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Kasuistika | Case report

Arrhythmogenic cardiomyopathy of left ventricle. A rare event, but possible

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Náhlá srdeční smrt
Synkopa

SOUHRN

Arytmogenní dyplazie pravé komory (arrhythmogenic right ventricular dysplasia, ARVD) je formou dědičné kardiomyopatie charakterizované nahrazováním svaloviny převážně pravé komory fibrózní a tukovou tkání. U osob s tímto postižením může dojít k rozvoji život ohrožujících komorových arytmií a srdečnímu selhání. Ještě častěji se s ní lze setkat u sportovců, kteří prodělají náhlou srdeční smrt. Postižena však může být nejen pravá, ale i levá komora. Popisujeme případ 38letého muže se dvěma epizodami synkop v anamnéze. Po echokardiografickém vyšetření byl pacient odeslán na vyšetření srdce magnetickou rezonancí. Morfologické zobrazení srdce prokázalo tukovou infiltraci epikardu postranní stěny levé komory (střední a apikální segmenty). Byla stanovena diagnóza izolované levostranné formy arytmogenní kardiomyopatie. Po implantaci kardioverteru-defibrilátoru byla zahájena farmakoterapie enalaprilem a bisoprololem.

Keywords:
Arrhythmogenic right ventricular dysplasia/cardiomyopathy
Cardiomyopathy
Implantable cardioverter defibrillator
Left sided arrhythmogenic cardiomyopathy
Sudden cardiac death
Syncope
Ventricular arrhythmia

ABSTRACT

Arrhythmogenic right ventricular dysplasia (ARVD) is a form of inherited cardiomyopathy characterized by fibro-fatty substitution mainly right ventricular (RV). Affected patients may succumb to life-threatening ventricular arrhythmias and heart failure. It is even more common among athletes who experience sudden cardiac death (SCD). The disease involvement is not limited only to the RV, but the left ventricle (LV) can also be involved. We have reported a case of a 38-year-old man, with two episodes of syncope in his history. After echocardiographic investigations, the patient was referred to cardiac magnetic resonance (CMR). Morphological images showed fatty infiltration of the epicardial layer of LV lateral wall (mid and apical segment). A diagnosis of 'Isolated Left-Sided Arrhythmogenic Cardiomyopathy' was made. An ICD implantation was performed, and a medical therapy with enalapril and bisoprolol was started.

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Introduction

Arrhytmogenic cardiomyopathy (AC) has originally been described as a disorder characterized by fibro-fatty replacement of the myocardium, primarily of the RV in association with ventricular arrhythmias, sudden death and progressive heart failure [1]. It is even more common among athletes who experience sudden cardiac death (SCD) [2]. However, similar histopathologic changes, although rare, are also found in the LV [3].

Case presentation

A 38-year-old man, competitive soccer player up to two years in advance. His clinical history was free from heart diseases but he had a history of psoriasis. He had two episodes of exertional syncope occurring during soccer competition. At first episode he presented to our Emergency Department where the first physical examination resulted normal. Blood and brain CT scan resulted also normal. ECG showed normal. No arrhythmias were found at 12-lead ECG telemetry during 12 h observation period. The transthoracic echocardiogram (TTE) showed a slightly reduced left ventricle ejection fraction (48%) and a lateral wall hypokinesia. Before discharge, the patient refused coronary angiography and he underwent dipyridamole stress echocardiography, resulted negative for myocardial ischemia [4–7]. Three months after, because of the second episode of exertional syncope, he was referred to our Emergency Department again. At first examination, physical findings were normal, BP was 130/80 mmHg but ECG showed an atrial fibrillation with average HR 160/m. A successful pharmacologic cardioversion (with flecainide) was performed. Blood and brain CT scan resulted again normal. TTE after SR restoration showed the same slightly reduced LVEF (45%) and a more clear hypokinesia of posterior and lateral LV wall. The patient underwent cardiac magnetic resonance (CMR). CMR showed the following findings. 1) The cine steady state free precession images disclosed hypokinesia of the mid-lateral LV wall without wall motion abnormalities of the right ventricle. 2) The black-blood proton-density weighted fast spin-echo images showed an irregularity of the boundary between the epicardium and fat of the mid and apical segments of the LV lateral wall (Fig. 1). This finding is consistent with epicardial fat infiltration. 3) The late gadolinium enhancement (LGE) images showed myocardial fibrosis of the mid and epicardial layers of LV lateral wall (Fig. 2). Overall these findings showed fibro-fatty infiltration of the LV lateral wall along with wall motion abnormalities. A diagnosis of 'Left-Sided Arrhythmogenic Cardiomyopathy' was made. The patient underwent ICD implantation and started medical therapy with bisoprolol 2.5 mg (BID) and enalapril 5 mg (OD) [8–14]. He's event-free until now.

Discussion

In our patient was made a diagnosis of 'Left-Sided Arrhythmogenic Cardiomyopathy'. Endomyocardial biopsy

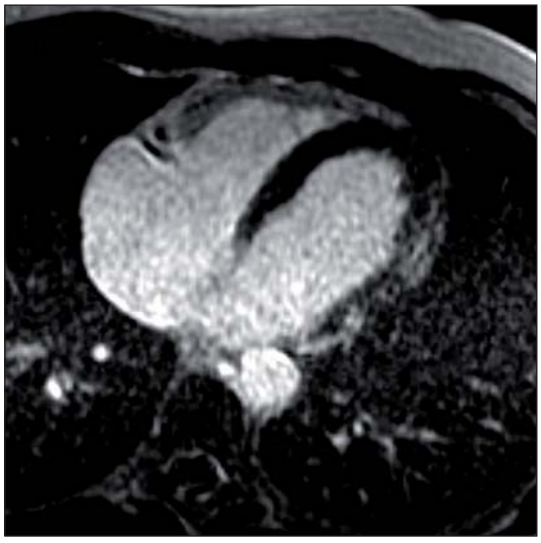

Fig. 1 – CMR: the black-blood proton-density weighted fast spin-echo image showed an irregularity of the boundary between the epicardium and fat of the mid and apical segments of the LV lateral wall. This finding is consistent with epicardial fat infiltration.

is the the gold standard as diagnostic method. However, it has some limitations: a) it is an invasive tool and b) the specimen is usually taken from the septum because of the high risk of perforation and cardiac tamponade in the case of the free wall. In our patient no endomyocardial biopsy was performed because the alterations were in the mid and apical segments of the LV lateral wall (Fig. 1). Several authors observed that arrhythmogenic cardiomyopathy is not limited to the right ventricle, but it can involve the entire myocardium [2,3,8,15,16]. Other possible diseases were considered, including coronary artery disease, storage or infiltrative disease (amyloidosis), cardiac sarcoidosis and arrhythmogenic right ventricular dysplasia/cardiomyopathy affecting dominantly the LV. M. Noorman et al. [1] described a case with a severe fibro-fatty replacement of nearly the entire right ventricular free wall in association with an involvement of the interventricular septum. In our case, CMR showed fibro-fatty infiltration limited to the mid and epicardial layers of LV lateral wall (Fig. 2).

Ischaemic aetiology was less probable (a dipyridamole stress echocardiography was negative). Patients suffering from cardiac amyloidosis usually demonstrate a different pattern [17], so it was an unlikely diagnosis. Typical symptoms of cardiac sarcoidosis, such as conduction system abnormalities, mediastinal lymphadenopathy, extracardiac manifestations and myocardial scar on CMR, were not present [18]. In conclusion the final diagnosis of arrhytmogenic left ventricle dysplasia/cardiomyopathy was made after the exclusion of these three possible diseases. To the best of our knowledge this is the first case of a arrhytmogenic left ventricle dysplasia/cardiomyopathy with the only involvement of the LV.

Conflict of interest
None.

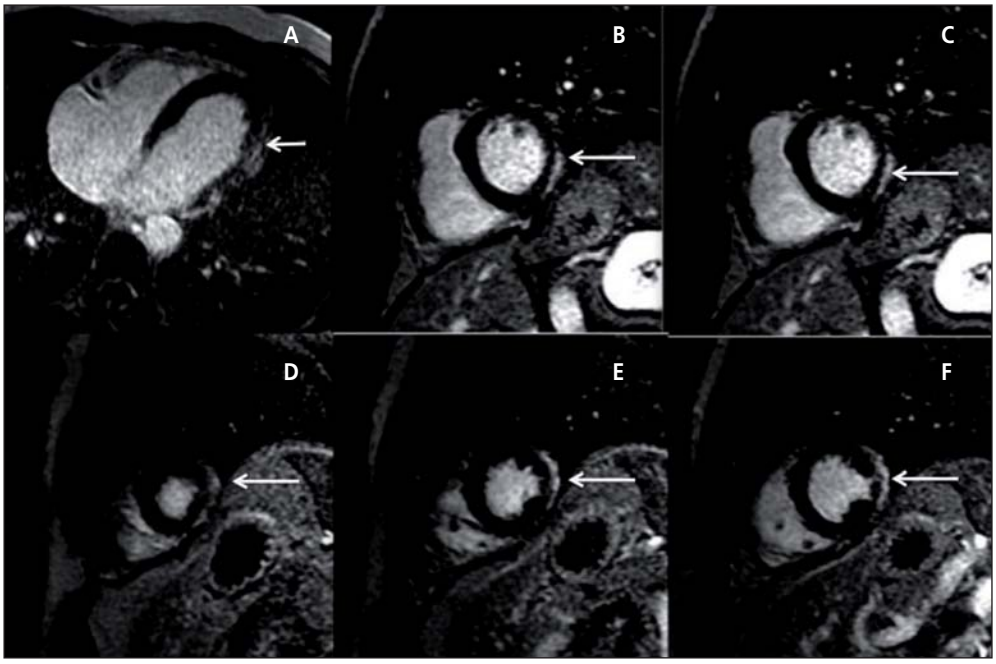

Fig. 2 – CMR. (A) The late gadolinium enhancement (LGE) image showing myocardial fibrosis of the mid and epicardial layers of LV lateral wall and different levels of short-axis. (B–F) Images showing myocardial fibrosis of the mid and epicardial layers of LV lateral wall.

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None.

Ethical statement
Authors state that the research was conducted according to ethical standard.

Informed consent
Informed consent was obtained from the patient.

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