the other capillary parameters (capillary ramification, microhemorrhages) between the two groups of patients.

Conclusions: In a limited cohort of MCTD patients with an average disease duration of 6.4 years and a follow-up of three years, the nailfold microangiopathy does not seem to be significantly progressive. Patients with MCTD seem to show less enlargement/giant capillaries, and larger absolute number of total and normal capillaries than SSc patients. Still difficult to identify a defined NVC pattern in MCTD patients.

REFERENCES:

Disclosure of Interest: None declared

AB0762
RELATIONSHIP OF THE SIX MINUTE WALKING TEST AND QUALITY OF LIFE IN PATIENTS WITH SYSTEMIC SCLEROSIS
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Background: The six minute walking test (6MWT) is a standardised measure of submaximal exercise capacity. It is a surrogate measure of heart and lung involvement. There are no studies on relationship between 6MWT and quality of life evaluated by Short form 36.

Objectives: To evaluate the relationships between the 6 min walking distance (6MWD) and each items of SF36

Methods: Fifty consecutive SSC patients were investigated. They underwent 6MWT and complete the SF-36 (assessed the 8 domains of the questionnaire as well as the physical component score-PCS and mental component score-MCS).

Results: 6MWD ranged from 253 to 582 (median 420); we listed the correlations from 6MWT and only the statistically significant features of SSC and the items of SF36

Age: median 48 (range 20–72); Rho = −0.51; p<0.00001
EScSG Activity Index: median 0.5 (range 0–5); Rho = −0.33; p<0.009
HAQ-DI: median 0.375 (range 0–2.75); Rho = −0.26; p<0.048
mRSS: median 2 (range 0–17); Rho=0.35; p<0.007
Pulmonary hypertension (echocardiography): median 30 (range 13–80); Rho = −0.26; p<0.048
SF36: PCS: median 43 (range 20–92); Rho 0.41; p<0.0016
PF: median 75 (Range 0–100); Rho = 0.40; p<0.002
GH: median 50 (range 10–92); Rho 0.43; p<0.0007

Conclusions: Our study first demonstrates that 6MWT is correlated to some aspects of quality of life as measured by SF36 in the SSC patients. This results must be considered when assessing 6MWT in SSC.

REFERENCES:

Disclosure of Interest: None declared

AB0764
COMPARISON OF DISEASE CHARACTERISTICS IN PATIENTS WITH JUVENILE-ONSET AND ADULT-ONSET PROGRESSIVE SYSTEMIC SCLEROSIS
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Background: Progressive systemic sclerosis (PSSc) has been known to affect mainly adults of 30–50 years of age. Juvenile –onset has been reported to be rare and associated with a different disease course as compared to adult-onset. The comparison of juvenile-onset and adult-onset form have been limited.1 These studies were coming from European and North American countries.1 As there would be also effects of ethnic differences, we aimed to assess clinical differences between the two forms of PSSc of paediatric and adult rheumatology centres of a tertiary centre, in Turkey.

Methods: Adult onset patients were defined as those who were registered and followed as ‘scleroderma’ at the departments of adult and paediatric rheumatology at Cerrahpasa Medical Faculty, Istanbul, between 2005 and 2017. Only those with at least 2 follow-up visits were included. Patients’ charts were re-evaluated retrospectively.

Abstract AB0764 – Table 1

<table>
<thead>
<tr>
<th></th>
<th>Adult onset, n=137</th>
<th>Juvenile onset, n=26</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at diagnosis, mean/SD</td>
<td>43.6±14.0</td>
<td>11.4±3.2</td>
<td>-</td>
</tr>
<tr>
<td>Follow-up duration, med. [QR], years</td>
<td>5 [2–0–7]</td>
<td>4 [2.5–6]</td>
<td>NS</td>
</tr>
<tr>
<td>Male/Female</td>
<td>20/117</td>
<td>2/24</td>
<td>NS</td>
</tr>
<tr>
<td>Familial history of chronic inflammatory diseases, n (%)</td>
<td>20 (14.6)</td>
<td>4 (15.4)</td>
<td>NS</td>
</tr>
<tr>
<td>Sclerodactyly, n (%)</td>
<td>128 (93.4)</td>
<td>25 (96.2)</td>
<td>NS</td>
</tr>
<tr>
<td>Raynaud phenomenon, n (%)</td>
<td>135 (98.5)</td>
<td>24 (92.3)</td>
<td>NS</td>
</tr>
<tr>
<td>Digital ulcers, n (%)</td>
<td>55 (41.4)</td>
<td>14 (54.0)</td>
<td>0.001</td>
</tr>
<tr>
<td>Intestinal lung disease, n (%)</td>
<td>71 (52.2)</td>
<td>6 (24.0)</td>
<td>0.009</td>
</tr>
<tr>
<td>PAH, n (%)</td>
<td>20 (14.9)</td>
<td>0</td>
<td>0.045</td>
</tr>
<tr>
<td>Arthritis/heart failure, n (%)</td>
<td>14 (10.4)</td>
<td>1 (4.0)</td>
<td>NS</td>
</tr>
<tr>
<td>Joint involvement, n (%)</td>
<td>20 (14.9)</td>
<td>13 (50.0)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Skeletal muscle involvement/mopathy, n (%)</td>
<td>10 (7.5)</td>
<td>7 (28.0)</td>
<td>0.002</td>
</tr>
<tr>
<td>Gastrointestinal system involvement, n (%)</td>
<td>42 (31.8)</td>
<td>8 (32.0)</td>
<td>NS</td>
</tr>
<tr>
<td>Arterial hypertension, n (%)</td>
<td>24 (18.2)</td>
<td>0</td>
<td>0.015</td>
</tr>
<tr>
<td>APA positivity, n (%)</td>
<td>119 (90.0)</td>
<td>18 (75.0)</td>
<td>0.007</td>
</tr>
<tr>
<td>DMARD use, n (%)</td>
<td>90 (65.7)</td>
<td>25 (96.2)</td>
<td>0.002</td>
</tr>
<tr>
<td>Vasculators, n (%)</td>
<td>113 (82.5)</td>
<td>13 (50.0)</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

Results: There were 140 patients with scleroderma in the adult outpatient clinic records and 51 in the paediatric clinic records. Of these patients, 3 (2%) adults
and 25 (49%) paediatric patients had localised scleroderma (p<0.001). We studied the remaining patients (adults: n=137, juvenile: n=28) who had systemic pattern. Male/female ratio, median follow-up duration, familial history of chronic inflammatory diseases and the frequency of sclerodactyly, digital ulcers, Raynaud phenomenon, arhythmia/heart failure and gastrointestinal involvement were similar between two groups (table 1). The frequency of interstitial lung disease, pulmonary artery hypertension, and serum ANA positivity were significantly more common in the adult onset group. Whereas joint and muscle involvements were significantly more common among juvenile onset patients. DMARD use was significantly more common in the juvenile group while the use of vasodilators was more frequent among adults.

Conclusions: Our results are online with previous reports: juvenile onset patients seem to have a milder form of disease. Major organ involvement as defined interstitial lung disease and pulmonary artery hypertension was more common among adult onset patients. On the other hand, as expected, joint involvement and myopathy were major causes of morbidity in the juvenile group. Contrary to that previously reported, cardiac involvement was not common in the juvenile group.

REFERENCES:

Disclosure of Interest: None declared

AB0765 DEVELOPMENT AND ASSESSMENT OF A STRUCTURED TRAINING PROGRAM FOR PATIENTS WITH SYSTEMIC SCLEROSIS
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Background: Structured patient education programs are a key element of patient care in many chronic diseases. They are often based on the Stanford University chronic disease self-management program and aim to empower patients and to improve compliance and coping abilities. However, in every condition self-management education programs yield the expected benefit.1

Objectives: To develop a structured patient education program for SSc patients and to prove training-specific effects on patients’ quality of life and disability.

Methods: We developed a structured patient education program. The content of the program was created by a team of rheumatologists and dermatologists. The program consists of three modules focusing on general knowledge about the disease, gastrointestinal involvement, digital ulcers (DU), skin and wound care and a patient diary on disease symptoms. Patients were either included in the intervention or in the control group. Disease symptoms and severity as well as clinical parameters were assessed at baseline (intervention and control), at the follow-up visit at month 3 (intervention only) and at the final follow-up visit at month 6. In the intervention group satisfaction with the education program was analysed.

Primary outcome measures were SHAQ-SF, 12-BFI, SHAQ-DU. Secondary outcome measure was the satisfaction survey. For comparisons between different times analysis of variance for repeated measures was used. For description of cohorts Mann-Whitney Wilcoxon test was used.

Results: 58 SSc patients were included, 27 received the educational program (intervention group) and 31 patients served as a control group. Both groups were matched regarding demographics and disease subtype. Incidence of DUs was significantly higher in patients from intervention group resulting in a more frequent administration of vasoactive therapiies. SHAQ-SF, 12-BFI, SHAQ-DU were comparable between control and intervention group. However, patients in the intervention group rated the training program as helpful and reported an increase in knowledge about their disease afterwards. A positive impact of the training program on SHAQ-SF, 12-BFI, SHAQ-DU was observed in individual patients.

Conclusions: Patients who participated in the training were overall satisfied with the program. However, no significant effects on quality of life after the intervention were observed. One reason for this finding might be the disease duration (mean 11.5 years). This needs to be further analysed in a consecutive study considering patients with shorter disease duration.

REFERENCE:

Disclosure of Interest: None declared

AB0766 INITIAL CHARACTERISATION OF WOMEN WITH BREAST IMPLANTS IN A GROUP OF PATIENTS WITH SYSTEMIC SCLEROSIS REFERRED FOR AUTOLOGOUS HSCT
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Background: The causal relationship between breast implants (BI) and systemic sclerosis (SSc) is still strongly contested.

Objectives: To add further input to this medical controversy, we studied the initial clinical characteristics of patients with breast implants and systemic sclerosis that are referred to our centre for autologous hematopoietic stem cell transplant.

Methods: From 163 patients, with the diagnosis of systemic sclerosis (SSc), limited SSc, CREST, Morphea or scleroderma sine scleroderma, referred to our centre for autologous hematopoietic stem cell transplantation, 132 were found to be females. To identify those with breast implants (BI) or have a history of breast implants, we performed a systemic chart review for all patients. Once the patients with actual breast implant devices or have history of breast implants were identified, alive patients were contacted to check the type of their breast implants (silicone vs saline), the year of insertion, the local complications, whether they were removed or replaced and the year of removal and replacement, and the type of replacement if applicable. Clinical and biological data were collected for all patients and were compared between those who have breast implants or history if breast implants and those who do not have.

Results: From 132 patients with SSc or SSc variants, thirteen had history of BI (9.8%). In 12 of the breast augmentation therapy preceded the development of SSc, with median time between BI insertion and the emergence of initial symptoms of SSc of 12 years (range 7–29). The remaining patient showed acceleration of her disease after BI surgery. Surprisingly, in all 12 patients for whom we could know the type of initial implants, the prostheses were saline. When we compared the clinical characteristics of those with BI and those without. Patients with BI appeared to have higher age (mean 49.95 vs 44.42 years, p=0.012, shorter time from initial symptoms to diagnosis (mean 4.76 vs 12.24 months, p<0.001), more frequently positive ANA (13/13 vs 89/114, p=0.06) and more frequently positive anti RNA polymerase III (7/10 vs 20/78, p=0.004).

Conclusions: Our data may support the hypothesis of a possible association between BI and SSc. Furthermore, these results raise questions regarding the safety of saline breast prosthesis. Finally, our finding may indicate a possible difference in the initial characteristics of SSc patients with BI and those without.

Disclosure of Interest: None declared

AB0767 EFFICACY OF SUBCUTANEOUS TOCLIZUMAB IN PATIENTS WITH RHEUMATOID ARTHRITIS AND SYSTEMIC SCLEROSIS OVERLAP SYNDROME
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Background: Systemic sclerosis (SSc) is a connective tissue disease that develops sclerotic changes in the skin and visceral organs. SSc is a disease of uncertain etiology, but is characterized by fibrosis of the skin, connective tissue, cardiac, gastrointestinal, and renal complications contribute to patient morbidity and decreased survival1. And patients present with stiffness of the limbs because of joint swelling and joint swelling in the skin and periartricular connective tissues. Interleukin-6 (IL-6) is a pleiotropic factor that plays a major role in inflammation; furthermore, IL-6 overexpression and pathogenicity in SSc have been demonstrated2, 6. IL-6 expression is reportedly high in both the skin and serum of SSc patients3, and its elevation depends on the skin score. And it is a candidate factor that can reproduce the pathological conditions of SSc as well as RA.

Objectives: We report the cases of rheumatoid arthritis (RA) patients with SSc who was administered anti-interleukin-6 receptor antibody tocilizumab (TCZ).

Methods: Two RA with refractory SSc patients were administered tocilizumab at 162 mg/kg twice a month for 12 months. RA disease activity is evaluated by DAS28-ESR and CDAI. Skin condition of SSc is evaluated by pinching the skin according to the modified Rodnan total skin score (mRTSS).

Results: They were both female, and age at the time of SSc diagnosis was 74 (patient 1) and 51 (patient 2) years old. The time lapse since SSc diagnosis was at first visit and 14 years, respectively. And it since RA diagnosis was 14 years and 6 years, respectively. Tocilizumab was administered at 162 mg every 2 weeks, which is equal to the dosage used for RA. Administration of prednisolone at 5 mg/day and DMARDs were continued. Overall, TCZ was well tolerated, and both patients experienced a general improvement in coping with normal daily activities. During the 12 month tocilizumab therapy, both RA disease activity and mRTSS decreased. The patient global assessment improved by 70 (75 to 5) and 44 (68 to 24) in patients 1 and 2 in 12 months, respectively. In RA disease activity, DAS28 decreased from 5.66 to 1.73 in 12 months in patient 1 and 7.14 to 4.43 in...
OSTEOMYELITIS COMPLICATING DIGITAL ULCERS IN SYSTEMIC SCLEROSIS

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Background: Skin ulcers are a frequent manifestation of systemic sclerosis (SSc). Skin ulcers are painful, represent a cause of disability and heavily affect patients’ quality of life. The presence of local infection may be responsible for osteomyelitis (OM) of the underlying bone. If gangrene develops, surgical amputation may be required. At the moment is not clear if there are predisposing factors to osteomyelitis development.

Objectives: To describe a population of SSc patients affected by cutaneous ulcers and osteomyelitis

Methods: We collected data of SSc patients satisfying the 2013 ACR criteria for SSc referring to our outpatient clinic from January 1st 2016 to December 31st 2017. The patient’s data were evaluated on the basis of individual clinical records, including demographic, clinical and serological findings. Cutaneous ulcers were defined as epithelial loss and loss of dermis; post-traumatic skin lesions were excluded. In cases suspected of infection, microbiological investigations were carried out. We have diagnosed OM by clinical, radiological and laboratory means, in particular the presence of pain, swelling, fever, erythema, purulent secretions, blood chemistry alterations and typical radiological characteristics at either plain X ray and/or MRI. Statistical analysis was performed using STATA software for descriptive analysis and groups comparisons. Given the low number of events only univariate analysis was conducted.

Results: A total of 189 patients were enrolled in the study. Of them, 21 (11.1%) were males, mean age was 64.39±12.5 years and median disease duration 11.59 (5.6–19.3) years. A diffuse cutaneous (dcSSc) involvement was present in 50 (28.5%), limited cutaneous (lcSSc) in 131 (69.3%) and a limited disease in 8 patients (ISSC) (4.2%). Digital ulcers (DU) were present in 29 patients (15.3%) and in 5 cases (2.6%) were complicated by the occurrence of OM. The pathogenesis responsible of the infections were isolated in 3/5 (60%) cases and were represented by: Methicillin-sensitive Staphylococcus aureus (2 cases) and P. aeruginosa, also multisensitive. OM affected the third finger of right hand in 2 (40%) patients, the second finger of right hand in 1 (20%) patient and the third finger of left hand in 2 patients (40%). In 2 cases (40%) surgical amputation had to be performed. Patients with OM were significantly younger (54.9±16.07 vs 64.65±12.34, p=0.0432) and had higher CRP levels than the rest of the patients (1.27±0.59 vs 0.42±0.74, p=0.0061). In patients with DU, the only predictive factor for the development of OM was the total number of ulcers in the single patient (OR 2.27, 1.39–3.71, p<0.001) while no significant influence was found for other demographic or disease specific parameter.

Conclusions: OM is a severe complication of DU in SSc. In most cases the aetiology agents are community-acquired pathogens. SSc patients with OM were younger but did not show any other obvious distinguishing feature. The number of ulcers in the single patients were predictive of OM development. Further and larger studies are needed to address this aspect of the microvascular involvement of SSc.

Disclosure of Interest: None declared


AB0769

SCLERODERMA MIMICS IN COHORT FROM AN EUSTAR CENTRE

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Background: The differential diagnosis of systemic sclerosis (SSc) can be sometimes challenging, especially when you have symmetrical skin thickening, Raynaud’s phenomenon (RP) or acroosteolysis. When symptoms and signs are unclear, patients should be referred to a specialist centre for assessment to differentiate between scleroderma and its mimics.

Objectives: Assessing the types of scleroderma mimics presenting in a tertiary care centre and outlining the diagnosis difficulties.

Methods: We evaluated a cohort of 140 patient admitted in our clinic with the suspicion of SSc from January 2007 until December 2017. 130 of them are with SSc and 10 patients with scleroderma mimics. The patients were evaluated for quality and distribution of skin involvement, the presence of systemic complications, the presence of scleroderma specific antibodies and the capillaroscopic pattern. If they have not met any criteria for SSc, they underwent further specific investigations.

Results: From the 140 patients evaluated, 10 (7.14%) were with scleroderma mimics. All these 10 patients were admitted in our clinic with the suspicion of SSc. 3 of them had severe RP, one had acroosteolysis and 6 had symmetric skin thickening. There were 4 males and 6 females. All the patients had no organ involvement (pulmonary arterial hypertension or pulmonary fibrosis), normal capillaroscopic pattern and negative antinuclear antibodies and negativespecific scleroderma antibodies. The patients with RP had no skin thickening or other clinical or laboratory changes and the diagnostic was primary RP. The patient with acroosteolysis had no skin sclerosis or RP and after genetic testing a diagnosis of Hajdu-Cheney syndrome was made. The 6 patients with skin thickening had no RP. These 2 patients with scleroderma had no underlying gammopathy or infections. The patient with eosinophilic fasciitis had extended skin thickening with eosinophilia ant typical aspect on skin biopsy but the skin test was negative. The patients with EA and PAH had no skin lesions and no diagnostic changes. The 1 patient with UIP presented with RP like symptoms and diagnostic of pulmonary arterial hypertension was established.

Conclusions: Even though are rare, scleroderma mimics can be a challenging diagnostic even in tertiary care centre and sometimes diagnostic can be delayed. A correct diagnostic is necessary to avoid unnecessary immunosupresion.

REFERENCES:


Disclosure of Interest: None declared


AB0770

18 FDG PET/CT PREDICTS DECLINE IN FUNCTIONAL RESPIRATORY TESTS IN SYSTEMIC SCLEROSIS PATIENTS BUT NOT IN RHEUMATOID ARTHRITIS PATIENTS

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Background: Interstitial lung diseases (ILD) is a frequent complication in connective tissue diseases (CTD) such as rheumatoid arthritis (RA) or systemic sclerosis (SS), but the lung is the only affected organ in the idiopathic pulmonary fibrosis (IPF). Non-specific interstitial pneumonitis (NSIP) is the more frequent form in SS while usual interstitial pneumonitis (UIP) predominates in RA patients and in the IPF form. Some studies suggested that 18-FDG-PET/CT could help to detect zones of activity in lung tissue in IPF and this in turn could predict the disease

Disclosure of Interest: None declared


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