Valvular, Myocardial, Pericardial, Pulmonary, Congenital Heart Disease –
Myocardial Disease – Clinical: Arrhythmogenic Right Ventricular Cardiomyopathy

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Myocarditis and arrhythmogenic right ventricular cardiomyopathy: a diagnostic challenge

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Background: Current arrhythmogenic right ventricular cardiomyopathy (ARVC) diagnostic criteria are mostly based on ventricle function and dimension. Previous studies have reported a significant overlap between ARVC and chronic myocarditis, at non-invasive assessment.

Purpose: Tto compare biopsy-proven ARVC and myocarditis patients, in order to identify clinical, imaging and invasive electroanatomic voltage mapping (EVM) differences between the two groups.

Methods: Patients with borderline diagnosis of ARVC or suspected myocarditis underwent compete assessment with cardiac magnetic resonance (CMR). All patients underwent endomyocardial biopsy (EMB) with targeted tissue sampling guided by EVM. All patients with an histological diagnosis of myocarditis or ARVC were included.

Results: 83 patients were included, divided into 35 (42.2%) ARVC and 48 (57.8%) myocarditis. Among ARVC patients, 25 (71.4%) had right dominant ARVC, 5 (14.3%) left dominant patter and 5 (14.3%) bi-ventricular involvement. Nine patients (23.1%) with suspected clinic diagnosis of ARVC before EMB, received and histological diagnosis of myocarditis. Two (5.7%) patients with suspected myocarditis were proven to have ARVC. When comparing patients with ARVC and patients with myocarditis, univariate analysis showed that age, sex, family history, arrhythmic disorders at presentation and ECG abnormalities were similar between the two groups (P>0.05 for all the variables). There was also no significant difference with regards to bi-ventricular function and dimension at CMR evaluation. More patients with myocarditis resulted positive at late gadolinium enhancement (LGE) evaluation, although non-significantly (P=0.082). Oedema was more frequently present in patients with myocarditis (P=0.01), while adipose tissue infiltration and segmental wall motion abnormalities were more often observed in patients with ARVC (P=0.002 and P<0.001 respectively). At EVM analysis, a significant greater number of patients had a pathological uni- and bi-polar EVM (P<0.05 in all cases) and the scar-area was greater in patients with ARVC: 18.8 vs 11.0 cmq (P=0.041).

Conclusion(s): A significant number of patients who received a clinical diagnosis of Myocarditis or ARVC according to current guidelines, were subsequently reclassified after histological analysis. Patients with ARVC and myocarditis were not distinguishable on the basis of clinical features and ventricular function and dimensions. Conversely, tissue analysis with CMR demonstrated how patient with ARVC had less oedema, more adipose tissue infiltration and had more extensive scar at EVM evaluation.