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THE REMITTING SERONEGATIVE SYMPTOMATIC SYNOVITIS WITH PITTING EDEMA SYNDROME (RS3PE): REVIEW OF TEN YEARS AT A REFERENCE HOSPITAL


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 years (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 22.22% were from rural areas.

Conclusions: RS3PE must be contemplated in elderly patients presenting with completely abrogates T follicular helper (Thf)-cells dependent germinal center reactions leading to de-novo plasmablast differentiation.

Objectives: In the present work we aim to study the effects of B-cell depletion therapy with rituximab on circulating Tfh cells and on the levels of CXCL13 - a chemotactic factor for B-lymphocytes produced by Tfh cells - in patients with IgG4-RD.

Methods: Thirty patients with IgG4-RD, diagnosed according to the "Consensus Statement on the Pathology of IgG4-RD" and fulfilling the "2019 ACR/EULAR Classification Criteria" were included in the present study. Ten patients with relapsing disease were treated with the anti-CD20 monoclonal antibody rituximab (two 1g infusions 15 days apart). Peripheral blood mononuclear cells and serum were collected before rituximab and three months after infusion. Tfh cells subsets in the peripheral blood were measured by flow cytometry and CXCL13 plasma levels were measured by ELISA assay.

Results: No changes in total Tfh cells and Tfh cells subsets were observed in three months after rituximab neither in absolute counts nor in percentage of CD4+ T cells. In particular, no difference in Thf1, Thf2, Thf17, T follicular regulatory and highly functional Tfh cells counts was observed before and after treatment. The serum level of CXCL13 was significantly higher in active untreated IgG4-RD patients compared to healthy controls (151.94 pg/ml vs 66.98 pg/ml, p value = 0.0026), but was not affected by rituximab treatment (p value = 0.41).

Conclusion: In relapsing patients with IgG4-RD rituximab does not affect circulating Tfh cells numbers and serum levels of CXCL13. Persistence of Tfh cells after rituximab and reconstitution of germinal center reactions likely drives IgG4-RD flare.

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OCULAR SCLERAL PATHOLOGY. UNDERLYING DISEASES AND SYSTEMIC TREATMENT. STUDY OF 175 PATIENTS FROM A SINGLE UNIVERSITY CENTER


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 all 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±4.2 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 IMID were 43.4% (Table 1). The most important underlying IMID were spondyloarthritides and inflammatory bowel disease, without significant differences between scleritis and episcleritis. Gout and hepatitis with 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: 10.1136/annrheumdis-2021-eular.2731 on 19 May 2021. Downloaded from http://ard.bmj.com/ Ann Rheum Dis: first published as 10.1136/annrheumdis-2021-eular.2731 on 19 May 2021. Downloaded from http://ard.bmj.com/

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OCULAR SCLERAL PATHOLOGY. UNDERLYING DISEASES AND SYSTEMIC TREATMENT. STUDY OF 175 PATIENTS FROM A SINGLE UNIVERSITY CENTER


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bDMARDs. Systemic glucocorticoids and Methotrexate were used more frequently in scleritis (Table 1). The main indication for biologic therapy was related to underlying IMID in both groups, but 7 bDMARDs in scleritis were indicated for systemic and ocular compromise. BVCA and IOP improved significantly after systemic treatment in scleritis (Figure 1).

Table 1. Underlying diseases and systemic treatment.

<table>
<thead>
<tr>
<th>UNDERLYING DISEASE</th>
<th>Overall</th>
<th>Scleritis</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (years), mean ± SD</td>
<td>48.9 ± 14.2</td>
<td>47.8 ± 14.3</td>
<td>0.061</td>
</tr>
<tr>
<td>Sex (women), n (%): Female</td>
<td>106 (60.6)</td>
<td>81 (60)</td>
<td>0.920</td>
</tr>
<tr>
<td>UNDERLYING DISEASE</td>
<td>Overall</td>
<td>Scleritis</td>
<td>p</td>
</tr>
<tr>
<td>Idiopathic, n (%)</td>
<td>41 (23)</td>
<td>36 (27)</td>
<td>0.436</td>
</tr>
<tr>
<td>Infectious, n (%)</td>
<td>11 (6.3)</td>
<td>7 (5.4)</td>
<td>0.276</td>
</tr>
<tr>
<td>Systemic lupus erythematosus, n (%)</td>
<td>10 (5.8)</td>
<td>7 (5.4)</td>
<td>0.678</td>
</tr>
<tr>
<td>Rheumatoid arthritis, n (%)</td>
<td>10 (5.8)</td>
<td>7 (5.4)</td>
<td>0.678</td>
</tr>
<tr>
<td>Granulomatosis with polyangiitis, n (%)</td>
<td>7 (4)</td>
<td>3 (2.4)</td>
<td>0.400</td>
</tr>
<tr>
<td>Relapsing polychondritis, n (%)</td>
<td>6 (3.4)</td>
<td>4 (3)</td>
<td>0.621</td>
</tr>
<tr>
<td>Systemic sclerosis, n (%)</td>
<td>5 (2.9)</td>
<td>2 (1.5)</td>
<td>3 (7.5)</td>
</tr>
<tr>
<td>Uveitis, n (%)</td>
<td>3 (1.7)</td>
<td>1 (0.8)</td>
<td>0.704</td>
</tr>
<tr>
<td>SYSTEMIC TREATMENT</td>
<td>Overall</td>
<td>Scleritis</td>
<td>p</td>
</tr>
<tr>
<td>Methotrexate, n (%)</td>
<td>72 (41)</td>
<td>37 (27.4)</td>
<td>35 (75)</td>
</tr>
<tr>
<td>Non-methotrexateDMARD, n (%)</td>
<td>39 (22.3)</td>
<td>17 (23.2)</td>
<td>22 (55)</td>
</tr>
<tr>
<td>TNFαDMARD, n (%)</td>
<td>35 (20)</td>
<td>20 (28.6)</td>
<td>15 (30)</td>
</tr>
<tr>
<td>Non-TNFαDMARD, n (%)</td>
<td>27 (15.4)</td>
<td>19 (14.1)</td>
<td>8 (20)</td>
</tr>
</tbody>
</table>

POS1359 REGIONAL DIFFERENCES IN DISEASE CHARACTERISTICS OF FAMILIAL MEDITERRANEAN FEVER IN TURKEY: PRELIMINARY REPORT


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.

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. Clinical features and characteristic of the patients including disease duration, medications, comorbidity conditions, and attack characteristics, amyloidosis, acute phase reactants, FMF gene mutations, arthritis, sacroilitis, and febrile myalgia were recorded. PRASS disease activity score, FMF-Qol, HAQ, and FAQ were assessed.

Results: A total of 281 patients with FMF (195 women, 86 men) were enrolled in the study. The mean age of the patients was 9 (SD: 12.4) 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 Anatolia (19.5%, 18%, and 5.4%). While fever and arthritis were more common in the eastern, pleuritis and sacroilitis were more common in the central Anatolia. Peritonitis and erysipelas like erythema rates 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

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