AB0638

ABSENCE OF LUNG INVOLVEMENT IN SYSTEMIC SCLEROSIS PATIENTS WITH SILICONE IMPLANTS

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Background: Although many cases have been reported for the last 3 decades, the correlation between silicone breast implants (SBI) and systemic sclerosis (SSc) is yet unclear.

Objectives: To assess the prevalence of breast implants in a well characterized SSc cohort followed at a dedicated tertiary center and to define characteristics that might differ between SSc patients with implants and those without.

Methods: A prospectively maintained database was interrogated regarding the prevalence of breast implants and the disease characteristics. Female patients aged 20 to 55y (215 patients) followed at our center between 2003-2018 were included. SBI cases were identified and their characteristics tabulated. Each SBI patient was matched with 2 controls matched for age, disease duration and type of disease. We used descriptive statistics, multivariate comparisons and multivariate analysis for correlations.

Results: 11 pts with SBI were found, 6 with diffuse (median age 37.5, range 32-55, median disease duration 3.5 y, range 1.5-8y, median mRSS 13.5, range 8-26) and 5 with limited SSC (median age 38, range 33-45, median disease duration 8y, range 3-22y, mRSS 2, range 2-5). The prevalence of SBI in the 20 to 55 y cohort is 5%, similar to the estimated prevalence in the general population. All had their implants instilled before the diagnosis of SSC (median 10y, range 4-13y). Four pts were first diagnosed with SSC within a year from diagnosis of silicon leakage. RNA polymerase 3 was positive in 3 patients, Scl70 in 3 others, anticentromere in 1 patient and 4 others were positive only for antinuclear antibody. There were no differences in the prevalence of digital ulcers, gastrointestinal involvement, skin score or the serologic profiles between the two groups. However, none of the SBI pts developed interstitial lung involvement, compared to 72% of the matched diffuse patients (p=0.001). There were neither renal crises in the SBI group (compared to 5 cases in the matched group) nor cardiac involvement.

Conclusion: The temporal relation and the different clinical features might generate a theory that silicone might induce a particular type of SSC, as the idea that silicone protects against lung involvement seems unreasonable. We plan a further prospective study.

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AB0639

PREDICTION OF MAJOR VASCULAR COMPLICATIONS IN SYSTEMIC SCLEROSIS (SSc)

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Background: in SSc, Digital ulcers (DU), scleroderma renal crisis (SRC) and pulmonary arterial hypertension (PAH) are major vascular complications (MVCs). Among the variables used to follow-up SSc patients, nail-fold videocapillaroscopy pattern (NVCp), estimated pulmonary systolic arterial pressure (sPAP), diffusion capacity of carbon oxide (DLco) and renal resistive index (RRI) on renal vascular Doppler ultrasound have previously shown to be predictive for MVCs development. Moreover, circulating endothelin-1 (ET1) levels have been associated with MVCs.

Objectives: to identify clinical, instrumental and laboratory predictors of MVCs development in SSc.

Methods: SSc patients fulfilling the 2013 ACR/EULAR criteria were enrolled from two SSc-care units. Data regarding clinical manifestations, instrumental and laboratory evaluation for renal, cardiac and cardiovascular involvement were collected. ET1 serum levels were measured with human ET-1 ELISA kit.

Results: 380 patients [aged 57 (46-68) years, 12% males] were enrolled in the study, with 42.4% prevalence of MVCs at baseline. Previously known associations were significantly confirmed between presence of MVCs and NVCp, sPAP, DLco, RRI. The levels of ET1 were increased in the SSc MVCs without reaching a statistical significance.

In the whole population, modified Rodnan skin score (mRSS) [OR 1.081 (95% CI 1.032-1.132), p=0.001], presence of telangiectasias [OR 2.246 (95% CI 1.201-4.201), p=0.11] and presence of dyspnoea [OR 2.100 (95%CI 1.029-4.286), p=0.041] were predictive for MVCs at baseline on

multivariate regression analysis. During a 3-years follow-up, 21 patients developed a new MVCs and sPAP was the only independent predictor [HR 1.057 (1.016-1.100), p=0.006] at multivariate analysis. When evaluating only MVCs naive patients (n=179), 15 new MVCs developed during the follow-up and joint involvement was the only independent predictors [HR 8.355 (1.741-44.233), p=0.037].

Conclusion: The data show that the most stringent factors predicting MVCs are sPAP and joint involvement in MVCs complicated and naive patients, respectively. These 2 items, together with mRSS and telangiectasias, should always be considered in SSc long-term follow up to shape up a tailored therapeutic approach.

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AB0640

SURVIVAL AND MORTALITY PREDICTORS IN SYSTEMIC SCLEROSIS: RESULTS FROM A MONOCENTRIC ITALIAN COHORT

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Background: Systemic sclerosis (SSc) is a connective tissue disease with a poor prognosis and disease-related pulmonary fibrosis, pulmonary arterial hypertension (PAH) and cardiac involvement accounted for most deaths (1). However, a decrease in mortality is seen in the last decades thanks to the opportunity of new screening and therapeutic strategies (2).

Objectives: To estimate the global survival and any predictor of mortality in a monocentric cohort of SSc patients with a 10-years follow-up.

Methods: We performed a retrospective analysis examining the medical records of our longitudinal SSc cohort with a median (IQR) follow-up of 56 (25-82.5) months in our Scleroderma Unit 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 modeling.

Results: We collected data from 347 SSc patients (female n. 319 (91.9%); mean (SD) age at diagnosis 50.6 (15.9) years, median (IQR) disease duration 8.75 (4.1-14.9) years; diffuse cutaneous involvement (dsSSc) n.57 (16.4%) patients) fulfilling the 1980 ARA and/or 2013 ACR/EULAR classification criteria. All patients were positive for ANA, while anti-Topo-I were found in 145 (41.8%), CENP-B in 139 (40.1%) patients. Fifteen (4.3%) patients were positive to other autoantibodies (Anti-RNA polymerase III, anti-Pm/Scl) while anti-ENA were negative or unknown for 48 (13.8%) SSc patients. Interstitial lung disease (ILD) was present in 158 (45.5%), pulmonary arterial hypertension, diagnosed by right heart catheterism, (Group 1 PAH) was found in 17 (4.9%), and 22/347 (6.3%) patients presented pulmonary hypertension combined with ILD (Group 3 PH-ILD). The overall survival rates were 89.1% and 87.3% at 5 and 10 years, respectively. As reported in Figure 1, the 5-years global survival was significantly impaired by the presence of PAH or PH-ILD, while isolated ILD did not impact the survival. The univariate and multivariate analysis showed that dsSSc, higher age at diagnosis, delayed referral to our Scleroderma Unit, body mass index and absence of heart (H) or lung (L) involvement (No H/L) were independent predictors of 5-years mortality (Table I). While gender, disease duration, specific autoantibodies, basal Rodnan skin score, smoking, renal or gastrointestinal comorbidities, NYHA functional class, steroid or immune-suppressive treatments did not reach the statistical significance.

Conclusion: Our study demonstrated a global 10-years survival rate over 85%, and the presence of PH with or without ILD represents the main negative predictor. The rapid referral to a specialized centre for SSc and early treatment with effective agents for PH could further improve prognosis.

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Abstract AB0640 Figure 1. Kaplan-Meier curves for 5-years survival in SSc patients (Log-rank 33.40, $p < 0.001$). Abbreviations: No H/L: absence of heart or lung involvement; ILD: interstitial lung disease; PH-ILD: Pulmonary hypertension associated to ILD; PAH: pulmonary arterial hypertension.

Table I. Statistically significant prognostic factors for 5-years mortality at univariate and multivariate analysis (Cox proportional hazards modelling).

	UNIVARIATE ANALYSIS			MULTIVARIATE ANALYSIS		
	HR	95%CI	P	HR	95%CI	P
dcSSc	3.45	1.33-8.95	0.01	4.02	1.86-8.71	0.0001
Age at diagnosis	2.63	1.01-6.75	0.04	2.71	1.12-6.61	0.03
Delay in referral	2.64	1.00-6.84	0.05	2.73	1.11-6.7	0.03
BMI	0.90	0.82-0.99	0.03	0.91	0.84-0.98	0.02
No H/L Involvement	0.45	0.21-0.97	0.04	0.17	0.06-0.5	0.001

Abbreviations: dcSSc: diffuse cutaneous involvement; BMI: body mass index; H/L: heart or lung involvement; HR: hazard risk.

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AB0641 VITAMIN D SERUM LEVELS AND THE RISK OF DIGITAL ULCERS IN SYSTEMIC SCLEROSIS: A LONGITUDINAL STUDY

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Background: Vitamin D is known to influence the immune system in many different diseases although longitudinal studies on systemic sclerosis (SSc) are still missing.

Objectives: this study wants to investigate if variations in vitamin D serum levels (25OHD) over time affect digital ulcers (DU) in SSc.

Methods: this is a retrospective study on 65 patients. Data on disease characteristics and 25OHD were collected in 2011 and 2016.

Results: The mean age of our cohort was 58 (12) years with a mean disease duration of 9.5 (5.3) years. Most of our patients had a limited subset (69.2%). At baseline 50.8% and 41.5% after 5 years had 25OHD < 30 ng/mL. Supplemented patients (8750 IU/week) at baseline were 39 (60.0%) and 45 (69.2%) at the end of follow up. Nevertheless, 31 (47.7%) had a decrease of 25OHD in 5 years. In univariate analysis, patients with incident digital ulcers (DU) had a decrease in 25OHD as compared to patients with no incident DU (-17.4 (37.0) vs. 13.0 (89.5), $p = 0.018$). No differences in 25OHD variations were found for other disease characteristics. In multivariate analysis correcting for previous DU and mRSS at baseline, patients with a decrease in 25OHD had an increased risk of developing DU (OR (IC95%): 16.6 (1.7 to 164.5), $p = 0.017$).

Conclusion: In this study we have shown that a decrease in 25OHD increases the risk of developing DUs and that vitamin D supplementation with the doses currently recommended may be insufficient in SSc. Further studies in wider cohorts are needed to confirm these results and to evaluate the effectiveness of a more aggressive or different vitamin D supplementation.

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AB0642 DESCRIPTION OF SAE1/2 ANTIBODY IN A DERMATOMYOSITIS COHORT

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Background: Myositis specific antibodies have gained special importance in last years. Its knowledge has allowed stratifying patients in different clinical phenotypes, predicting with greater accuracy prognosis and establishing a clinical attitude to follow. SAE1/2 antibody (anti-small ubiquitin-like modifier activating enzyme) was first described in 2007 in patients with amyopathic dermatomyositis with cutaneous and digestive involvement. Its prevalence ranges from 8% in European cohorts to 3% in Asians.

Objectives: Describing myositis specific antibodies (MSA) in a cohort of inflammatory myopathies. To characterize the clinical phenotype of SAE1/2 antibody and to compare it with the rest of MSA.

Methods: Patients diagnosed of dermatomyositis in a tertiary hospital from 1978-2018, according to the criteria of Bohan and Peter (1975) and according to Dalakas classification criteria (2015). Clinical and analytical data, including the immunological profile were collected, as well as the treatments received and the evolution of the disease.

Results: Out of 46, 41 dermatomyositis had positive antinuclear antibodies (ANA). 55% percent had ANA titles ≥ 640 , being the fine speckled pattern the most frequent (43%) followed by coarse speckled and homogeneous (13.6% each).

72% had MSA, the most frequent being antiJo1 (27.3%) followed by MDA5 (18.2%) and SAE1/2 (15%). Up to 40% had two or more antibodies, being the association with antibodies Ro52 and Ro60, the most frequent.

5 patients presented positivity against SAE antibody. In comparison to the rest of MSA, 80% presented with cutaneous debut ($p = 0.00$), being the most frequent manifestations heliotrope erythema ($p = 0.00$), Gottron papules ($p = 0.10$) and skin rash ($p = 0.00$). 60% had pathological capillaroscopy compared to 15% ($p = 0.00$). Muscular balance was preserved in 60% of patients. Sixty percent had dysphagia vs 9% ($p = 0.00$). Two of them had lung involvement (alveolar hemorrhage and rapidly progressive pneumonitis). In comparison with SAE negative group, patients presented more pulmonary hypertension (44.5vs34 mmHg), without reaching significant differences. No significant differences were found between muscle enzyme levels' neither acute phase reactants.

As complications, one patient presented a myocarditis with quickly rapidly progressive pneumonitis that required high doses of corticotherapy and another one alveolar hemorrhage, which was treated the same way. All patients required corticotherapy at doses of mg/kg, requiring two of them to be treated with DMARDs (methotrexate and dolquine) and one of them with immunoglobulins due to cutaneous involvement.

Mortality rate was 40% due to rapidly progressive lung affection and cardiorespiratory arrest.

Conclusion: Higher prevalence of SAE1/2 antibody in our cohort may be explained due to the use of amplified myositis antibodies kit that specifically includes this antibody. These patients typically present with cutaneous involvement and dysphagia but also lung affection as a complication. Studies with larger samples should be performed in order to know the prognosis of this antibody specificity.

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