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Cardiac fibrosis and microvascular damage detected by cardiac mr are a hallmark of systemic sclerosis heart involvement

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Introduction

Systemic sclerosis (SSc) is an autoinmune disease characterized by tissue fibrosis and microvascular damage. Heart involvement has been described in 37-80% of patients; prevalence rates vary depending on the diagnostic method and the type of patients included in the studies.

Purpose

To determine the prevalence of heart involvement in a cohort of patients with SSc, to describe the patterns of heart involvement in these patients using cardiac MRI, and to correlate these data with disease subsets and target organ involvement.

Methods

We included SSc patients from the SSc cohort of our Institution. They underwent clinical evaluation, EKG, CPK, CPK-MB, CRP, ESR, specific SSc autoantibodies, coronary angiotomography (GE, 64 multidetectors) and cardiac MRI (1.5 T... GE, sequencies in T1 GRE Delay Enhancement to evaluate myocardial damage, resting and post-adenosine myocardial perfusion - TIGRE-echoplanar-imaging) was analyzed semiquantitatively with signal intensity curves.

Results

We have included 43 SSc patients (24 patients with diffuse cutaneous SSc (dcSSc): 56% and 19 patients with limited cutaneous SSc (lcSSc): 44%; 42 of them were female); mean age 41 years, mean time of evolution: 10.2 years; mean LVEF: 58.7%; 51% showed subendocardic perfusion defects; 54% showed myocardial fibrosis (10% with patchy distribution, 40% in bands, 15% subendocardic, 30% with mixed patterns and 5% transmural), with similar prevalence in dcSSc and lcSSc; percentage of cardiac fibrosis was significantly higher in dcSSc (7.6%) than in lcSSc (2.3%; p = 0.05); 90% showed diastolic dysfunction. Ninety two percent of coronary angiotomographies were normal (mean Ca score 3.7); 26.5% of patients had abnormal EKGs (50% of dcSSc vs 5.6% of lcSSc, p = 0.006). Cardiac fibrosis was directly associated with cardiac microvascular damage (p = 0.03), lower LVEF (62 vs 55%, p = 0.002) and inversely associated with vascular (41 vs 59%, p = 0.03) and muscular involvement (26 vs 73%, p = 0.05). Microvascular damage was associated with higher thickness skin scores (18 vs 11 points, p = 0.03). There was no association of cardiac fibrosis with abnormal calcium score. There were no other associations with clinical or laboratory values.

Conclusion

Patients with systemic sclerosis show preserved systolic function, high frequency of cardiac fibrosis, diastolic dysfunction and subendocardic concentric perfusion defects, related to microvascular damage. Cardiac MRI is a sensitive, noninvasive, useful method to detect heart involvement in SSc.