Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.2317

POST355
THE REMITTING SERONEGATIVE SYMPTOMATIC SYNOVITIS WITH PITTING EDEMA SYNDROME (RS3PE): REVIEW OF TEN YEARS AT A REFERENCE HOSPITAL
S. García Pérez1, J. L. Modesto Dos Santos2, J. Mendizábal3, M. López I. Gómez2, G. Sada2, J. J. Restrepo Vélez3, J. J. Elejade5, L. Garrido5, Couril1, V. Aldasoro1, I. Pianigau Ziaude3, R. Gutierrez3, L. Horcada1, F. J. Annichérico5, N. Del Val del Amo2, M. C. Fito Manteca1, J. Sanchez Alvarez1.1 Complejo Hospitalario de Navarra (CHN), Rheumatology, Pamplona, Spain; 2Complejo Hospitalario de Navarra (CHN), Internal Medicine, Pamplona, Spain

Background: The Remitting Seronegative Symmetrical Synovitis with Pitting Edema Syndrome (RS3PE) is a rare rheumatological disease, considered a benign process.

Objectives: This study aims to describe its clinical features and serological markers, and also to analyze its possible association with neoplasms.

Methods: An observational retrospective study was performed to assess demographic and clinical characteristics of patients diagnosed from RS3PE at a reference hospital amongst the Rheumatology and Internal Medicine departments, from 2010 to 2021.

Results: Twenty-seven patients were included, with a mean age of 82.74 y.o. (IC95%: 80.45-85.04; range 66 to 93) and a 51.85% proportion of males. Only 22.22% were from rural areas.

All patients presented bilateral hand edema although some associated feet edema (40.74%) or morning stiffness (70.37%). Blood tests demonstrated anemia in 44.44% of patients. Inflammatory markers were elevated, such as CRP (mean 151.49 mg/L, IC95% 138.42-164.56; range 12.17-404.97, erythrocyte sedimentation rate (33.74 mm/hour, IC95% 24.22-43.26) and fibrinogen (531.96 mg/dL, IC95% 482.91-580.30). Only a few patients presented any autoimmune serological marker such as antinuclear antibodies (18.18%) or rheumatoid factor (8.70%).

X-ray screening was realized to 22 patients. 14 showed of osteoarthritis radiologic presentation, 4 had radiological findings of chondrocalcinosis and one of them presented both. Only one patient had bone erosion.

All but one patient received low-dose corticosteroids, with a good and rapid response in all cases. Three patients received treatment with methotrexate (2) or leflunomide (1).

Conclusion: RS3PE must be contemplated in elderly patients presenting with bilateral hand pitting edema and articular symptoms. No specific biomarkers have been described, but inflammatory reaction is often found in the absence of rheumatoid arthritis biomarkers. Rapid response to corticosteroids is prevalent. Only two neoplasms were detected during follow-up.

REFERENCES:

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.2715

POST357
OCULAR SCLERAL PATHOLOGY. UNDERLYING DISEASES AND SYSTEMIC TREATMENT. STUDY OF 175 PATIENTS FROM A SINGLE UNIVERSITY CENTER
L. Sanchez-Bibao1, V. Calvo-Rio2, J. L. Martin-Varias3, C. Alvarez-Reguera4, A. Herrera-Moran5, I. Gonzalez-Mazon6, R. Demetrio-Pablo6, M. A. Gonzalez-Gay7, R. Blanco1, H.U. Marques de Valdecilla, Rheumatology, Santander, Spain; 2H. Sierrallana, Rheumatology, Torrelavega, Spain; 3H. Sierrallana, Rheumatology, Torrelavega, Spain; 4H. U. Marques de Valdecilla, Ophthalmology, Santander, Spain

Background: Ocular scleral pathology (OSP) includes episcleritis and scleritis. Episcleritis is generally a benign disease with a self-limited course, while scleritis is a more severe ocular condition. In some severe and refractory cases systemic therapy may be required.

Objectives: In a wide series with OSP our aim was to assess a) underlying diseases and b) systemic treatment.

Methods: Study of unselected consecutive patients studied in a single University Hospital during the last ten years with: a) episcleritis and b) scleritis diagnosed by clinical features and slit-lamp (Watson and Hayreh criteria). Best corrected visual acuity (BCVA) and intraocular pressure (IOP) were measured at diagnosis and after systemic treatment.

Results: We studied 175 patients (106 women/ 69 men) /212 affected eyes with OSP (episcleritis=135; scleritis=40); mean age 48.9±14.2 years. OSP was unilateral in 138 (78.9%), recurrent in 74 (42.9%) and chronic in 21 (12%). Most of them were idiopathic (n=81, 46.3%) while associated with IMD were 43.4% (Table 1). The most important underlying IMD were spondyloarthritis and inflammatory bowel disease, without significant differences between scleritis and episcleritis. Granulomatosis with polyangiitis and systemic lupus erythematosus were more frequent in scleritis, not reaching statistical significance.

Regarding treatment, topical treatment was used in all patients. 41.1% received systemic treatment, including systemic glucocorticoids, cDMARDS and completely abrogates T follicular helper (Tfh)-cells dependent germinal center reactions leading to de-novo plasmablast differentiation.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.2271

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.2646

POST356
PERSISTENCE OF TFH CELLS AFTER RITUXIMAB IS ASSOCIATED WITH IG4-RELATED DISEASE RELAPSE
G. Mancuso1,2, T. Jofra3, M. Lanzillotta3, J. Gerosa4, G. Di Colò5, L. Dagna2, G. Fusteri1, E. Delia Torre1.1 Università Vita-Salute San Raffaele, Allergologia ed Immunologia clinica, Milano, Italy; 2Ospedale San Raffaele, Immunologia Reumatologia, Allergologia e Malattie Rare, Milano, Italy; 3Università Vita-Salute San Raffaele, Diabetes Research Institute (DRI), Milano, Italy; 4Università Vita-Salute San Raffaele, Internal medicine, Milano, Italy

Background: Clinical improvement after B-cell depletion with rituximab suggests a prominent pathogenic role of B-lymphocytes in Ig4-related disease (IG4-RD).

Ig4-RD, however, relapses in most cases together with re-expansion of clonally divergent plasmablasts indicating that treatment with rituximab does not
Table 1. Underlying diseases and systemic treatment.

<table>
<thead>
<tr>
<th>UNDERLYING DISEASE</th>
<th>Overall</th>
<th>Episcleritis</th>
<th>Scleritis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (years), mean ± SD</td>
<td>(n=175)</td>
<td>(n=135)</td>
<td>(n=40)</td>
</tr>
<tr>
<td>Sex (women), n (%)</td>
<td>48.9 ± 14.2</td>
<td>47.8 ± 14.3</td>
<td>52.6 ± 13.9</td>
</tr>
<tr>
<td>Underlying disease, n (%)</td>
<td>106 (60.6)</td>
<td>81 (60)</td>
<td>25 (62.5)</td>
</tr>
<tr>
<td>Idiopathic, n (%)</td>
<td>81 (46.3)</td>
<td>65 (48.1)</td>
<td>16 (40)</td>
</tr>
<tr>
<td>Infectious, n (%)</td>
<td>73 (4.6)</td>
<td>5 (3.7)</td>
<td>2 (5)</td>
</tr>
<tr>
<td>IMID, n (%)</td>
<td>76 (44)</td>
<td>57 (42.2)</td>
<td>19 (47.5)</td>
</tr>
<tr>
<td>Spondyloarthritis</td>
<td>21 (12)</td>
<td>17 (12.6)</td>
<td>4 (10)</td>
</tr>
<tr>
<td>Crohn's disease</td>
<td>16 (9.1)</td>
<td>14 (10.4)</td>
<td>2 (5)</td>
</tr>
<tr>
<td>Rheumatoid Arthritis</td>
<td>14 (8)</td>
<td>12 (8.9)</td>
<td>2 (5)</td>
</tr>
<tr>
<td>Granulomatosis with polyangiitis</td>
<td>7 (4)</td>
<td>3 (2.2)</td>
<td>4 (10)</td>
</tr>
<tr>
<td>Relapsing polychondritis</td>
<td>6 (3.4)</td>
<td>4 (3)</td>
<td>2 (5)</td>
</tr>
<tr>
<td>Systemic lupus erythematosus</td>
<td>5 (2.9)</td>
<td>2 (1.5)</td>
<td>3 (7.5)</td>
</tr>
<tr>
<td>Uveolitic colitis</td>
<td>3 (1.7)</td>
<td>2 (1.5)</td>
<td>1 (2.5)</td>
</tr>
<tr>
<td>Systemic treatment, n (%)</td>
<td>72 (41.1)</td>
<td>37 (27.4)</td>
<td>35 (87.5)</td>
</tr>
<tr>
<td>Methotrexate, n (%)</td>
<td>27 (15.4)</td>
<td>19 (14.1)</td>
<td>8 (20)</td>
</tr>
<tr>
<td>Non-methotrexate DMARD, n (%)</td>
<td>8 (4.6)</td>
<td>5 (3.7)</td>
<td>3 (7.5)</td>
</tr>
</tbody>
</table>

*p<0.05, p=0.05

Figure 1. BVCA and IOP at diagnosis and last visit.

Conclusion: OSP is a relatively frequent entity. It is necessary to exclude an underlying systemic disease to establish correct systemic treatment.

Disclosure of Interests: Lara Sanzíl-Blasco: None declared, Vanessa Calvo-Rio

Methods: BD patients who had treated with apremilast for active oral ulcers were included in the study. We investigated the improvement rate of oral and genital ulcers, skin lesions, arthritis. In addition, serum cytokines (IFN-γ, IL-10, IL-8, and TNF-α) before and after apremilast treatment were measured using a multiplex immunoassay (Luminex Assay, R&D Systems).

Results: Fourteen patients (3 males and 11 females) were enrolled in this study. The mean age was 46.6 ± 13.0 years and the mean duration of disease was 10.2 ± 8.8 years. All patients had active oral ulcers, five had genital ulcers, six had skin lesions, and four had arthritis. Three months after the treatment with apremilast, oral ulcers improved in 13 patients (92.9%). The improvement rates of genital ulcers, skin lesions and arthritis were 60%, 25% and 25%, respectively.

Changes in serum cytokines were different from those previously reported in PS. Adverse events were gastrointestinal symptoms such as nausea and diarrhea in 6 patients and sensorineural deafness in 1 patient. Medication was reduced in 2 patients, and discontinued in 1 patient due to nausea and diarrhea.

Conclusions: Apremilast is useful but only for active oral ulcers, and less useful for other lesions in BD patients. The effect of apremilast for other domain such as genital ulcers, skin lesions, arthritis, was not comparable to that of active oral ulcers. Additionally, BD may have different cytokine profile from PS and PfA.

REFERENCES:


Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.2772

POS1358

BACKGROUND

Familial Mediterranean fever (FMF) is a hereditary autoinflammatory disease accompanied by recurrent attacks of fever and serositis. It is prevalent among Mediterranean populations, mainly Turks, Armenians; Jews and Arabs. As genetic factors are variable in the population, environmental factors can also affect phenotypic characteristics.

OBJECTIVES: This study aimed to determine the geographic differences in disease characteristics and burden in patients with FMF in Turkey.

METHODS: Patients diagnosed with FMF according to the Tel-Hashomer criteria were included in this multi-center study. Patients were included from the different regions of Turkey. The study involves 31 different centers from 9 different regions.

RESULTS: A total of 281 patients with FMF (195 women, 86 men) were enrolled in the study. The mean age of the patients was 12.9 (3.0 to 14.4 years). While the patients in the eastern areas of Turkey were diagnosed earlier age (p<0.001), the patients in the western area had a longer diagnostic delay time (p=0.001). Patients enrolled from western regions tended to have higher ESR and PRASS scores than those from eastern and central Anatolian regions, but attack numbers per 6 months were similar among the regions. The highest proportion of patients who were M694V/M694V homozygous patients were in western, and then eastern and central regions. While fever and arthritis were more common in the eastern, pleuritis and scleritis were more common in the central Anatolia. Peritonitis and erysipelas like erythema rashes were similar among the regions. The majority of patients were receiving colchicine treatment in all three regions. FMF-QoL scores were highest in the eastern and lowest in the western (p=0.006). Patients enrolled in the central Anatolia region experienced more functional disability than those from the western and eastern regions (p=0.009). Anxiety and depression scores were similar between groups (p=0.385 vs p=0.549).

CONCLUSION: These findings suggest that patients with FMF have diversity concerning the age at diagnosis, diagnostic delay time, disease activity, quality of...