

Vasculitis

SAT0214

CLINICAL FEATURES OF ANEURYSMAL BEHÇET DISEASE

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Background: Arterial aneurysmal lesions are one of the unique features of Behçet's disease (BD). However, it is difficult to determine which BD patients will develop an aneurysm.

Objectives: In this study, we aimed to determine the differences between BD patients with and without an arterial aneurysm.

Methods: Data of the 23 BD patients with an arterial aneurysm was published as an abstract in 2013 (1). We retrospectively reviewed the medical records of 441 patients with BD according to International Study Group (ISG) criteria between January 2013 and June 2018. Totally, 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 carotid aneurysm included in the study. Data regarding demographic features, clinical, laboratory and vascular imaging findings were collected.

Results: A total of 39 BD patients (Male: 76.9%) with an arterial aneurysm were analyzed in this study. Mean age, mean age of diagnosis and a median follow-up of patients were 40.9±11.0 years, 29.7±7.7 and 71.8 (2-186) months, respectively. The mean age at the onset of the arterial aneurysm was 36.1±11.5 years. The median time lag between the onset of BD and detection of an aneurysm was 63.6 (0-482) months. The prevalence of male patients in BD with aneurysms, without aneurysms and only mucocutaneous were 76.9%, 52.3%, and 34.3%, respectively. Comparison of BD patients with and without an aneurysm shown in table 1. In multivariate analysis, having a venous thrombosis was the sole risk factor for the development of arterial aneurysm (OR 10.53, 95% CI 1.53-72.71) (Table). Distribution of aneurysms was shown in the figure. At the first diagnosis, a median number of the aneurysm was 1 (1-4), the median diameter of an aneurysm was 39 mm (10-80) and 16 (41%) patients had more than one aneurysm. Type of the firstly detected aneurysm was as follows; 38.5% saccular, 17.9% fusiform, 15.4% pseudoaneurysm and 2.6% dissecting aneurysms. Aneurysm type was not known in 25.6% of patients. The first diagnostic finding was an aneurysmal symptom in the 25% of BD patients.

Conclusion: Aneurysm was the diagnostic symptom in 25% of BD patients with an aneurysms. The median time lag between the onset of BD and detection of an aneurysm was almost 5 years. In aneurysmal BD, there is male sex dominance and having venous thrombosis is the most important risk factor. Although pulmonary arteries have an important role in the BD course, the involvement of the aorta and iliac arteries can be seen frequently.

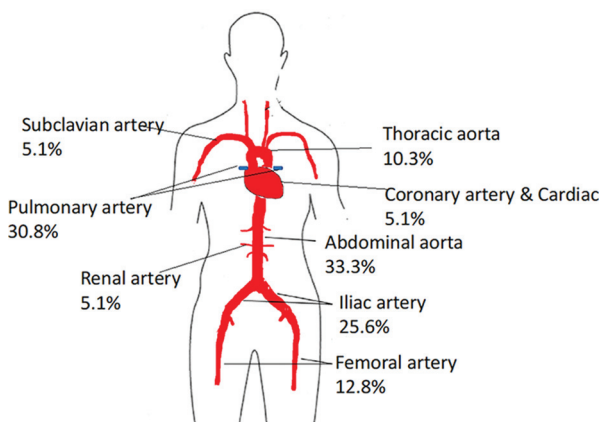


Figure.. Distribution of aneurysms

REFERENCES

[1] Kalyoncu U, et al. Ann Rheum Dis 2013;72 (abs)

Table.. Clinical features of Behçet disease patients with and without arterial aneurysm

	BD with arterial aneurysm, n=39	BD without arterial aneurysm, n=396	p
Age (mean±SD, year)	40.9±11.0	41.0±11.8	0.971
Age at diagnosis (mean ±SD, year)	29.7±7.7	29.2±9.1	0.504
Male	76.9	52.3	0.003
Smoke,%	36.7	48.0	0.235
Oral ulcers,%	100	99.7	1
Genital ulcers,%	80.6	74.5	0.425
Pathergy,%	34.5	44.8	0.303
Erythema nodosum,%	33.3	44.3	0.20
Papulopustular lesions,%	47.2	54.9	0.374
Ocular involvement,%	28.2	51.2	0.006
Arthritis,%	27.8	28.8	0.9
Gastrointestinal involvement,%	7.7	4.6	0.423
Neurologic involvement,%	5.1	18.7	0.033
Fever,%	24.3	NA	NA
Venous thrombosis,%	51.3	20.1	<0.001

BD: Behçet disease, NA: Non available

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SAT0215

APREMILAST IN REFRACTORY ORAL AND/OR GENITAL ULCERS IN BEHÇET'S DISEASE. MULTICENTER STUDY OF 49 CASES IN CLINICAL PRACTICE

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Background: Oral and/or genital aphthous ulcers are the most common symptoms of Behçet's disease (BD), and are often refractory to conventional treatment. The inhibitor of phosphodiesterase-4 Apremilast (APR) has demonstrated efficacy in the treatment of these manifestations.

Objectives: To assess the efficacy and safety of APR in BD patients with oral and/or genital ulcers refractory to conventional treatment.

Methods: National multicenter open-label study on 49 BD patients treated with APR at maintained standard dose of 30 mg twice daily, with the initial 5-day titration schedule in 38 cases. The main outcome was achievement of oral and/or genital ulcers remission.

Results: We included 49 patients (35 women/14 men), mean age of 44.5 ±13.4 years. Before APR, all patients had received several systemic conventional and/or biological drugs: oral corticosteroids (n=45), colchicine (n=48), NSAIDs (n=21), methotrexate (n=27), azathioprine (n=23), cyclosporine (n=9), dapsone (n=6), adalimumab (n=12), infliximab (n=8), tocilizumab (n=3), etanercept (n=3), sulfasalazine (n=2), cyclophosphamide (n=2) and/or others (pentoxifylline, thalidomide, mycophenolate mofetil, hydroxychloroquine, golimumab, 1 each). The main clinical symptoms for starting APR were oral (n=18) and genital (n=2) aphthous ulcers or both (n=29). After a mean follow-up of 8.3±6.8 months, most of the patients experienced main clinical improvement and prednisone dose was reduced or discontinued (TABLE). In this period of time, 31 patients developed any side-effect, most of them transitory: nausea (n=12), diarrhea (n=11),