Classification Criteria for Primary Systemic Vasculitis (DCVAS) study is a large multinational, observational case control study collecting detailed data from patients with primary vasculitides at inclusion.
Objectives: To describe the prevalence, associations with other disease characteristics and patterns of VN in patients with AAV at initial presentation.
Methods: Patients included in the DCVAS study and having completed 6 months follow-up until December 2016 were screened. All patients with a diagnosis of AAV confirmed by an independent expert team were included. VN was diagnosed by clinical features, neurophysiology and/or nerve biopsy. AAV organ manifestations were identified by described symptoms or by the items from the vasculitis damage index. Laboratory parameters and histology were retrieved from the database. Data were analysed descriptively.
Results: By Dec 2016, 1268 patients with a physician submitted diagnosis of AAV had their case summaries reviewed by an expert panel and the diagnosis of AAV confirmed in 839. 484 (58%) had GPA, 195 (24%) MPA, 150 (18%) EGPA and 10 (1.2%) an unclassified AAV. Of these patients, 247 (29.4%) had findings compatible with VN. Mean age in patients with and without VN was 58.7 (SD 15) and 55.4 (SD 17) years, respectively. 133 (53.9%) of patients with VN and 289 (48.8%) without VN were female. VN was diagnosed by biopsy in 5.7%, by the presence of mononeuritis multiplex in 10.1% and by the description of new onset peripheral neuropathy in the context of AAV in 84.2%. Frequency of VN was 19.4% in GPA, 24.1% in MPA and 68.0% in EGPA. 5.6% of patients had motor, 27.9% sensory, 36.4% had sensorimotor neuropathy, and 7.4% had neuropathy exclusively documented on VDI. VN was associated with older age (p=0.008), the presence of MPO-ANCA (p=0.005), skin ($p \leq 0.001$), musculoskeletal ($p \leq 0.001$), cardiac (p=0.001) and multiorgan (>5 organs) involvement (p=0.05) and with the absence of renal (p=0.002), gastrointestinal (p=0.03) and eye involvement ($p \leq 0.001$).
Conclusions: VN has a high prevalence in patients with AAV. In EGPA, more than half of the patients suffered from peripheral nerve involvement. A typical clinical scenario (e.g. older age, MPO positivity, skin and joint/muscle involvement) may help to identify patients at risk of neuropathy. DCVAS was not primarily designed to assess VN, therefore these data should be interpreted with caution.
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THU0307 NMR-BASED SERUM METABOLOMICS OF PATIENTS WITH TAKAYASU ARTERITIS (TA): RELATIONSHIP WITH DISEASE ACTIVITY

A. Jain[1], D. Kumar[2], R. Misra[1], P.A. Bacon[3], A. Guleria[4], D.P. Misra[4], S. Singh[4], D. Dubey[4], S. Chaurasia[4], S. Kumar[4], U. Kumar[4], S.K. Mishra[4], A. Zanwar[4], A. Rawat[4]. *[1]Clinical Immunology, SGPGIMS; [2]CBMR, Lucknow, India; [3]Department of Rheumatology, Birmingham, United Kingdom; [4]Affiliation not provided*

Background: Takayasu arteritis (TA) is a chronic large vessel vasculitis of unknown etiopathogenesis. The serological and radiological parameters currently used to assess the disease activity are not highly specific and there is a pertinent need for a biomarker discovery. In our previous study [1], NMR based serum metabolomics had revealed distinctive metabolic signatures in patients with TA compared to age/sex matched healthy controls. In this study we sought to investigate whether these distinctive metabolites correlate with disease activity.
Objectives: To identify the discriminatory serum metabolic profiles and their correlation with disease activity.
Methods: Patients with TA fulfilling ACR criteria were assessed for disease activity by ITAS 2010, with a score of 4 or more, considered as active. The serum metabolic profiles of active and inactive TA patients were obtained at 800 MHZ NMR spectrometer and were compared using multivariate orthogonal partial least-squares discriminant analysis (OPLS-DA) to identify metabolites that changed in response to disease activity [based on PLS-DA VIP (variable importance on projection) score >2.0 and permutation test, p-value <0.01].
Results: 88 patients were categorized into active (34) and inactive (54) groups. Median age in active and inactive groups was 25 years and 27 years respectively. Female to male ratio was 3.4:1 in the active group and 5:1 in the inactive group. Majority had class V disease. Mean duration of illness was 4.0±3.5 years in active TA and 6.5±5.5 years in inactive TA group. An exquisite separation in OPLS-DA score plot showed metabolic differences between active and inactive TA patients (Fig. 1A). The key metabolite entities identified with highest discriminatory potential (VIP score >2) were glucose, glutamine, glycine, N-acetyl glycoprotein (NAG), choline, and low/very-low density lipoproteins (LDL/VLDL). Of them glucose, glycine, and NAGs were elevated in the sera of active TA patients, whereas glutamine, choline and LDL levels were decreased in these patients. Receiver operating characteristic (ROC) curve analysis revealed NAG has the highest potential to discriminate active from inactive TA patients (area under the ROC curve was 0.75 (p-value <0.0001) (Fig. 1B, 1C).
Conclusions: The study revealed discriminatory metabolites between active and inactive TA patients and evaluated the possibility of NAG as a clinical biomarker for activity judgment in this disease. However, more work needs to be done to validate the results in a large cohort of patients in a longitudinal manner.

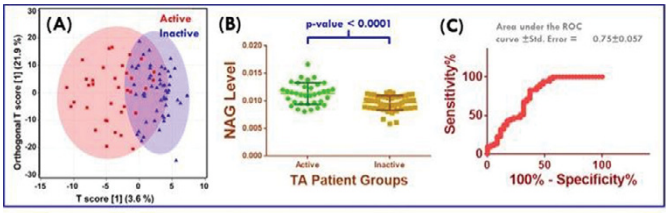

Figure 1: **(A)** 2D score plot obtained from OPLS-DA analysis of 1D ^{1}H NMR spectra. **(B)** Scatter plot showing the serum levels of NAG in active and inactive Takayasu arteritis (TA) patients. The center line refers to the median; whereas the dark lines above and below the center line indicate the 25th and 75th percentiles. **(C)** The receiver operating characteristic (ROC) curve analysis performed to evaluate the specificity, sensitivity, and area under ROC curve (AUC) of NAG peak, showing discriminatory potential of NAG metabolite, based on univariate (box plot) and ROC curve analysis.

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THU0308 EXTENSIVE ANALYSIS OF T CELL RECEPTOR GAMMA (TCRG) GENE REARRANGEMENTS REVEALS A SIMILAR REPERTOIRE IN EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (EGPA) AND IN HYPEREOSINOPHILIC SYNDROME (HES)

C. Baldini[1], S. Galimberti[2], E. Ciabatti[2], I. Petrini[3], G. Tarrini[2], M. Latorre[4], E. Elefante[1], F. Ferro[1], R. Grossi[2], N. Pisanti[2], M. Petrini[2], M. Mosca[1]. *[1]Clinical and Experimental Medicine, Rheumatology Unit; [2]Clinical and Experimental Medicine, Hematology Unit; [3]Translational Research and New Technology in Medicine, General Pathology; [4]Cardio-Thoracic and Vascular Department, University of Pisa, Pisa, Italy*

Background: Hypereosinophilia-associated syndromes are a heterogeneous group of diseases characterized by sustained and elevated blood eosinophilia with evidence of eosinophil-induced organ damage. Classically, Eosinophilic granulomatosis with polyangiitis (EGPA) and Hypereosinophilic syndrome (HES) present several overlapping clinical and laboratory features, making it challenging to correctly insert patients in restricted and well-defined categories with specific and more effective therapeutic approaches. Therefore, great efforts are ongoing searching for novel biomarkers able to differentiate these two disorders in daily practice.
Objectives: To detect T cell receptor gamma (TCRG) clonal rearrangements in EGPA and HES, comparing the frequency distribution of V region and J region segment utilization in the study population.
Methods: In this single center study, we included consecutive patients with a diagnosis of EGPA and HES. Inclusion criteria were: documentation of a persistent peripheral eosinophilic count of ≥1.5 x109/L and signs or symptoms of organ involvement. Clinical and laboratory data of the patients were collected. Sequence-based determination of the frequency distribution of TCRG Gene Rearrangements was performed using next-generation sequencing with the Illumina MiSeq (LymphoTrack TRG assay, Invivoscribe).
Results: We included 21 patients (9 with EGPA and 12 with HES). Four EGPA patients were MPO-ANCA positive. We detected TCRG clonal rearrangements in 44% (4/9) patients with EGPA and in 42% (5/12) patients with HES (p-value = n.s). No association was observed between TCRG clonal rearrangements and ANCA status in EGPA patients. Recurrent TCRG gene rearrangements were observed; in particular, Vg10JgP1 (5 cases) and Vg4Jg1/2 (4 cases) were detected in both EGPA and HES, whereas Vg9Jg1/2 (2 cases) and Vg10Jg1/2 (2 cases) were found only in patients with HES.
Conclusions: Even if preliminary, this study reveals a similar T cell receptor gamma repertoire in EGPA and HES, thus suggesting a possible antigen-driven inflammatory response underlying hypereosinophilia in both EGPA and HES. Moreover, our results would suggest that the TCR clonality cannot be used as a tool for the differential diagnosis between EGPA and HES.
Disclosure of Interest: None declared
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THU0309 TREATMENT WITH METHOTREXATE AND RISK OF RELAPSES IN PATIENTS WITH GIANT CELL ARTERITIS IN CLINICAL PRACTICE

D. Freites Núñez, J. Font, C. León, I. Morado, L. Rodríguez-Rodríguez, L. León, Z. Rosales, B. Fernández, J. Jover, L. Abásolo. *Reumatologia, Hospital Clinico San Carlos, Madrid, Spain*

Objectives: To assess the incidence rate of relapses and to analyze the risk of relapses in patients with *Giant Cell Arteritis* (GCA) treated with and without