Background: Cardiac involvement in systemic sclerosis (SSc) accounts for 26-36% of deaths. This most frequently manifests as ventricular rhythm disturbances (VRDs), eventually culminating in sudden cardiac death. However, no specific guidelines exist for implantation of cardioverter defibrillators (ICD) in SSc patients. Parametric cardiovascular magnetic resonance (CMR) indices of myocardial oedema and fibrosis like native T1/T2 mapping have been shown to be associated with prognosis in SSc patients with acute cardiac events and normal echocardiograms. However, their relationship with arrhythmogenicity per se has not been previously investigated in SSc.

Objectives: To investigate the relationship between parametric CMR indices and arrhythmogenicity in SSc patients.

Methods: We aimed to test the performance of the ‘old’ European Scleroderma Trials and Research Group (EUSTAR) Activity Index (old-AI) (1), the ‘new’ EUSTAR activity index (new-AI) (2), and the scleroderma activity index derived from the old-AI (Pecs-AI) (3). We compared the three indices to the disease activity based on the physician’s global assessment (PGA). We also assessed correlations with the change in modified Rodnan Skin Score (mRSS), FVC and CRP.

Methods: We evaluated 77 patients (50 diffuse /dSSc/ and 27 limited cutaneous SSc /lSSc/patients) from a single tertiary clinical center. Cohort enrichment was performed to increase the number of patients with early disease and dcSSc. Seventy-two patients were re-evaluated after one year. Nine patients had overlap syndromes: rheumatoid arthritis (n=3), Sjögren syndrome (n=2), polymyositis (n=2), and mixed connective tissue disease (n=2). The overall disease activity was evaluated using both the old and new indices (old-AI, Pecs-AI, new-AI) and the PGA of disease activity, based on the blinded evaluation of a single physician (LV). In addition to the minimal essential data from the EUSTAR database we also performed detailed assessment of the musculoskeletal involvement evaluating measures of hand function, DAS28 scores, and the Clinical Disease Activity Index (CDAI) (4).

Results: Three times more patients with active disease were identified by the new-AI compared to the old-AI at baseline investigation (n=37, 48.7%, vs. n=11, 14.3%). Two patients (18%) with active disease based on the old-AI were missed by the new-AI. Pecs-AI index identified 15 patients (19.5%) with active disease (cut-off >2.75 points). Active disease was equally frequent in dSSc and lSSc patients based on old-AI, but was more frequent in dSSc patients based on the new-AI in the whole cohort, and also after excluding overlap cases.

Patients with active disease based on the old-AI had more frequently rheumatoid factor (68% vs. 12/45, p=0.047), and DLCO<70% (11/11, vs. 36/65, p<0.01). Active disease based on the new-AI was associated with current cyclophosphamide treatment (9/37, vs. 2/39, p=0.023), and diabetes mellitus (7/30, vs. 0/39, p<0.01). The PGA correlated moderately at both baseline and one year follow-up examination with the old-AI (rho: 0.593, and rho: 0.589, respectively, p<0.001), the new-AI (rho: 0.429, and rho: 0.429, respectively, p<0.001), and the Pecs-AI (rho: 0.425, and rho: 0.583, respectively, p<0.001).

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