Methods: Twenty SSc-patients with indication for ILO-treatment (prophylactic or due to digital ulcerations (DU)) will be included in a prospective observational study. Patients will be offered to combine ILO with stellate blockade (ILOST).

Results: Patients were prospectively recruited between April 2008 and June 2019 based on the EUSTAR database and CRDC multi-center cohort from 154 patients.

Results: Compared to HC, women with SSc had significantly higher prevalence and greater severity of sexual dysfunction [FSFI total score (SSc: 18.4±3.3, HC: 25.5±2.5, p<0.0001) as well as in all subscales (p<0.0001 for all), BISF-W total score (SSc: 17.2±2.1, HC: 32.0±1.9, p<0.0001), dysfunction of pelvic floor (PIQoF-12 (SSc: 3.8±0.7, HC: 8.9±0.6, p<0.0001), PFIQ7 (SSc: 33.4±5.6, HC: 8.8±1.4, p<0.0001), and worsening sexual quality of life [SQoL-F (SSc: 55.3±3.3, HC: 82.1±2.1, p<0.0001)]. Men with SSc also reported more severe sexual dysfunction: [IIEF - erectile function (EF) (SSc: 15.4±3.0, HC: 26.2±2.1, p=0.004), IIEF - orgasmic function (SSc: 8.4±1.1, HC: 14.5±0.8, p=0.045), IIEF - intercourse satisfaction (SSc: 9.7±1.5, HC: 15.7±1.3, p=0.008)], MSFHD - erectile function (SSc: 9.0±1.2, HC: 12.9±0.9, p=0.005), MSHQ - Satisfaction (SSc: 19.2±2.1, HC: 26.6±0.9, p<0.001)], and worse sexual quality of life [SQoL-M (SSc: 68.5±7.4, HC: 86.7±6.2, p=0.023)].

Conclusions: Both women and men with SSc reported significantly impaired sexual function compared to HC with identical age. Worsen scores in SSc were associated with disease activity, increased systemic inflammation, health status, physical activity, fatigue and depression.

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Results: Antinuclear antibodies were detected in 2129 out of 2809 systemic sclerosis patients enrolled in the multi-center cohort and 4.2% of them were negative. There was significant difference between patients with negative and positive ANAs based on gender (253/50 vs 29/11746, p<0.001). The presence of Raynaud’s phenomenon is less common (71.8% vs 99.8%, p<0.001) in the ANA-negative patients. In addition, compared with ANA-positive patients, the incidence of certain critical organ involvements, including gastrointestinal reflux (5.6% vs 18.5%, p=0.002), intestinal lung disease (65.2% vs 73.9%, p=0.015) and pulmonary arterial hypertension (11.5% vs 29.2%, p=0.006) were significantly lower in ANA-negative patients than in the positive group. The proportion of IgG elevation, an indicator of disease activity and severity of inflammation, was significantly lower in the ANA-negative patients than that in the positive group (14.3% vs 41.2%, p<0.001), while no significant differences were found in other inflammatory indicators and skin scores.

Conclusion: This study describes the clinical features of SSC patients with negative ANAs, which have been rarely mentioned or focused in existing studies. Antinuclear antibody is proved to be strongly associated with the clinical manifestations of systemic sclerosis patients and ANA-negative SSC patients tend to be in relatively milder conditions, including a less common involvement of critical organs and a more temperate inflammatory severity.

References:

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**SEXUAL DYSFUNCTION IN FEMALE SCLERODERMA PATIENTS AND ITS CORRELATION WITH VASCULAR INVOLVEMENT**

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Background: Systemic Sclerosis (Scleroderma, SSC) is a autoimmune disease characterized by multiple organ dysfunction, which ultimately leads to multiple clinical and psychological complications. Among various complications of scleroderma, sexual dysfunction can be named as a major issue in both male and female patients, which has great impact on quality of life of the patients.

Objectives: Investigating the sexual dysfunction in scleroderma patients and its relation to their vascular involvements.

Methods: A case control study was done on 80 married female scleroderma patients with age between 20-60 years old. Eighty normal individuals adjusted for age, place of living and socioeconomic status were also recruited. Sexual performance in both groups was assessed using FSFI standardized questionnaire, which evaluated it in 6 domains of desire, arousal, lubrication, orgasm, satisfaction, and pain. Micro and macro-vascular involvements of the patients were also determined using Raynaud Condition Score, Echocardiography, physical exam for assessing their digital ulcers and reviewing their medical records for presence of past or present history of renal crisis and thromboembolic events.

Results: The total score of FSFI in the case group was significantly lower compared to control one (16.68 ± 6.35, 19.69 ± 6.01, P-value =0.001). The score was significantly lower in all domains of sexual dysfunction except for pain and lubrication. Moreover, the mean score of FSFI was also found to be significantly lower in limited form of the disease compared to diffuse one (14.6 ± 6.9, 18.1 ± 5.5, P-value 0.01). No significant association was found between vascular complications and sexual impairment of the scleroderma patients.

Conclusion: This study can be named as the first survey investigating the sexual dysfunction in Iranian female scleroderma patients and assessing its relation with vascular complication of the disease. Thus, it can be a guide for future studies on sexual dysfunction especially in societies with cultural limitations in discussing this issue.

References:

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**OUTCOME OF INTERSTITIAL LUNG DISEASE (ILD) IN ANTI-PM/SCL PATIENTS WITH SYSTEMIC SCLEROSIS: RESULTS FROM AN EUSTAR CASE-CONTROL STUDY.**

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Background: The main clinical associations of anti-PM/Scl in Systemic Sclerosis (SSc) so far reported include calcinosis, myositis and interstitial lung disease (ILD). Nevertheless, data regarding the long-term outcome of ILD in these patients are lacking. A single centre Spanish cohort reported a better functional outcome in 14 SSc-ILD patients anti-Pm/Scl+ as compared to 49 anti-Topo I after a mean follow-up of 7 years (1).

Objectives: To analyze the long-term outcome of ILD in a large multicentre EUSTAR study dedicated to anti-Pm/Scl SSc patients.

Methods: A case-control study within the EUSTAR cohort collected 165 anti-PM/Scl- SSc cases and 257 anti-PM/Scl+ SSc controls, matched for sex, cutaneous subset, disease duration, and age at onset. Data for ILD at HRCT were available for 162/165 cases and 249/257 controls. Data for pulmonary function tests (PFT) at the baseline (T0), 1 year after diagnosis (T1) and at the last visit (LV) were analyzed.

Results: A significantly higher frequency of ILD was reported in anti-PM/Scl+ cases vs anti-PM/Scl- controls (62.3% vs 39.4%, p<0.0001, OR 95%, CI 2.55, 1.70-3.83). Complete PFTs data were available for 81/101 ILD anti-PM/Scl+ cases and 78/98 anti-PM/Scl- ILD controls, with similar age at onset and female/male ratio and disease duration at LV (112±81 months vs. 115±64 months, p=0.77). Diffuse cutaneous involvement was less frequent in cases than in controls (272% vs. 44.9%, p=0.03).

In ILD cases, %pFVC tended to improve from T0 (85.1±18.3) to T1 (89.5±16.5, p=0.045) and to LV (87.9±16.9, p=0.057), while in ILD controls remained stable from T0 (90.4±18.5) to T1 (91.1±16.5, p=0.38) and significantly declined to LV (85.0±18.0, p=0.0002). %pDLCO remained stable from T0 (60.5±18.6) to T1 (60.1±17.6, p=0.87) and to LV (60.4±18.6, p=0.77) in ILD cases, while significantly declined from T0 (67.0±18.9) to T1 (62.7±18.2, p=0.0016) and to LV (59.6±18.4, p=0.0001) in the control group. Mean %pFVC and %pDLCO at the 3 time points were not significantly different between the two groups.

Delta %pFVC (LV-T0) was 2.85±11.3 for the anti-Pm/Scl+ group vs -5.42±13.4 in the control group (p=0.0004) with a significant smaller proportion of patients with FVC loss ≥10% from T0 to LV in the anti-Pm/Scl+ group (12.3% vs. 39.7%, p=0.001). Delta %pDLCO (LV-T0) was -0.13±10.8 for the anti-Pm/Scl+ group vs -7.36±10.8 in the control group (p=0.0015), with a significant smaller proportion of patients with DLCO loss ≥10% from T0 to LV in the anti-Pm/Scl+ group (13.6% vs. 42.3%, p=0.0001).

Conclusion: In this multicenter real-life study, the long-term pulmonary functional outcome in SSc-ILD patients with anti-Pm/Scl positivity seems to be more favorable than in patients without anti-Pm/Scl antibodies.