led to the myocardial infarction, and death. Pathological manifestation of cardiac involvement in PN included the left ventricular hypertrophy due to renovascular arterial hypertension in 26 cases (70.3%). In addition, interstitial myocarditis was observed in 4 cases (10.8%).

Conclusions: Our date suggest that cardiac involvement is common in polyarteritis nodosa (81.1%), and coronary vasculitis affecting medium and small-sized arteries with wide range of acute and chronic changes can be the life-threatening condition.

Disclosure of Interest: None declared

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THU0333 ARE THE DEMOGRAPHIC AND CLINICAL FEATURES OF POLYARTERITIS NODOSA SIMILAR BETWEEN THE UK AND TURKEY?

O. Karadag[1,2], A. Erden[1], E.D. Batu[1], S. Gopaluni[2], A. Sari[1], B. Armagan[1], I. Ertenli[1], S. Ozen[1], D. Jayne[2]. *[1]Hacettepe University Vasculitis Centre, Ankara, Turkey; [2]Vasculitis and Lupus Clinic Addenbrooke's Hospital, University of Cambridge, Cambridge, United Kingdom*

Background: Polyarteritis Nodosa (PAN) is the first described but the least frequent primary systemic vasculitis. Various subgroups of necrotizing vasculitis have separated out from PAN, and are now defined such as HBV-related, cutaneous PAN or monogenic forms of vasculitis. There is a paucity of information on the current phenotypes and, ethnic and geographic differences of PAN.

Objectives: This study compares the demographic and clinical features between two PAN cohorts.

Methods: A retrospective survey of databases from two vasculitis centres between 1990–2016 for PAN patients fulfilling the EMEA Vasculitis Classification algorithm. All paediatric patients met the Ankara 2008 (EULAR/PReS endorsed) criteria for childhood PAN. Patients with typical angiographic and/or histopathologic findings consistent with PAN were included. Demographics, and clinical characteristics, Disease Extent Index (DEI) and Vasculitis Damage Index were recorded. A subgroup analysis based on disease extent between the UK patients with Turkish (TR) patients was performed.

Results: 93 (M/F: 51/42) patients (UK: 47, TR: 46) were recruited into the study. The mean age was 46.7 (20.7) years. Three were HBV-related, 20 (21.5%) had paediatric onset, 16 (16.5%) were cutaneous PAN.

Turkish patients had younger disease of onset (28.7 (17.8) vs. 43.0 (18.0), $p<0.001$). Twelve (26%) of Turkish patients had a monogenic disease (FMF association in 7, DADA2 in 5).

Cutaneous PAN was more frequent in the UK group (12 pts vs. 4pts, p=0.031) whereas renal involvement was increased in the TR group (Table). In contrast to systemic involvement, female predominance was seen in cutaneous PAN (40.3% vs. 68.8%, p=0.037). DEI was similar in both systemic involved groups (6.1 (2.4) vs. 6.5 (2.3), p=0.428). No difference was found between paediatric and adult onset patients except for frequency of cutaneous lesions (100% vs. 64.3%, p=0.002).

During a median 67.5 (32–126) months follow up, 13 patients deceased. No difference was found between the groups regarding relapse rate, death and vasculitis damage index (Table).

Table: Demographic and clinical characteristics of PAN patients

	All patients n: 93	UK cohort (n:47)	Turkish cohort (n: 46)	p
Age at time of study	46.7 (20.7)	52.5 (19.6)	33.6 (17.1)	**<0.001**
Age at diagnosis, years	35.8 (19.2)	43.0 (18.0)	28.7 (17.8)	**<0.001**
Time to diagnosis, months	2 (1-6)	2 (1-5)	3 (2-8)	**0.041**
Sex, male, %	54.8	44.7	65.2	**0.047**
Any constitutional symptoms, %	87.7	87.1	88.1	0.898
Any cutaneous manifestations, %	71.7	68.1	75.6	0.426
Musculoskeletal manifestations, %	78.0	76.7	82.6	0.278
Neurologic manifestations, %	38.8	32.5	54.3	0.089
Testicular pain/tenderness (men only), %	20.3	28.5	16.7	0.310
Renal involvement, %	58.0	40.4	76.1	**<0.001**
Gastrointestinal manifestations, %	47.1	48.8	45.0	0.733
Follow up, months	67.5 (32-126)	79.0 (35.0-143.0)	65 (29.5-108)	0.669
Response to treatment, % • Complete • Partial • No response	 58.5 30.8 10.8	 60.4 30.2 9.4	 54.5 31.8 13.7	0.838
Any relapse,%	57.1	58.9	52.9	0.675
Death, %	14.0	12.7	15.2	0.733
VDI	1 (0-1)	1 (0-1)	1 (0-2)	0.632

Values are labeled as mean (SD or median (IQR 25%-75%), VDI: Vasculitis Damage Index

Conclusions: Even though, the Turkish group had a younger disease onset and higher additional monogenic disease; disease extent, relapse rate, death, and damage index were similar in both groups. Among the cutaneous PAN group there was a female predominance. A multi-centre GWAS study could highlight the impact of genetic background on disease presentation and severity.

Disclosure of Interest: None declared

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THU0334 EVALUATION OF SUBCLINICAL VASCULAR DAMAGE IN PATIENTS WITH POLYMYALGIA RHEUMATICA: A CROSS-SECTIONAL STUDY USING AN INTEGRATED, NON-INVASIVE APPROACH OF COLOR DOPPLER ULTRASOUND AND CARDIO-ANKLE VASCULAR INDEX (CAVI) MEASUREMENT OF ARTERIAL STIFFNESS

P. Sessa[1], I.M. Rutigliano[1], R. Scrivo[1], V. Silvestri[2], F. Ciciarello[3], G. La Torre[4], F. Conti[1], R. Priori[1], C. Alessandri[1], V. Riccieri[1], M. Di Franco[1], B. Gossetti[2], G. Valesini[1]. *[1]1) Department of Internal Medicine and Medical Specialties; [2]2) P. Stefanini General and Specialist Surgery Department; [3]3) Department of Cardiovascular Science; [4]4) Department of Public Health and Infectious Diseases, University of Rome la Sapienza, Roma, Italy*

Background: The association of chronic inflammatory rheumatic disorders with an increased risk of vascular disease, especially cardiovascular and cerebrovascular disease, is a consolidated matter, but data on polymyalgia rheumatica (PMR) are still inconsistent.

Objectives: The aim of our cross-sectional study was to investigate the presence of vascular damage in patients with PMR by analyzing subclinical vascular disease through validated, non-invasive cardiovascular disease markers.

Methods: We enrolled patients with PMR diagnosed according to the EULAR classification criteria and, as controls, patients with major cardiovascular risk factors (MCVRF) including hypertension, diabetes, hypercholesterolemia, cigarette smoking, and obesity. In all of them we performed color Doppler ultrasound to evaluate the common carotid intima-media thickness (IMT), the prevalence of carotid artery stenosis and of anterior-posterior abdominal aortic diameter (APAD); we also assessed the cardio-ankle vascular index (CAVI) to measure arterial stiffness and contextually the ankle-brachial index (ABI) to investigate the presence of lower-extremity peripheral arterial disease.

Results: Forty-eight patients with PMR and 56 with MCVRF were included. Demographic parameters were balanced between groups. A significant increase of IMT (1.03±0.23 *vs* 0.89±0.20; p=0.02), CAVI (8.59±1.23 vs 7.59±0.93; p=0.01) and APAD values (22.03±4.86 *vs* 19.14±4.65; p=0.03) was found in PMR patients with respect to MCVRF controls. No differences were reported with regards to the prevalence of carotid artery stenosis or ABI values between the two groups. No significant correlation between disease duration or duration of glucocorticoid treatment and IMT or CAVI values was found in PMR patients. Results of bivariate analysis showed a significant correlation between IMT and CAVI in both PMR and MCVRF patients (r^2=0.845 and 0.556, respectively; $p<0.001$).

Conclusions: Our study adds new information on cardiovascular risk in PMR patients, showing an increase in subclinical cardiovascular lesions and paving the way for further studies to define the utility and modality of cardiovascular screening for primary prevention in these patients.

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THU0335 ACETYLCHOLINESTERASE IS HIGHLY EXPRESSED IN THE INFLAMED VESSEL WALL OF PATIENTS WITH GIANT CELL ARTERITIS

P. Therkildsen[1], B.D. Nielsen[1], K.K. Keller[1], T. Steiniche[2], L.C. Gormsen[3], I.T. Hansen[1], E.-M. Hauge[1]. *[1]Department of Rheumatology; [2]Department of Histopathology; [3]Department of Nuclear Medicine and PET Centre, Aarhus University Hospital, Aarhus, Denmark*

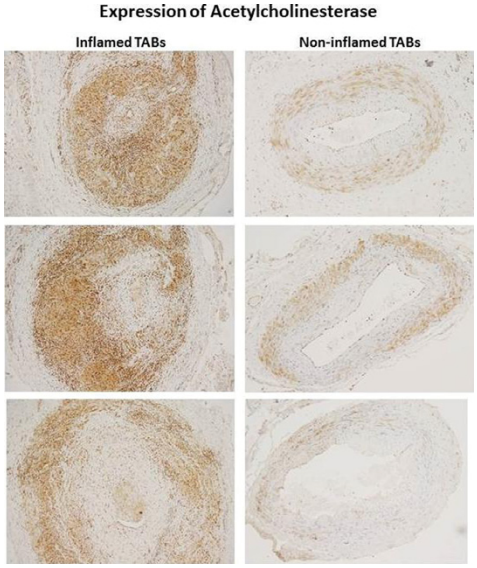
Background: The temporal artery biopsy (TAB) remains the gold standard in the diagnosis of giant cell arteritis (GCA) and is part of the ACR Classification criteria for GCA. However, TABs are false-negative in 10–60% of cases [1]. Cellular studies have shown that activated immune cells upregulate the acetylcholinesterase (AChE) expression [2]. If AChE is upregulated in the active GCA vessel wall, it may potentially improve the TAB as a diagnostic tool.

Objectives: To investigate the *in-situ* expression of acetylcholinesterase (AChE) in the vessel wall of patients with biopsy-positive GCA and compare to non-GCA patients.

Methods: In this histological case-control study, TABs from a total of 24 TAB-positive GCA and 44 TAB-negative non-GCA patients (21 patients with a final diagnosis of PMR, 23 patients with other diagnosis) were retrospectively selected from TABs performed between January 2012 and December 2015. A total of 295 TABs were assed for inclusion. Only positive TABs showing clear transmural inflammation were included. Patients treated with >7 days of prednisolone prior to the TAB were excluded. Clinical data were obtained from electronic patient records to confirm or dismiss clinical diagnosis. *TAB-HE-stains* were reviewed by a pathologist with expertise in vasculitis. Immunohistochemical methods were used to determine the AChE expression. The histological inflammation and AChE

expression were assessed and graded on 0–1-2 scale, blinded to histological and clinical data. Solitary AChE staining of the media was not included in the assessment.

Results: 24 positive and 44 negative TABs, with corresponding clinical positive and negative GCA diagnosis, were included in this study. We found that 10/24 positive TABs showed high AChE expression (grade 2) and 14/24 showed moderate AChE expression (grade 1). No AChE expression was observed outside the media in negative TABs from non-GCA patients (i.e. grade 0). The AChE expression was in 79% agreement with the degree of histological inflammation with a kappa value of 0.58. Prednisolone treatment for up to 7 days did not suppress the AChE expression. Neither the AChE expression, nor the histological inflammation showed correlation to any clinical or biochemical findings.



Conclusions: Our study shows that high to moderate AChE expression was observed in all 24 biopsies from TAB-positive GCA patients and that the AChE expression was in good agreement with the histological inflammation. No non-specific AChE expression was observed outside the media in any of the 44 TABs from TAB-negative non-GCA patients. This indicates that AChE could play a significant role in the inflammatory process in GCA and may be a potential biomarker in inflammatory diseases such as GCA.

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THU0336 GIANT CELL ARTERITIS (GCA) IN OCTOGENARIAN PATIENTS

R. Solans-Laqué[1], E. Fonseca[2], B. Escalante[3], A. Martinez-Zapico[4], G. Fraile[5], M. Conesa[6], M. Abdilla[7], M. Monteagudo[8], J. Lasso[9], E. Calvo[10], B. Gracia[3], M. Castillo[11], J. Mestre[1] on behalf of REVAS Study Group (GEAS-SEMI).
[1]Internal Medicine, Vall Hebron University Hospital, Barcelona; [2]Internal Medicine, Cabueñes Hospital, Gijon; [3]Internal Medicine, Lozano Blesa Hospital, Zaragoza; [4]Internal Medicine, Central Hospital (HUCA), Asturias; [5]Internal Medicine, Ramon y Cajal Hospital, Madrid; [6]Internal Medicine, Miguel Servet Hospital, Zaragoza; [7]Internal Medicine, Alzira Hospital, Valencia; [8]Parc Taule Hospital, Sabadell; [9]Internal Medicine, Alcañiz Hospital, Teruel; [10]Internal Medicine, San Jorge Hospital, Huesca; [11]Internal Medicine, Virgen Rocio Hospital, Sevilla, Spain

Background: GCA is the most common systemic vasculitis in elderly people. It is characterized by granulomatous involvement of the major branches of the aorta with predilection for the temporal arteries. Headache is the most frequent initial complaint, and permanent visual loss the most feared complication.

Objectives: to describe the initial manifestations and outcome of CGA in octogenarian patients, and to investigate if there are some differences compared to younger patients

Methods: demographic, clinical and histological features, treatment and outcome of all patients with biopsy-proven GCA recruited at 11 different Hospitals from Spain (REVAS Study) were analysed. Statistical analysis was performed using SPSS vs. 20

Results: among the 418 patients included in the study, 180 (27.5%) were older than 80 y at disease onset (mean age 83.77±3.1, range 80–92y). Cardiovascular risk factors (hypertension, hypercholesterolemia, diabetes) were more prevalent in octogenarian patients than in younger (73% vs. 50%, p=0.018). Headache was the most frequent initial complaint (78.5%), followed by polymyalgia rheumatica (49.6%), jaw claudication (48.7%) and toxic syndrome (43.5%). A total of 28 (24.35%) patients suffered permanent visual loss, due to anterior ischemic optic ischemic neuropathy (n=26) or central retina vein thrombosis (n=4). When compared with younger patients, neither headache, nor jaw claudication, nor scalp tenderness and polymyalgia, were more common in octogenarian patients. Only fever was less common in patients >80 y at diagnosis than in younger (27.7% vs. 72.3%, p=0.006), and permanent visual loss was most prevalent among octogenarian patients (24.3% vs. 15.7%, OR 1.73, 95CI 1.1–2.9, p=0.04). We found an inverse correlation between fever and permanent visual loss (p=0.001). Blindness was significantly correlated with jaw claudication (p=0.006) and amaurosis fugax (p=0.001). We did not found any significant correlation between blindness and vascular risk factors. Regarding to LAB, ESR was higher in octogenarian patients (p=0.006) as well as thrombocytosis >400.000 platelets/mm (p=0.026). No differences were found related to anaemia prevalence. Inflammatory infiltrate with lumen occlusion and giant cells were significantly more prevalent in temporal artery biopsies from patients with permanent bilateral visual loss (p=0.007 and p=0.01, respectively). No differences were found related to treatment. All patients received oral prednisone at tapering dosage, 85.9% calcium and vitamin D supplementation, and 60% bifosfonates. We did not found differences related to treatment-side effects (osteoporotic fracture, diabetes, infections) between octogenarian patients and the younger.

Conclusions: octogenarian patients with GCA have an increased risk of developing permanent visual compared to younger patients. The presence of jaw claudication and amaurosis fugax must prompt to initiate corticosteroid treatment quickly. No differences were found related to treatment side effects in octogenarian patients.

Disclosure of Interest: None declared

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THU0337 CLASSIFICATION OF ANTINEUTROPHILIC CYTOPLASMIC ANTIBODY-ASSOCIATED VASCULITIS AND CLINICAL IMPACT AND OUTCOME

S. Deshayes[1], A. Aouba[1], K. Kathy[2], D. Mariotte[2], T. Lobbedez[3], N. Martin Silva[1]. *[1]Department of Internal Medicine; [2]Department of Immunology; [3]Department of Nephrology, CHU Côte de Nacre, Caen, France*

Background: Antineutrophilic cytoplasmic antibody (ANCA)-associated vasculitis (AAV) have overlapping manifestations. Classifications based on clinical criteria or ANCA specificity have emerged to individualize homogenized group of patients in terms of clinical forms and outcomes.

Objectives: The aim of our study was to retrospectively re-evaluate the clinical impact and outcome of our monocentric AAV patients' cohort, according to classifications based on clinical criteria and/or ANCA specificity.

Methods: A retrospective monocentric study carried out in Caen university hospital led to identify proteinase-3 (PR3) or myeloperoxidase (MPO)-ANCA AAV patients (via an ELISA technique), respectively from March 1997 to June 2016, and from September 2011 to June 2016. Patients with eosinophilic granulomatosis with polyangiitis were excluded. AAV were classified between granulomatosis with polyangiitis (GPA) or microscopic polyangiitis (MPA), and limited or severe forms according to respectively European Medicines Agency vasculitis algorithm and WGET group. Categorical variables were reported as percentages and compared using Chi^2 or Fisher's tests according to expected frequencies. Continuous variables were expressed as means and analyzed using Student's t-test. Associations between survival, or relapse free survival, and AAV classifications were evaluated by the log-rank test. A p-value <0.05 was considered to be statistically significant.

Results: A total of 150 GPA/MPA were included.

Table 1

	anti-MPO GPA (n=20)	anti-PR3 GPA (n=74)	anti-MPO MPA (n=43)	anti-PR3 MPA MPA (n=13)
Male %	35	54	67	54
Renal involvement %	75	61	88	92
Ear-nose-throat involvement %	65	77	12	0
Lung involvement %	70	80	70	31
Skin involvement %	25	30	21	38
Neurological involvement %	45	24	23	31
Rheumatologic involvement %	55	62	37	54
Limited vasculitis, %	40	23	12	23
Relapse %	50	50	21	8
Death %	0	23	19	31
Initial BVAS, median ± SD	20±7.2	18.5±7.4	17±5.1	15±7.5
Age at diagnosis (years), median ± SD	66.5±11	60±17	62±13	70±21
Follow-up (months), median ± SD	49±47	55±65	50±37	33±108

As expected, ear-nose throat involvement were significantly higher in PR3-ANCA vs MPO-ANCA, and GPA vs MPA (p<0.001). Survival was significantly higher in