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FRI0394 NEUROPATHIC PAIN IS ASSOCIATED WITH SKIN THICKNESS IN SYSTEMIC SCLEROSIS PATIENTS

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Background: Systemic Sclerosis (SSc) is a connective tissue disorder characterized by fibrosis of skin and internal organs, vasculopathy and systemic inflammation. To date, few studies assessed pain in SSc patients, (1) namely the neuropathic component. Data is also scarce on defining possible neuropathic pain (NP) predictors in these patients.

Objectives: To determine if patients with SSc have a higher prevalence of NP compared with a group of age and sex matched controls and study possible associations between NP and SSc clinical variables.

Methods: The study evaluated patients diagnosed with SSc fulfilling the SSc classification criteria of the American College of Rheumatology (ACR), older than 18 years old and followed up at our Rheumatology Unit. Forty-eight patients with mean age of 56.98±12.73 years and mean disease duration of 9.77±6.12 years were included. Comparison group comprised 45 age and sex matched controls. Patients and controls were consecutively evaluated at our Unit and NP in 4 questions questionnaire (DN4) was used for assessing the presence of NP. In SSc patients, skin involvement was also evaluated clinically by the modified Rodnan skin thickness score (mRSS) ranging from 0 to 51. Hand mobility (HAMIS) and SSc Severity Scale (SScSS) were also calculated and relevant clinical variables of the disease were collected. Parametric and nonparametric tests were used for comparative analysis. Multivariate logistic regression was used to identify factors associated to NP. Statistical significance was defined as $P < 0.05$.

Results: In our study, prevalence of NP assessed by DN4 was significantly higher in SSc patients comparative to controls (56.2% versus 13.3%, $p < 0.001$). In addition to age and sex, presence of diabetes ($p = 0.541$) was also similar in both groups. In SSc group, patients with and without NP showed some statistically significant differences (table 1).

Table 1. Comparative analysis between SSc patients with and without NP

	NP positive patients (n=27)	No-NP patients (n=21)	P value
Median age, years (IQR)	61 (57–69)	52.38 (42.5–63.5)	0.032
Mean disease duration (years)	11.36±6.55	7.73±4.93	0.04
Diffuse/limited cutaneous subtype (n)	6/21	0/21	0.024
Mean mRSS	22.81±8.74	7.57±3.64	<0.001
Mean HAMIS	10.44±6.16	1.52±2.29	<0.001
Mean SScSS	10.00±4.32	3.90±2.07	<0.001
Digital ulcers n (%)	17 (63.0)	6 (28.6)	0.018
Calcinosis n (%)	15 (55.5)	6 (28.6)	0.062

Multivariate logistic regression revealed that only mean mRSS (odds ratio [OR] =1.90, 95% confidence interval [CI] 1.01 to 3.55, $p = 0.045$) was independently associated with the presence of NP.

Conclusions: To the best of our knowledge, this is the first study evaluating the prevalence of NP in SSc patients in comparison with age and sex matched controls. NP was significantly more prevalent in patients with SSc. Skin thickness assessed by mRSS was independently associated with the presence of NP.

References:

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FRI0395 INCIDENCE OF MYOSITIS-SPECIFIC AUTOANTIBODY (MSA) SPECIFICITIES IN SERA REFERRED TO NEW ZEALAND (NZ) MEDICAL LABORATORIES

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Background: Idiopathic inflammatory myopathies can be classified by clinicopathological phenotype into four major groups: overlap myositis (OM), dermatomyositis (DM), immune mediated necrotising myositis (IMNM) and inclusion body myositis (IBM). The different phenotypes associate with distinct MSA specificities although a variable percentage within each group are seronegative and the majority of IBM patients are seronegative¹. No commercial assay for the IBM associated specificity, cN1A, is available. In NZ testing for MSA is restricted to two laboratories: Waikato Hospital (WHL) and Canterbury Health Laboratories (CHL). Both laboratories use the commercial Euroimmun Euroline assay. In addition CHL has developed an ELISA for anti HMGCR autoantibodies and receives referrals from throughout NZ.

Objectives: To define the incidence and specificities of MSA in serum samples referred for testing in New Zealand.

Methods: For the period 3 November 2015 to 2 November 2016 each laboratory information system (LIS) was interrogated for requests for MSA. For the purposes of this report positive results were grouped into the specificities associated with each of the defined clinicopathological phenotypes.

DM: Mi2, Tif- γ , MDA 5 NXP2 SAE1
 OM: Ku, PM-Scl-100, PL75, Jo1, PL-7, PL-12, EJ, OJ
 IMNM: HMGCR, SRP

Results: Of screened sera (n=793, CHL=622, WHL=171) 11% were positive for MSA (88/793). Positive sera were distributed amongst three clinicopathological associated autoantibody specificities: DM 23% OM 50%, IMNM 27%. The most common serum specificities were HMGCR (n=24) and Jo-1 (n=13). No serum with specificities to SRP or OJ was detected. 22/24 sera positive for anti HMGCR had an elevated CPK concentration (CPK>2500u/l in 21/24 and CPK=544 in 1/24, reference range =66–220).

Conclusions: In New Zealand 11% of sera referred for MSA testing were positive. As a group the most common specificities were those associated with OM while the most common antigen specificity was to HMGCR. Anti-HMGCR antibodies were almost invariably associated with major elevations in serum CPK.

References:

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FRI0396 LEVELS PENTRAXIN 3 AND C1Q IN SYSTEMIC SCLEROSIS: ASSOCIATION WITH PULMONARY ARTERIAL HYPERTENSION AND CLINICAL FINDINGS

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Background: Systemic sclerosis (SSc) is an autoimmune rheumatic disease with unknown etiology that characterized fibrosis and vascular endothelial damage with obliteration of the microvasculature caused from exaggerated synthesis of collagen and extracellular matrix deposition in skin and internal organs. Pentraxin 3 (PTX3) accepted as a vascular inflammatory marker which secreted in early stage of endothelial dysfunction.

Objectives: In this study, we aimed to investigate plasma PTX3 and serum complement C1q levels and their relations with cytokines and pulmonary arterial pressure in SSc.

Methods: Fifty nine patients diagnosed SSc and 28 subjects without inflammatory rheumatic disease recruited to the study. Plasma PTX3, and serum TNF- α , IL-1 β , IL-4, IL-10, INF- γ , TGF- β and complement C1q measured in both groups. Clinical findings and pulmonary arterial pressure of the SSc patients assessed.

Results: Ages of fifty nine with SSc (54 female and 5 male) and 28 control subjects (23 female and 5 male) were 49,47±12.74 and 43,07±13,71 respectively. PTX3, IL-1 β and IL-4 levels found decreased in SSc patients. There were no differentiations between limited and diffuse cutaneous SSc subgroups in terms of PTX3, cytokines and C1q levels. Plasma PTX3 and serum IL-1 β concentrations found decreased only limited cutaneous SSc subgroup comparing controls. According to the immunosuppressive drugs use; PTX3 levels found decreased in using group and TNF- α ve IL-1 β levels found decreased in not using group comparing controls. Significant positive association found between PTX3 and C1q and all cytokines except TGF- β .

Conclusions: PTX3 suggested to be used in the pathogenesis and assessment of disease treatment efficacy of SSc, because of found decreased in SSc patients especially in limited cutaneous and immunosuppressive drugs using subgroups. Associations between C1q, and PTX3 and pulmonary artery pressure highlighted possible roles of both parameters in development of pulmonary hypertension. It is possible to develop preventive treatment strategies with better understanding of these associations. Advanced studies should be carried out for clarifying this condition.

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FRI0397 EVALUATION OF VITAMIN B12 DEFICIENCY AND ASSOCIATED FACTORS IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Vitamin B12 (Vit B12) deficiency is a common condition, which can be manifested with non-specific clinical features or with neurological and/or haematological abnormalities in severe cases. In systemic sclerosis (SSc)

gastrointestinal involvement (GI), nutritional status and medications may lead to Vit B12 deficiency.

Objectives: We aimed to investigate the frequency of Vit B12 deficiency and its determinants in SSc patients.

Methods: Sixty-two (90.3% female) SSc patients were enrolled in to the study. The nutritional status of patients was assessed with Malnutrition Universal Screening Tool (MUST). Serum Vitamin B12, homocysteine and Helicobacter Pylori Immunoglobulin G (H. Pylori IgG) levels were measured in all patients. Serum Vit B12 levels of patients were classified as; Low (<200 pg/ml), Borderline (200 - 300 pg/ml) and Normal (>300 pg/ml). Serum homocysteine levels of patients were classified as; Elevated (>9 µmol/L) and hyperhomocysteinemia (>15 µmol/L). H. Pylori IgG antibody level >5 U/ml considered as positive. Serum Vit B12 level <200 pg/ml or being on Vit B12 replacement therapy was considered as B12 deficiency.

Results: The mean age of the patients was 50.2 (12.5) years and mean disease duration was 12.0 (7.5) years. Forty-four (71.0%) patients were limited and 18 (29.0%) patients had diffuse SSc. The mean serum Vit B12 level of the patients was 323.6±291.5 pg/ml. Seventeen (27.4%) patients had normal, 23 (37.1%) patients had borderline and 22 (35.5%) patients had low serum Vit B12 level. Forty-four (71.0%) patients were considered as Vit B12 deficient; 22 had serum Vit B12 level <200 pg/ml (4 of these patients were on vitamin B12 replacement therapy), 22 were already on Vit B12 replacement therapy and Vit B12 level ≥200 pg/ml. The mean homocysteine levels were higher in the group with Vit B12 <200 pg/ml as compared to other groups (p=0.005). In the group with Vit B12 level <200 pg/ml, 33.3% (7/21) of the patients had hyperhomocysteinemia and 76.2% (16/21) had elevated homocysteine levels (Table). Fifty-one (82.3%) patients had GIS involvement and 16 (25.8%) patients had medium-high risk MUST score. H. Pylori IgG antibody was positive in 40 (64.5%) patients. There were no statistically significant differences between the patients with and without Vit B12 deficiency regarding to age, mean disease duration, hemoglobin level, GI involvement, medium-high risk MUST score, H. Pylori IgG antibody positivity and other clinical features (p>0.05 for all).

	Vit B12 level			p
	>300 pg/ml n=16	200–300 pg/ml n=22	<200 pg/ml n=21	
Homocysteine µmol/L, mean (SD)	9.1 (3.4)	10.4 (3.0)	14.1 (6.5)	0.005
Homocysteine >9 µmol/L, n (%)	7 (43.8)	16 (72.7)	16 (76.2)	0.084
Homocysteine >15 µmol/L, n (%)	1 (6.3)	0	7 (33.3)	0,004

Homocysteine level was not measured in 3 patients.

Conclusions: SSc patients are at risk for Vit B12 deficiency. Using homocysteine level seems to be unpractical for confirmation of Vit B12 deficiency in a complex disease such as SSc because its level is influenced by many factors. Patients with SSc should be closely monitored for Vit B12 deficiency and replacement therapy should be planned if necessary.

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FRI0398 INFLAMMATORY MYOPATHIES WITH CAMPTOCORMIA OR DROPPED HEAD SYNDROME ARE ASSOCIATED SCLEROMYOSITIS WITH LATE ONSET AND DELAYED DIAGNOSIS

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Background: Severe weakness of axial muscle leads to dropped head syndrome or camptocormia. The signification of these symptoms has not been studied in inflammatory myopathies (IM).

Objectives: To assess the signification of dropped head syndrome and/or camptocormia in patients with IM.

Methods: All practitioners of the French Myositis Network and the Club Rhumatisme et Inflammation (>1000 physicians) were invited to report their patients suffering from IM (myopathy with myositis specific autoantibody and/or typical muscle biopsy according to the ENMC criteria) with camptocormia and/or dropped head syndrome. These axial IM cases were included only if no other explanation of axial weakness was found. IM patients without axial involvement (non-axial IM group), were randomly selected from the participative centers, and included as control patients (ratio 1:2). Clinical, serological, muscle pathological features, management and outcomes were studied using a standardized form.

Results: Twenty patients (sex ratio 2.6) with axial involvement (camptocormia: 60%, dropped head syndrome: 40%) were included. Compared with the control group, these axial IM-patients were older (64.05±11.64 y. vs. 48.22±18.28, p<0.005) and diagnosis of IM was delayed (16.4±4.5 months vs. 8.5±2.7, p<0.05).

All patient except one had also proximal weakness of the limbs. CK blood level was 2794±870 UI/L, which was similar to the controls (2864±559 UI/L). According

to the ENMC classification, non-specific myositis was the most frequent finding on muscle biopsy (n=5/15, 30% vs. n=1/24, 4% in the non-axial group, p<0.05) and dermatomyositis (DM) pattern tended to be less frequent in axial-IM patients (3/12 20% vs. 11/24, 46%, p=0.10).

Most of the patients (75%) had also extra muscular involvement including acrosyndrome (45%), interstitial lung disease (35%), sclerodactyly (30%), telangiectasia (25%), digital tip ulcer (10%), sclerodermy (5%). By contrast, no patient had polyarthritides (vs. 20% in the controls, p<0.05). DM rash was hardly threefold less frequent in axial IM patients (15% vs. 42%, p<0.05).

Auto-antibodies associated with scleromyositis were the most frequent in axial-IM patients (30% vs. 10.3%, p=007, mainly anti-PM/ScI). One patient (5%) had cancer within the 3 years before or after IM diagnosis (NS vs. controls).

Thus, most frequent diagnosis in axial IM-patients were scleromyositis (35% vs. 5% in the controls, p<0.05) and inclusion body myositis (20% vs. 2.6% in the controls, p<0.05). DM was twofold less frequent than in control (10% vs 41%, p<0.05). Other IM subtypes were not statistically different from the control groups. Except patients with diagnosis of sIBM, all axial IM-patients received corticosteroids with another immunomodulatory drugs (median number 2, range 1–5). Half of the axial-IM patients received intravenous immunoglobulin. After a mean follow up of 68.4±3.76 months all patients had improvement, including in axial weakness, except in patients with sIBM. One patient died from ischemic cardiomyopathy.

Conclusions: In IM, camptocormia and dropped head syndrome are associated with late onset scleromyositis and sIBM with delayed diagnosis.

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FRI0399 EFFICACY OF AN INTENSIVE 24-WEEK PHYSIOTHERAPY PROGRAMME IN PATIENTS WITH SYSTEMIC SCLEROSIS - PRELIMINARY DATA FROM A SINGLE-CENTER CONTROLLED STUDY

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Background: Involvement of skin and musculoskeletal system in systemic sclerosis (SSc) leads to loss of function, disability and reduced quality of life. Data on efficacy of non-pharmacologic care in SSc is very limited due to variety in studied interventions/outcomes.

Objectives: To address the limitations of existing studies, and evaluate the effect of a controlled, long-term (24-week intervention, 24-week follow-up), intensive (1h physiotherapy + 0.5h occupational therapy twice weekly, and home-exercise for 0.5h 5x weekly), tailored physiotherapy programme on function/impairment of hands/face, and quality of life/disability in cohorts with a substantial number of SSc patients.

Methods: All patients fulfilled ACR/EULAR 2013 criteria, had skin involvement of hands/mouth, and were consecutively recruited from 2014 to 2016 at the Institute of Rheumatology in Prague. Both groups received educational materials and instructions for home exercise at baseline, however, only intervention group underwent the intensive physiotherapy programme. At months 0,3,6,12 all patients were assessed by a physician (physical examination, mRSS-Modified Rodnan's skin score, EUSTAR SSc activity score, Medsger SSc severity score), and a physiotherapist blinded to intervention [validated measurements (dFTP-delta finger to palm, inter-incisor/inter-lip distance, grip strength using Baseline dynamometer); tests (HAMIS-Hand Mobility In Scleroderma)], patients filled out patient reported outcomes/questionnaires (CHFS-Cochin Hand Function Scale, MHISS-Mouth Handicap In SSc Scale, HAQ, SHAQ, SF-36) and provided blood for routine laboratory analysis and biobanking. Normality of data was tested, inter-group analysis was performed with 2-way ANOVA, and intra-group analysis by Friedmann's test with Dunn's post hoc test.

Results: 25 SSc patients (22 female/3 male, 14 limited cutaneous (l)SSc/11 diffuse cutaneous (dc)SSc, median of age 54.0 and disease duration 7.0 years, mRSS 12) were recruited into the intervention group (IG) and 29 patients into the control group (CG) (25 female/4 male, 16 lSSc/13 dcSSc, median of age 49.0 and disease duration 5.0 years, mRSS 11). Compared to observed statistically significant improvement in dFTP, grip strength, HAMIS, inter-incisor and inter-lip

Parameter (unit)	Intervention group	Control group	Intra-group analysis (Friedmann-Dunn)		Inter-group analysis (ZWA)
	Mean ± SEM	Mean ± SEM	Intervention gr.	Control group	
dFTP (cm)	m0: 5.7 ± 0.5	m0: 6.6 ± 0.5	m0-m3: p<0.001	m0-m3: p<0.01	p<0.0001
	m3: 6.2 ± 0.5	m3: 6.1 ± 0.5	m3-m6: p<0.05	m3-m6: p<0.05	
	m6: 6.8 ± 0.6	m6: 5.8 ± 0.4	m0-m6: p<0.001	m0-m6: p<0.001	
Hand grip strength (kg)	m0: 17.2 ± 1.8	m0: 16.6 ± 1.3	m0-m3: p<0.05	m0-m3: p=NS	p<0.0001
	m3: 19.2 ± 1.9	m3: 14.9 ± 1.4	m3-m6: p=NS	m3-m6: p=NS	
	m6: 19.7 ± 1.9	m6: 13.9 ± 1.9	m0-m6: p<0.001	m0-m6: p<0.01	
HAMIS	m0: 9.8 ± 1.3	m0: 3.9 ± 1.1	m0-m3: p<0.01	m0-m3: p<0.01	p<0.0001
	m3: 7.1 ± 1.2	m3: 6.4 ± 1.2	m3-m6: p<0.01	m3-m6: p<0.001	
	m6: 4.1 ± 0.9	m6: 9.3 ± 1.1	m0-m6: p<0.001	m0-m6: p<0.001	
Inter-incisor distance (mm)	m0: 30.6 ± 1.6	m0: 32.9 ± 1.3	m0-m3: p<0.01	m0-m3: p<0.001	p<0.0001
	m3: 33.3 ± 1.6	m3: 30.2 ± 1.4	m3-m6: p=NS	m3-m6: p=NS	
	m6: 36.2 ± 2.0	m6: 29.8 ± 1.4	m0-m6: p<0.001	m0-m6: p<0.001	
Inter-lip distance (mm)	m0: 39.2 ± 1.6	m0: 41.7 ± 1.1	m0-m3: p<0.01	m0-m3: p<0.05	p<0.0001
	m3: 42.4 ± 1.7	m3: 39.9 ± 1.2	m3-m6: p=NS	m3-m6: p=NS	
	m6: 44.6 ± 1.8	m6: 40.0 ± 1.3	m0-m6: p<0.001	m0-m6: p<0.001	

Acronyms: SEM, standard error of the mean; Friedmann, Friedmann's test; Dunn, Dunn's post hoc test; ZWA, two way ANOVA; dFTP, delta finger to palm; HAMIS, Hand Mobility In Scleroderma; m0, month 0 (= at the baseline); m3, month 3 (= in the middle of intervention period); m6, month 6 (= at the end of intervention); p, p-value; NS, not significant