AB0789

HOW DOES THE DURATION OF THE DISEASE INFLUENCE THE QUALITY OF LIFE?

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Background: It is noticed that over the last decades the prognostic in patients with Idiopathic Inflammatory Myopathies (IIM) – a group of autoimmune disease characterised by muscle involvement, has improved along with increasement of disease duration and thus affects the quality of life.

Objectives: To assess the patient's quality of life related to the duration of the disease.

Methods: We performed a cross-sectional study from December 2015 to December 2017, the patients included fulfilled the Bohan and Peter¹ criteria for IIM. Demographic and clinical data were collected using a special questionnaire. Consistent with the objective the study group was divided in two subgroups by disease duration 1-less than 24 months and second subgroup more than 2 years. In order to estimate the quality of life (QoL) was applied Short Form-8 with 8 items for 8 domains and two components: mental and physical. Statistical data was analysed using MedCalc software version 12.

Results: There were 67 patients enrolled in the study, including 51 females and 16 males with a F:M ratio of 3.2:1, mean age 53.1±12.5 (range 25–78). The disease mean disease duration was 8.3±5.3 (range 0.5–12) years, there were 16 patients in the subgroup with the disease duration less than 2 years. The mean physical component was 36.48±9.05 and the mental component – 41.69±9.62 points, determined as reduced quality of life. Regarding the QoL of patients from subgroup 1, we found the physical component – 38.15±8.83 and the mental was 40.95±9.22 points. In the second subgroup we appreciated the physical and the mental component – 35.77±9.14 and 42.01±9.86 points, respectively. It was identified moderate correlation ($r=0.49$ $p<0.005$) between the both domains of the QoL and disease duration till 2 years, for the duration of more than 2 years we found moderate correlation ($r=0.51$ $p<0.005$) with mental compound and a weak one for physical domain ($r=0.24$ $p<0.005$).

Conclusions: Patients with idiopathic inflammatory myopathies had reduced quality of life by both domains. Disease duration in patients with early idiopathic inflammatory myopathies – less than 2 years, has a greater impact on patient's quality of life.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.5450

AB0790

FREQUENCY OF OSTEOPOROSIS IN POSTMENOPAUSAL WOMEN WITH SYSTEMIC SCLEROSIS

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Background: The prevalence of low bone mass or osteoporosis in patients with systemic sclerosis (SSc) varies significantly between studies performed in different countries and vary from 3% to 51% (Omar MA et al. 2013).

Objectives: To determine the frequency of osteoporosis (OP) in postmenopausal women with SSc in comparison with healthy control.

Methods: 163 postmenopausal women were enrolled in the study: 83 with SSc (mean age 58.5±8.1 years, mean disease duration 9.7±6.8 years) and 80 healthy control (mean age 59.2±6.6 years). Demographic characteristics and risk factors for OP in study groups are summarised in table 1. BMD was measured at lumbar spine, femoral neck and total hip by dual energy X-ray absorptiometry (DXA, Hologic 4500A). BMD decreasing grade was determined in according to WHO criteria.

Results: BMD in women with SSc was significantly lower than in control group at any site: lumbar spine – 0.832±0.119 vs 0.867±0.098 g/cm² ($p=0.031$); femoral neck – 0.633±0.098 vs 0.739±0.114 g/cm² ($p<0.0001$), and total hip – 0.752±0.116 vs 0.839±0.119 g/cm² ($p<0.0001$). Frequency of OP in SSc group was 32%, in control group – 13% ($p<0.002$). Mean age at menopause was less in SSc women than in control ($p<0.05$).

OP was significantly more often in SSc patients taking oral glucocorticoids compared to those without glucocorticoid therapy (39% and 20%, respectively, $p<0.05$). Low BMD was associated with age and interstitial lung disease in SSc women ($p<0.05$). At the same time no associations were found out among low BMD and disease duration, daily and cumulative doses of glucocorticoids. 23 (28%) of patients had osteoporotic fracture, among them 8 (10%) of women had two or more fractures in the anamnesis. The most frequent localizations of the fractures were distal forearm and vertebrae: 7 (32%) and 5 (23%) patients, respectively.

Abstract AB0790 – Table 1. Demographic characteristics of the study groups and risk factors of osteoporosis

Factor affecting BMD	SSc (n=83) Mean±SD or%	Control (n=80) Mean±SD or%
Age (years)	58.5±8.1	59.2±6.6
Body height (m)	1.61±0.06	1.59±0.06
Body weight (kg)	67.7±14.3	66.5±11.7
Body mass index (kg/m ²)	26.1±5.2	25.7±5.7
Age at menopause (years)	46.4±5.4*	49.7±3.0
Postmenopause duration (years)	12.2±7.2*	9.5±7.0
Smoking et presence time	3 (3.6%)	5 (6.3%)
Low energy fracture	23 (28%)	16 (20%)

* $p<0.05$ compared vs control group

Conclusions: OP in postmenopausal women with SSc were detected among 1/3 of cases, significantly exceeded its frequency in healthy control. Risk factors for OP were age, oral glucocorticoids and interstitial lung disease.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.2855

AB0791

SPECIFIC FEATURES OF SKIN INVOLVEMENT IN PATIENTS WITH SYSTEMIC SCLEROSIS AND ASSOCIATED PULMONARY ARTERIAL HYPERTENSION

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Background: Pulmonary arterial hypertension, associated with systemic sclerosis (SSc-PAH), is a severe irreversible manifestation of the disease, potentially fatal in its' late stage. It was shown that survival in SSc-PAH pts is much worse than in pts with idiopathic pulmonary arterial hypertension (IPAH). Unfavourable outcomes due to late recognition can be explained by predominance of subtle, clinically poor manifest SSc types, especially in terms of cutaneous and vascular syndromes.

Objectives: To assess the clinical features and survival rates in pts with systemic sclerosis sine scleroderma (ssSSc), associated with PAH.

Methods: 14 pts with ssSSc-PAH were analysed in comparison with 54 pts with clinically manifest skin involvement SSc-PAH (3 pts with diffuse (dcSSc-PAH) and 51 pts with limited cutaneous involvement (lcSSc-PAH)), and 48 pts with IPAH.

Results: Pts with IPAH were younger than both type SSc-PAH – 37 (28; 44), 48 (37; 56) and 54 (48; 62) y, respectively. In SSc-PAH pts with skin involvement and the diagnosis of PAH was established earlier (within 18 (10; 44) mo) than in pts with ssSSc (23 [15; 47] mo), although differences are not statistically significant. The PAH functional class was slightly higher in ssSSc-PAH, than in IPAH and SSc-PAH, the differences are not significant. Raynaud's phenomenon (RP) was present in all SSc-PAH pts, although in cutaneous SSc pts digital ischaemic lesions were more frequent (51% vs 14%, $p=0.03$), as well as contractures (53% vs 7%, $p=0.006$). There were no other differences in clinical features between the groups. Anticentromere antibodies (ACA) were present in 7 (50%) pts with ssSSc-PAH and in 36 (65%) pts with cutaneous SSc. Anti-topoisomerase-I antibodies (anti-Scl-70) were found only in 2 pts with lcSSc. More than 1 type of autoantibodies was detected in the majority of SSc pts. A wide range of antinuclear ABs was found in pts with ssSSc-PAH with prevailing ACA (in 7 pts), as well as anti-Sm, anti-La ABs, anti-nucleosome ABs (in one case), anti-Ro ABs (in 5 pts), anti-RNP-70 ABs – in 4 pts, anti-dsDNA ABs – in 2 pts, RF – in 3pts. SSc diagnosis was established according to ACR-EULAR 2013 classification criteria. The following diagnostic criteria were present in ssSSc-PAH pts: RP (in all pts), ulcers (3), scars (2), telangiectasia (10), PAH (14), SSc-associated ABs (7), capillaroscopic lesions (12). The mean total score was 11 (9;12) while ≥ 9 scores are required for SSc diagnosis, 100% pts with ssSSc-PAH met ACR-EULAR 2013 criteria, thus, justifying the SSc confirmation in this group of pts. There were significant differences in survival rates between IPAH pts and pts with various types of SSc-PAH (log-rank test, $p=0.06$). 5 year survival in ssSSc-PAH was somewhat lower, than in SSc-PAH – 50.6% vs 64.9%, respectively; IPAH pts had the best survival rates of 82.5%, and these differences are close to significant.

Conclusions: Clinical features and survival ssSSc-PAH are very similar to those in pts with cutaneous SSc-PAH with the exception of skin involvement and associated symptoms (digital ischaemic lesions and contractures). Rheumatologists

should be aware of such specific features as similar survival rates in cutaneous and ssSSc pts, and late recognition of PAH in pts with ssSSc, as well as its similarity with IPAH.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.5517

AB0792

AUTOIMMUNE AND INFLAMMATORY DISTURBANCES IN PATIENTS WITH PULMONARY ARTERIAL HYPERTENSION ASSOCIATED WITH SYSTEMIC SCLEROSIS

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Background: The prognosis of patients with pulmonary arterial hypertension associated with systemic sclerosis (SSc-PAH) is significantly worse, than other forms of PAH, and mechanisms of this phenomenon are unknown. Therefore, the isolation of autoimmune disorders is of great importance for early diagnosis and differential diagnosis, as well as the search for new therapeutic targets.

Objectives: To identify the autoimmune disorders in patients with SSc-PAH.

Methods: The study includes 52 pts with idiopathic pulmonary arterial hypertension (IPAH), 51 pts with SSc-PAH, 65 pts with SSc without PAH. Serum concentrations of the C-reactive protein (CRP), anticentromere antibodies (ACA) and antibodies to topoisomerase-I (anti-Scl-70) were routinely measured. The control group consists of 146 volunteers. Statistical analysis includes univariable logistic regression, ROC analysis and Kaplan-Mayer method.

Results: The average age of patients with IPAH was 37.9±10.5 years, SSc-PAH – 52.3±12.7 years, SSc without PAH – 51.2±13.2. Patients did not differ in functional class (FC), which was the main criterion of comparability. Mean values of FC in groups with SSc-PAH and IPAH also did not differ (2.7±0.8 and 2.6±0.7, respectively). ACA was associated with a 15.2-fold increased odds of developing PAH in SSc (OR 15.2, 95% CI 5.4–43.0), on the contrary, presence of anti-Scl-70 associated with low risk of PAH (OR 0.5, 95% CI 0.01 to 0.21). The level of CRP in the serum was significantly higher in patients with PAH than in the control group: 4.1 (1.9, 10.0) and 0.61 (0.25, 1.9), $p < 0.000001$, and also in comparison with patients without PAH (1.9 (0.8, 6.4), $p = 0.02$). In pts with PAH, the level of CRP correlated with FC and right atrium pressure and 6 min walk test distance. The level of CRP was significantly higher in patients with FC III-IV compared with FC I-II and in non-surviving patients. The Kaplan-Mayer analysis showed that pts with CRP level more than 4.75 mg/L at the time of diagnosis of PAH had a significantly lower survival rate (median 48 months) than pts with normal values (median 91 months) ($p < 0.005$), with 67% sensitivity and 61% specificity.

Conclusions: SSc-PAH is a unique phenotype combining the manifestations of SSc and PAH, the pathogenetic mechanisms of which modify the course of these states. It is based on a feature of autoimmunity with the predominance of ACA and a low occurrence of anti-Scl-70. An increase in the concentration of CRP, as well as the effect of its baseline level on survival, attests to the significant role of autoimmunity and inflammation in the pathogenesis of this fatal SSc complication.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.7170

AB0793

DESCRIPTIVE ANALYSIS OF A COHORT OF PATIENTS DIAGNOSED WITH INFLAMMATORY MYOSITIS IN A TERTIARY HOSPITAL

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Background: Inflammatory myopathies are a group of rare systemic diseases characterised by muscle weakness and inflammation. Clinical manifestations, course and prognosis of these pathologies are very heterogeneous.

Objectives: The aim is to describe the main characteristics of patients diagnosed with inflammatory myositis fulfilling Bohan and Peter criteria.

Methods: Descriptive analysis of a cohort of 34 patients from the same hospital with follow-up between January 2010 and December 2017. We recorded demographic characteristics, clinical manifestations, treatment, comorbidity and mortality.

Results: 34 patients (73% female) were recruited with an average age at diagnosis of 56.3 years^{27–83} among adults and 10 years^{4–15} among children. Most of them were Caucasian (94%). 18% were smokers and 15% previous smokers. The most frequent type was dermatomyositis (DM) (40%) followed by antisynthetase syndrome (ASS) (15%), necrotizing myopathy (12%), inclusion body myopathy (12%), overlap myositis (9%) and polymyositis (9%). 2 patients (out of 4) with necrotizing myopathy were treated with statins.

Clinical manifestation included muscle weakness (84%) and skin manifestations (48%) mainly among DM patients. 8 patients (24%) showed interstitial lung disease (4 non-specific interstitial pneumonia, 3 usual interstitial pneumonia and 1 cryptogenic organising pneumonia), especially among patients with overlap syndrome (n=3), DM (n=2) and ASS (n=2). Pulmonary hypertension occurred in 7 patients (21%), 30% among patients with overlap myositis associated to systemic sclerosis. The rest of extramuscular manifestations are expressed in the table 1. Muscle biopsy was performed in 57% of patients (77% compatible with myopathy). MRI was carried out in 45% (100% with active myositis). EMG was performed in 94% of patients with myopathic findings in 67% of them. 20 patients (60%) presented positive antinuclear antibodies, being the most frequent antiPML-SCL (18%), antiJo1 (18%), antiRo (12%) and anti-MDA5 (9%).

All patients were treated with corticosteroids. Only 2 responded to corticosteroids in monotherapy. More than 90% needed additional immunosuppressive treatment and 65% received 2 or more immunosuppressants. The most commonly used drugs were methotrexate (72%), rituximab (28%), azathioprine (25%), immunoglobulins (21%) and cyclophosphamide (21%). Only in 12% treatment could be stopped because of sustained remission.

3 cases of cancer (9%) were reported: myelodysplastic syndrome, lung neoplasm (in the case of paraneoplastic myositis) and lymphoma.

During the follow-up period 4 deaths were registered (12%) due to infections and cancer.

38% of patients required a multidisciplinary approach.

	n	%	Dermatomyositis	Polymyositis	Inclusion Body Myopathy	Necrotizing Myopathy	Antisynthetase syndrome	Overlap syndrome
Raynaud	7	21,2%	2	0	0	1	2	2
Cardiac Manifestations	7	21,2%	1	2	1	2	1	0
Arrhythmia	4	12,2%	1	1	1	0	1	0
Cardiomyopathy	1	3%	0	0	0	1	0	0
Heart attack	1	3%	0	1	0	0	0	0
Aortic stenosis	1	3%	0	0	0	1	0	0
Gastrointestinal Manifestations	6	18,1%	1	1	2	1	1	0
Dysphagia	3	9,1%	1	1	0	0	1	0
Chronic gastritis	2	6%	0	0	1	1	0	0
Constipation	1	3%	0	0	1	0	0	0
Severe Infections	5	15,1%	3	1	0	0	1	0
Hematologic Manifestations	5	15,1%	0	0	2	1	1	1
Anaemia	4	12,1%	0	0	2	1	1	0
Anaemia + thrombocytopenia	1	3%	0	0	0	0	0	1
Arthritis	4	12,1%	1	0	0	0	1	2

Conclusions: Inflammatory myopathies have frequent multiorgan involvement and represent a heterogeneous group of systemic diseases as shown in our registry and in the literature. Most patients need chronically combined immunosuppressive treatment and few achieve sustained remission. In consequence the collaboration of several specialties is necessary for the diagnosis and management of these pathologies.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.2639

AB0794

MYOSITIS DAMAGE INDEX IN A MYOSITIS POPULATION COHORT

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Background: Idiopathic inflammatory myopathies (IIM) are a heterogeneous and uncommon group of diseases characterised by muscular involvement and many systemic manifestations. They usually have a chronic course, and despite treatment they often develop functional impairment. MDI-MYODAM a score to assess damage has been recently developed. Few publications in this regard have reported a prevalence of 92%. There are no studies in Mexican-mestizo population describing the extent of damage and its main determinants.

Objectives: To investigate the prevalence of damage measured with the MDI-MYODAM tool in patients with IIM and its relationship with characteristics of the disease and treatment

Methods: A cross-sectional study was conducted in an IIM cohort from a national reference hospital. MDI questionnaire was applied to all patients. Demographic, disease characteristics, IIM subgroup (dermatomyositis, polymyositis and overlap), comorbidities (Charlson index), medical treatment were collected. Descriptive statistics were applied. Bivariate Pearson correlation test was conducted to