FRI0229

THE IMPACT OF SYSTEMIC SCLEROSIS ON BODY IMAGE PATIENTS

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Background: Satisfaction with body image has a major impact in quality of life. Systemic sclerosis (SSc) is a can result in disfiguring physical changes.

Objectives: Our aim was to determine the impact of systemic sclerosis on body image using the Satisfaction with Appearance Scale (SWAP). (1)

Methods: Cross-sectional study including patients satisfying the 2013 American College of Rheumatology criteria for SSc diagnosis, aged ≥ 18 years, treated in a tertiary Rheumatology Department. Demographic and clinical data were collected from Reuma.pt and clinical records. All patients provided informed consent and fulfilled SWAP questionnaire, which consists of 14 questions in 4 subscales: satisfaction with facial appearance, satisfaction with non-facial appearance, social discomfort due to appearance and perceived social impact of appearance. Patients rate each item on a numerical rating scale from 1 (strongly disagree) to 7 (strongly agree). Scores for the facial and non-facial appearance range from 0-24 and scores for the social discomfort and perceived social impact subscales range from 0-18. Total SWAP score can range from 0-84 and higher values indicate greater dissatisfaction with appearance and poorer body image. A descriptive analysis was used to summarize demographic and clinical data; categorical variables were described using frequencies; and continuous data using mean and standard deviation. Correlation between variables [Rodnan, age, disease duration, Hospital Anxiety and Depression Scale (HADS) and Short Form Health Survey (SF36)] and SWAP score was tested with Pearson or Spearman coefficient, as appropriated. Scores of SWAP and its subscales in preclinical, limited and diffuse forms of SSc were compared using ANOVA test. Analyses were performed with SPSS Statistics, V.21 and $p < 0.05$ was considered statistically significant.

Results: We enrolled 38 patients, 84.2% (n=32) female, with mean age 60.3 ± 14.5 years and mean disease duration 13.3 ± 6.5 years. All but one were caucasian. Fifty percent (n=19) had a limited form, 26.3% (n=10) had preclinical scleroderma and 23.7% (n=9) had a diffuse form of SSc. Regarding the autoantibody profile: 63.2% (n=24) had anti-centromere antibodies, 28.9% (n=11) had anti-Scl-70 antibodies, 5.3% (n=2) had anti-PM antibodies and 2.6% (n=1) had no positive antibodies. The median of Rodnan scores was 4 (IQR 0-9). The total mean SWAP score was 44.8 ± 12.5 with worse results at "Satisfaction with facial appearance" subscale (mean score 14.4 ± 6.1). There is no statistically significant difference in the SWAP score (or its subscales) between the three diagnosis subtypes. No statistically significant correlation was found between the total and subscale SWAP scores and any of the continuous variables considered and no statistically significant difference was found between the different forms of SSc.

Conclusion: We found no significant differences between preclinical, limited or diffused SS. SWAP scores were not significantly correlated with the total Rodnan score, age or disease duration. Contrary to our expectations SWAP did not show any relationship with depression, anxiety (HADS) or quality of Life (SF-36). However, our sample is too small to support definite conclusions. Further studies assessing body image in SSc and its impact in quality of life are warranted to support the holistic care of these patients.

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- [2] 10.1037/0278-6133.22.2.130;
- [3] 10.3899/jrheum.141482.

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THE 2009-2019 SURVIVAL AND MORTALITY PREDICTORS IN A LARGE MULTICENTRE SYSTEMIC SCLEROSIS COHORT

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Background: Systemic sclerosis (SSc) is one of the connective tissue diseases with the poorer prognosis and disease-related causes, particularly pulmonary fibrosis, PAH and cardiac involvement, accounting the most deaths.

Objectives: This multicentre study aimed to evaluate the global survival and any predictor of mortality in a large multicentric cohort of SSc patients.

Methods: We performed a retrospective analysis examining the medical records of our longitudinal SSc cohorts with a median (IQR) follow-up of 11 (6-18) years from 3 Scleroderma Units since January 2009. All clinical, laboratory and instrumental findings have been recorded and analyzed using Chi-squared tests, Kaplan-Meier curves, log-rank tests, and Cox proportional hazards models.

Results: Data from 750 SSc patients (91.9% female; mean (SD) age at first Non-Raynaud symptom 48.4 (15.3) years, median (IQR) disease duration 3 (0-8) years; diffuse cutaneous involvement 162 (21.6%) patients) fulfilling the 1980 ARA and/or 2013 ACR/EULAR classification criteria, were collected. All patients were positive for ANA, anti-Topo-I Abs were found in 235 (31.3%) and Cemp-B Abs in 300 (40%) patients. 98 (13.1%) patients were positive to other Abs (Anti-RNA polymerase III, anti-Pm/Scl) and anti-ENA were negative/unknown for 117 (15.6%) patients. Interstitial lung disease (ILD) was present in 202 (26.9%), pulmonary arterial hypertension (PAH) was found in 29 (3.9%), and 50/750 (6.7%) patients presented pulmonary hypertension combined with ILD (PH-ILD). The overall 10-years survival was 93.1% and, it was significantly impaired by the presence of ILD, PAH or PH-ILD [Figure]. The univariate analysis showed that female gender, higher age at first Non-Raynaud symptom, earlier referral to a tertiary Scleroderma center, absence of any ENA antibodies, and PH-ILD presence were survival predictors. After multivariate analysis the significance of PH-ILD was lost [Table]. Disease duration, basal Rodnan skin score, smoking, renal or gastrointestinal comorbidities, NYHA functional class, steroid or immune-suppressive treatments did not reach the statistically significance.

Conclusion: Our study demonstrated a global 10-years survival rate over 93%. Male patients and rapid evolution of Non-Raynaud symptoms represent the main death predictors in our SSc cohort. A rapid referral to a tertiary rheumatological centre and early treatment with effective agents are associated to a better prognosis.

Figure. Kaplan-Meier curves for 5-years survival in SSc patients (Log-rank 8.96, $p=0.03$).

Table. Prognostic factors for 10-years survival at univariate and multivariate analysis.

	UNIVARIATE ANALYSIS			MULTIVARIATE ANALYSIS		
	HR	95%IC	P	HR	95%IC	P
Female gender	0.35	0.15-0.81	0.01	0.31	0.15-0.66	0.002
Age at first Non-Raynaud symptom	1.07	1.04-1.1	0.001	1.08	1.05-1.11	0.001
Time referral to a tertiary SSc centre	0.83	0.76-0.92	0.001	0.84	0.77-0.93	0.001
Absence of any ENA antibodies	0.08	0.01-0.62	0.01	0.09	0.01-0.71	0.02
PH-ILD presence	2.6	1.01-6.82	0.04	2.4	0.93-6.1	0.069

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TREATMENT OF DIGITAL ULCERS IN PATIENTS WITH SYSTEMIC SCLEROSIS: PROSPECTIVE COHORT STUDY

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Background: Digital ulcer (DU) is a common clinical manifestation in patients with systemic sclerosis (SSc). About 70% of patients with SSc experience DUs during the first 10 years, which limit daily activities and may result in digital gangrene or amputation. Several vasoactive/vasodilating agents have been suggested for treatment, but few studies have compared the efficacy of those drugs.

Objectives: The objective of our study was to compare the efficacy of medical treatment for SSc related-DUs, focusing on endothelin receptor antagonist (ERA) and phosphodiesterase-5 inhibitors (PDE5inh).

Methods: In this prospective observational cohort study, we recruited patients who had one or more active SSc-related DUs and newly started or changed a medical treatment for SSc-related DUs from 13 medical centers in South Korea. The primary outcome was to compare the time to resolution of cardinal DU (CU) according to the treatments. The secondary outcomes included changes in the size or number of CU and changes in the number of DUs. CU was defined as the most clinically significant DU chosen by the investigators. Patients were followed up at every 4 weeks after enrollment until 12 weeks and finally at 24 weeks.

Results: Seventy-one patients were enrolled. Seven patients were excluded due to follow-up loss or withdrawal of consent. A total of 64 patients were analyzed. Seventy-eight percent (n=50) were female. The mean age at enrollment was 49.6 ± 11.6 year-old, and the mean disease duration was 7.1 ± 5.9 years. Twenty-eight patients (43.8%) were limited SSc. Forty-nine patients (n=76.6%) started ERA treatment (bosentan=49). Eleven patients (17.2%) started PDE5inh treatment (n=9 for sildenafil, n=1 for udenafil, and n=1 for tadalafil). Four patients who started medication other than ERA or PDE5inh classified as other treatment groups. Seventeen patients (26.6%) were on background calcium channel blockers (CCBs). CU healed in 25 patients (39.1%) at 12 weeks and 43 patients (67.2%) at 24 weeks. The mean time to heal CUs were 54.4 ± 22.7 days at 12 weeks and 91.6 ± 49.2 days at 24 weeks. Time to heal CU was comparable among patients on ERA, PDE5inh, and others (p=0.53, figure 1). The CU area was comparable among the three groups at baseline, 12, and 24 weeks. The mean area of CU in patients on ERA at baseline at 12, and 24 weeks was 21.3±19.4 mm², 8.2±14.6 mm², and 4.6±7.7 mm², respectively. The mean area of CU in patients with PDE5inh at baseline at 12, and 24 weeks was 26.2±28.1 mm², 3.5±3.6 mm², and 1.3±4.3 mm². New DUs developed in 4 patients (8.3%) in ERA, whereas 4 patients (40.0%) in PDE5inh at 4 weeks. The use of ERA was significantly associated with less new DUs development than the use of PDE5inh at 4 weeks follow-up (RR for developing new DU patients on ERA, 0.21; 95% CI 0.06-0.70; p=0.02). At 24 weeks follow-up, none of the patients on CCB developed new DUs.

Conclusion: The time to heal CU for ERA and PDE5inh users was comparable in the current study. ERA treatment was associated with reduced new DU occurrence compared with PDE5inh treatment. None of the patients with CCB treatment developed new DU development at 24 weeks.

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Figure 1

FRI0232

ALTERED FIBRIN CLOT PROPERTIES IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Vasculopathy in Systemic sclerosis (SSc) is connected with the activation of coagulation. However, the fibrinolytic activity still remains unclear since the most preliminary evidences are discordant (1, 2).

Objectives: To assess the haemostatic function, fibrin clot density and clot lysis time in SSc patients and healthy controls (HC) to determine their relation to disease findings.

Methods: Patients who fulfilled the 2013 ACR/EULAR SSc criteria and have never been treated with endothelin receptor antagonist, phosphodiesterase 5 inhibitors or prostanoids were eligible. Our study included 58 SSc patients [36 limited (lcSSc) and 22 diffuse cutaneous SSc (dcSSc)] and 46 sex/age-matched HC. Clinical evaluation of patients was performed, including high-resolution CT (HRCT), pulmonary function tests and the revised EUSTAR activity index. The interstitial lung disease (ILD) group (n = 15) was defined as moderate or severe changes on HRCT, with a forced vital capacity (FVC) < 85% predicted, without evidence of significant pulmonary arterial hypertension. The serum concentration of ICAM1 and von Willebrand factor antigen (VWF) were measured by ELISA. Haemostatic potential parameters; including overall haemostasis (OHP), overall coagulation (OCP) and overall fibrinolysis (OFF) potential, were assessed and endogenous thrombin potential (ETP) was determined. Maximum absorbance (Cmax), reflects the fibrin clot density and clot lysis time (Lys50t0), reflects fibrinolytic susceptibility, were calculated from OHP and OCP curves (3). Fibrin structure was visualised using scanning electron microscopy (SEM).

Results: The OFF value was significantly decreased, Lys50t0 prolonged (p<0.05), while OHP and ETP were increased (p<0.05) in patients. In dcSSc group ETP, OHP, Cmax and Lys50t0 were higher compared to HC (p<0.05). In SSc group, a positive association was found between coagulation parameters (OCP, OHP, Cmax) and the erythrocyte sedimentation rate (ESR), fibrinogen and ICAM1 (respectively p<0.05). Lys50t0 was positively correlated with ICAM1, ESR and VWF (respectively p<0.001, p<0.05, p<0.05). An inverse correlation was found between Cmax and both the diffusing capacity of the lungs for carbon monoxide (r=-0.408, p<0.01) and FVC (r=-0.318, p<0.01). Increased Cmax was found in ILD respect to HC (p<0.01). Denser plasma clot was associated with active disease (p<0.01). Longer Lys50t0 was observed in pitting scars group (p<0.01). Prolonged Lys50t0 was independently predicted by ICAM1 (OR 1.12, 95% CI 1.03-1.2, p<0.01).

Conclusion: Our results provide evidences of denser plasma fibrin clot among patients with lung involvement and impaired fibrinolysis, selectively presented among SSc patients with pitting scars. Thus, these patients might be at risk for thrombotic complications. Raised ICAM-1 levels could reflect impaired fibrinolysis, giving insight the important role of this molecule in endothelial homeostasis.

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