

Neuro-Behcet's Masquerading as Movement Disorder

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Abstract

Background: Behçet's disease was described as a three-symptom complex comprising uveitis, oral aphthous and genital ulcerations. It is a multi-systemic, recurrent, inflammatory disorder of unknown etiology. CNS involvement occurs in 10-25% of the patients with Behçet's disease. Movement disorders have rarely been reported in neuro-Behçets. **Case Report:** Here in, we are reporting a rare case of neuro-Behçets presenting as acute chorea and dystonia. **Conclusion:** We are presenting case with movement disorder and neuropsychiatric manifestation in form of depression, dystonia, chorea in Behçet's disease.

Keywords: Behçet's Syndrome, Chorea, Dystonia, Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infections.

Introduction

In 1937 Hulusi Behçet first described the eponymous entity of a systemic vasculitis affecting veins and arteries of all sizes. The inflammation in the venous and arterial blood vessels of different sizes explains the manifold signs and symptoms of Behçet's disease. Apart from recurrent oral and genital ulcers; inflammation of the eyes, arthritis, central and peripheral nervous system damages, gastrointestinal and rarely cardiac involvement are other manifestations [1,2]. CNS involvement in Behçet's disease is about 10-25% [4], of which presentation can be either parenchymal (brainstem meningoencephalitis, cranial nerve palsy, epilepsy and peripheral neuropathy) or non-parenchymal (meningitis, cerebral venous thrombosis). Movement disorder is very rare in neuro-Behçets (NB), with few case reports in available literature.

Case Report

A 25 year old male with no significant pre-morbid illness came to our hospital with complains of abnormal movements in left sided limbs for one

week duration which became painful for the last one day. He was also complaining of knee joint pain and tiredness. No history of fever, loss of consciousness, seizures, headache, limb weakness, numbness, bowel or bladder incontinence, recurrent oral or genital ulceration, eye infections or multiple joint pain could be elicited.

He was also suffering from depression for the last six months and on treatment for the same. He had a past history of road traffic accident in 2016. On examination he is moderately built with normal general examination and no neuro-cutaneous markers. Nervous system examination showed generalized chorea along with dystonic posturing involving all four limbs, more predominantly in left sided limbs with bilateral extensor plantars. Other systemic examination findings were non contributory. Rheumatic chorea/ Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infections (PANDAS)/ autoimmune encephalitis were considered initially.

On evaluation, all basic hematological investigations were normal. Lumbar puncture and cerebro-spinal fluid analysis were non-contributory. MRI brain showed multifocal asymmetric discrete and partly confluent lesions in bilateral basal ganglia, thalami, brain stem and corona radiata with mesodiencephalic preponderance with patchy areas of enhancement in these lesions. Hence CNS vasculitis secondary to neuro-Behcet's or demyelinating disorder were considered at this point. His vasculitis profile (ANA, ANCA, APLA) were sent and they all came negative. Additional investigation like pathergy test was performed to confirm the diagnosis of Behcet's, which came negative. Finally HLA typing was done and HLA-B52 came out to be positive. Cardiology consult was sought and it was normal.

The patient was treated with intravenous pulse steroids for five days followed by tapering dose of steroids along with azathioprine, haloperidol and supportives. Gradually patient started improving, later on discharged with tapering dose of steroids and azathioprine and followed up as outpatient.

Discussion

Behcet's disease (BD) is now known to have a wide systemic spectrum. The age of onset is 24-54 years with the mean age of 32.6 years [3]. Males are more frequently affected than females. The mean duration of illness is 6.5 years [3]. The major diagnostic criteria include oral ulceration (75%), genital ulceration (71%), eye lesion (28.6%), and skin lesions. Minor criteria include arthritic, neurologic (50%), gastrointestinal and vascular symptoms such as thrombophlebitis (42.9%) [3]. Central nervous system (CNS) involvement occurs in 10-25% of the patients with BS [4]. The presentation can be either parenchymal (brainstem meningoencephalitis, cranial nerve palsy, epilepsy and peripheral neuropathy) or non-parenchymal (meningitis, cerebral venous thrombosis). The most common neuro-pathologic findings in neuro-Behcet's disease are focal necrotic lesions in the brain.

In the absence of ocular lesions, oral and genital ulceration, the evaluation of neurologic signs which occur during the course of Behcet's disease is challenging. Movement disorders have rarely been reported in NB [5,6]. In a study of Benamour *et al.* [7], 925 patients with BS were analyzed, and 154 of them had NB in which only one patient had chorea. A literature review by Kuriwaga *et al.* showed that the onset of chorea in cases of BS ranges from the time of onset of BD to 31 years after the onset of the disease [5]. The clinical picture, pathology and brain MRI may not be concordant in cases with NB [8]. Autopsy studies have showed that in NB disease, low-grade chronic lymphocytic meningo-encephalitis with widespread peri-venular neutrophilic or lymphocytic and plasmocytic cuffing with multifocal necrotic foci are observed. These changes are mostly seen in the brain stem and basal ganglion region and to a lesser extent in the spinal cord [9,10]. Although basal ganglion is frequently involved in neuro-Behcet's disease, movement abnormalities are rare.

Usually, the few cases of NB with chorea in the literature reveal high intensity on T2-weighted images and on FLAIR sequence in the periventricular white matter and basal ganglia. FLAIR sequence demonstrating lesions are clearer than the T2-weighted MRI [5,6]. Arber *et al.* studied epidemiological data, family history, clinical data, and HLA typing in three groups of patients with Behcet's syndrome and found that HLA-B51 and B52 were present in 63% and 21%, respectively, of the patients [11]. 95% of the affected family members were either B51 or B52 positive. In our patient HLA-B52 was positive.

In the case of Kurikawa *et al.* [5], treatment with prednisolone resolved the chorea, suggesting that chorea was caused by an autoimmune mechanism. In our case, a modest improvement in chorea was observed after treatment with both the immunosuppressants and azathioprine, and it is hard to differentiate which one made the difference.

Conclusion

We are presenting this case who presented to us with movement disorder and neuropsychiatric manifestation in form of depression, dystonia, chorea without other features of Behcet's disease. The diagnosis is made based on the characteristic imaging finding and HLA-B52 positivity.

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References

- Mendes D, Correia M, Barbedo M, Vaio T, Mota M. Behcet's disease a contemporary review. *J Autoimmun.* 2009;32:178-188.
- Koseoglu E, Yildirim A, Borlu M. Is headache in Behcet's disease related to silent neurologic involvement? *Clin Exp Rheumatol.* 2011;29(Suppl. 67): S32-37.
- Kyung DP, Dongsik B, Eun SL, *et al.* Clinical study on death in Behcet's disease. *J Kor Med Sci.* 1993;8:241-245.
- Ildan F, Gocer AI, Bagdatoglu H, Tuna M, Karadayi A. An intracranial arterial aneurysm complicating Behcet's disease. *Neurosurg Rev.* 1996;19:53-56.
- Kuriwaka R, Kunishige M, Nakahira R, Inoue H, Higashi T, Tokumoto Y, *et al.* Neuro-Behcet's disease with chorea after remission of intestinal Behcet's disease. *Clin Rheumatol.* 2004;23:364-367.
- Kimura N, Sugihara R, Kimura A, Kumamoto T, Tsuda T. A case of neuro-Behcet's disease presenting with chorea. *Rinsho Shinkeigaku.* 2001;41:45-49.
- Benamour S, Naji T, Alaoui FZ, El-Kabli H, El-Aidouni S. Neurological involvement in Behcet's disease. 154 cases from a cohort of 925 patients and review of the literature. *Rev Neurol (Paris).* 2006;162:1084-1090.
- Akman-Demir G, Serdaroğlu P, Tasci B. Clinical patterns of neurological involvement in Behcet's disease: evaluation of 200 patients. The Neuro-Behcet Study Group. *Brain.* 1999;122:2171-2182.
- Sugihara H, Muto Y, Tsuchiyama H. Neuro-Behcet's syndrome: report of two autopsy cases. *Acta Pathol Jpn.* 1969;19:95-101.
- Hadfield MG, Aydin F, Lippman HR, Sanders KM. Neuro-Behcet's disease. *Clin Neuropathol.* 1997;16:55-60.
- Arber N, Klein T, Meiner Z, Pras E, Weinberger A. Close association of HLA-B51 and B52 in Israeli patients with Behcet's syndrome. *Annals of the Rheumatic Diseases.* 1991;50:351-353.