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THU0287 ANEURYSM IN BEHÇET'S DISEASE: MANAGEMENT, PROGNOSIS AND MORTALITY

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Background: Arterial aneurysms are one of the unique features of Behçet's disease (BD). Although arterial aneurysms are relatively rare, they carry risk of rupture and have poor prognosis.
Objectives: To evaluate the medical and interventional therapies and long-term outcomes of BD patients with extracranial arterial aneurysms.
Methods: We retrospectively reviewed the medical records of 441 BD patients according to ISG criteria between 2013 and 2018 at the Hacettepe University Vasculitis Center (HUVAC). We determined 45 BD patients with an arterial aneurysm. Six patients with isolated carotid and/or cranial arterial aneurysms who were followed by other clinics excluded from the analysis. Overall, 39 BD patients with an extracranial and extra carotidal aneurysm included in the study. Data regarding demographic characteristics, clinical, laboratory and vascular imaging findings, history of medical treatments and outcomes were collected. Vascular intervention of patients were grouped as surgery or endovascular intervention. Radiological response after vascular intervention divided into 3 groups as regression, stable and progression according to radiological evaluation. Regression was defined as a decrease or disappearance of the aneurysm. Stable was defined as no change in the aneurysm diameter. Progression was defined as an increase in diameter of a previous aneurysm or occurrence of a new vascular event. We colligated the stable and regression groups and compared with the progression group. Relapse was defined the progression of the aneurysm which underwent an intervention.
Results: A total of 39 BD patients (Male, 80%) with an arterial aneurysm were analyzed in this study. Mean age and mean age of diagnosis patients were 40.9±11.0 and 29.7±7.7 years, respectively. The clinical features of BD was as follows; oral ulcer 100%, genital ulcer 80.6%, acneiform lesion 47.2%, erythema nodosum 33.3%, pathergy positivity 34.5%, arthritis 27.8%, fever 24.3%, uveitis 34.5%, neurological involvement 30.6% and, gastrointestinal involvement 7.7%. Distribution of arterial aneurysms were 5.1% subclavian, 5.1% coronary artery & cardiac, 38.8% pulmonary, 10.3% thoracic aorta, 33.3% abdominal aorta, 5.1% renal, 25.6% iliac and 12.8% femoral. Induction and maintenance treatments are shown in figure 1. In our cohort, 25 (64%) patients needed a vascular intervention, including 9 (23%) open operations and 16 (41%) endovascular interventions. Of 7 (18%) patients had to perform an urgent open operation or endovascular intervention but we didn't find any relation between the progression of an aneurysm and urgent intervention. First induction treatment options and prognosis of arterial aneurysms shown in figure 2. At median 71.8 months follow up, 7 (18%) patient died in the BD aneurysm group. The rate of progression of the aneurysm was 20% in BD patients with pulmonary artery involvement and 64% in BD patients with aortic and its branch involvement (p=0.038). In the absence of medical induction treatment, 5 patients had progression, 1 had regression and 1 had no follow-up.

Abstract THU0287 – Table 1.

	Induction treatment, n=39 (%)	Maintenance treatment*, n=39 (%)
No treatment, n (%)	7** (17,9)	3 (7.6)
Colchicine	32 (82)	15 (38.4)
Medium-high dose steroid, n (%)	31 (79.4)	11 (28.2)
Cyclophospamide, n (%)	18 (46.1)	9 (23.0)
Interferon-α, n (%)	21 (53.8)	18 (46.1)
Azatiopurine, n (%)	5 (12.8)	11 (28.2)
TNFi, n (%)	1 (2.5)	5 (12.8)
Anticoagulant n (%)	12*** (30.7)	7 (17.9)

*Some patients have received more than one type of immunosuppressive in maintenance therapy
** These patients were followed by rheumatology department after the surgical or endovascular intervention.
*** Patients already using anticoagulants due to venous thrombosis

Abstract THU0287 – Table 2.

Figure 2. Evaluation of response according to vascular intervention and induction treatments *

	Stabile/Regression, n=18	Progression, n=18	p
Vascular intervention			
Done, n=23 (%)	10 (43.5)	13 (56.5)	0.298
Not done, n=13 (%)	8 (61.5)	5 (38.5)	
Induction treatment			
Cyc	2	1	
Cyc+IFN-α	10	5	
IFN	3	2	
Anti-TNF	0	1	0.315
Azatiopurine	2	3	
No treatment	1	5	
Only steroid	0	1	

*3 patients have no follow-up data
Cyc: Cyclophospamide, IFN: Interferon-α

Conclusion: Arterial aneurysms in BD have poor prognosis and high mortality. Glucocorticoids, cyclophosphamide and interferon-α are the most preferred treatments for induction and maintenance therapy for aneurysm of the BD. Approximately 2/3 of the patients required a vascular intervention. Clinical and radiological progression was more prominent in the aorta and its branches. As expected, there is more progression in interventional procedures without medical treatment.
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THU0288 LONG-TERM FOLLOW-UP OF ANTI-IL6-RECEPTOR TOCILIZUMAB IN REFRACTORY UVEITIS IN PATIENTS WITH BEHÇET'S DISEASE. MULTICENTER STUDY OF 14 PATIENTS IN CLINICAL PRACTICE

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Background: Ocular involvement in Behçet's disease (BD) is a potential severe and disabling complication. Anti-TNF-α agents have shown an improvement of visual outcome in BD-related uveitis refractory to conventional immunosuppressive (IS) drugs. However, these drugs do not achieve control of intraocular inflammation in all patients or are not well tolerated. Tocilizumab (TCZ) has shown efficacy in different refractory ocular inflammatory diseases.
Objectives: To assess the efficacy of long-term therapy with TCZ in refractory uveitis associated to extraocular manifestations (EOM) due to BD.
Methods: Multicenter study of patients with BD refractory to standard systemic treatment.
Results: We followed up 14 patients (9 men/5 women) (26 affected eyes); mean age 40.8±19.5 years. Pattern of ocular involvement: panuveitis (10; 4 with retinal vasculitis), anterior (3) and posterior (1) uveitis; 8 recurrent and 6 chronic; 9 with cystoid macular edema. At TCZ onset the following EOM were present: oral and/or genital ulcers (10), arthritis (6), folliculitis/pseudofolliculitis (6), erythema nodosum (3), livedo reticularis (1), intestinal affection (1) and neurological involvement (3).
Before TCZ, they had received corticosteroids (13 intraocular, 12 oral and 12 iv), conventional IS drugs and biologic agents: methotrexate (11), cyclosporine (8), azathioprine (10), colchicine (1), cyclophosphamide (2), mycophenolate mofetil (1), adalimumab (10), infliximab (6), golimumab (3), canakinumab (1), or etanercept (1). TCZ was used in monotherapy (7) or