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SMOKING HISTORY AND SCLERODERMA CLINICAL TRIALS CONSORTIUM DAMAGE INDEX (SCTC-DI) AS INDEPENDENT PREDICTORS OF MORTALITY IN SYSTEMIC SCLEROSIS IN A SINGLE-CENTRE ITALIAN COHORT

M. Breda, M. G. Lazzaroni, F. Franceschini, P. Airò. 1ASST Spedali Civili, 2MD Anderson Cancer Center, 3Division of Pulmonary Medicine, Mayo Clinic, Rochester, United States of America
4Heart Center Kantonsspital Luzern, Department of Cardiology, Lucerne, Switzerland; 5Mayo Clinic, Department of Medicine, Division of Rheumatology, Rochester, United States of America; 6Centre de Référence Maladies Neuro-Musculaires, Assistance Publique-Hôpitaux de Paris, Groupe Hospitalier Pitié-Salpêtrière, DHU RMN, Paris, France; 7Clinique médicale, St-Mandé, France; 8Centre de Référence Maladies Neuro-Musculaires, Assistance Publique-Hôpitaux de Paris, Groupe Hospitalier Pitié-Salpêtrière, DHU RMN, Paris, France; 9 Assistance Publique-Hôpitaux de Paris, Hôpital Avicenne, INSERM U1272, Université Sorbonne Paris Nord, Service de Pneumologie, Bobigny, France

Background: Autoantibodies permit to classify and subgroup connective tissue diseases (CTD) in homogeneous groups of patients in terms of phenotype and prognosis. Anti PM-ScI antibodies have been associated with different CTD categories such as: idiopathic inflammatory myositis (IIM), systemic sclerosis (SSc), Sjögren's syndrome (SS), systemic lupus erythematosus (SLE), mixed connective tissue disease (MCTD) or unclassified connective tissue disease (UCTD).

Objectives: To determine clinical spectrum of anti-PM-ScI associated disease and if it an homogenous condition.

Methods: This multicentric (four hospitals) observational and retrospective study included all consecutive patients with positive testing for anti-PM-ScI antibodies on immunoblot assay and connective tissue disease (2011-2020). Epidemiological, biological, clinical and radiological data were collected in standard form as well as patient's outcome.

Results: One hundred twenty height patients (female n=96;75%) were included. Median (quartiles) age at diagnosis was 50 (18;84) (IQR) and follow-up duration of 7 (3.75-12) years. Seventy-six (59.3%) patients were simple anti-PM-ScI positive and 47% were associated with other antibodies: anti-SSA/Ro52 (n=13; 10.92%), SSC associated antibodies (n=21; 16.4%), anti-dsDNA for (n=9; 7%), anti-RNP (n=6; 4.7%) and anti-CCP antibodies (n=6; 4.7%). Most patients had cutaneous involvement (n=106; 83%) with skin thickening (n=47; 36%), mechanics hands (n= 28; 22%), calcinosis (n=26; 20.3%) and subcutaneous edema (n=20; 15.62%). Vascular involvement was frequent with Raynaud phenomenon (n= 89; 69%), telangectasia (n=36; 28%), skin ulcers (n=27; 21%), pulmonary hypertension (n=8126; 6.7%) and scleroderma renal crisis (n=2; 1.5%). A majority of patients also displayed an interstitial lung disease (ILD) (n=83; 65.8%); nonspecific interstitial pneumonia (92.7% and/or organizing pneumonia (25.3%). ILD was characterized by a subacute onset in 3781 (45.7%); median (quartiles) forced vital capacity (FVC) and total lung capacity (TLC) at diagnosis of 88% [73-105] and 79.5% [68.5-101] respectively. Sixty patients (47%) had muscular sign including myalgia (47%), elevated CPK capacity (TLC) at diagnosis of 88% [73-105] and 79.5% [68.5-101] respectively. Anti-cardiolipin antibodies were found in 37/81 (45.7%); median [quartiles] forced vital capacity (FVC) and total lung capacity (TLC) at diagnosis of 88% [73-105] and 79.5% [68.5-101] respectively.

In the Cox-regression analysis smoking history and severe organ damage at diagnosis were independent predictors of mortality both in the Australian derivation cohort and in the Canadian validation cohort (1). Several independent predictors of mortality in SSC have been reported, but only limited data are available on the role of smoking.

Objectives: To evaluate smoking history and SCTC-DI as independent predictors of mortality in SSC in a single centre Italian cohort.

Methods: A retrospective analysis was performed on patients prospectively followed in our centre from 1989 to 2019, with at least 2 evaluations and/or cause of death available. Organ damage was evaluated through the SCTC-DI (0-55 scale; severe damage=12), while comorbidities through the Charlson Comorbidity Index (CCI). Survival analysis was performed with Kaplan-Meier curves and with Log-rank test to compare different subsets. Cox-regression analysis was performed to identify baseline independent predictors of mortality.

Results: 648 SSc patients were 99% Caucasian, 90% female and had a median age at diagnosis of 55.5 years (IQR: 45.0-65.6); 19% had diffuse cutaneous involvement. ACA was positive in 52%; anti-Topol in 22% and anti-RNA Polymerase III in 5%.

Median SCTC-DI at diagnosis was 2 (0-4) (n=560); ever smokers were 27%. During the follow-up 240 patients died after a median period of 10.2 years (4.9-16.8). The cause of death was related to SSC in 41% (n=65; most frequent causes pulmonary arterial hypertension (n=35) and interstitial lung disease (n=30) and to other diseases in 40% (n=95; most frequent cause cancer (n=40)), while was undetermined in 19%.

Overall survival at 5, 10, 15, 20 years was 87.8% (SE 1.3%), 75.6% (1.9), 63.8% (2.3), 47.7% (2.8), respectively and was higher in females vs. males (p<0.001) and in ACA vs ACA- (p=0.05), but did not differ between cutaneous subsets. 111 patients were lost at follow-up (17%).

In the Cox-regression analysis smoking history and severe organ damage at diagnosis were independent predictors of death, while age at diagnosis, female gender, diffuse cutaneous subsets, ACA positivity and CCI were not (Table 1).

Conclusion: Smoking history and severe organ damage were independent predictors of death in a large Italian single centre SSC cohort. SCTC-DI was confirmed as a useful tool to predict mortality at every timepoint.

REFERENCES:

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