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Acknowledgments: None

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.6149

AB0614

METHOTREXATE DOESN'T LOWER THE RISK OF DEVELOPING INTERSTITIAL LUNG DISEASE IN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES WITH JO-1 ANTIBODIES.

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Background: In patients with idiopathic inflammatory myopathies (IIM) most commonly found autoantibody against histidyl-tRNA synthetase (anti-Jo-1) is associated with development of interstitial lung disease (ILD), which has been recognized as a serious mortality factor.

Objectives: To assess if methotrexate as an initial steroid sparing agent lowers the risk of developing ILD in anti-Jo-1 positive patients diagnosed with IIM.

Methods: Medical records of IIM patients treated in a referral clinic in capital city of Poland between 2008 and 2018 were reviewed. Inclusion criteria were: fulfillment of ACR/EULAR 2017 classification criteria for IIM, positivity of anti-Jo-1 antibodies in the EUROLINE test, introduction of corticosteroids equivalent to ≥ 0.5 mg of prednisone. Exclusion criteria: insufficient data on disease course, history of IIM <18 months.

Results: 29 patients were included for this analysis. ILD was present at the onset in 52% (n:15) patients. Other 14 patients were treated initially with corticosteroids ≥ 0.5 mg/kg along with methotrexate up to 25mg/week. In all 14 patients methotrexate was well tolerated and led to successful reduction of steroid dose. However, ILD attributed to the primary disease appeared in follow up in 50% (n:7) of these patients (medium 36 months), which resulted in alteration of treatment. In 7 patients ILD didn't develop.

Conclusion: Our study shows that methotrexate in dose up to 25mg/week doesn't lower the risk of developing ILD in Jo-1 positive IIM patients in the long term suggesting that other medication should be used as a first line treatment for this group.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.5028

AB0615

MORTALITY AND CAUSES OF DEATH AMONG ROMANIAN SYSTEMIC SCLEROSIS PATIENTS

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Background: Systemic sclerosis (SSc) is associated with an increased risk of death compared to the general population. Survival in SSc patients has improved significantly over the last 20 years with a decrease in renal involvement as a cause of early death and an increase in death caused by cardiopulmonary involvement (1,2). Causes of death in SSc patients have not been described in a Romanian cohort so far.

Objectives: To study the causes of death in patients with SSc prospectively followed-up from 2002 to 2018 in a single tertiary centre from Romania.

Methods: The cohort consists of 197 patients who fulfill the American College of Rheumatology /EULAR 2013 criteria for SSc. We examined the data of patients who had died during follow up. Patients were reviewed at least twice a year and the cause of death was classified as SSc-related or nonSSc-related.

SSc-related deaths were then attributed to the major organ involved. A univariate Cox proportional hazard (PH) regression was used to examine the association between each variable and mortality. Variables reported in the literature to associate with mortality were considered in the multiple Cox PH regression model.

Results: Of 197 SSc patients (87.8% females), 47.7% had diffuse SSc and 52.2% had limited SSc. The mean age at diagnosis was 47 (SD 12) years and mean follow up duration was 6.75 years. There were 41 deaths (20.8%). Survival rate was substantially lower in men ($P < 0.003$). The mean age at the time of death in those with diffuse SSc was lower compared to limited SSc (55.8 years vs 68.7 years). Sixty percent of deaths were SSc-related (pulmonary cause [n=11], cardiac cause [n=6], gastrointestinal involvement [GI, n=3], renal crisis [n=2] and others [n=4]). Deaths not related to SSc were associated with cancer and infections. Age at onset of Raynaud phenomenon [HR 1.05], male gender [HR 3.41], diffuse SSc [HR 0.48], presence of tendon friction rub [HR 4.54], digital ulceration [HR 3], esophagitis [HR 2.07] and cardiovascular involvement [HR 3.68], use of corticosteroids [HR 2.13] and cyclophosphamide [HR 2.02] were associated with poor prognosis in multivariate analysis.

Conclusion: In our cohort the main causes of death were lung and cardiovascular involvement. Deaths occurred early after the onset of the disease and the survival rate was significantly reduced among men. Multivariate analysis showed that age at onset of Raynaud phenomenon, male gender, diffuse disease form, presence of tendon friction rub, digital ulceration, esophagitis and cardiovascular history, use of corticosteroids and cyclophosphamide were independently associated with mortality.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.5438

AB0616

REDUCED BONE MINERAL DENSITY IN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES: A LONGITUDINAL STUDY

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Background: Reduced bone mineral density (BMD) leads to fragility fracture which is associated with a significant morbidity and excess mortality [1,2]. Patients with idiopathic inflammatory myopathies (IIM) should be at a heightened risk of reduced BMD as a result of the systemic inflammation, reduced mobility and corticosteroid use [3]. A previous cross-sectional study demonstrated a high prevalence of osteoporosis (23.7%) and osteopenia (47.4%) in a cohort of IIM patients [4]. However, longitudinal data are lacking.

Objectives: To assess the BMD of IIM patients longitudinally and to investigate the factors associated with accelerated bone loss.

Methods: This is a single centered observational study. Existing adult Chinese patients with IIMs who had serial BMD measurements done were recruited. The diagnosis of IIMs was based on the Bohan and Peter's criteria with definite or probable cases being included [5]. Patients with clinically amyopathic disease must have the typical Gottron's papules or heliotrope rash as determined by rheumatologists or dermatologists, and with no symptoms or signs of muscle involvement according to Sontheimer [6]. BMD was measured by dual energy X-ray absorptiometry (DEXA). Clinical variables thought to be associated with bone health were documented.

Results: All together 28 patients were studied. The mean age of the patients at disease onset was 46.1 years (S.D. 12.2). There was a female predominance (92.9%). The subgroups of IIMs were: dermatomyositis (39.3%), polymyositis (25.3%), clinically amyopathic dermatomyositis (21.4%) and immune mediated necrotising myopathy (14.3%). Only a minority of the patients smoked (7.1%) and none of them drinks regularly. About one fifth of the patients were underweight. All patients have been exposed to systemic corticosteroid, while 82.1% of them were still on it between the two scans with 32.1% even on high dose (>0.5 mg prednisolone/kg/day). Three out of the 28 patients (10.7%) was found to be osteoporotic at baseline and 17 patients (60.7%) were osteopenic. Follow-up DEXAs were performed mostly 5 to 10 years after the initial scan. Despite 8 patients (28.6%) were given active anti-osteoporotic medications, the bone health deteriorated significantly. The mean baseline neck of femur BMD dropped from 0.711 to 0.657 g/cm² ($p=0.042$) on follow-up, while the total

lumbar BMD from 0.951 to 0.905 g/cm² ($p=0.036$). The T-score in 11 patients (39.3) reached osteoporotic range at the second DEXA. Together with the patients with osteopenia, 78.6% of the IMM patients had reduced BMD at the follow-up scan. Actually, 5 patients (17.9%) already had one episode of fragility fracture. The use of high dose corticosteroid in between the 2 scans was found to be associated with a greater degree of mean BMD loss in the hip (-0.171 vs -0.007 g/cm², $p=0.007$).

Conclusion: Reduced BMD is prevalent in patients with IIM. Follow-up study revealed significant worsening of bone health. High dose corticosteroid use might be especially detrimental. Liberal assessment of BMD and use of anti-osteoporotic drugs in IIM patients are advisable. Prompt use of steroid-sparing agents to minimize steroid exposure may also be helpful.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.6163

AB0617

OPTICAL COHERENCE TOMOGRAPHY IN THE ASSESSMENT OF SKIN FIBROSIS IN SYSTEMIC SCLEROSIS: A CROSS-SECTIONAL STUDY

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Background: Previous studies have shown that Optical Coherence Tomography (OCT) is a reliable biomarker of skin fibrosis and significantly correlates with the severity of the skin involvement in Systemic Sclerosis (SSc) ^{1,2}.

Objectives: Aim of this cross-sectional study was to evaluate the performance of skin OCT to discriminate between SSc and healthy controls (HC) and to compare results with the current gold standard, the modified Rodnan skin score (mRss), in a different SSc study cohort.

Methods: Dorsal forearm skin of consecutive diffuse cutaneous SSc (dcSSc) patients and matched-HC was scanned by an investigator blinded to the clinical data using Vivosight scanner (Michelson Diagnostics, Kent, UK). Minimum Optical Density (MinOD), Maximum OD (MaxOD) and OD at 300 micron-depth (OD300) were measured. Clinical involvement was assessed by a blinded operator using the mRss and results were compared with imaging data. Statistical analysis was performed using GraphPad Prism software V.7.0.

Results: A total of 88 OCT images were obtained from 22 dcSSc patients [20 Female, mean age 49 (± 11) years, 12 with < 5 years disease duration] and 22 HC [20 Female, mean age 50.7 (± 6.7) years]. All OCT measures (MinOD, MaxOD and OD300) were significantly lower in SSc patients than in HC ($p=0.011$, $p<0.0001$, $p<0.0001$ respectively). MaxOD and OD300 were significantly different between the four groups (0-3) of patients based on the mRss at the site of analysis ($p=0.035$, $p=0.001$ respectively). Skin OCT showed a good performance in discriminating SSc skin vs HC (overall AUC 0.72, 0.8 and 0.89 for MinOD, MaxOD and OD300 respectively).

Conclusion: These results confirm in a cohort different from those of the previous studies that skin OCT is able to reflect the severity of skin involvement in SSc. Longitudinal studies are needed to validate its potential as surrogate outcome measure of skin fibrosis in SSc patients.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.4726

AB0618

COMPREHENSIVE ANALYSIS OF AUTOANTIBODY PROFILE IN A TURKISH SYSTEMIC SCLEROSIS COHORT

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Background: Serum autoantibodies closely reflect patterns of organ involvement and disease progression in systemic sclerosis (SSc). The entire autoantibody profile is less well defined in many cohorts and the data regarding their clinical associations and frequencies is limited.

Objectives: To determine the autoantibody profile of patients with SSc, as well as their clinical associations, in well-characterized inception- cohort with disease duration less than 3 years.

Methods: Serum samples of 100 patients out of 105 enrolled in the study were analyzed for ANA patterns with indirect immunofluorescence (IIF) assay using HEp-20-10/primate liver mosaic IIFT kit. Sera of 96 patients were subjected to commercial line immunoassay to quantify autoantibodies against 13 different autoantigens.

Results: 92 (92%) out of 100 patients were positive for ANA by IIF (Table 1). The speckled staining was the most pattern followed by nucleolar in 10 patients, centromere in 4, reticular in 1, nuclear in 2 and homogenous in 1. All patients ($n=96$) patients were positive for at least 1 autoantibody by immunoblotting (Table 2). Twenty-two (49%) of patients with anti-Topo I, 12 (44%) of the patients with anti-CENP and 4 (22%) of the patients with anti-RNAPIII were single positive. There was no difference in terms of the clinical findings when the patients with single and coexpression of these antibodies were compared. The distributions of the most frequent autoantibodies are shown in Figure 1. Interstitial lung disease was more frequent in the patients positive for anti-Topo I (78.8%) and anti-RNAPIII (27.3%). One of the two patients with breast cancer was anti-RNAPIII positive and none of the patients have diagnosed scleroderma renal crisis. Anti-Topo I was more common in patients with dcSSc (75%) and anti-CENP in lcSSc (46.4%).

Table 1. Demographic, clinical and laboratory characteristics of the SSc patients.

Sex	
Female N, %	91 (86.7%)
Male N, %	14 (13.3)
Female-to-male ratio	
Age, mean $\pm$ SD years	48.6 $\pm$ 12.7
Disease duration, mean $\pm$ SD years	2 $\pm$ 1.4
Disease classification N, %	
Diffuse	39 (36.5%)
Limited	65 (62.5%)
Sine scleroderma	1 (1%)
Interstitial lung disease	37 (34.3%)
Pulmonary arterial hypertension	3 (2.8%)
Scleroderma renal crisis	0
Digital ulcer	14 (13.3%)
Raynaud phenomenon	105(100%)
Telangiectasia	31 (28.8%)
Calcinosis	1 (1%)
Malignancy	3 (2.9%)
Antinuclear antibody profile N*, %	
Positive	92 (92%)
Staining pattern	
Speckled	65 (65%)
Nucleolar	13 (13%)
Centromere	29 (29%)
Homogeneous	3 (3%)
Reticular	3 (3%)