

Neuro-Behcet's Disease

A clinical review

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ABSTRACT

Objective: To assess the neurological complications of Behcet's disease among Jordanian patients, and to relate these complications with the presence of antiphospholipid antibodies.

Methods: Twenty-two patients with Neuro-Behcet's disease admitted to King Hussein Medical Center, Amman, Jordan between January 1997 and August 1999, were evaluated clinically and by neuroimaging techniques. There were 17 males (77%), 5 females (23%), with a mean age of 36 years (range 18-51). Antiphospholipid antibodies were tested in all the patients.

Results: Brain infarcts were seen in 18 cases; 12 in the anterior fossa, 6 in the posterior fossa, and 2 patients had spinal cord ischemia. Five patients had sinus thrombosis; 4

in the superior sagittal sinus, one in the sigmoid sinus, all were proved by magnetic resonance venogram. All patients were tested for antiphospholipid antibodies and showed negative results.

Conclusion: Brain infarcts are the most common neurological complications of Behcet's disease, less common is sinus thrombosis, and we found no link between these complications and the presence of antiphospholipid antibodies.

Keywords: Behcet's disease, Neuro-Behcet's, sinus thrombosis.

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Behcet's disease is a systemic disease of unknown etiology, producing vasculitis, and manifests mainly with ocular or as oro-genital ulceration. Other system involvement occurs in about 70% of cases, these include pulmonary, joint, cardiac and central nervous systems.¹ Neurological complications of this disease range between 4-29% of cases, and it occurs more with chronic cases,¹ and behaves as a relapsing-remitting disorder, or with progressive course.² Some reports³⁻⁸ indicate a relation between Neuro-Behcet's disease and the presence of antiphospholipid antibodies. In this paper we are trying to ascertain any relation between Neuro-Behcet's disease and the presence of antiphospholipid antibodies.

Methods. Between January 1997 and August 1999, 22 patients with neurological complications of Behcet's disease were evaluated in the Neurology Department at King Hussein Medical Center, Amman, Jordan. All fulfilled the criteria for definite Behcet's disease. The medical records of these patients were reviewed. A special record abstract form was specially designed for the purpose of this study. The age of the patients ranged between 18 and 51 years, with a mean age 36 years. Seventeen cases were male (77%) and 5 were female (23%) with a ratio of 3:1. The disease duration ranged between 2 and 15 years (average 6.5 years), most of the patients were on colchicine, and few patients were on oral

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Table 1 - Distribution of neuro-presentation among the group study.

Type of presentation	Number of cases (%)
Weakness	16 (77)
Ataxia	3 (14)
Diplopia	2 (9)
Headache	7 (32)
Isolated sensory symptoms	3 (14)
% = percentage *totals do not add up to 100	

steroids. Kappa statistics were used to test agreement in antiphospholipid antibodies tests between 2 different laboratory readers.

Results. All patients were referred to the Neurology Unit for further assessment after they presented with neurological manifestations. All patients were evaluated clinically and neuroimaging studies were carried out according to their clinical presentation. Weakness was the most common presenting symptom in the study group, this was seen in 16 cases. Out of these 16 cases, 12 presented with hemispheric manifestations either hemiparesis or hemiplegia, which was relapsing-remitting in 10 cases, and alternating in 6 cases. In all of these cases the disease was active, in the form of oro-genital ulcers and red eyes. Two cases showed brainstem syndrome and both presented with diplopia together with weakness, one showed 6th nerve palsy, and the other 3rd nerve palsy with contralateral weakness. Two patients presented with spinal cord syndrome, one had paraparesis, and one presented with tetraparesis, together with sensory findings and sphincter disturbances. Patients with weakness

Table 2 - Distribution of radiological findings among the study group.

Type of presentation	Number of cases (%)
Ischemic Infarct	20 (80)
Hemisphere	12
Brainstem	2
Cerebellum	2
Spinal Cord	2
Sinus Thrombosis	5 (20)
Superior Sagittal Sinus	4
Signoid Sinus	1
% = percentage	

showed brain hemispheres, brainstem, spinal cord presentations in 12,2,2 cases respectively. Three cases presented with ataxia, one was progressive type and 2 were stable after the initial presentation, and showed improvement later on. Seven cases presented with headache, in 5 of these vomiting was associated, and from these 5 cases 4 showed papilledema, and all these 5 cases proved to have sinus thrombosis. Ten cases presented with sensory complaints, isolated in 3 cases, or with other motor complaints in the remainder of the cases. The distribution of neuro-presentation among the study group is shown in Table 1. The commonest presenting symptom in the study group was weakness, however diplopia was the least common. Neuroimaging studies, including computerized tomography (CT) and magnetic resonance imaging (MRI) were carried out in all cases. Patients presented with, either hemiparesis or hemiplegia, brain CT scan was carried out in all these cases and 10 out of 12 showed evidence of ischemic infarct in one or more of the hemispheres, compatible with the clinical presentation, for the 2 cases in which brain CT scan was negative, for brain MRI confirmed the presence of ischemic infarct in the appropriate area. In the 5 patients with either cerebellar or brainstem symptoms and signs, brain CT scan showed only ischemic infarct in the cerebellum in one case, and in the remainder of the cases, only brain MRI showed ischemic infarct in the brainstem or cerebellum. In all patients who presented only with headache, all 7 cases brain CT scan was normal. Brain MRI and magnetic resonance venogram (MRV) showed 5 cases of sinus thrombosis, 4 in the superior sagittal sinus, and one in the sigmoid sinus, the distribution of radiological findings are presented in Table 2. The most common complication was ischemic infarcts and constituted about 80%, sinus thrombosis was only 20%. Antiphospholipid antibodies were observed for in all study groups, and in all of these cases it was negative. In 10 of the 20 cases the test was carried out in 2 different laboratories to show test agreement, where Kappa statistics were 100%.

Discussion. Behcet's disease is a chronic multi-system vasculitic illness, which has certain criteria for diagnosis, with geographic variation.⁹ Central nervous system involvement occurs late in the disease, and rarely in early stages.² Our study showed that ischemic infarcts were seen in 80% of cases, more in the brain hemispheres (70%) less in the brainstem and cerebellum (20%), and rarely in the spinal cord (10%). Symptoms and signs of increased intracranial pressure were seen in 22% of cases, while similar studies conducted in Iraq showed brain infarcts in 65% of cases, more in the brainstem, and less commonly papilledema (27%).

Other studies showed 51% central nervous system vascular lesions, mainly in brainstem and basal

ganglia, spinal cord involvement was seen in 14% of cases,¹⁰ with similar results found in other studies.¹¹ Magnetic resonance imaging remains the gold standard for the investigation of the neurological manifestation of Behcet's disease.⁹⁻¹² The pattern of findings led to differentiation from multiple sclerosis much easier and earlier.¹² Our findings confirmed the fact that brain MRI is more sensitive than brain CT scan in visualizing ischemic infarcts, especially for posterior fossa lesions.

Many previous reports revealed different frequencies of positive anticardiolipin antibodies in Neuro-Behcet's cases, by inducing intravascular clotting.⁶ Some reports showed 40% positivity of IgG anticardiolipin antibodies,³ others showed 25.5% positive results of IgG, and IgM type.^{5,6} Other reports showed no evidence of these antibodies in Neuro-Behcet's disease,¹³ which is consistent with our findings in which negative results were obtained among the study group.

Neurological manifestations of Behcet's disease are serious and relatively common, and it is more common in males than females probably because the disease itself is more common in males. The brain MRI is the preferable type of neuroimaging study in Neuro-Behcet's disease, and MRV in suspicious cases of sinus thrombosis. The neurological complications of this disease are probably related to the activity of the disease, in which suppression of the activity may delay or prevent these complications. In this study there was no association between the neurological complications and the presence of antiphospholipid antibodies.

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