Abstract: P114

Left ventricle involvement detected by cardiac MRI in arrhythmogenic right ventricular cardiomyopathy: a case series

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Background: arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) is an inherited myocardial disorder, with mutations in desmosomal protein genes, characterized by ventricular arrhythmias and predisposing to sudden cardiac death, particularly in young patients and athletes. Although structural abnormalities include loss of myocytes and fibro-fatty replacement predominate in right ventricle (RV), it is well recognized that left ventricle (LV) involvement is common, with biventricular involvement particularly in advanced disease. For this reason, LV involvement was originally considered only an end-stage complication of ARVC, based mostly on echocardiographic and pathology studies. The application of Cardiovascular Magnetic Resonance (CMR) in ARVC patients and family members rewrote the natural history of the disease. CMR is an ideal technique in ARVC workup, as it provides comprehensive information on cardiac morphology, function and tissue characterization in a single investigation. CMR evidence of LV involvement (fat infiltration, late gadolinium enhancement, LGE) is a strong independent predictor of cardiac events in patients with definite, borderline or possible ARVC diagnosis and has high negative predictive value for hard cardiac events.

Purpose: to present three different cases of ARVC with left ventricular involvement.

Methods: retrospective case series of three patients admitted to our Cardiac Magnetic Resonance (CMR) unit after ablation of sustained ventricular tachycardia with no previous history of cardiac disease. The first patient (Fig.1, case 1) shows biventricular LGE and right ventricle thrombus with one major CMR criteria for ARVC according to the 2010 modified ARVC Task Force criteria; the second patient (Fig. 1, case 2) has extensive subepicardial left ventricle LGE and fatty infiltration of the lateral wall with no CMR evidence of right ventricle involvement; the third patient (Fig. 1, case 3) has one minor CMR criteria for ARVC according to the 2010 modified ARVC Task Force criteria and left ventricle subepicardial and intramyocardial septal and inferior LGE. They all have genetic testing for inherited cardiomyopathies including analysis for known ARVC genes that were positive.

Conclusions: These cases confirm that ARVC may lead to an extreme phenotypic variability and it can present with early LV involvement, dominant or isolated LV disease, which can be easily detected on CMR. Left ventricle involvement is not included in 2010 modified ARVC Task Force criteria and it could cause CMR misdiagnosis of ARVC.
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