SAT0239

PREDICTION OF LONG-TERM EVOLUTIONARY PROFILES IN EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (CHURG-STRAUSS) BASED ON BASELINE AND FOLLOW-UP CHARACTERISTICS

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Background: Eosinophilic granulomatosis with polyangiitis (EGPA) (Churg-Strauss) is a small-vessel necrotizing vasculitis characterized by blood and tissue eosinophilia and asthma. Glucocorticoids (GCs) control the disease, but GC-dependence is frequent. Evolving concepts distinguish vasculitis-related symptoms from asthma and/or ENT manifestations. That distinction has become even more important since the development of B-cell and eosinophil-targeted therapies.

Objectives: This study aimed to describe and identify characteristics predicting long-term EGPA outcomes.

Methods: We set up a multicenter European cohort that included 636 EGPA patients. Based on recent consensus, we distinguished 4 EGPA-evolutionary profiles: GC-dependent asthma and/or ENT manifestations (requiring prednisone >7.5 mg/d), ≥1 vasculitis relapse(s) (excluding asthma and/or ENT flares), both phenotypes, and complete remission (no GC-dependent asthma/ENT signs, no vasculitis relapse and prednisone <5 mg/d at last follow-up). Baseline and follow-up characteristics predicting those outcomes were analyzed.

Results: After median follow-up of 63 (IQR 30-110) months, 35.8% had GC-dependent asthma and/or ENT manifestations, 12.9% had ≥1 vasculitis relapse(s), 14.3% had both phenotypes, 14.6% were in complete remission, 14.4% were in partial remission and 7.8% had not reach remission.

Patients with GC-dependent asthma/ENT manifestations were younger at diagnosis ($p<0.0001$), had more frequent GC-treated asthma before overt EGPA ($p=0.002$), had more ENT manifestations ($p=0.01$) and less frequent MPO-ANCA ($p<0.0001$). Their daily GC dose was higher at every time point ($p<0.0001$), and they had more frequent active asthma at last follow-up ($p<0.0001$).

Patients with vasculitis relapse(s) had more frequently neurological manifestations at diagnosis ($p=0.002$) and MPO-ANCA positivity ($p<0.0001$), and less frequently pulmonary infiltrates ($p=0.031$). Median time from diagnosis-to-1st vasculitis relapse was 25 (11-60) months. During follow-up, their daily GC dose was lower than those with GC-dependent asthma/ENT manifestations, but similar to those in complete remission. At last follow-up, neurological sequelae tended to be more frequent ($p=0.06$).

Finally, patients in complete remission were older ($p<0.0001$), had more fever ($p=0.03$), less GC-treated asthma ($p=0.002$) and ENT manifestations at diagnosis ($p=0.01$), lower daily GC dose during follow-up ($p<0.0001$), lower eosinophils count at 6 months ($p=0.002$) and less frequent sequelae ($p=0.003$).

Conclusion: EGPA seems to evolve toward distinct phenotypic profiles, which could be identified using baseline and follow-up characteristics. Early identification of those profiles could allow guided choices of the best therapeutic option.

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SAT0240

DEVELOPMENT AND PRELIMINARY VALIDATION OF THE BEHÇET'S SYNDROME OVERALL DAMAGE INDEX (BODI)

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Background: Irreversible organ damage is considered a core outcome by the OMERACT working group. However, no specific tools are currently available to detect and measure damage accrual in Behçet's syndrome (BS).

Objectives: To develop and preliminarily validate the Behçet's syndrome Overall Damage Index (BODI). NCT03803462.

Methods: A preliminary version of the instrument (p-BODI) was developed by reviewing pre-existing tools [e.g. Vasculitis damage index (VDI)] and through an extensive literature review. p-BODI was then reviewed and implemented by a multi-rounds Delphi process, involving an international and multidisciplinary (5 rheumatologists, 4 internist, 1 ophthalmologist, 1 neurologist) panel of experts in BS management and a patients' delegate. A group of clinicians (CG), not involved in the BODI development, was asked to independently score a set of clinical vignettes, in order to test the instrument reliability, after a training process consisting of a user manual and a video-tutorial. Then, Cohen's K and Intra-class correlation coefficient (ICC) between assessors and gold standard were calculated. Afterwards, BODI validation was conducted according to the OMERACT Filter 2.0 in a multicenter BS cohort.

Results: Starting from a list of 120 candidate items, the final version of BODI consisted of 4 overarching principles, 30 items and 12 sub-items (each of them scores one point) grouped in 8 domains (figure).

In terms of reliability, the mean K coefficient was 0.84 (95%CI 0.78 to 0.90) and the ICC was 0.88 (95%CI 0.80-0.95).

Validation cohort consisted of 228 BS patients (49.1% males), with a median (IQR) age and disease duration of 46.9 (35.5-55.0) and 11.7 (5.8-20.7) years, respectively. Overall, prevalence of any BODI damage (BODI ≥1) was 56.1% with a median score of 1.0 (0-2.0). In regard of construct validity, BODI score significantly correlated with VDI (Spearman's rho 0.693, $p<0.001$). Besides, BODI score did not correlate with BDAF (rho 0.016, $p=0.807$), contrary to VDI (rho 0.141, $p=0.034$). Such results support the validity of BODI, unlike VDI, in discriminating damage from current disease activity in BS. On multiple regression analysis, factors independently associated to higher BODI damage score were male gender (β coefficient 0.143; $p = 0.014$), longer disease duration (β 0.221; $p < 0.001$), past major organ involvement (β 0.377; $p < 0.001$) and required use of anti-TNF α inhibitors (β 0.222; $p < 0.001$).

Full agreement among the CG was reached in judging BODI as a credible, comprehensive, easy to use, timesaving and acceptable instrument.

Conclusion: BODI is the first tool specifically developed to assess damage in BS. Preliminary data encourage further validation of BODI in more extended and multi-ethnic BS cohorts before being applied in clinical practice and as a therapeutic outcome.

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SAT0241 HEALED TEMPORAL ARTERITIS. ANALYSIS OF PATIENTS REFERRED TO RHEUMATOLOGY WITH A DIAGNOSIS OF TEMPORAL ARTERITIS

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Background: The diagnosis of giant cell arteritis (GCA), or temporal arteritis (TA), is a clinical one. Temporal artery biopsy is usually obtained to strengthen support for treatment. Unfortunately a biopsy does not always confirm the diagnosis; results can be reported as active, healed, or negative even in patients with all other features of giant cell arteritis. Histopathologic features suggestive of resolving inflammation are evident in healed TA, but these are non-specific and can also be seen in other conditions that are unlikely to benefit from prolonged steroid treatment. It is not clear if a pathological diagnosis of healed TA helps to support treatment which may be associated with significant toxicity.

Objectives: We analyzed clinical features of patients with "healed" TA and compared them to patients with active and negative biopsies with the goal of identifying any features that may aid treatment decisions.

Methods: Retrospective analysis of 49 patients with established TA diagnosis, seen at a rheumatology practice in Houston, Texas from 8/2007 to 12/2018. Fifty seven percent were referred by Ophthalmology, the rest by other specialties. Patients were divided into 3 groups based on biopsy: Active, Healed, and Negative. Clinical features analyzed at the time of presentation were gender, age of onset, fulfillment of the American College of Rheumatology (ACR) 1990 criteria for the classification of Giant Cell Arteritis and the 2016 revised ACR criteria for early diagnosis of Giant Cell Arteritis (rACR), and the presence of vision abnormalities, headache, temporal artery tenderness/induration, jaw claudication, fever, anemia, sedimentation rate at onset, and flares during follow up. Descriptive data was reported as means $\pm$ standard deviation for continuous variables and as proportions for categorical variables; analyzed with ANOVA and chi squared, respectively. Post hoc analysis with Bonferroni corrections and Fisher's exact tests were used. All analyses were

performed on Stata version 15 (StataCorp LP, College Station, TX, USA). A p-value of ≤ 0.05 was considered statistically significant.

Results: Thirty-two (65%) of the 49 patients were female and 17 (35%) were male. Biopsy reports showed active disease in 14 (28.5%), healed in 31 (63.3%) and negative in 4 (8.2%) patients. Average age was 72.7 years ± 6.3 , 73.0 ± 9.1 , 61.25 ± 1.3 years in these 3 groups, respectively (p value 0.008). The Active versus Healed groups were statistically different with respect to age (p 0.028), those meeting ACR and rACR criteria (p 0.029 and p 0.002), and the incidence of jaw claudication: 8 active (57.1%) versus 5 healed (16.1%) patients (p 0.05). There was no statistically significant difference in ophthalmologic manifestations, headache, temporal artery palpation, fever, anemia, and flares. In addition, 90% of patients reported side effects from steroids.

Conclusion: The percent of healed TA was 63.3%, higher than in other studies. Patients with healed TA were less likely to fulfill ACR and rACR criteria for giant cell arteritis. Further studies are needed to identify the subset of patients with healed TA who would have a clear benefit from high dose steroids, that would outweigh the treatment side effects. At the end of the day, the diagnosis of TA remains a clinical one and early involvement of Rheumatology may help avoid unnecessary biopsies and treatment. Ultrasound, MRA, and PET-CT scan may be able to fill the gap when biopsy fails to confirm the diagnosis.

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SAT0242 SENSITIVITY OF TEMPORAL ARTERY BIOPSY IN GIANT CELL ARTERITIS: SYSTEMATIC LITERATURE REVIEW AND META-ANALYSIS OF CLINICAL DATA

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Background: The role of temporal artery biopsy (TAB) as a reference test for the diagnosis of giant cell arteritis (GCA) is currently questioned by the use of non-invasive imaging techniques such as temporal artery ultrasonography (TA-US). Although TAB is highly specific, a subset of patients with a clinical diagnosis of GCA does not show the characteristic histopathological signs. The lack of knowledge of the proportion of GCA cases with positive findings on TAB hampers comparisons of the sensitivity of TAB and imaging tests for diagnosing GCA.

Objectives: We performed a systematic literature review and meta-analysis to estimate the sensitivity of TAB in GCA and to identify factors that may influence the estimate.

Methods: We searched MEDLINE via PubMed, EMBASE and CENTRAL databases for articles reporting TAB in GCA that were published from 1990 to 2017, with no language restriction. Eligibility criteria included studies with ≥ 30 GCA cases fulfilling the original or modified 1990 ACR classification criteria for GCA. From eligible publications, two independent researchers extracted the main methodological, geographic, demographic, and clinical characteristics and the number of TAB-positive cases among all cases with interpretable results for TAB. By meta-analysis, we computed the pooled proportion of TAB-positive GCA cases by using a random-effects model with a binomial-normal distribution and assessed heterogeneity by the I^2 statistic. Subgroup and meta-regression analyses were used to examine the effect of 16 covariates (e.g., geographic, demographic, clinical and study descriptors) on TAB positivity.

Results: Among 3820 screened publications, 32 independent studies (3092 GCA patients in total) were used for the analysis. The pooled proportion of TAB positivity was estimated at 77.3% (95% confidence interval 71.8–81.9%), with high between-study heterogeneity ($P=90\%$). Subgroup analysis suggested a potential influence of year of publication (Table). This result was confirmed by univariate ($P=0.0008$) and multivariate meta-regression ($P=0.0004$). No other analyzed covariate significantly influenced the sensitivity of TAB in GCA.

Conclusion: The 77% estimated sensitivity of TAB in GCA indicates that it is not inferior to that of TA-US (1). The decline in TAB-positive GCA cases over time could reflect an increasing propensity of clinicians to accept GCA diagnosis in the absence of proof by TAB. The unexplained high between-study heterogeneity could also result from differences in TAB sampling, processing or interpretation.

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