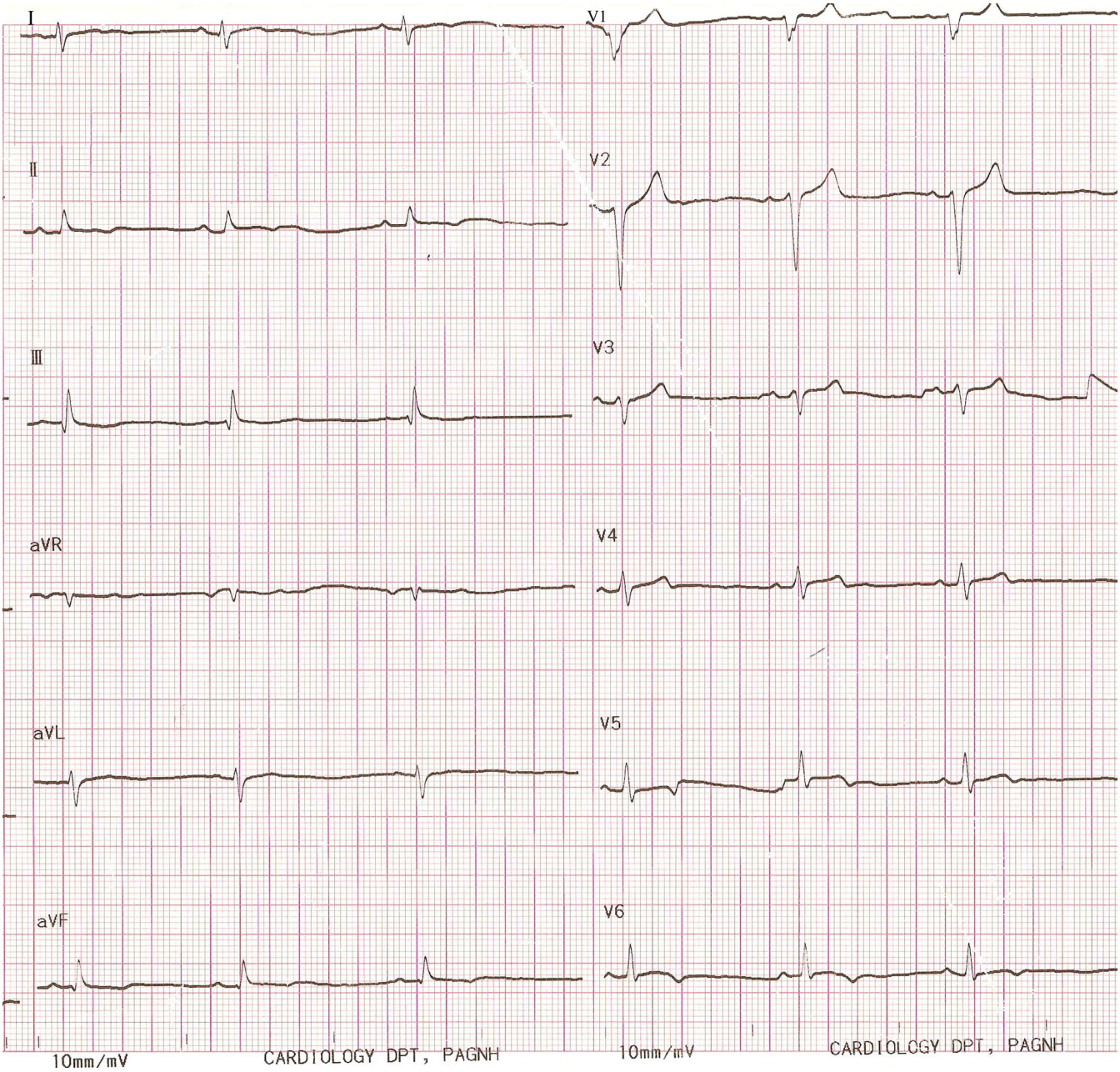

Fig. 1. Recording from 12-lead ECG (25 mm/s, 10 mm/mV). There is low QRS voltage in limb and left precordial leads, T-wave inversion in leads V5 and V6, and flattening T waves in leads II, III and aVF; fragmented QRS complex is observed in lead V1.

right ventricular outflow tract at parasternal long axis view; left ventricular ejection fraction was 60%. Regional akinesia, dyskinesia or aneurysms were not observed in both ventricles. Mild regional hypokinesia of right and left ventricular apex were the only echocardiographic findings. Exercise testing was negative for myocardial ischemia while Thallium-scan was suspicious of ischemia of the left ventricular apex. Thus, coronary angiography was performed revealing no significant stenoses. On electrophysiological study, ventricular tachycardia was not inducible. At this point the patient fulfilled three minor Task Force Criteria for the diagnosis of ARVC (repolarization abnormalities in leads V4-V6, terminal activation duration of QRS $\geq$ 55 ms, and > 500 ventricular extrasystoles on 24h holter monitoring) thus he did not receive a definite diagnosis of ARVC.[5] The patient was discharged on metoprolol 25 mg twice daily and was recommended of an annual follow-up, unless new symptoms occur. Unfortunately, he died suddenly six months later.

On postmortem examination, the weight of the heart was 550 grams and mild right ventricular dilatation was observed. Macroscopic pathology of myocardium revealed fibrotic subepicardial and midwall bands on anterolateral and postero-apical left ventricular walls as well as on the interventricular septum. Histological findings included myocyte loss with fibro-fatty replacement and myocyte abnormalities, such as myocytolysis, cytoplasmic vacuolization, dysmorphic nuclei, small empty cells with peripheral nucleus and perinuclear halo (Fig. 2). Elements of myocardial inflammation were absent. Similar findings