laboratory and management characteristics along with the clinical course of Budd-Chiari syndrome (BCS) associated with Behçet's disease (BD).
Methods: Sixty patients with BD with BCS (40 male, 20 female) were identified in 23 rheumatology centers (Group I). A total of 169 consecutive patients (100 male, 69 female) with BD who did not have clinically apparent BCS during the follow-up were evaluated as the control group (Group II).
Results: Comparison of the demographic and clinical findings between the Group I and the Group II were as follows: The mean age of disease onset was 23.1 +/- 6.7 years vs. 26.8±0.6 years (p=0.013), mean age at diagnosis was 27.2±0.9 vs. 30.4±0.6 years (p=0.008), arthritis was 10% vs. 28.4% (p=0.002), papulopustular skin lesion was 48.3% vs 69.2% (p=0.003), central nervous system (CNS) involvement 10% vs. 3% (p=0.03), cardiac involvement was 16.7% vs. 2.4% ($p<0.001$), superficial thrombophlebitis was 23.3% vs. 4.7% ($p<0.001$), and deep vein thrombosis was 58.3% vs. 15.4% ($p<0.01$). On diagnosis 50% of BD patients with BCS were classified as Child-Pugh A. Inferior vena cava obstruction was observed in 38.3% and portal vein thrombosis was seen in 3.3% of the patients with BCS. Mortality in BCS patients with BD was 18.3%. BCS related treatment after diagnosis in patients with BD were as follows: 71.7% of patients were treated with monthly cyclophosphamide intravenous pulses, 53.3% received intravenous pulse corticosteroids, 55.9% used azathioprine, 54.2% had warfarine treatment, and 50.8% were treated with low molecular weight heparin.
Conclusions: This study shows a higher frequency of cardiac and CNS involvement, superficial thrombophlebitis, papulopustular skin lesion, deep vein thrombosis in BD patients with BCS. Arthritis was observed less common in BD patients with BCS. The mean age onset was lower in patients with BCS. Medical treatment with immunosuppressive agents and anticoagulation appears to be the treatment of choice in BD patients with BCS. The majority of the patients with BCS were Child–Pugh class A on diagnosis. The inferior vena cava is frequently involved and, often associated with deep vein thrombosis and cardiac involvement.
Disclosure of Interest: None declared
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THU0330 MYCOBACTERIAL CORD FACTOR ANALOG INDUCES HIGH IL-6 SECRETION AND MINCLE RECEPTOR EXPRESSION IN PATIENTS WITH TAKAYASU ARTERITIS

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Background: Association between Takayasu Arteritis (TA) and Tubcerculosis (TB) has been in vogue for years. Scientific evidence for this association is limited.
Objectives: To study pro-inflammatory cytokine responses and mRNA expression in patients with TA on exposure to Trehalose-6,6-dibehenate (TDB), a synthetic analogue of mycobacterial cord factor in comparison with healthy controls.
Methods: Patients with type V TA satisfying 1990 ACR criteria and age and sex matched healthy controls were recruited. 10 ml blood was collected in heparin coated tube. PBMCs were separated by density gradient centrifugation. PBMCs were cultured with and without 5μg/ml and/or 50 μg/ml of TDB for 48 hours in RPMI medium at 5% co2 incubator. IL-6, TNF-αand IL-17 were measured in cell culture supernatant. RNA was extracted from the cells and quantification of gene expression was performed using sequence specific primers with SYBR green in Step One Plus™ Real-Time PCR Systems (Applied Biosystems).
Results: Twenty two TA patients and 21 age and sex matched healthy controls were recruited. Baseline characteristic of cases included median (range) age:26.5 years (15–49), male:femaleratio of 5:17, ITAS-CRP2010: 7 (1–26) and DEITAK: 7.5 (1–21). Both patients and controls showed response by secreting IL-6 and TNF-α upon stimulation withTDB. Relative induction of IL-6 was significantly higher in TA [31.88 (0.74–168)] as compared to healthy controls [1.931 (0.644–8.21); $p<0.002$]. IL-17 was undetectable even upon TDB stimulation. Relative mincle expression was significantly upregulated by TDB in TA1.03 (0.623–1.346) fold, as compared to healthy controls 0.43 (0.36–0.57) fold ($p<0.05$). Relative gene expressions of IL-6, IL-8, TNFα, IFN-gand BCL-10 were not significantly different between TA and healthy controls.
Conclusions: Stimulation with TDB led to higher secretion of IL-6 and increased mincle expression in PBMCs of TA patients as compared to healthy controls.
Disclosure of Interest: None declared
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THU0331 MYELOPEROXIDASE DEPLETION IS A SELECTIVE FEATURE OF TAKAYASU ARTERITIS AND PREDICTS THE CLINICAL OUTCOME

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Background: Takayasu's arteritis (TA) is a rare large-vessel vasculitis characterized by persistent remodelling of the vessel walls. The role of innate immune cells in TA is poorly understood. Biomarkers to be used for assessment of TA activity and clinical outcome are missing. A trimodal pattern of myeloperoxidase (MPO) distribution (simultaneous occurrence of neutrophils with complete depletion, reduced and normal content of the enzyme) is detectable in the very early phase of acute myocardial infarction and has been associated with a burst of neutrophil interaction with activated platelets[1]
Objectives: Here, we aimed at characterizing the phenotype of neutrophils and platelets in patients TA and correlating these biological findings with clinical data.
Methods: neutrophil MPO expression has been studies in 93 subjects, including 21 TA patients, 20 age- and sex-matched healthy donors that served as controls, 25 patients with chronic stable atherosclerosis (CSA), eight patients with giant cell arteritis (GCA), five patients with gramulomatosis with polyangitis (GPA), four patients with eosinophilic GPA (EGPA) and ten patients with polymyalgia rheumatica (PMR). Blood samples were collected and processed as described[1–3]. Neutrophil MPO content, platelet P-selectin and High-Mobility Group B1 (HMGB1) in platelet derived microparticles (PDμP) release were assessed by flow cytometry. TA patients were then followed up for a median time of 3.43 years.
Results: The trimodal distribution of neutrophils MPO content was identified in 17/21 TA, 1/8 GCA, 0/10 PMR, 0/25 CSA, 0/5 GPA and 0/4 EGPA patients; moreover in 0/20 healthy controls (ANOVA, F=15.22, $p<0.0001$). Neutrophil MPO content and the presence of neutrophils with complete MPO depletion in TA patients correlated inversely with i) platelet P-selectin expression ($p<0.01$), ii) the fraction of $HMGB1^{+}$-PDμP ($p<0.01$) and iii) disease duration, implicating an association of platelet and neutrophil activation with the disease natural history. In fact, the Kaplan Mayer analysis reveals that TA patients that had neutrophils with complete MPO depletion are at a significantly increased risk of ischemic complications during the follow up (long-rank=4.864, p=0.027, HR=36.15 abd CI-95%=0.15–7.51).
Conclusions: Neutrophils and platelets are significantly activated in TA. Paroxysmal neutrophils activation, leading to complete MPO depletion, is a common feature and predict ischemic events and as such the clinical disease outcome.
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THU0332 CARDIAC INVOLVEMENT IN POLYARTERITIS NODOSA: A RETROSPECTIVE PATHOLOGICAL STUDY OF 37 AUTOPSY CASES

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Background: Cardiac involvement is a significant cause of the death and disability in systemic vasculitis. The comprehensive analysis of histopathologic findings in polyarteritis nodosa (PN) can help better understand mechanisms of excessive cardiovascular risk in patients with systemic vasculitis.
Objectives: To investigate the variation of cardiac pathological findings in autopsy cases of PN.
Methods: A retrospective analysis of cardiac pathological changes was performed in 37 autopsy cases of PN over period of 15 years. There were 28 males and 9 females, ranging from 23 to 85 years. The median age at disease onset was 35, 7 years; the median duration of disease was 2.1 years with a range of 2 month to 7 years.
Results: The destructive-productive vasculitis of the coronary arteries have been found in 30 cases (81.1%) of PN. The vessels most affected were myocardial, and epicardial medium and small-sized arteries. Histologically, the remodelling of coronary artery tree in PN had a wide range of acute and chronic changes that likely were depended on the duration of the disease. There were three types of pathological findings: (1) arteritis with predominantly destructive changes; (2) arteritis with predominantly proliferative changes; (3) destructive-productive arteritis. The early phase of PN has been characterized by abnormalities of vessels wall from mucoid swelling to fibrinoid necrosis. The most common was panarteritis with involvement of all layers of the arterial wall, and inflammatory responses with intramural, and perivascular infiltrates, mainly composed of lymphocytes, and neutrophils. However, endo-, meso-, and periarteritis also were observed in some cases. The most remarkable lesions in coronary arteries were the nodules visible to the naked eye along coronary arteries. They were present in 9 cases (30%), and can be described as well-demarcated nodular thickening of the artery walls with size from 0.1 to 0.4 cm. The pathological basis of nodules are areas of focal inflammation, destruction and aneurysmal dilatations of artery wall. In cases of chronic course of PN, the intimal hyperplasia due to proliferation of endothelial cells have been observed. Interestingly, that this productive endarteritis with luminal narrowing of coronary arteries was common in patients who were exposed to different types of occupational xenobiotic (silica dust, pesticides, insecticides, solvents, heavy metals). In 5 (16.7%) cases, luminal occlusion due to intimal proliferation, fibrosis and thrombus formation

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?

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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

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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

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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