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Hemiballismus Secondary to Neuro-Behçet Disease: Case Report

Nöro-Behçet Hastalığına Bağlı Hemiballismus: Olgu Sunumu

Turgay Demir, Şebnem Bıçakçı, Miray Erdem

Çukurova University Faculty of Medicine, Department of Neurology, Adana, Turkey

Summary

Behçet's Disease is a multisystemic, inflammatory, recurrent disorder with oral and genital ulcerations along with cutaneous and ophthalmic symptoms. Central nervous system involvement is called Neuro-Behçet's Disease. Extrapyramidal symptoms are rare in Neuro-Behçet's Disease. We report here a case with Neuro-Behçet disease presented with acute hemiballismus. (Turkish Journal of Neurology 2014; 20:60-2)

Key Words: Extrapyramidal, hemiballismus, Neuro-Behçet

Özet

Behçet Hastalığı oral ve genital ülserler, cilt ve oftalmolojik bulguların gözlendiği multisitemik, inflamatuvar, tekrarlayıcı bir hastalıktır. Nöro-Behçet ise santral sinir sistemi etkilenmesinin olduğu hastalığın klinik formlarından birisidir. Nöro-Behçet olgularında ekstrapiramidal bulgular oldukça nadir görülmektedir. Biz bu yazıda akut hemiballismus ile prezente olan Nöro-Behçet olgusunu sunmak istedik. (Türk Nöroloji Dergisi 2014; 20:60-2)

Anahtar Kelimeler: Ekstrapiramidal, hemiballismus, Nöro-Behçet

Introduction

The etiology of Behçet disease (BD), a chronic, recurrent, systemic vasculitis, is still unknown. In addition to the mucocutaneous lesions, eye, vascular, articular, gastrointestinal, urogenital, pulmonary and neurological involvements constitute the disease's clinical profile (1). In the 20-40 age group, it is seen more commonly in men than women. There is a geographical difference in the prevalence of the disease. This prevalence, being higher in the Mediterranean and Middle Eastern countries and Japan, was seen to be 13.5/100.000 in Japan and 0.64/100.000 in England. In Turkey, the prevalence was estimated as 8-42/10.000 (1-6).

Central nervous system (CNS) involvement in BD has been shown to range between 2.2% to 50% of the cases (7). Usually 5 years after the onset of the systemic findings does the CNS involvement takes place. This condition is named as neurobehçet syndrome (NBS). The most common neurological finding seen in NBS cases is headache (8). Pyramidal findings, memory problems, disinhibition, apathy, sphincter involvement and cerebellar syndrome are the primary clinical findings in that order (9). Extrapyramidal system, peripheral nervous system and primary muscle bundle involvement are rarely seen in NBS cases (10). Here, we present a case of BH that has been followed for 3 years. The case came to the emergency policlinic with involuntary movements in the left upper and lower extremities and loss of balance. This case is reviewed in the context of the literature on NBS spectrum.

Case

The 32 year old male patient came to the emergency policlinic with involuntary movements on the left arm and leg and loss of balance that started 24 hours ago and got progressively worse. Three years before this visit, he was diagnosed with BD after the examination made to investigate his oral and genital ulcerations. For the past 6 months, he was given 40 mg intravenous

Address for Correspondence/Yazışma Adresi: Turgay Demir MD, Çukurova University Faculty of Medicine, Department of Neurology, Adana, Turkey
Phone: +90 322 338 60 60-3206 E-mail: drtdemir@gmail.com
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methylprednisolone once for the impaired vision on his left eye, and then was put on 24 mg/day oral methylprednisolone and tetracycline. He did not report fever, headache, trauma, antipsychotic drugs or toxic substance use prior to the onset of his complaints.

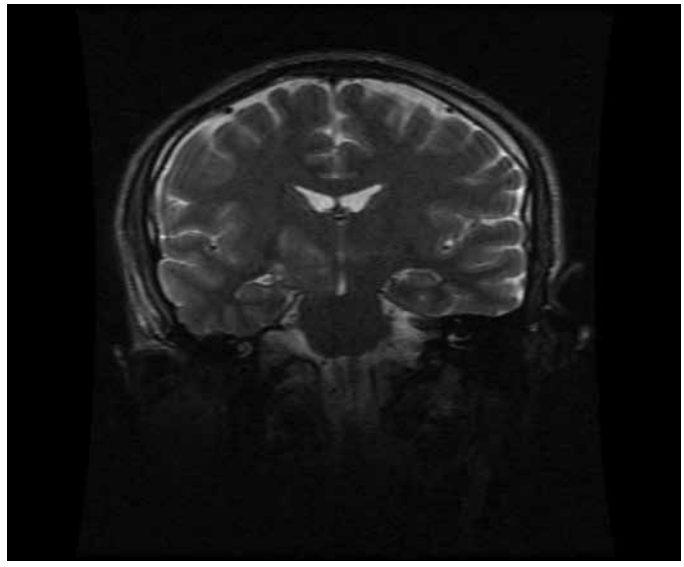
In the neurological examination, he was moderately drowsy with dysarthric speech and intact time-person-location orientation; there was optic atrophy on the left in the fundoscopic exam, nasolabial sulcus was undefined, bilateral hand squeeze was minimally weak. There were ballistic movements on the left upper extremity, impaired finger-nose test and knee-heel test on the left, hypotonia on the left upper and lower extremities and a tendency to fall during walking. His electrocardiography sinus was rhythmical and normocardic, his blood biochemistry and hemogram values were normal and his uncontrasted cerebral computerized tomography (CT) did not show any pathologies. In the cerebral magnetic resonance imaging (MRI), there was a signal pathology that indicated a late, subacute infarction starting from the brainstem and moving on to the insular cortex, involving thalamus and basal ganglia (Figure 1). Carotid and vertebral artery color Doppler ultrasonography and transthoracic echocardiography were normal. There were diffuse basal rhythm disturbances and slow activity paroxysms of subcortical nature in his electroencephalography.

The case was evaluated as parenchymal NBS. He was given 7 day, 1 gr/day intravenous methylprednisolone and 10 mg/day oral haloperidol. His neurological examination after 24 hours showed a decrease in his findings, especially the involuntary movements. In the follow-up examination a week after, there was a marked improvement for the extrapyramidal and cerebellar findings on the left upper and lower extremities. After consulting with the rheumatology clinic and taking his clinical findings into consideration, he was started on cyclophosphamide on the 10th day of his admission. He was switched to oral methylprednisolone and it was planned that he would stop using it completely. In the one-month follow-up, all of the findings except or the left ophthalmic atrophy were seen to be improved; his follow-up cerebral MRI showed a marked improvement for the lesions (Figure 2).

Discussion

The cerebral involvement in NBS can be either parenchymal or non-parenchymal. It appears at five to ten years after the disease onset, which corresponds to the late stage, in 2.2%-50% of all Behçet patients regardless of gender. In the parenchymal involvement, the findings are often related to the involved areas while in the non-parencymal involvement, the most common indications are venous sinus thrombosis, cerebral venous infarcts, increased intracranial pressure syndrome (IIPS) or epileptic seizures due to either conditions (11). Our case consulted for optic system problems described in the medical history and, acutely developing extrapyramidal and cerebellar involvement findings. The condition was diagnosed as parenchymal involvement secondary to NBS after the clinical and radiological evaluation.

Neurobehçet syndrome progressing with extrapyramidal findings is rarely reported in the literature, with these few instances being individual cases (9,12,13). The first time was when Bogdanova et al. reported pseudobulbar palsy, mild and asymmetrical bilateral pyramidal syndrome, rigidity, bradykinesia, mask-like face and impaired postural reflexes, postural tremor on the extremities, myoclonic jerks on the facial muscles and tongue and multifocal hyperintense small, bilateral lesions in the periventricular white matter, brainstem and basal ganglia seen in the cerebral MRI of a patient with BD who developed parkinsonism (12). Kimura et al. reported concentration difficulty, emotional lability, amnesia, bilateral upper extremity and cervical chorea, bilateral pyramidal findings along with bilateral caudate nucleus atrophy and diffuse white matter lesions hypointense in T1 and hyperintense in T2 adjacent to lateral ventricle in the cerebral MRI during the neurological workup of a 46 year old male patient who consulted for involuntary movements on the upper right extremity (13). Revilla et al. described chorea and jaw dystonia in a NBS case and found hyperintense lesions in the brainstem and bilateral basal ganglia and deep white matter in the T2 sequence (9).



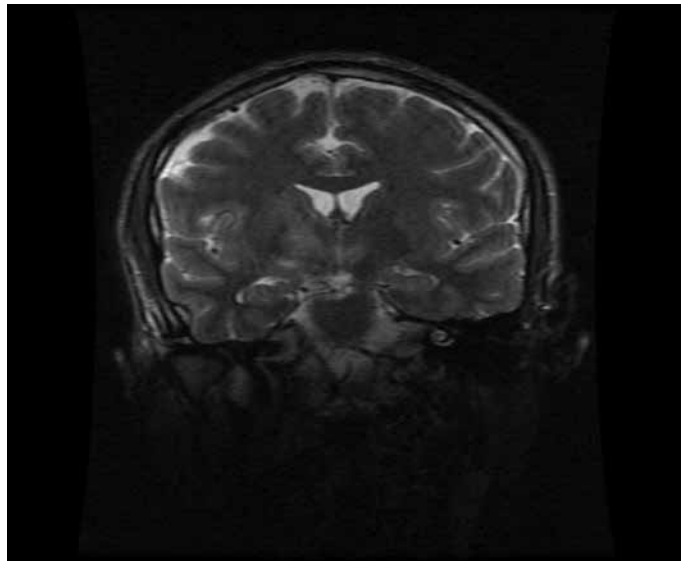


Figure 1 and 2. Pre-treatment coronal T2 MRI

All three of these cases showed basal ganglia, deep white matter and brainstem involvement in the cerebral MRI. Our case also showed wide-spread signal pathology starting from brainstem and radiating towards basal ganglia and right temporal lobe, terminating at around Sylvian fissure. Al-Araji and Lee showed thalamus and basal ganglia involvement with the rates 33% and 43% respectively but they did not report any extrapyramidal system involvement clinically (7,14). These results suggest that basal ganglia involvement is not the only underlying reason and that there could be several other mechanisms at play. SPECT studies showed that perfusion changes could be present in the absence of clinical and cerebral MR evidence (15). This finding suggests the existence of a subclinical involvement. In addition, the researchers also proposed that the cerebral damage is not only caused by vascular factors but also the neutrophilic and eosinophilic inflammation of the brain tissue (15).

Improvements in the clinical as well as radiological findings are commonly seen in NBS. Gerber et al. reported improvement in the 35% in the cerebral lesions and complete resolution in 40% of the cases (16). In our case, it was noteworthy that the MRI lesions that were correlated with the clinical findings were significantly improved at the 1-month follow-up (Figure 2).

In conclusion, NBS should be taken into consideration in the differential diagnosis of the acute-subacute developing extrapyramidal system involvement in the younger cases and such cases should be evaluated clinically and radiologically.

References

1. Alpsoy E, Akman A. Behçet's disease: an algorithmic approach to its treatment. Arch Dermatol Res 2009;301:693-702.
2. Chamberlain M A. Behçet's syndrome in 32 patients in Yorkshire. Ann Rheum Dis 1977;36:491-499.
3. Demirhindi O, Yazıcı H, Pinyıldız P, Dayıoğlu N, Tüzün Y, Altaç M, Or H, Müftüoğlu A, Yurdakul S, Pazarlı H. Silivri Fener Köyü ve Yöresinde Behçet Hastalığı sıklığı ve bu hastalığın toplum içinde taranmasında kullanılabilecek bir yöntem. CTFD 1981;12:509-514.
4. Yurdakul S, Günaydın İ, Tüzün Y, Tankurt N, Pazarli H, Ozyazgan Y, Yazici H. The prevelance of Behçet's syndrome in a rural area in Northern Turkey. J Rheumatol 1988;15: 820-822.
5. İdil A, Gürler A, Boyvat A, Caliskan D, Ozdemir O, Isik A, Tunçbilek A, Koçyigit P, Calikoglu E. The prevalance of Behçet's disease above the age of ten years. The results of a pilot study conducted at the Park Primary Health Care Center in Ankara, Turkey. Opthalmic Epidemol 2002;9:325-331.
6. Azizlerli G, Köse AK, Sarıca R ve ark. Prevalance of Behçet's disease in Istanbul, Turkey. Int J Dermatol 2003;42:803-806.
7. Al-Araji A, Sharquie K,Al-Rawi Z. Prevalance and patterns of neurological involvement in Behcet's disease: A prospective study from Iraq. J Neurol Neurosurg Psychiatry 2003;74:608-613.
8. Monestero R, Mannino M, Lopez G, Camarda C, Cannizzaro C, Camarda LK, Ciccia F, Triolo G, Camarda R. Prevalence of headache in patients with Behçet's disease without overt neurological involvement. Cephalalgia 2003;23:105-108.
9. Saylan T, Mat C, Fresko İ, Melikoğlu M. Behcet's Disease in the Middle East. Clin Dermatol 1999;17:209-223.
10. Revilla FJ, Racette BA, Perlmutten JS. Chorea and Jaw-Opening Dystonia as a manifestation of NeuroBehcet's syndrome. Mov Disord 2000;15:741-744.
11. Akman-Demir G, Serdaroglu P, Tasçi B. The Neuro-Behçet Study Group. Clinical patterns of neurological involvement in Behçet's disease : evaluation of 200 patients. Brain 1999;122:2171-2181.
12. Bogdonova D, Milanov I, Georgiev D. Parkinsonian syndrome as a neurological manifestation of Behçet's disease. Can J Neurol Sci 1998;25:82-85.
13. Kimura N, Sugihara R, Kimura A, Kumamoto T, Tsuda T. A case of neuro-Behçet's disease presenting with chorea. Rinsho Shinkeigaku 2001;41:45-49.
14. Lee SH, Yoon PH, Park SJ, Kim DI. MRI Findings in Neuro-Behcet's Disease. Clin Radiol 2001;56:485-494.
15. Vignola S, Nobili F, Picco P, Gottorno M, Buoncompagni A. Brain Perfusion Spect in juvenile neuro-Behcet's Disease. J Nucl Med 2001;42:1151-1157.
16. Gerber S, Biondi A, Dormont D, Wechsler B, Marsault C. Long-term MR follow-up of cerebral lesions in neuro-Behçet's disease. Neuroradiology 1996;38:761-768.