AB0804 THE RELATIONSHIP BETWEEN AUTOANTIBODIES AND CLINICAL SYMPTOMS IN PATIENTS WITH INFLAMMATORY MYOPATHY

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Background: Myositis-specific autoantibody (MSA) and myositis-associated autoantibody (MAA) are often detected in dermatomyositis (DM) and polymyositis (PM), and are useful diagnostic markers. In addition, autoantibodies are reportedly related to clinical symptoms, including skin findings, muscle symptoms, and interstitial lung disease, and provide information useful for medical treatment. However, the clinical significance of the presence of multiple MSA/MAA and anti-Ro52 autoantibodies has not been determined.
Objectives: The purpose of our study was to clarify the clinical significance of multiple MSA/MAA and anti-Ro52 autoantibodies.
Methods: We enrolled 58 patients diagnosed with DM and PM at Kagawa University Hospital. PM and DM were diagnosed according to the Bohan and Peter criteria.[1] The patients were analysed for MSA (anti-Mi-2, anti-Jo-1, anti-SRP, anti-PL-7, anti-MDA5, anti-TIF1γ, anti-PL-12, anti-EJ, and anti-OJ) and MAA (anti-RNP, anti-Ku, and anti-PM-Scl) by ELISA (MESACUP anti-ARS, MDA5 and TIF1γ test, MBL, Nagoya, Japan) and Line blot (EUROLINE Myositis Profile 3, EUROIMMUN, Lübeck, Germany). We extracted MSA/MAA(+) and anti-Ro52(+) patients and analysed the association between the presence of multiple MSA/MAA or anti-Ro52 autoantibodies and patients' clinical features (skin findings such as Gottron papule, heliotrope rash and mechanic's hand, interstitial lung disease, malignancy, and arthritis).
Results: 53/58 patients were positive for MSA/MAA, followed by anti-Ro52 (n=27), anti-PL7 (n=12), anti-Jo1 (n=8), anti-PM-Scl75 (n=7), anti-Ku (n=6), anti-SRP (n=4), anti-EJ (n=4), anti-TIF1γ (n=4), anti-MDA5 (n=4), anti-Mi2 (n=3), anti-PL12 (n=1), anti-PM-Scl100 (n=1), and five cases were MSA/MAA(-). Five patients had multiple MSA/MAA (anti-PL-7+anti-Jo-1, anti-PL-7+anti-PL-12, anti-PL-7+anti Ku, anti-Ro52 +anti-Jo1+anti-PM-Scl75, and anti-Ro52 +anti-Jo1+anti Ku). In patients with DM/PM, anti-Ro52(+) patients were complicated by interstitial lung disease (ILD) and had a higher KL-6 value at onset ($p<0.05$). Notably, 17/27 patients with anti-Ro52 autoantibodies were also anti-ARS(+) and all patients with anti-ARS autoantibodies had ILD. In contrast, among the anti-ARS(−) patients (n=35), there was no significant correlation between anti-Ro52(+) and ILD. Therefore, anti-ARS may be a greater risk factor for ILD than anti-Ro52. Furthermore, patients with multiple MSA/MAA had a higher incidence of rapidly progressive ILD ($p<0.05$). There was no correlation between multiple MSA/MAA or anti-Ro52 autoantibodies and CK or muscle symptoms.
Conclusions: We hypothesise that the presence of multiple MSA/MAA may be useful for predicting ILD. However, in this study, anti-Ro52 autoantibodies did not correlate with ILD and myositis.

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AB0805 CLINICAL FEATURES AND OUTCOME IN SCLERODERMA RENAL CRISIS

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Background: Systemic sclerosis (SSc) is a chronic, autoimmune disease of the connective tissue that involves skin, subcutaneous tissue, muscles and joints, as well as the internal organs: kidneys, lungs, heart. Depending on the extent it can occur as limited (lcSSc) or diffuse (dcSSc) clinical variant. About half of the patients with renal involvement the clinical manifestation is limited to a moderate increase in serum creatinine, mild proteinuria, and moderate hypertension. The most serious complication remains sclerodermal renal crisis (SRC). There was significant heterogeneity in definitions. As a rule for SRC was characterised by new-onset severe hypertension, hypertensive encephalopathy and seizures, acute kidney injury with oliguria, proteinuria and erythrocyturia, and microangiopathic hemolytic anaemia with thrombocytopenia.
Objectives: The aim of our study was to analyse the clinical features and outcome of patients with scleroderma renal crisis.
Methods: We retrospectively reviewed the medical records of patients with scleroderma renal crisis between January 2002 and December 2016. Conditional logistic regression and multivariate analysis was performed to determine factors independently associated with outcome in patients with SRC.
Results: 18 (14 females and 4 males, median age 47,3±14,6 years) patients diagnosed with SRC were included in the study. The mean duration from first symptom of systemic sclerosis (Raynaud's phenomenon, cutaneous sclerosis, arthralgia/arthritis, puffy hands, interstitial lung disease, pulmonary arterial hypertension, digestive hypomotility and etc.) to SRC attack was 4,47±2,9 years. 13 SRC patients belonged to dcSSc, and 5 patients – to lcSSc. Among SRC patients, 9 were negative of anti-centromere antibodies (ACAs). All these 18 patients had hypertension and renal insufficiency, including 4 dialysis dependent after the onset of SRC and 6 with thrombotic microangiopathy. There were 6 patients receiving renal biopsy (the pathological findings were mainly summarised as intimal thickening and stenosis of renal arterioles). Among 16 patients with long-term followed-up, 15 patients received angiotensin converting enzyme inhibitors (ACEI), 8 patients died, 1 patient was dialysis dependent. Survival from first symptom in those patients with Raynaud's phenomenon mode of onset was higher at 5 years and at 10 years than those with onset as non-Raynaud's phenomenon ($p < 0,05$). In multivariate analysis, factors related to mortality in SRC were older age at onset, male gender, treatment with corticosteroids, dcSSc subset, interstitial lung disease, pulmonary arterial hypertension, heart involvement, and the mode of onset with non-Raynaud's phenomenon, especially in the form of pulmonary involvement. The mode of onset should be considered an independent prognostic factor in SRS. The treatment of SRC relies on aggressive blood pressure control with an ACEI, combined with other antihypertensive drugs if needed.
Conclusions: SRC usually occurred at the early course of SSc. dcSSc was more frequent than lcSSc. ACAs were found in 50% of SRC patients. The use of ACEIs was still the cornerstone of SRC treatment.

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AB0806 THE IMPACT OF DYSPHAGIA IN IDIOPATHIC INFLAMMATORY MYOSITIS: AN ONLINE SURVEY OF HIGHLY-SPECIALISED CENTRES

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Background: Dysphagia represents a frequent and disabling symptom in patients with idiopathic inflammatory myopathies (IIM). Despite this, there are not widely accepted diagnostic and therapeutic guidelines for dysphagia in IIM.
Objectives: This study is aimed at surveying the approach to dysphagia in IIM patients in European and worldwide referral centres.
Methods: An anonym on-line survey was designed and clinicians from European Union (EU) countries, and countries from North and Central America and South Korea were invited via email to participate. The questionnaire included 11 items about the characteristics of the hospital of the responder, the evaluation of the impact of dysphagia on disease severity assessed on a visual analogue scale (10 VAS), the techniques used for the assessment of dysphagia, the use of validated patients reported outcome (PRO) and the therapeutic approach of dysphagia.
Results: Between December 2017 and January 2018, 52 clinicians from different centres working in 21 different countries (18 EU countries, 3 non-EU countries) completed the survey. The total numbers of patients followed in the participating centres were 3817 with an average number of patients followed in each centre of 75 (±83). The majority of centres followed only adult patients,[45] 2 only paediatric patients (<18 years) and 5 both. The impact of dysphagia on disease severity was considered severe by all the participants with a mean VAS score of 7.3. All but one centre routinely ask the patients for the presence of dysphagia during the clinical examination. The assessment of dysphagia is performed using validated PRO questionnaires in only 7 centres (SWAL-QOL 2 centres, EAT-10 2 centres, MDADI 2 centres, UCLA-GIT 1 centre); 2 centres evaluate dysphagia using a graduate dysphagia scale by means a 10 cm VAS, and 2 routinely screen patients by a functional test (time necessary to drink a glass of water).
In all the centres an instrumental evaluation of esophageal motility is performed: esophageal manometry in 24 centres, videofluoroscopic swallowing exam in 17, esophageal barium x-rays in 16, and pHmetry and functional endoscopy in 14 each. Esophageal scintigraphy and esophagogastroscopy were respectively performed in 5 and 3 centres. The presence of dysphagia greatly influences the therapeutic approach to IIM in 49 centres leading to an increase of the corticosteroids dosage (11 centres), a change of the immunosuppressive treatment[12] or the initiation of intravenous immunoglobulins (IvIg).[26]
Conclusions: This study suggests that the approach to dysphagia is variable, but dysphagia has an impact on IIM patients and influences the therapeutic

decisions of the treating physician. Different therapeutic strategies are adopted regarding the treatment, and in many cases highly expensive drugs as Ivlg are prescribed. Further studies and recommendations are certainly needed for homogenising the patient management for this severe aspect of the disease.
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AB0807 PLATELET INDICES AS MARKERS OF INFLAMMATION IN SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is a connective tissue disease involving multiple organs with an unknown etiology. Platelet function may be associated with endothelial dysfunction and immune regulatory mechanisms. Recently, an increased tendency to platelet aggregation and enhanced platelet activation have been described in SSc patients, suggesting a role for platelets in the disease itself.[1 2]
Objectives: To evaluate platelet indices in systemic sclerosis (SSc) patients and identify their clinical significance as novel inflammatory biomarkers in correlation to markers of endothelial dysfunction: vascular endothelial growth factor (VEGF) and flow mediated dilatation (FMD).
Methods: Thirty-five SSc patients were enrolled in addition to thirty-five age and sex matched healthy volunteers as controls. All patients and controls underwent full medical history taking, thorough clinical examination, assessment of severity extent of skin sclerosis using the modified Rodnan skin score, erythrocyte sedimentation rate (ESR), C- reactive protein (CRP), complete blood count with special consideration to mean platelet volume (MPV), platelet distribution width and platelets count, assay for serum VEGF concentration, and brachial FMD assessment by colour duplex sonography.
Results: There was a highly significant decrease in the mean MPV in SSc patients compared to the controls (8.65±0.6 fl vs. 9.55±0.52 fl). There was a significant increase in the mean platelet count in SSc patients compared to controls ($331.63±64.66 \times 10^3$/ml vs. $297.80±44.48 \times 10^3$/ml). In SSc patients, a significant negative correlation was found between the mean MPV and each of ESR, CRP and VEGF (r=−0.42,–0.368 and −0.55 respectively, p<0.05); and a significant positive correlation was found between the mean MPV and mean FMD (r=0.378, p<0.05). Linear regression test, showed an association between mean MPV and each of ESR and CRP (t=−3.312,–2.92 respectively, p<0.05).
Conclusions: MPV levels could be an easily measurable parameter to reflect the inflammatory condition and disease activity in systemic sclerosis patients.

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AB0808 MULTICENTRIC STUDY OF SYSTEMIC SCLERODERMA IN TUNISIA

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Background: Scleroderma is an autoimmune connective tissue disorder which is characterised by fibrosis of visceral organs, skin and blood vessels. This condition can be localised or systemic. Its estimated prevalence is 250 cases in a million and it is more common in women than in men.
Objectives: The aim of this retrospective multicenter study was to analyse demographic, clinical, laboratory features and outcome of SSc in Tunisia throughout 8 Departments of Internal Medicine and to compare them with those of other geographic groups.
Methods: One hundred and eight cases of SSc were recorded (American College of Rheumatology criteria) during a 15 years period. They were 93 women and 15men with an average age at SSc onset of approximately 46.9 years.[17–75]
Results: Only 18 patients had limited cutaneous SSc, 11 patients had a CREST syndrome. Our Tunisian patients were characterised by a high frequency of cutaneous signs: sclerodactily (80.6%), proximal sclerosis (81%), telangiectasia (34.3%), Raynaud's syndrome (91%), pigmentation disorders (36%), ulceration (24%) and subcutaneous calcification (4.6%). The other clinical manifestations were dysphagia in 54% (n=59), pulmonary involvement in 55.6% (n=60), cardiac manifestation in 27% (n=29), arthritis in 19.4% (n=21), renal involvement in 13.8% (n=15), and neurological involvement in 12 cas. 78.7% were antinuclear antibody (ANA) positive, 25% were Antitopoisomerase-I antibodies (anti-Scl-70 antibody) positive and 10% with anti-centromere antibodies.
SSc was associated to Sjögren's syndrome n=27, systemic lupus erythematosus (n=8), Polymyositis (n=5) and Rheumatoid arthritis (n=1)
Treatment regimen included, calcium channel blockers (65.7%), steroids (47%), Colchicine (47.2%), D penicillamine (21%), immuosuppressive therapy was added in patient with partial control (n=13). Median follow-up period was 3.7 years. With the above treatment protocol, (29.6%) patients achieved disease control on treatment, (43.3%) had partial control while (26.8%) showed no response or progressive disease, Six patients died, three of them with scleroderma renal crisis.
Conclusions: The findings of this study documents The high frequency of extensive cutaneous sclerosis, Potential limitations and biases in our study need discussion. Specific recruitment of patients in tertiary referral centres may be the source of selection bias. Patients were evaluated by different doctors. The therapeutic management and outcome monitoring were heterogeneous. This study remains the most representative of Tunisian Scleroderma patients recruited from all parts of Tunisia

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AB0809 A NEW SCORE TO PREDICT DIGITAL ULCERS COMBINING CLINICAL DATA, IMAGING AND PATIENT HISTORY IN SYSTEMIC SCLEROSIS

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Background: Ischaemic complications such as digital ulcers (DU) are a common complication in systemic sclerosis (SSc) patients.
Objectives: The aim of this study was to combine clinical characteristics and imaging methods to a composite predictive score.
Methods: Seventy-nine SSc patients received clinical examination and their patient history was taken. Furthermore, we performed nailfold capillaroscopy (NC), colour Doppler ultrasonography (CDUS) and fluorescence optical imaging (FOI) of the hands at baseline. Newly developed digital ulcers over a period of approximately 12 months were registered. We used criteria with significant (p<0.5) OR values above 3.5 in regard to the development of these new DU to create the score (**CIP-DUS**, **c**linical features, **i**maging, **p**atient history – **d**igital **u**lcer **s**core)
Results: Twenty-nine percent of the patients developed new DU during follow-up (48.1% diffuse SSc, 18.4% limited SSc). The following criteria were used: SSc diffuse subtype (OR 4.127, p=0.0087), modified Rodnan skin score >8 (OR 9.429 [95%CI: 3.0–29.2], p<0.0001), pulmonary arterial hypertension (OR 6.854 [95% CI: 1.6–9.7], p=0.0088), present digital ulcers or pitting scars at baseline (OR 15.71 [95%CI: 3.3–74.3], p<0.0001), history of digital ulcer or pitting scars (OR 36.15 [95%CI: 2.1–626.9], p<0.0001), NC pattern (OR 18.6 [95%CI: 1.1–326.4], p=0.0035), reduced capillary density (n<7/mm) in digit III of the right hand in NC (OR 9.0 [95%CI: 1.1–73.6], p=0.0266), missing initial enhancement in FOI in digit III of the right hand (OR 3.857 [95%CI: 1.2–12.8], p=0.0323), percentage of pathologic (i. e. narrowed or occluded) vessels>35% in CDUS (OR 4.286 [95%CI: 1.5–12.4], p=0.0099). Criteria with greater OR should impact the score to a higher degree so we appointed three points to dichotomous criteria with OR >10, two points for criteria with OR between 5–10, and one point for criteria with OR <5. Regarding the NC pattern, 3 points were given to patients with late pattern, 2 points for active and 1 point for early pattern.
Best results were found for a cut-off of >10 points with obtained sensitivity levels of 95% and specificity levels of 74% in regard to new DU (AUC=0.8687, p<0.0001). In the absence of CDUS and FOI data, specificity levels dropped slightly to 72% with unchanged sensitivity values of 95%.