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-TIF1 γ , 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 TIF1 γ 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-TIF1 γ (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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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.6512

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 scleroderma 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 $\pm$ 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 $\pm$ 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 SRC. 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.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.1693

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 ($\pm$ 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 esophagogastrosocopy 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 treatment¹² 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