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, antinuclear antibody 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.

Disclosure of Interests: None declared

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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/ScI) 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.

REFERENCES

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