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Background: Hydroxychloroquine (HCQ) is a well-tolerated drug that contributes to downregulating the immune response against autoantigens and it has been used in several autoimmune diseases. In systemic sclerosis (SSc) it is used to treat inflammatory arthritis without proof of efficacy.

Objectives: Our aim was to evaluate the use of HCQ and its impact on Health Assessment Questionnaire disability index (HAQ-DI) and the Cochin Hand Function Status (CHFS). In a large SSc cohort compared to a propensity matched group of SSc patients not using HCQ.

Methods: SSc patients from the European Scleroderma Trials and Research (EUSTAR) data base treated with HCQ for at least 6 months were evaluated. Demographic and clinical data, concomitant drugs, duration of HCQ treatment and reasons for its discontinuation, HAQ-DI and CHFS (at least 2 evaluation) were recorded and were the outcome variables of interest. Statistical analysis was performed using propensity score matching for age, gender, disease duration, corticosteroids, immunosuppressives, vasoactive drugs, DMARDs in a 3:1 control:HCQ ratio. Standard descriptive statistics and Student's t-test and Chi-square test were used to assess the propensity-matched groups.

Results: 1,636 of 17,805 SSc patients (9.2%) were treated with HCQ for at least 6 months; out of these 3% (50/1636) had at least a baseline and follow-up HAQ-DI evaluation, (and 44/1636 (2.7%) had at least a baseline and follow-up CHFS evaluation. Propensity matching assured that pts were matched for demographic variables such as gender (mean on HCQ vs no HCQ:females:92.0 vs 85.3), age(49.8 vs 49.97yrs) disease duration(8.3 vs 9.1 yrs), limited disease(55.3 vs 62.6%) as well as background medications (P>0.1-0.9). We did not find any significant changes in HAQ or CHFS (difference in slope) over 365 days of treatment, comparing the HCQ-treated group to the non-HCQ treated patients (p=0.240 for both (Figure 1).

Conclusion: Results from the EUSTAR registry showed that HCQ was used by 9.2% of SSc patients. HCQ use did not improve the HAQ or CHFS, comparing HCQ users to non-HCQ users.

Figure 1. HAQ trend and CHFS trend over the time

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POS0322

CORRELATION BETWEEN QUANTITATIVE COMPUTED TOMOGRAPHY AND DISEASE ACTIVITY IN SYSTEMIC SCLEROSIS

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Background: High Resolution Computed Tomography (HRCT) is the gold standard to evaluate Interstitial Lung Diseases (ILDs) extent and severity. Quantitative Computed Tomography (QCT) is a promising tool as it provides an operator-independent assessment of ILD extent. Even if there are emerging data on QCT in Systemic Sclerosis (SSc), its correlation with disease activity (DA) has not been yet studied.

Objectives: To evaluate the correlation between QCT score and DA in an Italian cohort of SSc patients.

Methods: A multicentric, observational study was conducted in three Italian rheumatological centers.

Adult SSc patients classified according to the ACR/EULAR 2013 criteria [1] were assessed with pulmonary function tests, HRCT and for DA. CT images were analyzed quantitatively with the densitometric radiomic data obtained through a free open software – Mean lung attenuation (MLA), Standard Deviation (SD), Kurtosis, Skewness and Lung volume. DA assessment was conducted according to EUSTAR index [2]: a score ≥ 2.5 was considered indicative of high disease activity.

Age below 18 and pregnancy were considered exclusion criteria. We used Student's T test to evaluate the means of the parameters, Pearson's r test for correlations, receiver operating characteristics curve to define the cutoff values of the significant details, and linear regression with collinearity test to define the role of the details. P value <0.05 was considered statistically significant.

Results: Sixty patients were enrolled (male 8, female 52), with mean age 53.2 years (SD 15.6) and mean disease duration 5.3 years (SD 4.2). QCT indexes distribution was different in high DA vs low DA SSc patients. In particular mean lung attenuation (MLA, -834.7 vs -812.1, p = 0.03), standard deviation (95.9 vs 102, p = 0.03), skewness (2.2 vs 1.7, P = 0.006) and kurtosis (5.5 vs 3.3, p = 0.009) of the parenchymal lung and skewness (3.1 vs 2.8, p = 0.03) of the whole lung were statistically different. DA correlates with MLA (r = 0.28, p = 0.003), standard deviation (r = 0.21, p = 0.02), skewness (r = -0.32, p = 0.001) and kurtosis (r = -0.29, p = 0.001) of the parenchymal lung and MLA (r = 0.25, p = 0.006), skewness (R = -0.27, p = 0.003), kurtosis (r = -0.21, P = 0.02) of the whole lung. The skewness of the parenchymal lung was the QCT index with the best performance in identifying high DA SSc patients (cutoff value ≤ 1.85 ; area under the curve 0.74, p = 0.005; sensitivity 79.5%, specificity 68.7% accuracy 76.6%).

Conclusion: To our knowledge, this is the first study which correlate the QCT score with DA in SSc patients. Our results suggest that QCT can identify SSc patients with high DA score. This could open a scenario of new applications as an operator-independent contribution in DA scores with a potential role in clinical practice. Further studies are needed to confirm the data and to better identify the most suitable parameters for the purpose.

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POS0323

ANTI PM-SCL ASSOCIATED AUTO IMMUNE DISEASES: MULTICENTRIC COHORT OF 128 PATIENTS

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

Objectives: To determine clinical spectrum of anti-PM-Scl 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-Scl 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-Scl positive, and 40.7% 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%), telangiectasia (n=36; 28%), skin ulcers (n=27; 21%), pulmonary hypertension (n=8/120; 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 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. Sixty patients (47%) had muscular sign including myalgia (47%), elevated CPK (n=51; 40%) and muscular weakness (Medical Research Council score <4) (n=19/124;15%). Finally, fifty-three (41.7%) had gastroesophageal reflux. Thirty-nine patients (30.4%) experienced at least one muscular or ILD relapse and 6 (4.84%) died during follow-up (2 breast cancer, 1 pneumonia, 3 unknown etiology). Concerning patients' prognosis, relapses were associated with skeletal (n=29, 74.4% vs n=32, 35.96%, p < 0.001) or cardiac muscle involvement (n=7, 18.4% vs n=2, 2.5%, p=0.007), and subacute ILD (n=19, 65.5% vs n=18, 34.62%, p= 0.05) with organized pneumonia pattern (n=11, 32.3% vs n=10, 13.9%, p=0.05). Strikingly, ILD occurred mainly in men (90.6% vs 57.2%, p < 0.001) and was associated with anti-Scl-70 positivity (n=14, 16.67% vs 0%, p= 0.01). Muscle involvement was associated arthralgia (n=46, 76.67% vs n=34, 50.75%, p=0.005), respiratory signs at diagnosis: dyspnea NYHA ≥3 (n=46, 75.41% vs n=30, 44.78%, p < 0.001), sub-acute ILD (n=24, 61.54% vs n=13, 30.95%, p=0.011) with lower FVC (73% [64;88] vs 98 [76;105], p < 0.001). Ulcers were associated with Anti-Scl70 positivity (n=9, 33.33% vs n=5, 4.95%, p < 0.001), Raynaud phenomenon (n=27, 100% vs n=62, 61.39%, p < 0.001), digestive involvement (n=20, 74.07% vs n=34, 33.66%, p < 0.001), ILD with chronic onset (n=15, 78.95% vs n=29, 46.77%, p=0.027) and increased incidence of deaths (n=4, 16% vs n=2, 2.02%, p= 0.01).

Conclusion: Conducted on the largest cohort of Anti-PM-Scl patients, this study highlights two main phenotypes that determine different outcome and prognosis. One was associated with muscular disease and subacute onset ILD with more frequent relapses. The second with a vascular phenotype associated with chronic ILD, digestive involvement, chronic evolution and increased incidence of death. This could lead to a reclassification of PM-Scl associated auto immune diseases.

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POS0324

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

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Background: Systemic Sclerosis (SSc) is characterized by increased mortality and organ damage accrual. A composite SSc Damage Index was recently developed by the Scleroderma Clinical Trials Consortium (SCTC-DI) and was demonstrated as a predictor 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 indeterminate 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<0.0001) and in ACA+ vs ACA- (p=0.005), 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 (score>12) were identified as 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

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Characteristics	P-value	Hazard ratio	Confidence interval 95%
Female gender	0.9726	0.992	0.619-1.590
Diffuse cutaneous subset	0.7407	1.036	0.729-1.366
Ever smoker	0.0005	1.545	1.211-1.972
SCTC-DI at diagnosis >12 (severe damage)	0.0353	3.450	1.089-10.929
ACA positivity	0.7407	1.046	0.801-1.366
Age at diagnosis	0.4961	1.005	0.991-1.018
Charlson Comorbidity Index (age excluded)	0.1555	1.094	0.966-1.238

Table 1. Cox-regression analysis for any cause of death.

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POS0325

RADIOMIC BIOMARKER OF PULMONARY VASCULAR RELATED STRUCTURES PREDICTS MORTALITY IN SYSTEMIC SCLEROSIS

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Background: Quantitative computed tomography (QCT) extracts features from high-resolution CT scans and quantifies lung parenchymal and vascular abnormalities which may not be discernable by qualitative review. The threshold values