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POS0830

FACTORS INFLUENCING PATIENT-REPORTED OUTCOMES IN ANCA-ASSOCIATED VASCULITIS

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Background: Patient-reported outcomes (PROs) are currently poorly integrated in the clinical evaluation of disease activity in patients with ANCA-associated vasculitis (AAV).

Objectives: To assess the distribution of the Patient Global Assessment (PtGA) in patients with AAV in stable remission, and to identify correlates of PtGA; to assess the discordance between PtGA score and PhGA.

Methods: Patients with a diagnosis of AAV [eosinophilic granulomatosis with polyangiitis, granulomatosis with polyangiitis, microscopic polyangiitis] in stable, complete remission (defined by a BVAS=0) and with a Physician Global Assessment (PhGA)=0 were included. A questionnaire including several aspects of disease captured by PROs was collected. PtGA on a 0-100 mm visual analogue scale (VAS) was assessed, with higher scores representing higher/worse levels of disease activity. Similarly, VAS for pain, chronic damage according to the patient’s opinion, general health (GH), fatigue, and sleep quality were collected. The worst symptom in the patient’s opinion affecting the overall assessment of disease activity was recorded. The Cragg Hurdle model was used to assess the predictors of PtGA.

Results: 65 patients were included, female 57%, mean age 61±12 years. Mean vasculitis damage index (VDI) was 4.4 ± 2.3, with 45% of patients having a VDI ≥ 5. Despite having been classified as being in remission, PtGA was elevated in 37% of patients. We explored several correlates of PtGA. Higher degree of damage accrual (VDI) did not influence the patient’s evaluation of current disease activity. Similarly, we did not identify a correlation between patient age, educational level, number of organ-systems involved, number of comorbidities, the number of previous major or minor relapses, higher disease duration, nor the type of AAV diagnosis (figure 1, panel A). Only sex significantly correlated with PtGA scores: 19 (51%) of female patients reported an elevated PtGA compared to only 5 (18%) of male patients (p=0.0099). PtGA resulted to be significantly correlated with other (mostly modifiable) PROs including VAS pain, perception of the level of chronic damage accrual, GH, and fatigue (figure 1, panel B). The agreement between patients’ and physicians’ assessments of disease activity was 63%. Patients reported pain, followed by chronic respiratory symptoms to be the worst-experienced ongoing manifestations affecting their evaluation of disease activity.

Conclusion: A significant proportion of patients with AAV considered to be in remission by the physician still declares to have persistent aspects of uncontrolled disease. PtGA is significantly influenced by persistent pain and fatigue, which warrant better assessment in the future.

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POS0829

SPECIFICITY OF PANCA AUTOANTIBODIES IN AUTOIMMUNE DISEASES

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Background: The clinical significance of pANCA by indirect immunofluorescence is well-established. However, their clinical utility is sometimes hindered by the fact that pANCA are also detected in various autoimmune diseases. Myeloperoxidase (MPO) is considered as the major autoantigen recognized by pANCA in ANCA-associated vasculitides (AAV) and predominantly in microscopic polyangiitis (MPA). However, information regarding the targets of pANCA in other autoimmune diseases is still elusive.

Objectives: To investigate the specific autoantigens recognized by pANCA in autoimmune diseases.

Methods: Sera from all patients that were found positive for pANCA in the diagnostic laboratories of the Department of Pathophysiology, School of Medicine, National and Kapodistrian University of Athens and the Department of Immunology and Histocompatibility, Evangelismos General Hospital, Athens, Greece during the last two years were studied. The pANCA+ sera were evaluated for positivity pANCA by indirect immunofluorescence with a title ranging from 1/10 to 1/1000. All sera were tested on a panel of antigens including: MPO, elastase, cathepsin G and bactericidal/permeability increasing protein (BPI) by a commercially available multiplex ELISA (ANCA profile ELISA, Euroimmun, Lubeck, Germany).

Results: A total of 82 patients were included in the study. All patients had positive pANCA by indirect immunofluorescence with a titre ranging from 1/100 to 1/640. According to respective classification criteria, 21 patients had systemic vasculitides (15 MPA, 1 granulomatosis with polyangiitis; GPA, 1 Behcet’s disease; BD, 1 aortitis, 2 Henoch-Schonlein purpura; HSP and 1 cryoglobulinemic vasculitis; CV), 29 had systemic lupus erythematosus (SLE), 6 antiphospholipid syndrome (APS), 8 Sjögren’s syndrome (SS), 2 rheumatoid arthritis (RA), 1 systemic sclerosis (SSc), 1 Hashimoto thyroiditis and 1 sarcoidosis. The specificities of pANCA in each entity are shown in the following table.

<table>
<thead>
<tr>
<th>Autoimmune Diseases</th>
<th>Antigens recognized by pANCA+ sera</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>MPO</td>
</tr>
<tr>
<td>Vasculitides</td>
<td>MPA</td>
</tr>
<tr>
<td></td>
<td>GPA</td>
</tr>
<tr>
<td></td>
<td>BD</td>
</tr>
<tr>
<td></td>
<td>Aortitis</td>
</tr>
<tr>
<td></td>
<td>HSP</td>
</tr>
<tr>
<td></td>
<td>CV</td>
</tr>
<tr>
<td>SLE</td>
<td>6.9 (2/29)</td>
</tr>
<tr>
<td>APS</td>
<td>16.6 (1/6)</td>
</tr>
<tr>
<td>SS</td>
<td>0 (0/8)</td>
</tr>
<tr>
<td>RA</td>
<td>50 (1/2)</td>
</tr>
<tr>
<td>SSc</td>
<td>100 (1/1)</td>
</tr>
<tr>
<td>Hashimoto</td>
<td>0 (0/14)</td>
</tr>
<tr>
<td>Sarcoidosis</td>
<td>0 (0/1)</td>
</tr>
</tbody>
</table>

Conclusion: pANCA positive staining in AAVs is highly restricted to MPO specificity. On the contrary, pANCA staining pattern in other autoimmune diseases, involves unknown autoantigens that are under investigation in our laboratory.
Early Infectious Risk in Patients with Newly-Diagnosed Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis According to the Remission-Induction Therapy: A French Monocentric Retrospective Study Including 145 Patients

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Background: Few comparative data exist on early infections second- ary to remission-induction therapy (RIT) with rituximab versus cyclo- phosphamide in newly-diagnosed ANCA-associated vasculitis (AAV) patients.

Objectives: We compared and analyzed the rate and predictors of severe infections in such patients within the first six months following RIT.

Methods: We included, from the databases of Caen University Hospital, all consecutive adults newly-diagnosed with granulomatosis with poly- angiitis or microscopic polyangiitis between January 2006 and December 2013. We compared the survival without severe infections (WSI) and the survival without infection of any severity (WIOAS) within 6 months from the RIT, and used a multivariate cox analysis to identify predictors of infection.

Results: We included 145 patients, 27 in rituximab group and 118 in cyclo- phosphamide group. Patients in the rituximab group more frequently had pneumococcal vaccination (p=0.01) and creatinine level >150 µmol/L, while other characteristics, including Birmingham Vasculitis Activity Score, were comparable between both groups.

Overall, 37 severe infections and 65 infections of any severity were recorded. The survival WSI was similar in both groups (p=0.69), but survival WIOAS was lower in rituximab group (p=0.005).

In multivariate analysis, risk factors at diagnosis for severe infections were chronic urinary tract disease, dialysis and absence of prophylaxis with trimethoprim-sulfamethoxazole (p<0.01 each).

Conclusion: The survival WSI within the 6 months following RIT was sim- ilar in patients with newly-diagnosed AAV treated by rituximab or cyclo- phosphamide, but survival WIOAS appeared to be lower within the 6 months following rituximab despite a better pneumococcal vaccination coverage.

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Figure 1. Comparison of the 6-month survival without severe infection (A) or without infection of any severity (B) in patients with newly-diagnosed ANCA-associated vasculitis treated by rituximab or cyclophosphamide

A Novel Grey Scale and Power Doppler Ultrasonographic Score for Idiopathic Inflammatory Myopathies: Siena Myositis Ultrasound Grading Scale

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Background: No clear-cut guidelines exist about the use of diagnostic procedures for idiopathic inflammatory myopathies (IIM) and only scanty and conflict- ing data report the use of ultrasound (US).

Objectives: We aimed to assess if grey-scale (GS) and Power Doppler (PD) US, graded with a 0-5-points-scale, may be a reliable tool in a cohort of patients affected by IIM.

Methods: We prospectively collected, since July to October 2020, all patients referred to Vasculitis and Myositis clinic, Rheumatology Unit, Uni- versity of Siena, for suspected IIM, as well as patients with a previous, defi- nite diagnosis of IIM and evaluated during follow-up or referred from other centers for a second opinion. All patients underwent US examination of both thighs in axial and longitudinal scans. Edema and atrophy, both assessed in GS, and PD, were graded with a 0-3-points-scale. Spearman test was used to identify the correlations between US and clinical and serological variables.

Results: A total of 18 patients was included. Four of them were evaluated twice, at baseline and within 3 months of therapy. Muscle edema was found to be directly correlated with physician global assessment (PhGA), serum myo- globin and PD and negatively with disease duration. PD score was positively correlated to PhGA and negatively to disease duration. Muscle atrophy directly correlated with Myositis Damage Index and patients’ age. The single-thigh sub-analysis evidenced a direct correlation between PD score and Manual Muscle Test.

Conclusion: In our cohort, we found that edema and PD are strictly related to early, active myositis, suggesting that an inflamed muscle should appear swollen, thickened and with Doppler signal. Conversely, muscle atrophy reflects the age of the patient and the overall severity of the disease. Such findings shed a new, promising, light in the role of US in diagnosis and mon- itoring of IIMs.

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Scleroderma, myositis and related syndromes