Objectives: To analyze demographic feature, disease manifestations and laboratory findings of patients with PR3-ANCA positive GPA in comparison with patients ANCA-negative or MPO-ANCA positive GPA.

Methods: This is a retrospective analysis of 57 patients with GPA from a single center in Ukraine observed from 2010 till the end of 2019. The clinical and demographic data, initial Birmingham vasculitis activity score (BVAS/WG), erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were compared between patients with PR3-ANCA positive GPA and ANCA-negative or MPO-ANCA positive GPA.

Results: Of the 57 patients analyzed, 24 (64.9%) had PR3-ANCA–positive GPA, 6 (16.2%) had MPO-ANCA–positive GPA and 7 (18.9%) had ANCA-negative GPA. ANCA–negative GPA patients were younger at diagnosis compared to PR3-ANCA–positive and MPO-ANCA–positive patients (36 versus 47 and 49 years; p = 0.04). The gender ratio was similar in patients with PR3-ANCA–positive GPA and patients with MPO-ANCA–positive GPA or ANCA-negative GPA (33% vs 36% male, p = 0.61). The ocular manifestations - conjunctivitis/episcleritis (15% vs 50%) and ear involvement - otitis, mastoiditis (0% vs 33%) occurred more often in patients with PR3-ANCA–positive GPA (p<0.05), whereas sensory peripheral neuropathy (54% vs 21%) and Raynaud’s syndrome (31% vs 0%) were more frequent in compared group (p<0.05). ANCA-negative patients with GPA had lower, but not significant, initial BVAS/WG score than PR3-ANCA–positive or MPO-ANCA-positive patients with GPA (179 versus 23.5 and 24.8; p=0.20). There were no significant differences between groups in ESR or CRP levels and in the frequency of involvement of other organs and systems.

Conclusion: We demonstrate clinical differences between PR3-ANCA–positive patients with GPA and MPO-ANCA–positive or ANCA-negative patients with GPA. The eye and ear involvement are common for patients with PR3-ANCA–positive GPA and patients with MPO-ANCA–positive GPA or ANCA-negative GPA. Characterized by higher frequency of sensory peripheral neuropathy and Raynaud’s syndrome.

References:

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.3456

INVESTIGATION OF PERMANENT ORGAN DAMAGE IN GIANT CELL ARTERITIS: DISEASE FLARES ARE ASSOCIATED WITH INCREASED DAMAGE SCORES

B. Ince1, S. Artan2, Y. Yağcıklıya, B. Artım-Esen1, A. Gümüld, M. L. Ocağ1, M. İnce1. Istanbul University, Istanbul Faculty of Medicine, Dept of Internal Medicine, Division of Rheumatology, Istanbul, Turkey; Istanbul University, Istanbul Faculty of Medicine, Istanbul, Turkey

Background: Development of organ damage is a major concern in patients with systemic vasculitis. Treatment may also contribute to this important outcome. Scoring systems has been developed to evaluate organ damage in systemic vasculitis and specifically for large vessel vasculitis (1).

Objectives: We aimed to investigate permanent organ damage and determining factors in our giant cell arteritis GCA cohort.

Methods: Organ damage detected at the time of diagnosis and / or follow-up and irreversible for at least 3 months in GCA patients followed up between 1998-2018 were recorded by using Vasculitis Damage Index (VDI) and Vascular Vasculitis Damage Index (LVVID) form patients records of our vasculitis clinic. In the statistical evaluation, chi-square, students t-test and logistic regression analysis were used.

Results: Eighty-nine patients (64% women, mean age 67.9 ± 9.1) were included in the study, the mean follow-up duration was 61.6 ± 56.8 months. All organ damage findings according to both VDI and LVVID are shown in Table-1. In this cohort, cardiovascular damage items and diabetes mellitus were prevalent at baseline. At least one damage item was present in 53 (59.5%) according to VDI; 54 (%50.7) according to LVVID and agreement was high between two damage indices (kappa=0.97). Forty-seven of patients (52%) had a damage item presumably with contribution of GC treatment e.g. locomotor system findings, hypertension, diabetes and cataract; 12 (13.5%) had damage items related to disease (total or partial vision loss, ischomic optical neuropathy). Mean time to diagnosis and initial symptoms was longer in patients with permanent vision loss (10.2±4.3 vs. 5.2±1.2 months p=0.006). The presence of damage was associated with flares in univariate and multivariate analysis (29/45 vs. 2/35 p>0.001 ORs = 19 *95% GA 4.2– 87.9). All patients who had a flare during the first year (n = 15) developed signs of damage at follow-up. No association was found between the development of organ damage and the age of diagnosis, the time between first complaint and diagnosis, presence of cranial, ophthalmologic findings, PET-CT positivity, cumulative steroid dose, and DMARD use.

Conclusion: In our study, permanent organ damage was analysed by using different indices. In this patient population baseline cardiovascular damage and diabetes mellitus were expected but information for osteoporosis was lacking. More than half of the patients had damage and significant part of the present items was considered due to corticosteroid treatment. The most common damage item developed was osteoporosis. There was a very good agreement between the two indices, despite few specific items in LVVID. The striking relationship of disease flare with damage and frequency of visual problems despite treatment indicate the necessity of new treatment strategies.

References:

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.6379

ANALYSIS OF 89 PATIENTS WITH GIANT CELL ARTERITIS FROM TURKEY: PET-CT AS AN EMERGING METHOD FOR DIAGNOSIS AND HIGH FLARE RATE WITH STANDARD CARE

B. Ince1, S. Artan2, Y. Yağcıklıya, B. Artım-Esen1, A. Gümüld, M. L. Ocağ1, M. İnce1. Istanbul University, Istanbul Faculty of Medicine, Dept of Internal Medicine, Division of Rheumatology, Istanbul, Turkey; Istanbul University, Istanbul Faculty of Medicine, Dept of Internal Medicine, Istanbul, Turkey.

Background: The prevalence of giant cell arteritis (GCA) in Turkey has been reported lower than other European countries and the information on clinical patterns, diagnostic modalities, treatment and prognosis of GCA are limited (5).

Objectives: We aimed to analyse our GCA cohort from a large outpatient clinic for the last 20 years.

Methods: Data of the GCA patients followed up at least 6 months in our vasculitis clinic between 1998 and 2018 evaluated retrospectively according to EULAR 2018 GCA clinical research recommendations (2). Chi-square, students t-test, logistic regression analysis and Kaplan-Meier test were used for statistical analysis.

Results: Eighty-nine patients with adequate follow-up data (64% female, mean age 67.9 ± 9.1) were analysed. Median follow up duration was 46 months (3-256) and mean time to diagnosis after presenting symptom (TTD) was 5.9±1.2 months (0-60). Polymyalgia rheumatica was found in 36 (40.4%) patients. The clinical findings of the patients are shown in Table-1. Mean TTD was longer in patients with acute vision loss (AVL) (11±4 vs. 4.8±1.1 months p=0.002). Mean CRP was 90.7±82 (8-343) mg/L and ESR was 103.7±25 (52-138) mm/h at the time of diagnosis. Mean age was lower (63±2 vs 69±1 p=0.01); mean CRP (141,8±107,3 vs. 76,6±67,9 mg/dl p=0,023) and ESR (120,8±25,1 vs. 99,3±23,4 mm/h p=0.004) was higher in patients without cranial symptoms (extracranial GCA group). PET-CT findings compatible with large vessel vasculitis were present in 64% (34/53). Sixteen of 19 (%84,2) patients in the extracranial GCA group had positive PET-CT. Temporal artery (TA) biopsy positivity was 64% (34/53). Sensitivity of ACR 1990 Criteria was 77.5% and GIACTA study inclusion criteria was 58.4% in this cohort at diagnosis. Fulfillment of GIACTA criteria was still present in 12 (13.5%) patients after six months of follow up. Treatment data was shown in Table-2. Total flare rate was 34.8% and flare rate was lower in the extracranial GCA group (320 vs. 28/69 p=0.035 OR=0.78 %95 CI 0.64 – 0.96). Reduced survival was observed in cases diagnosed older than 65 years (168.8±23,9 vs 209±17,3 months p=0,015).

Conclusion: The analysis of the largest single center cohort from Turkey confirmed that delayed diagnosis is associated with vision loss. A subgroup of patients without apparent cranial symptoms but positive PET-CT findings is delineated. These patients are younger, present with higher inflammatory response and fewer relapses. The sensitivity of ACR criteria in our cohort is less than 80%. High flare rate especially in GCA patients with cranial symptoms and GIACTA criteria fulfillment after 6 months of treatment in more than 10% of the patients show a need for new treatment options.

References:

Background: Giant cell arteritis (GCA) is the most common type of large vessel vasculitis. Typically it presents in patients over the age of 50 with a combination of temporal headaches, scalp tenderness, jaw claudication, raised inflammatory markers and visual disturbance. The diagnosis of GCA is often challenging and there is a difficult balance of over and under investigation. There have been several proposed scoring systems to help clinicians risk stratify patients who may present with suspected GCA. One such scoring system, published in 2017, showed clinical utility in a large international multi-centre study. Following analysis frequent, followed by the presence of murmurs and the difference in blood pressure, claudication being less frequent in the upper and lower limbs.

Conclusion: patients in this series are characterized by having an extensive disease partly due to a late diagnosis, with a high percentage of complications associated with vascular stenotic compromise, which generates morbidity and impact on the quality of life.

References:

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.6605

Table 1: Clinical characteristics

<table>
<thead>
<tr>
<th>Systemic / Extracranial Findings</th>
<th>Cranial Findings</th>
<th>Ophthalmologic Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fatigue</td>
<td>78 (%76)</td>
<td>20 (%)12</td>
</tr>
<tr>
<td>Weight loss</td>
<td>47 (%32,8)</td>
<td>9 (%1,1)</td>
</tr>
<tr>
<td>Arthralgia</td>
<td>43 (%48,3)</td>
<td>16 (%1,1)</td>
</tr>
<tr>
<td>Fever</td>
<td>35 (%12,4)</td>
<td>16 (%4,5)</td>
</tr>
<tr>
<td>Vascular murmur</td>
<td>3 (%3,4)</td>
<td>16 (%4,5)</td>
</tr>
<tr>
<td>Pericardial effusion</td>
<td>2 (%2,2)</td>
<td>16 (%4,5)</td>
</tr>
<tr>
<td>Extraventric Extracardiac</td>
<td>1 (%1,1)</td>
<td>16 (%4,5)</td>
</tr>
</tbody>
</table>

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.1163

Table 2: Treatment data of GCA cohort

| Initial glucocorticoid (GC) dosage (mg) | 46.7±20.1 |
| Pulse GC treatment                   | 3±12.3   |
| MTX usage                            | 6±6.0    |
| bOMARD usage                         | 7±6.0    |
| Acetyl salicylic acid usage           | 6±6.0    |

Figure 1. Ing et al’s Nomogram of parsimonious model. by logistic regression on data from 530 biopsies, Ing et al. developed a parsimonious prediction model comprising 5 candidate criteria: age, jaw claudication, ischemia-related loss of visual acuity, platelet count and logCRP (Figure 1). [1]

Objectives: Increasingly, ultrasound doppler imaging is recognised and accepted as satisfactory means of confirming the diagnosis of GCA, with the presence of the halo sign characteristic for GCA. The aim of our study was to determine whether this GCA prediction model accurately predicts positive temporal artery biopsies in a large, real world UK cohort. In addition, we assessed whether this model accurately predicts positive temporal artery ultrasounds.

Methods: A retrospective cohort study was performed using electronic medical records of patients referred for temporal artery biopsy (TAB) and temporal artery ultrasound (USTA) for suspected GCA. All TAB performed at the Royal Wolverhampton NHS Trust between June 2014 - June 2018 and all USTA performed between January 2015 - January 2019 were analysed. Patients who undergo USTA for suspected GCA at our centre routinely have bilateral temporal and axillary arteries scanned. Patients were excluded if they already had a previous diagnosis of GCA (and the clinical question was suspected flare), or if there was insufficient information available.

Results: The total number of patients who underwent a confirmatory diagnostic test (either TAB or USTA) for suspected GCA was 187. Thirteen of these patients met the exclusion criteria, the remaining 174 patients were included for analysis. 126/174 patients underwent a TAB, 63/174 had an USTA. 15/174 had both these were included in the USS cohort because for all these patients the ultrasound was the first diagnostic test performed (Table 1). Our results appear to closely mirror the original multi-centre results with regards to prediction of biopsy positive GCA, with the cediles closely following those in the inclusion cohort. 0% of the ‘low’ risk probability biopsy cohort were misclassified - none had a positive biopsy. However, 8% of the ‘low’ risk probability ultrasound cohort were misclassified - 2 had a positive ultrasound.

Nomogram: probability of giant cell arteritis

Figure 1. Ing et al’s Nomogram of parsimonious model.