

Cardiac magnetic resonance features of left dominant arrhythmogenic cardiomyopathy: differential diagnosis with myocarditis

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Objectives: To identify potential imaging features at cardiac magnetic resonance (CMR) specific for left-dominant arrhythmogenic cardiomyopathy (LDAC) diagnosis.

Materials and methods: Between January 2011 and May 2016, we considered 36 consecutive stable patients with a recent diagnosis of significant VA and ECG morphology consistent with a LV origin, detection of potential LV arrhythmic substrate at CMR, undergoing a clinically-indicated LV endomyocardial biopsy. Exclusion criteria were history of known cardiac disease, contraindications to CMR and impaired CMR image quality. After application of these criteria, in 9 patients endomyocardial biopsy showed tissue abnormalities consistent with the diagnosis of LDAC. From the same CMR-endomyocardial biopsy registry, a second group of 9 consecutive patients with a histological diagnosis of previous myocarditis were identified.

Results: Mid-wall LGE in the interventricular septum was detected in 5 myocarditis, without findings in LDAC group ($p=0.03$), whereas subepicardial LGE at the level of posterolateral wall of LV was detected in 8 cases of LDAC vs. 2 cases of myocarditis ($p=0.02$). Fat infiltration, and particularly subepicardial posterolateral fat infiltration, was found in all LDAC patients vs. one myocarditis only ($p<0.01$). No differences in other CMR findings or in any clinical or echocardiographic parameters were found between patients with a biopsy consistent with LDAC vs. patients in whom biopsy suggested myocarditis.

Conclusions: In patients with significant VA and ECG morphology consistent with a LV origin, identification of morpho-functional involvement of the subepicardial layer of LV posterolateral wall at CMR (LGE, fat infiltration, wall dyskinesia) is consistent with a diagnosis of LDAC.