Vasculitis - large vessel vasculitis

AB0576 SERUM 25-HYDROXYVITAMIN D WAS CORRELATED WITH THE DISEASE ACTIVITY IN TAKAYASU'S ARTERITIS

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Background: 25-Hydroxyvitamin D [25(OH)D] deficiency has been linked to autoimmune disease. However, the relationship between 25(OH)D and Takayasu's arteritis (TAK) disease activity remains largely unknown[1]
Objectives: This study aimed to investigate the association between serum 25(OH)D and disease activity in TAK patients.
Methods: A total of 117 patients with TAK (TAK group) and 75 healthy people (control group) were enrolled in our study. Fifty-nine of the patients were re-evaluated after six months of therapy. The serum 25(OH)D levels were compared between the groups. The correlations between the 25(OH)D levels and TAK disease activity were analyzed.
Results: The serum 25(OH)D levels were markedly lower in the TAK patients than those in the healthy subjects [(12.16±4.9) μg/L vs (21.97±8.33) μg/L] (P<0.001). Serum 25(OH)D in all TAK patients was negatively correlated with IL-6 (r=-0.214, P=0.035). The serum 25(OH)D levels in the TAK patients were significantly higher after six months of therapy with glucocorticoid and immunosuppressive agents than those at baseline [(12.16±4.9) μg/L vs (18.01±7.32) μg/L] (P<0.001). Finally, we showed that elevated changes in the 25(OH)D levels before and after treatment were positively correlated with reduced changes in the NIH, ITAS2010 and ITAS-A scores (r=0.490, P<0.001; r=0.496, P<0.001; and r=0.306, P=0.018, respectively).
Conclusion: The serum 25(OH)D levels were significantly downregulated. The changes in 25(OH)D were positively correlated with changes in disease activity.
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AB0577 CLINICAL MANIFESTATIONS AND VARIANTS OF THE COURSE OF TAKAYASU ARTERITIS IN A KYRGYZ COHORT

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Background: The lack of a unified clinical classification and standardized methods of diagnosis creates additional difficulties for timely verification of the diagnosis in real clinical practice. According to a number of studies, there are some differences in phenotypic manifestations among patients with Takayasu arteritis (TAK) of different ethnicity, which determined the objectives of this study.
Objectives: To study the features of the course of the TAK in the Kyrgyz cohort (KC) of patients.
Methods: The angiographic classification of R. Moriwaki et al. was used to determine the anatomical type of vascular bed lesion [1]. The TAK activity was evaluated according to BVAS index, developed by R. Luqmani et al. R. [2], Jefferson's classification was used to characterize the clinical stage of TAK [3]. K. Ishikawa's classification was used to detect the severity of complications and assess the forecast of the disease [6]. All patients underwent a standard laboratory tests, with the mandatory determination of highly sensitive CRP (hs-CRP) and interleukin 6 (IL6). Degree of peripheral vessels lesion was assessed according to data from Duplex Doppler ultrasound scanning of arteries in the color Doppler imaging mode.
Results: The study included 130 patients who were diagnosed with TAK, verified according to criteria of the American College of Rheumatology (ACR, 1990) [4,5] and who received treatment at the NCCIM from January 2014 to November 2021. The majority of patients were women (93.08%), of Kyrgyz nationality (90.77%), of young age - 32 years old [23; 41], with a disease duration of 6.5 years [3.0; 11.0], with the onset of the disease at the age of 23 [18; 31]. By anatomical type of vascular lesion, the patients were distributed as follows: type I was detected in 18 (13.85%), II (a and b) - in 24 (18.46%) patients, III - in 2 (1.54 %), IV - in 5 (3.85%) and V - in 81 (63.3%) patients, which indicated the prevalence of generalized and diffuse lesions of all parts of aorta. According to the BVAS index, most patients had a severe exacerbation - 74 cases (57%), mild exacerbation observed slightly less (42 cases or 32.3%), partial remission occurred in 13 cases (10%) and complete remission in only 1 case (0,77%). By clinical classification, the majority of patients (119 or 91.54%) had vascular stage; none of the patients had the criteria for the prevasculitis stage. In the overwhelming majority of patients (43.84%) nature of vascular lesion was represented by isolated stenosis while a combination of stenosis with occlusive changes was observed in almost 40% of patients.
Conclusion: In the KC of patients with TAK, the overwhelming majority of cases was among women (90.77%) of young age with the onset of the disease at 23 years. Generalized lesions of the arterial bed (63.3%) and severe exacerbation (57%) were mainly observed. The vascular stage predominated (in 91.54% of cases), with the prevalence of stenotic and occlusive changes (40%).
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AB0578 C-REACTIVE PROTEIN TO ALBUMIN RATIO IN BEHÇET'S DISEASE

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Background: Behcet's Disease (BD) is a systemic variable vessel vasculitis that involves the skin, mucosa, joints, eyes, vascular, nervous and the gastrointestinal system.
Objectives: There is no specific laboratory test for the diagnosis and following of Behçet's disease (BD).This study aims to evaluate the relationship between C-reactive protein (CRP)/albumin ratio (CAR) and other hematological parameters in BD.
Methods: A total of 200 participants (100 BD patients and 100 healthy control) were recruited from the rheumatology outpatient clinic in this cross-sectional study. The laboratory data were obtained from the hospital database. Laboratory tests were conducted to measure complete blood count, erythrocyte sedimentation rate (ESR), CRP, albümin, CAR, neutrophil to lymphocyte ratio (NLR), and platelet to lymphocyte ratio (PLR). Laboratory findings of BD patients and healthy controls were compared and evaluated. BD Activity scores (BDCAF/Behcet's Disease Current Activity Form) were calculated. The inclusion criteria were fulfilling the international diagnostic criteria of BD, being over 18 years and accepting to participate in the study.
Results: In the BD group, there were 42 male and 58 female participants with a mean age of 42.49 ± 13.15 and in the healthy control group, 44 male and 56 female participants with a mean age of 44.90 ± 10.98. The age and gender distributions of the groups were as similar (P: 0.16 and P: 0.78, respectively). NLR, CRP, ESR and CAR values were significantly higher in patient group than in the healthy controls (respectively p:0.022, p: 0,013, p:0.001, p:0,011). BDCAF score varied between 0 and 4 with a mean of 1.55± 0.64. A statistically significant correlation was observed between BDCAF and CRP, ESR and CAR (p<0.0001).
Conclusion: In this study, a significant correlation was found between CAR and BDCAF, NLR, CRP, albumin and ESR. CAR can be useful in the diagnosis and following of BD patients. However, further studies are needed.
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AB0579 EVALUATION OF PATIENTS WITH PEDIATRIC BEHÇET'S DISEASE: A TERTIARY CENTER EXPERIENCE

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Background: Behçet's disease (BD) is an systemic inflammatory vasculitis characterized by recurrent oral aphthae and genital ulcers. In the course of the disease, skin, eye, musculoskeletal, nervous system and gastrointestinal system involvements can be detected.
Objectives: To evaluate the clinical, laboratory and radiological findings of pediatric cases diagnosed with BD.
Methods: Fifty patients (0-20 years old) who were followed up with the diagnosis of BD at the Pediatric Rheumatology outpatient clinic of Uludag University Faculty of Medicine between January 2011 and July 2021 were included in our study. The patients were diagnosed according to the diagnostic criteria of the International BD Study Group.
Results: Twenty-four (48%) of the patients were male and 26 (52%) were female. The mean age at the diagnosis of patients was 9 ±4.55 years (10.75±4.55 years in boys, 12.35±3.65 years in girls). Twenty patients (36.3%) had a family history of Behçet's Disease. Oral aphthae were present in 96% (n=48) patients, while genital ulcers were in 32% (n=16) (Table 1). Of the nine patients with uveitis, 6 had panuveitis, 2 had anterior uveitis and 1 had posterior uveitis. . There was no difference in the distribution of symptoms according to the gender of the patients. HLA-B51 allel was found in 39 (78%) and ANA was positive in 14 patients (28%). Immunological tests showed that serum immunoglobulins were low in 11 (32.3%) of 34 patients. Low IgG levels were detected in 6 patients, low IgM levels were in 3, and low IgA levels were in 2. Thrombus was presented in three cases (thrombus in the right ventricle in one case, in the intracranial transverse sigmoid sinus and left jugular vein in two cases). The most commonly used drug for aphthae was colchicine (n=45, 82%). The use of biological agents according to patient manifestations is shown in Table 2. In the follow-up, clinical findings improved in 35 patients (70%) Complete improvement was detected only with biological agents in 8 patients with uveitis. One patient was operated due to the development of complicated cataracts secondary to uveitis. Three patients were diagnosed with Covid-19, one of them was followed without treatment, while two of them were treated with favipiravir at home. Three patients with Covid-19 infection were using only colchicine treatment.

Table 1.

Findings at the time of diagnosis	n	%
Uveitis	9	18
Skin findings	18	36
Gastrointestinal tract involvement	19	38
Central nervous system involvement	6	12
Vascular involvement	3	6
Arthritis	17	34

Table 2.

Findings Biological Agents	Uveitis (n)	Arthritis (n)	Venous thrombus (n)	Central nervous system involvement (n)
Azathioprine	2	2	3	1
Methotrexate	2	2	-	-
Adalimumab	3	1	-	-
Infliximab	3	1	-	-

Conclusion: Behçet's disease is rare in childhood. Although it is not common, life-threatening complications can be observed. To reduce morbidity and complications, physicians should be aware of manifestations and rare clinical pictures of the BD.
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AB0580 BEHÇET'S DISEASE IN OSLO NORWAY IN PERIOD 1999 – 2021

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Background: Bechet's disease (BD) is a multisystemic, inflammatory disease, characterized by variety of clinical manifestations, including mucocutaneous, ocular, vascular, neurological (CNS) and gastrointestinal (GI) involvements. The disease can be seen at any age and occurs across gender, ethnic groups, and geographical areas. However, there are only a few studies from northern Europe.
Objectives: To study the demographics and clinical feature of patients with Bechet's disease (BD) in Oslo Norway.
Methods: In this retrospective study we reviewed hospital records from Oslo University Hospital (OUS), a referral hospital for systemic inflammatory vasculitis in Norway. All patients living in Oslo with ICD-10 code M35.2 were identified, and their BD-related data recorded. Those fulfilling either the International Study Group criteria (ISG) or International Criteria for Bechet's disease (ICBD) were included. Patients were defined as non-Norwegian when none of the parents was of Norwegian ancestry. The differences between groups were compared by Chi-square test using the Statistical Package for the Social Sciences (SPSS).
Results: We identified a total of 58 patients fulfilling the inclusion criteria (Table 1). Among them, 30 (52%) were females and 25 (43%) of Norwegian ancestry. Mean age at onset was 26 years, and at diagnosis 32 years.

Table 1. Demographics of Behcet`s patients in Oslo Norway

Counts (%)	Norwegian ancestry	Non-Norwegian ancestry	All (%)
All (%)	25 (43)	33 (57)	58 (100)
Male (%)	8 (29)	20 (71)	28 (48)
Female (%)	17 (57)	13 (43)	30 (52)
ISG Criteria (%)	20 (47)	23 (53)	43 (100)
ICBD Criteria (%)	25 (43)	33 (57)	58 (100)
Age at diagnosis mean (SD) years	33 (11.85)	32 (9.36)	32 (10.41)
Age at onset mean (SD) years	24 (9.34)	27 (9.52)	26 (9.42)

The most prevalent clinical manifestations were oral ulcers (n=58, 100%), genital ulcers (n=49, 84%) and skin lesions (n=40, 69%). Ophthalmic complications (n=21, 36%), arthritis (n=20, 34%), vascular involvement (n=7, 12%), CNS (n=3, 5%) and gastrointestinal (GI) complications (n= 4, 7%) were also present. There was no significant difference of clinical features between patients with Norwegian ancestry and those with non-Norwegian ancestry (data not shown).

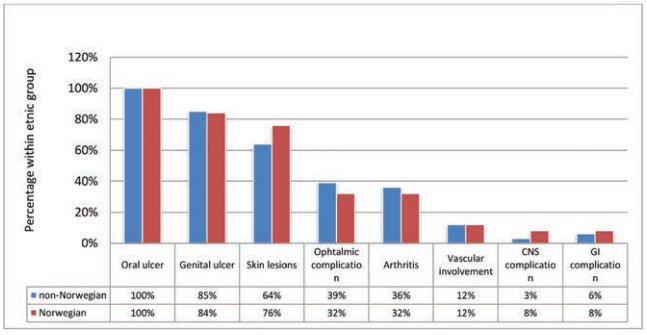


Figure 1.

Conclusion: BD was most prevalent among patients of non-Norwegian ancestry. We found a wide spectrum of clinical manifestations occurring during the disease course. Mucocutaneous lesions were most frequent, but potential serious manifestations were also present. These manifestations were similarly distributed between patients with Norwegian and non-Norwegian ancestry (Figure 1).
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AB0581 TRACHEAL STENOSIS; AN IMPORTANT INVOLVEMENT IN GRANULOMATOSIS WITH POLYANGIITIS

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Background: Granulomatosis with polyangiitis (GPA) is a granulomatous systemic vasculitis of unknown etiology that can affect many organs. Approximately 90% of patients with GPA have upper respiratory tract involvement, including the nasal cavity, sinuses, ear and trachea. Subglottic stenosis (SGS) may develop in some patients due to tracheal involvement.
Objectives: It is aimed to evaluate the general characteristics, treatments and disease prognosis of our GPA cases with tracheal stenosis as it is a significant cause of morbidity and mortality.

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