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AB0648 CORRELATES OF FATIGUE IN ANCA-ASSOCIATED VASCULITIS

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Background: Fatigue is a common symptom among patients with ANCA-associated vasculitis (AAV) identified as the greatest burden of their disease.[1 2] Research revealed associations between fatigue and bio-psychosocial factors but not with clinical factors.
Objectives: To assess fatigue and its correlates among patients with granulomatosis with polyangiitis (GPA) and microscopic polyangiitis (MPA).
Methods: 37 patients (44% women; mean age 52.3 years; range 18–85) with GPA (27 patients) and MPA (10 patients) hospitalised in 3 clinical centres completed Multidimensional Fatigue Inventory-20 (MFI-20).[3] Anxiety and depression were assessed by the Hospital Anxiety and Depression Scale (HADS). Socio-demographic data including age, sex, education, marital and occupational status were recorded. Disease characteristics included its' duration, severity, activity, organ involvement and laboratory data.
Results: The mean age was 52.3 years (range 18–85 years), and the mean disease duration was 43.1 months (range 1–248). 4 patients had limited type of the disease, 13 – early systemic, 16 – systemic and 4 – severe type of the disease. 75% of patients had active disease as defined by Birmingham Vasculitis Activity Score, BVASv3 (mean BVASv3 in active patients 12.2). 8% of patients were not taking steroids. 40,5% of patients had CRP >5 mg/, 43% had anaemia, 10% – thrombocytosis and 43% had renal insufficiency. Mean score of MFI-20 was 57 points (range 31–100). There were no differences in MFI-20 overall score between groups according to sex, education, marital and occupational status. No significant associations between fatigue and disease-related factors as well as steroid dose were observed. Depression (r=0,79, p<0,00000) and anxiety (r=0,63, p<0,00002) were strongly correlated with MFI-20 overall score.

Abstract AB0648 – Table 1. Socio-demographic characteristics of the study sample.

Variable	n (%)
Education	
primary	6 (16)
occupational	3 (8)
secondary	12 (32)
higher	16 (43)
Marital status	
married	29 (78)
divorced	3 (8)
free state	4 (11)
widower	1 (3)
Occupational status	
employed	18 (49)
unemployed	3 (8)
pensioner	8 (22)
annuitant	5 (13)
student	3 (8)

Conclusions: Since fatigue was not related to clinical characteristics, tiredness declared by the patient should not be interpreted as a symptom of active disease. There were no connexions between fatigue and socio-demographic variables. The strong association between fatigue and depression and anxiety levels indicates the necessity to supplement the standard drug therapy and/or cognitive-behavioural therapy aimed at reducing anxiety and improving mood.

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AB0649 USE OF BIOLOGICAL DMARDS IN PATIENTS WITH PRIMARY VASCULITIS; RESULTS FROM TURKBIO REGISTRY

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Background: Untreated, The systemic vasculitides can be devastating, with high rates of morbidity and mortality. Recently, most of biological agents have evaluated in clinical trials, and management of systemic vasculitis has been revolutionised over the last decade.[1 2]
Objectives: Here, we report the frequency of using and switching rate of biological agents in different types of primary vasculitis patients.
Methods: TURKBIO registry is the Turkish version of Danish DANBIO rheumatological database which has been established in 2011. All patients with primary vasculitis who received biological agents registered in TURKBIO registry between dates of October 2011 and January 2018 were included in this study. The demographic data, the date of starting to use of biological drug, frequency of using and switching biological agents were collected.
Results: As of January 2018, 108 primary vasculitis patients were recruited (mean age: 38.4±10.9 [min-max: 19–67]; female 48%); 48 patients (44%) of them had Behcet's disease (BH), 35 (32%) had Takayasu arteritis (TA), 24 (22%) had granulomatosis polyangiitis, and one of them had microscopic polyangiitis. The most commonly used biological agents in current treatment were as follows: 75% of patients received infliximab (INF) and 15% received adalimumab (ADA) in BD patients; 48.6% received tocilizumab (TCZ), 23% received INF and 20% received ADA in TA patients; all patients with granulomatosis with polyangiitis (GPA) were treated with rituximab. The switching rate was 54% in TA patients, 27% in BD patients, and 4% in GPA patients. The most frequent switching was found at INF (28/78) and ADA (9/23) which was the most commonly used agent in TA and BD. The lowest switching rate was TCZ (2/17) in TA patients (table 1).

Abstract AB0649 – Table 1. Demographical features and managements of patients with primary vasculitis

n (%)	**All Patients n:108**	**BD n=48**	**TA n=35**	**GPA n=24**
Mean Age year mean±sd (min-ma)	**38.4±10.9 (19–67)**	**37±8.3 (19–54)**	**35.7±11.1 (20–59)**	**47.7±11.1 (23–67)**
Gender (F)	**52(48)**	**11 (23)**	**32 (91)**	**9 (37.5)**
Current Treatment	19 (18)	2 (4)	17 (49)	0
Tocilizumab	25 (23)	0	0	24 (100)
Rituximab	2 (2)	0	2 (6)	0
Certolizumab	44 (41)	36 (75)	8 (23)	0
Infliximab	14 (13)	7 (15)	7 (20)	0
Adalimumab	3 (3)	2 (4)	1 (3)	0
Etanercept	1 (1)	1 (2)	0	0
Golimumab				
Last Drug Survival mean±sd (min-max) (month)	**25.2±17.9 (2–88)**	**26.8±16.2 (3–77)**	**28±21.7 (2–88)**	**22.3±15.9 (2–60)**
Switching Rate	**33 (31)**	**13 (27)**	**19 (54)**	**1 (4)**
Tocilizumab	3	0	2	1
Infliximab	28	10	18	0
Adalimumab	9	4	5	0
Etanercept	4	3	1	0

Conclusions: This is the first evaluation of primary vasculitis patients who used biological agents from TURKBIO registry. According our data, there was high switching rate with anti-TNF agents in TA patients. The limitation of this study was the low number of the patients with primary vasculitis who used biological agents.

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AB0650 THROMBOTIC MICROANGIOPATHY ASSOCIATED TO ANCA-POSITIVES VASCULITIS: A FRENCH RETROSPECTIVE CASE CONTROL STUDY AND LITERATURE REVIEW

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Objectives: In this large nationwide French case-control study, we describe the features of TTP and ANCA-positive vasculitis; compare to vasculitis without associated TTP; describe the outcome and treatments.
Methods: We collected all cases with TTP and associated vasculitis. We conducted a literature review using PubMed, Web of Science, congress posters from January 2005 to August 2017 of PTT and ANCA-positive vasculitis.
A control group of MPA without TTP during all the disease foillow-up was extracted from the Saint Antoine and Montfermeil Hospital patients with vasculitis. Firstly we compared our PTT cases and the literature review cases and secondary with a control group ANCA vasculitis without MAT.
Results: 8 patients with MAT secondary to ANCA associated vasculitides were included in our French series 75% of Women with a median age 45 years,[21–76] positive ANCA in 50% and 37.5% is MPO, BVSA score at 16, FFS at 1. In 10 literature cases 90% Women, a median age 60 years [17–77], positive ANCA in 100% and 30% is MPO, BVSA score at 39, FFS at 1.
The clinical features at the diagnosis of vasculitis were fever (n=5; 62%), ENT involvement (25%), kidney cresescnt glomerular involvment (n=6; 75%) with kidney failure in joint involvement (n=2; 25%) polyarthralgia type, with gastrointestinal involvement type mesenteric ischemia and pericarditis and lung involvement (nodules and alveolar haemorrhage) (n=1; 12.5%), in one case each Median C-reactive protein levels were at 49 mg/L [1–204], with creatininemia at 170 mg/dl [80–588]. ANCA were present in 4 patients (50%), MPO in 3 cases (37.5%). The time between the diagnosis of vasculitis and TTP was 9 months [0–51].
TTP features were: Hb at 7.8 g/dl [4.8–10], LDH 1658 [777–3110], platelets 33000 [3000–1 25 000], haptoglobin at 0 [0–0.05], creatinine at 205 [80–757]. ADAMTS 13 levels were at 10 [1–138], and x patients have normal ADAMTS13 levels, with anti ADAMTS 13 antibodies in 1 case. Plasma exchanges were done in all patients with median of 10 exchanges [4–23], 37.5% are dialyzed.
Conclusions: In comparison to our cases, the literature patients have similar organs involvement, but median creatininemia levels and BVAS levels were higher in the literature cases. Considering TTP features, our cases have less frequent active vasculitis, less important creatinine levels and thus less recurs to kidney dialysis.
In vasculitis associated with TTP, there was no significant differences in organ involvements, BVAS and FFS scales values, laboratory data and ANCA levels. Only creatininemia as expected was higer in vasculitis associated with TTP (225 Mmol vs 150 Mmol, p=0.0044).
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AB0651 FREQUENCY OF PULMONARY HYPERTENSION IN BEHÇET'S DISEASE

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Background: Behçet's disease (BD) is a systemic vasculitis that involvement of pulmonary arteries can be seen.
Objectives: The aim of this study was to determine the frequency and the causes of pulmonary hypertension (PH) in patients with BD.
Methods: We studied consecutively 154 BD patients who were fulfilled the International Study Group criteria for diagnosis of BD. All patients were evaluated with transthoracic echocardiography (TTE) for the presence of PH. BD patients were categorised according to the involved organs in 5 groups: group 1 mucocutaneus and articular, group 2 ocular, group 3 vascular, group 4 gastrointestinal and group 5 neurologic involvements. The presence of PH was defined as estimated sPAB ≥40 mmHg, by TTE. Every subject evaluated by a detailed medical history and physical examination was performed. Additional laboratory results were obtained from hospital file records.
Results: The mean age (SD) and the median (min-max) disease duration of the patients were 41.8±12.6 years and 126 (6–540) months respectively. PH was detected in 17 (11%) BD patients. Only 9 (52%) patients were symptomatic (NYHA FC >1). Left sided heart disease (Group II: 9 (52%) patients) was the leading cause of PH. Four (23%) patients had group IV PH and 75% (3/4) were symptomatic. Diastolic dysfunction (DD) was found in 32 (20.8%) patients and only 1 patient had systolic dysfunction. The number of patients with DD was significantly higher in patients with PH as compare to patients without PH (8 (47.1%) vs 24 (17.6%), **p=0.005**). There were no difference in demographic and clinical features of patients with and without PH. Only acneiform lesion was more frequently in patients without PH as compare to patients with PH, **p=0.047** (table 1). There were no differences in frequency of PH in BD groups (table 2).

Abstract AB0651 – Table 1. Demographic and clinical characteristics features of Behçet disease

	All	sPAP ^{3}40 mm HG	sPAP<40 mm HG
Female, n (%)	62 (40.3)	6 (35.3)	56 (40.9)
Age, mean, SD	41.8±12.6	48.1±14.6	41.0±12.1
Diseae duration (month), median (min-max)	126 (6–540)	168 (12–540)	120 (6–480)
Smoking, n (%)	76 (49.4)	7 (41.2)	69 (50.4)
Never smoker	47 (30.5)	5 (29.4)	42 (30.7)
Current smoker	31 (20.1)	5 (29.4)	26 (19.0)
Ex-smoker			
Oral ulcer, n (%)	154 (100)	17 (100)	137 (100)
Genital ulcer, n (%)	104 (67.5)	11 (64.7)	93 (67.9)
EN, n (%)	64 (41.6)	7 (41.2)	57 (41.6)
Papulo-pustular lesion, n (%)	35 (22.7)	3 (22.7)	32 (23.4)
Acneiform lesion*, n (%)	105 (68.2)	8 (47.1)*	97 (70.3)*
Articular, n (%)	33 (22.7)	4 (23.5)	31 (22.6)
Uveitis, n (%)	75 (48.7)	9 (52.9)	66 (48.2)
Pathergy, n (%)	40 (26)	6 (35.3)	34 (24.8)
Vascular, n (%)	48 (31.2)	9 (52.9)	23 (31.1)
Neurologic, n (%)	18 (11.7)	2 (11.8)	16 (11.7)
Gastrointestinal, n (%)	12 (7.8)	1 (5.9)	11 (8.0)

sPAP: Systolic pulmonary artery pressure, EN: Erythema nodosum

Abstract AB0651 – Table 2. Behçet disease group distribution in the pulmonary hypertension patients

Behçet disease (n)	Mucocutaneus and articular (39)	Ocular (75)	Vascular (48)	Gastrointestinal (12)	Neurologic (18)	p
sPAP ^{3}40, n (%)	2 (5.1)	9 (12)	9 (18.8)	1 (8.3)	2 (11.1)	0.413

sPAP: Systolic pulmonary artery pressure

Conclusions: The most frequent cause of PH in BD patients was group II PH. There were also significant number of patients with group IV PH. Pulmonary endarterectomy may be an option of treatment in inactive patients with group IV PH.
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AB0652 PET/MR IN LARGE-VESSEL VASCULITIS: CLINICAL VALUE FOR THE DIAGNOSIS AND ASSESSMENT OF DISEASE ACTIVITY

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Background: The diagnosis and the activity determination could be challenging in large-vessel vasculitis (LVV).
Objectives: The aim of this study was to analyze the value of hybrid PET/MR in LVV.
Methods: All consecutive patients with LVV who underwent PET/MR were included. PET/MR patterns were defined as inflammatory in the case of positive PET (grade=3) and abnormal MR (stenosis and/or wall thickening) and fibrous in the case of negative PET (grade 1 or 2) and abnormal MR.
Results: Thirteen patients with median age at 67 years (23–87 years) and 10 (77%) females were included, and underwent 18 PET/MR scans. Eleven PET/MR performed at diagnosis (n=4) or relapse (n=7) and 7 in patients in remission. 8/18 (44%) had PET/MR inflammatory pattern and 3/18 (17%) had fibrous pattern. PET/MR were normal in 2/10 (20%) cases of TA versus 5/8 (62%) cases of GCA

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