Table 1. Demographic, clinical and laboratory characteristics of patients with Schnitzler’s syndrome

<table>
<thead>
<tr>
<th>Pts</th>
<th>The age (y)</th>
<th>The age at onset (y)</th>
<th>Diagnosis delay (y)</th>
<th>ESR (&lt;15 mm/h)</th>
<th>CRP (&lt;6 g/l)</th>
<th>M-gradient (g/l)</th>
<th>Anakinra</th>
<th>Canakinumab</th>
<th>The treatment duration (y)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>51</td>
<td>40</td>
<td>4</td>
<td>31</td>
<td>107</td>
<td>7.1</td>
<td>0</td>
<td>1</td>
<td>5</td>
</tr>
<tr>
<td>2</td>
<td>52</td>
<td>38</td>
<td>22</td>
<td>40</td>
<td>29</td>
<td>5.7</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>3</td>
<td>36</td>
<td>29</td>
<td>3</td>
<td>140</td>
<td>44</td>
<td>78</td>
<td>0</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>4</td>
<td>51</td>
<td>53</td>
<td>2</td>
<td>192</td>
<td>96</td>
<td>5.1</td>
<td>1</td>
<td>0.5</td>
<td>0.5</td>
</tr>
<tr>
<td>5</td>
<td>69</td>
<td>66</td>
<td>2</td>
<td>49</td>
<td>96</td>
<td>5.1</td>
<td>1</td>
<td>1</td>
<td>1</td>
</tr>
</tbody>
</table>

Abbreviations: Pts – patients, y- years, ESR – erythrocyte sedimentation rate, CRP – C-reactive protein

monoclonal IgM secretion was revealed in 5 pts, IgMx and IgM– in 1 and IgGx and IgG–. 1 NO NLRP3, TNFRSF1A gene mutations were identified. Prior to the diagnosis, all pts were treated with glucocorticoids with a transient clinical response and a disease relapse after reducing the dose or stopping the treatment. 2 pts failed to respond to methotrexate and 1 – to hydroxychloroquine. 4 pts were prescribed with 150 mg canakinumab, a monoclonal antibody targeting IL-1, subcutaneously once every 8 weeks. The treatment duration varied from 6 months to 5 years. 2 pts, who initially received daily 100 mg anakinra subcutaneously for 2 to 3 months with a positive response, were further treated with canakinumab. During the treatment with canakinumab, all pts rapidly responded with a complete resolution of fever, rash, arthralgias and bone pains, an overall health improvement and a normalization in ESR and CRP levels. The therapy was well tolerated. In 1 patient, the intervals between canakinumab injections were prolonged to 5 months without any evidence of relapse. During this period, the male patient became a parent to a healthy child.

Conclusion: In rheumatology practice SchS can be misdiagnosed with AOSD. AOSD patients should be tested for monoclonal gammopathy. IL-1 inhibitors are a highly effective and well-tolerated treatment option for SchS. In SchS patients with a complete response to canakinumab, injection intervals can be individualized.

Disclosure of Interests: None declared

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AB1058

JOINT HYPERMOBILITY SYNDROME AND PRIMARY OPEN-ANGLE GLAUCOMA

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Background: Eye symptoms: myopia, prolapso of the upper eyelid, epiblepharon in the upper eyelid are small diagnostic criteria for joint hypermobility syndrome (JHS).

There are few publications in the literature on the relationship between JHS and primary open-angle glaucoma (POAG).

It is known that in the development of JHS, the distribution of collagen of types I and III with the predominance of collagen of type III is important, the latter is encoded by the COL3A1 gene. When using POAG in the connective tissue of the middle and deep layers of the sclera by the immunohistochemical method, intense focal accumulation of type I and III collagen was previously revealed, and in the layers of the sclera’s own substance, type III collagen, unusual for it.

Objectives: To study articular and extraarticular clinical manifestations, instrumental, laboratory signs, as well as to conduct molecular genetic studies on the carriage of the Coll3A1 gene in patients with a diagnosis of POAG and compare them.

Methods: Nine consecutive patients with an established diagnosis of POAG (burdened heredity by glaucoma) with arthralgia were sent for consultation to the ‘anaSTILLs’ study (anakinra in Still’s disease) was designed to further evaluate efficacy and safety of anakinra in patients with Still’s disease across all age groups.

Discipline of Interests: None declared

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AB1059

A RANDOMIZED, PLACEBO-CONTROLLED STUDY OF ANAKINRA IN PATIENTS WITH STILL’S DISEASE

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Background: Adult-onset Still’s disease (AOSD) and systemic juvenile idiopathic arthritis (SJIA) are rare autoinflammatory disorders associated with an activated IL-1 pathway, characterized by spiking fever, rash, arthritis, lymphadenopathy, hepatosplenomegaly and serositis. There is a growing understanding that SJIA and AOSD are one disease with different ages of onset, i.e. Still’s disease.

The anaSTILLS study (anakinra in Still’s disease) was designed to further evaluate efficacy and safety of anakinra in patients with Still’s disease across all age groups. The primary objective was to demonstrate efficacy of anakinra versus placebo as assessed by ACR30 response with absence of fever at Week 2. Secondary objectives included: early onset of efficacy, sustained efficacy, time to study drug discontinuation, safety, pharmacokinetics, clinical signs and biomarkers.

Methods: ‘anaSTILLS was a randomized, double-blind, placebo-controlled, 12-week study including patients with active and newly diagnosed (6 months) Still’s disease according to adapted ILAR criteria if <16, or Y amaguchi criteria, if ≥16 years of age at disease onset. Patients were randomized to anakinra 2mg/g (max 100mg/day), 4mg/kg (max 200mg/day) or placebo.

Results: 12 patients were randomized and received study drug: 6 anakinra (2mg/kg n=2, 4mg/kg n=4) and 6 placebo, the study was terminated early due to slow recruitment. 1 patient on placebo had lymphoma, not Still’s disease, and was excluded; thus in total 11 patients were analyzed for efficacy. 8 were children [median (range) age=4.0 (1-11) years] and 3 were adults [median (range) age=32.0 (25-51) years]. 55% were male and the mean symptom duration was 74.2 days. All patients on anakinra but none on placebo achieved ACR30 response according to adapted ILAR criteria if <16, or Yamaguchi criteria, if ≥16 years of age at disease onset. Patients were randomized to anakinra 2mg/g (max 100mg/day), 4mg/kg (max 200mg/day) or placebo.

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ACR30/50/70/90 responses in the anakinra group were sustained throughout the study period. Patients in the anakinra group had a prompt and persistent decrease in CRP and ferritin levels at Week 1, which was not observed in the placebo group. There were no unexpected safety findings. All anakinra patients developed anti-drug antibodies (ADAs) at some timepoint during the study. ADAs were persistent throughout the treatment period, except in one patient. Titers were low to moderate. One placebo patient had low ADA titers at one occasion. No neutralizing antibodies were observed and the ADAs did not appear to impact clinical efficacy or safety.

Conclusion: Anakinra is superior to placebo in the treatment of Still’s disease. ADAs occur frequently but do not appear to adversely impact efficacy or safety. These results confirm the benefits of anakinra treatment in patients with active, newly diagnosed Still’s disease across ages.

Figure 1: Individual ACR90 response with absence of fourier and treatment duration over time and ACR90/50/70 at week 2

Figure 2: Time to study drug discontinuation, Kaplan-Meier plot

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