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 Peter[1] 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 maore 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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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% (Omair 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±0119 vs 0.867±0,098 g/sm^2 (p=0.031); femoral neck – 0.633±0.098 vs 0.739±0.114 g/sm^2 (p<0.0001), and total hip – 0.752 ±0116 vs 0.839±0,119 g/sm^2 (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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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 $\geq$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 ssSS-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