Anti-desmoglein2 autoantibodies are present in patients with cardiac sarcoidosis and correlate with cardiac inflammation

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BACKGROUND Arrhythmogenic right ventricular cardiomyopathy (ARVC) has several phenocopies such as cardiac sarcoidosis (CS), idiopathic outflow tract ventricular tachycardia (OT-VT) and myocarditis. Differentiation between these entities can be challenging. Recently, we have identified diagnostic anti-desmoglein-2 autoantibodies (anti-DSG2 Abs) in patients with ARVC.

PURPOSE We sought to examine whether anti-DSG2 Abs are also present in clinical phenocopies of ARVC.

METHODS Anti-DSG2 Abs in sera of 25, 19 and 22 patients with sarcoidosis, OT-VT and myocarditis, respectively, were assessed by western blots and ELISA. Clinical and imaging parameters, as well as conventional biomarkers were correlated to detected anti-DSG2 Ab intensity levels.

RESULTS Anti-DSG2 Abs, at various intensities, were identified in 6/25 (24%) patients with sarcoidosis, all presenting with CS, but were absent in patients with OT-VT and myocarditis. Cardiac 18F- fluorodeoxyglucose positron emission tomography (18F-FDG PET) was positive in all sarcoidosis patients with positive anti-DSG2 Abs, corresponding to a median PET maximum standardized uptake value (SUVmax) of 5.65 [IQR: 5.15 – 10.9]. In sarcoidosis patients without anti-DSG2 Abs, the SUVmax values were significantly lower with a median of 0 [IQR: 0 – 4] (p = 0.011). The Pearson correlation coefficient (R) was 0.188 (p = 0.039) indicating a positive correlation between cardiac 18F-FDG uptake and anti-DSG2 Abs. No significant correlation was detected for any of the other clinical parameters and biomarkers.

CONCLUSIONS In addition to being present in ARVC, anti-DSG2 Abs are also found in CS, a common phenocopy of ARVC; conversely, anti-DSG2 Abs are absent in idiopathic OT-VT and myocarditis. Anti-DSG2 Ab levels positively correlate with myocardial disease activity in CS as indicated by cardiac 18F-FDG PET scanning.

Abstract Figure. Central illustration