Comparison of the Rituximab (RTM) in Monotherapy Regimen and Mycophenolate Mofetil (MMF) Efficacy and Safety in Systemic Sclerosis (SSc) with Interstitial Lung Disease (ILD)

O. Konueva1, L. Ananieva1, L. Garzanova1, O. Desinova1, O. Ovsyannikova1, M. Starovoytova1, I.V. Nasonova Research Institute of Rheumatology, Moscow, Russian Federation

Background: Although ILD occurs in the majority of patients with SSc, treatment options for this manifestation is empirical and at present consists of cyclophosphamide or MMF. However, the immunosuppressants (IS) use leads to rather limited improvement of ILD and is associated with many adverse reactions. The search for novel, more efficacious agents has been continued, such as attracting much attention RTM.

Objectives: To compare the impact of MMF and RTM a single-agent therapy on SSc clinical manifestation and activity, and the safety of these agents in the open-label prospective non-randomized study.

Methods: 80 patients with the confirmed SSc diagnosis and ILD evidence based on MSCT findings were enrolled into the study. All patients received low and moderate-dose glucocorticoids regimen. Group A(n=35) received RTM as a single therapy agent for 13.3±2.3 months at total dose 1.33±0.5g (the patient's average age was 45.0±15.15 years, with female proportion 80%; SSc duration 6.3±3.3 years; diffused/localized forms 1:1). Group B(n=36) received MMF for 12.4±6 months at total dose 0.6±0.5g (the average age 45±13.5 years, females 91%, SSc duration 7.1±5 years, diffused/localized forms 1:1). The time courses of FVC, DLCO, modified skin count (mRss, points), activity index (EScSG, points), and cardiac rhythm and conductivity disorders (ECG) were assessed into the study.

Results: In Groups A and B the therapy was associated with significant decrease in mRss (p=0.02 and 0.09, respectively) and EScSG (p=0.00017 and 0.000165, respectively). Reducing the number of patients with cardiac conductivity disorders was observed only in MMF-treated patients (p=0.03). Evaluation of FVC time course revealed significant FVC increase only in Group A (p=0.002), with median increment about 5%. In Group A 10% FVC increase was found in the third of the patients thus exceeding respective parameter in Group B (p=0.2). The patient percentage with FVC decrease by ≥10% did not differ significantly between groups. During the follow-up period no change of the other studied parameters was observed.

The therapy was better tolerated in RTM-treated group: during RTM therapy no change of the other parameters was observed. The therapy was better tolerated in RTM-treated group: during RTM therapy no change of the other parameters was observed.

Conclusion: Both agents effectively alleviated skin induration and EScSG, and significantly improved FVC. However, only RTM use revealed significant FVC increase including clinically significant FVC increase. RTM was slightly better tolerated compared to MMF. The study findings substantiate potential use of RTM both as a first-line agent for ILD treatment in the patients with SSc, and in the event of IS inefficacy of poor tolerability. The MMF use is more preferable in both as a first-line agent for ILD treatment in the patients with SSc, and in the event of IS inefficacy of poor tolerability. The MMF use is more preferable in the patients with less pronounced ILD and cardiopathy.

Disclosure of Interests: None declared

Prognostic Factors for Steroid-Free Remission in Patients with Idiopathic Inflammatory Myopathies: Importance of Anthropometric Measurements

J. S. Lee1, S. H. Nam2, S. J. Choi3, W. J. Seo2, S. Hong2, C. K. Lee1, B. Yoo1, J. S. Oh1, Y. G. Kim1, University of Ulster College of Medicine, Al- Asan Medical Center, Division of Rheumatology, Department of Internal Medicine, Seoul, Korea, Rep. of (South Korea); 2Seoul Veterans Hospital, Division of Rheumatology, Department of Internal Medicine, Seoul, Korea, Rep. of (South Korea); 3Asan Medical Center, Department of Biomedical Informatics, Seoul, Korea, Rep. of (South Korea)

Background: Several studies have been conducted on factors associated with mortality in idiopathic inflammatory myopathies (IIM), but few studies have assessed prognostic factors for steroid-free remission in IIM.

Objectives: We investigated the various clinical factors, including body measurements, that affect IIM treatment outcomes.

Methods: Patients who were newly diagnosed with IIM between 2000 and 2018 were included. Steroid-free remission was defined as at least three months of normalisation of muscle enzymes and no detectable clinical disease activity. The factors associated with steroid-free remission were evaluated by a Cox regression analysis.

Results: Of the 106 IIM patients, 35 displayed steroid-free remission during follow-up periods. In the multivariable Cox regression analyses, immunosuppressants’ early use within one month after diagnosis [hazard ratio (HR) 6.21, 95% confidence interval (CI) 2.61–14.74, p < 0.001] and sex-specific height quartiles (second and third quartiles versus first quartile, HR 3.65, 95% CI 1.40–9.51, p = 0.008 and HR 2.88, 95% CI 1.13–7.32, p = 0.027, respectively) were positively associated with steroid-free remission. Polymyositis versus dermatomyositis (HR 0.21, 95% CI 0.09–0.53, p = 0.001), presence of dysphagia (HR 0.15, CI 0.05–0.50, p = 0.002) and highest versus lowest quartile of waist circumference (WC) (HR 0.24, 95% CI 0.07–0.85, p = 0.027) were negatively associated with steroid-free remission.

Conclusion: The early initiation of immunosuppressant therapy, type of myositis and presence of dysphagia are strong predictors of steroid-free remission in IIM; moreover, height and WC measurements at baseline may provide additional important prognostic information.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.3010

In Myositis Patients, Sjögren’s Syndrome is Associated with Inclusion Body Myositis and with Anti-CN1A Antibodies Indepenently of the Myositis Subtype

D. Levy1, B. Nespoli1, M. Giannini2, R. Felten3, C. Varoquier1, M. Rinagel2, A. S. Korgaonkar4, V. Poindron1, T. Martin1, F. Maurier5, H. Cherfell6, B. Bouldoire1, B. Hervier7, C. Lenormand7, L. Arnaud7, B. Geny1, J. Sibilia1, J. E. Gottenberg1, A. Meyer1, 1Strasbourg, Strasbourg, France; 2Hôpitaux Universitaires, Strasbourg, France; 3Hôpital Privé, Metz, France; 4Pontarlier, Pontarlier, France; 5Colmar, Colmar, France; 6Paris, Paris, France

Background: Myositis are characterized by weakness and muscle inflammation. They encompass heterogeneous conditions, which include dermatomyositis (DM), inclusion body myositis (IBM) and polymyositis (PM) according to the EULAR/ACR 2017 criteria. We recently recorded a high prevalence of IBM in a cohort of primary Sjögren’s syndrome (SS) (1). The signification of SS in the setting of myositis is unanswered.

Objectives: To refine the signification of SS in the setting of myositis.

Methods: Among a monocentric myositis cohort (according to the EULAR/ACR 2017 criteria), SS patients (according to the ACR/EULAR 2016 criteria) were identified (myositis/SS+ group) and compared to myositis patients without SS (myositis/SS- group).

Results: Among 414 myositis patients, SS criteria were available for 96 patients. Thirty-two (33%) presented SS. Patients with SS tended to be more frequently women (F/M ratio 9.7 vs 3.0, p = 0.07). Age at diagnosis of myositis was similar in both groups (53 years [range 21-74] vs 53 years [range 16-77], p = 0.51). Myositis subtypes reparation (as defined by EULAR/ACR 2017 criteria) was different in myositis/SS+ and myositis/SS- groups (p = 0.021), IBM being four-fold more prevalent in myositis/SS+ group (25% vs 6%, p = 0.018). Accordingly, the delay between the first muscle symptoms and myositis diagnosis was longer in myositis/SS+ group (7 months [0-336] vs 4 months [0-122], p = 0.041). Moreover, anti-cN1A antibodies, myositis-specific antibodies were less frequently found in myositis/SS+ patients than in myositis/SS- ones (16/32 [50%] vs 46/64 [72%], p = 0.035).

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.4235
Anti-cNIA antibodies were more prevalent in myositis/SS+ patients (33% vs 5.8%; p = 0.0032). However, in myositis/SS- group, anti-cNIA was frequent in each of the EULAR/ACR 2017 myositis subtypes and the association between SS and anti-cNIA positivity was maintained in a multivariate analysis adjusted with the diagnosis of IBM (p = 0.023).

Seven of the myositis/SS+ patients (22%) had systemic involvement typical of SS (vs 6% of the myositis/SS- patients; p = 0.12) including polynuropathy (6% vs 6% [10%] and 2 cyclophosphamidic vasculitis [1%] vs 1% [6%]). In addition, 2 (6%) myositis/SS+ patients developed a lymphoma (one diffuse large lymphoma of the parotid and one non-Hodgkin lymphoma), vs none of the myositis/SS- patients (p = 0.011). Only one (3%) of the myositis/SS+ patients developed myositis-associated cancer (diagnosed within 3 years of myositis diagnosis) versus 6% of the myositis/SS- patients (p = 0.66).

Aside hydroxychloroquine, more frequently used in myositis/SS+ group (38% vs 16%, p = 0.018), no significant difference was found in the management of the patients taking into account the myositis subtype.

Conclusions: Myositis patients with SS have more frequently IBM than myositis patients without SS. They also have more frequently anti-cNIA antibodies, independently of the myositis subtype. They might develop systemic complications of SS.

References:

Disclosure of Interests:
Dan LEVY: None declared, Benoit Nespola: None declared, Coralie Varoquier: None declared, Anne-Sophie Korgaonkar: None declared, Vincent Poindon: None declared, Thierry Martin: None declared, Francois Maurier: None declared, Hassam Chereih: None declared, None declared, None declared, None declared.

Scientific Abstracts

DOI: 10.1136/annrheumdis-2020-eular.5990

Conclusion: Only 28.7% of the patients that were MA/MAA positive had a diagnosis of IBM. Other autoimmune diseases and ILD were commonly found in this group of MA/MAA positive patients.

References:

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.5990

Background: Idiopathic inflammatory myopathies (IIM) are a group of immune-mediated diseases characterized my muscle weakness, skin rash and systemic involvement. Myositis-specific antibodies (MSA) and myositis-associated antibodies (MAA) play a major role in IIM diagnosis, classification and prognosis. Nevertheless, MSA/MAA testing is not standardized and there very few studies addressing their relationship with other diseases.

Objectives: To describe a cohort of patients tested positive for MAA/MAA, and to explore its relationship with IIM and other autoimmune diseases.

Methods: We retrospectively review all the serum samples obtained from patients tested for MSA/MAA during 2019 in the Immunology department of Ramón y Cajal University Hospital, Madrid, Spain. These antibodies were tested by spe- cimen IGM antibody (EUROLINE: Autoimmune Inflammatory Myopathies 16 Ag) with highly purified MAA/MAA. Positivity was established according to absorbance titer and adjusted by positive control of each test (arbitrary units, AU). Patients were diagnosed with IIM according to their clinician diagnosis. Diagnosis and classification were confirmed by an independent rheumatologist (JL) according to current understanding of IIM classification.

Results: Three-hundred-seventy-five samples were tested for MSA during the study period. Two-hundred-seventy-nine were negative for all antibodies tested. Ninety-six samples were positive for one or more MSA/MAA, corresponding to 74 patients (11 patients had 2 different samples). Forty-nine (66.2%) of the patients who tested positive were female and 25 (33.8%) were male. Mean age was 56.65 years. Only 22 patients (29.7%) had a confirmed diagnosis of IBM, 24 (32.4%) had a diagnosis of other autoimmune disease, and 11 (14.9%) were diagnosed with interstitial lung disease (ILD) (Figure 1). Six ILD patients had anti-PM-Scl or anti-Ku antibodies, which are associated with scleroderma or overlap-CTD myositis, nevertheless, they remained classified as ILD as no other features were described in this group.

Seventeen patients were positive for more than 1 MAA or MSA, including 14 patients positive for anti-Ro-52. Antibody titer was higher in the IIM group compared to non-MSA group (59.59 vs 44.16, p=0.015). Anti-M2-4 was positive in 4 ILD without any other myositis features, and high titer anti-SSP (n=4, mean 59.75 AU) was found in primary biliary cirrhosis (PBC) patients. Additionally, 5 patients positive for anti-Jo-1 using ELIA (Thermo Fisher) were diagnosed with antisynthetase syndrome. IIM diagnosis and its relationship with antibody titer is represented in table 1.

Background: Interstitial lung diseases (ILD) could originate from idiopathic cause or secondary to connective tissue diseases (CTD). The most common causes of gated associated ILD (CTD-ILD) include mixed connective tissue disease, systemic sclerosis (SSc), inflammatory myositis such as polymyositis (PM) and dermatomyositis (DM). Our preliminary data had demonstrated that ILD is not uncommon in patients with rheumatoid arthritis (RA) and Sjögren’s syndrome.