the associations with joint and muscle involvement, lung fibrosis, and intestinal symptoms were confirmed (table 1).

Table 1 Results of the univariable and multivariable analysis adjusted on sex, age at disease onset and disease duration (n=8142 patients). Results are presented as number/number available data (%) unless stated otherwise.

Conclusions: In the largest series of anti-PM-Scl positive patients so far reported, well-known clinical associations were confirmed. Moreover, scleroderma renal crisis was more frequent than in the antibody-negative patient controls (which included a majority of anticientromere-positive patients, and a relatively small number of anti-RNA polymerase III-positive patients). However, this association was probably explained by covariates, such as joint and muscle involvement, or lung fibrosis. A possible role of corticosteroid therapy might therefore be suspected.

REFERENCE:

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Disclosure of Interest: None declared


VERTEBRAL FRACTURE PREVALENCE AND MEASUREMENT OF THE SCANOGRAPHIC BONE ATTENUATION COEFFICIENT ON CT SCAN IN 70 PATIENTS WITH SYSTEMIC SCLERODERMA

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Background: Osteoporosis screening is not systematic in scleroderma patients but some studies demonstrated a similar risk between rheumatoid arthritis and systemic sclerosis.1–14 Thoracic and/or TAP (thoraco-abdomino-pelvic) CT (Computed Tomography) scans are classically performed in the follow-up of scleroderma, mainly to evaluate lung involvement.

Objectives: To study vertebral fracture (VF) prevalence and the scanographic bone attenuation coefficient of the first lumbar vertebra (SBAC-L1) on CT scans in systemic scleroderma patients. Secondary objectives are to study specific risk factors of SBAC-L1 £145 Hounsfield Units (HU) and to evaluate SBAC-L1 measurements reproducibility.

Methods: This monocentric retrospective study included patients followed from 2000 to 2014 and fulfilling ACR/EULAR 2013 criteria for systemic scleroderma and who underwent a thoracic or TAP CT scan. Osteoporotic risk factors, Dual Energy X-ray Absorptiometry (DXA) measurements and clinical characteristics were collected. For CT scan, the VFs were determined according to Genant’s classification on sagittal sections. The SBAC-L1 was measured in Hounsfield Units (HU) on axial section of L1 in a Region of Interest drawn in trabecular bone. Intra- and inter-reader reliabilities for SBAC-L1 were calculated. An SBAC-L1 £145 HU (fracture threshold) was used to define patients at risk of VF.4 Predictive factors for VF or SBAC-L1 £145 HU were studied.

Results: A total of 70 patients were included (mean age: 62.3 ±15.6 years, women 88.5%, diffuse scleroderma 22.9% (n=16)) in the study. Sixty patients (85.7%) presented with at least one clinical risk factor for osteoporosis. Eighteen patients (25.7%) received vitaminocalcic supplementation and 10 (14.3%) received antiresorptive therapy. DXA was only performed on 30 patients (42.8%) and 35 patients (50%) presented a SBAC-L1 £145 HU. SBAC-L1 measurements were highly reliable (Kappa >0.9 for both intra- and inter-reader reliability). For the univariate analysis, a SBAC-L1 £145 HU was significantly associated with age (OR=1.09, CI 95%: 1.04–1.13), calcinosis (OR=6.3, CI 95%: 1.61–24.75) and periarticular calcifications (OR=3.22, CI 95%: 1.06–9.77). For the multivariate analysis, age (especially patients older than 63 years), calcinosis andacro-osteolysis were independently associated with a SBAC-L1 £145 HU.

Conclusions: On a large sample of scleroderma patients with clinical risks of osteoporosis, only 42.8% were screened for DXA and 16.7% of them were osteoporotic. The VF prevalence on CT scan was 4.3% and the SBAC-L1 measurement identified 50% of the population at the fracture threshold. The presence of calcinosis, periarticular calcifications oracro-osteolysis should lead to an osteoporosis screening, especially for patients under 63 years old.

REFERENCES:

Disclosure of Interest: None declared


WHOLE BODY DISTRIBUTION AND CLINICAL ASSOCIATIONS OF TELANGIECTASIA IN SYSTEMIC SCLEROSIS: A CROSS-SECTIONAL STUDY

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Background: Telangiectasia (TA), one of the diagnostic criteria for systemic sclerosis (SSc), could be a clinical marker for the severity of vasculopathy, including pulmonary hypertension (PH).

Objectives: We designed a cross-sectional study: (i) to describe the whole-body distribution of TA, (ii) to assess the associations between the whole-body number of TA and the characteristics of patients, (iii) to determine whether the number of TA may be useful to discriminate SSc-PH patients.

Methods: Patients were included in the National Referral Centre for Rare Systemic And Autoimmune Diseases if they fulfilled the 2013 ACR/EULAR criteria for SSc. They were excluded if they had received laser treatment. The whole-body number and distribution of TA were recorded at inclusion. The associations were studied using univariate, adjusted and multiple linear regressions.

Results: 106 patients were enrolled, including 12 with PH. The median (interquartile range) number of TA was 30 (82.7). Their distribution was: 37.2% on the face, 33.2% on the upper limbs including 26.4% on the hands, 28.1% on the trunk including 17.1% for the upper part of the trunk, and 1.5% on the lower limbs. Using multivariate linear regression model, the whole-body telangiectasia number was independently associated with male gender (percentage change (95% CI)= +144.4% (7.5; 455.9), p=0.003), pulmonary hypertension (+162.8% (5.6; 553.8), p=0.033), history of pulmonary embolism (+336.4% (39.0; 1270.1), p=0.012), glomerular filtration rate (–1.6% (3.2; –0.1) per 1 ml/mn/1.73m 2 increase, p=0.038) and soluble endoglin (+28.2% (1.2; 62.5) per 1 ng/nl increase, p=0.039). The ROC analyses assessing the ability of telangiectasia to discriminate the presence of pulmonary hypertension revealed that the area under the curve was significant for the telangiectasia number on the whole body (0.77 (0.57; 0.88), on the hands and face (0.81 (0.57; 0.91)) and on the hands (0.77 (0.57; 0.89)).
ARTERIAL HYPERTENSION (PAH): A RETROSPECTIVE MONOTHERAPY VERSUS COMBINATION THERAPY IN SSc-patients, TA were predominantly located on the face, hands and the upper part of the trunk. They may reflect the vasculopathy of SSc and could represent a clinical biomarker for vascular disease, particularly for PH, one of the most severe vascular complications of the disease.

REFERENCES:

CONCLUSIONS: In SSc-patients, TA were predominantly located on the face, hands and the upper part of the trunk. They may reflect the vasculopathy of SSc and could represent a clinical biomarker for vascular disease, particularly for PH, one of the most severe vascular complications of the disease.

THU0419 ASSOCIATION OF INFLAMMATORY MARKERS C-REACTIVE PROTEIN AND ERYTHROCYTE SEDIMENTATION RATE WITH PULMONARY FUNCTION TESTS AND EUROPEAN SCLERODERMA STUDY GROUP ACTIVITY INDEX (ESCSG-AI) IN SYSTEMIC SCLEROSIS – ASSOCIATED INTERSTITIAL LUNG DISEASE IN FOLLOW UP STUDY

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Background: Inflammatory markers are very important to assess severity and activity of SSc-ILD, but its role needs further investigation.

Objectives: To assess inflammatory markers of SSc such as hsCRP and ESR and compare with lung function test and ESCSG-AI in the long-term follow up study.

Methods: It was a longitudinal study involving 77 pts with SSc-ILD (mean age was 46.2±13.4; 69% have limited subset of the disease; 93% were female). The mean duration of follow up was 58.9±11.4 months. Pts. were investigated with HRCT twice (at first visit and at the end of the study) and according the CT changes were divided into 3 groups: the 1st group (16 pts) with improvement; 2nd group (39 pts) without any changes and 3rd group (22 pts) with worsening of fibrosis. Other data collected including biological results (high-sensitivity C-reactive protein (hsCRP) and erythrocyte sedimentation rate (ESR)).

RESULTS: There were no significant differences between groups related to sex, frequency of diffuse form and duration disease. Mean levels of hsCRP and ESR didn’t change significantly during the follow up. In all pts the mean levels of hsCRP and ESR correlated directly with each other at first visit and at the end of the study (R=0.45 and R=0.4 (p<0.001) accordingly). We compared the mean levels of hsCRP and ESR with mean dates of FVC, DLCO and ESCSG-AI score in first visit and the end of follow up. Mean levels of hsCRP inversely correlated with mean dates of FVC (% of predicted), composite score (ESCG-AI).

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CONCLUSIONS: In our group of pts. the hsCRP has proven to be an accurate reflection of disease severity especially in pts with progression of ILD.