NÖRO PSİKİYATRİ ARŞİVİ

Letter to the Editor / Editöre Mektup

Arch Neuropsychiatry 2017; 54:376-378 • DOI: 10.5152/npa.2017.18121

A Patient with Neuro-Behçet's Syndrome Presenting with Peripheral Nerve Involvement

Periferik Sinir tutulumuyla prezente Nörobehçet Sendromlu Ender Bir Olgu

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Dear Editor,

We observed the case of a male patient with Neuro-Behçet's syndrome with peripheral nerve involvement 5 weeks ago. Here we would like to comment about this observed rare case.

Behçet's disease (BD) is regarded as an auto-inflammatory process characterized with dominancy of pro-inflammatory cytokines in an immunological manner. Vascular, parenchymal, and meningeal involvement of the central nervous system is commonly observed in BD (1). The brainstem region comprises the most commonly affected structures in clinical picture, named as Neuro-Behçet's syndrome (NBS). Moreover, spinal cord lesions, hemispheric lesions, and meningoencephalitis could occur in patients with NBS. Peripheral neuropathy could also exceptionally occur (1,2,3,4,5). The exact underlying mechanism of peripheral neuropathy in BD is still unknown. One of the possible mechanism is ischemic nerve injury secondary to vasculitis of the vasa nervorum; another mechanism is neuropathy due to the side effects of the used drugs (e.g., colchicine) (5). In most of the patients, peripheral neuropathy is prominent in the lower extremities (1). Although sensorial findings in patients with NBS generally occur as hemihypoesthesia or hypoalgesia, hyperesthesia or hyperalgesia could also occur. In this paper, the case of a patient with NBS with a complaint of allodynia on his left foot, diagnosed as vasculitic neuropathy, is presented.

A 27-year-old man presented to us with a chief complaint of serious burning sensation and pain on his left leg that began 10 days ago. He could not sleep or leave his home because of these symptoms. Regarding his past medical history, he was diagnosed with BD 10 years ago. He also has a history of peripheral vascular disease and is a smoker. Azathioprine and cyclosporine had been used for 4 years for the management of BD, but azathiopirine was changed to cyclophosphamide after the development of uveoretinitis. The cyclophosphamide treatment was also discontinued after 7 months upon the recurrence of lower respiratory tract infections. During neurological examination, the muscle strengths were 4/5 on proximal and distal muscles of the left upper extremity, 4/5 on proximal muscles of the left lower extremity, and 3/5 on distal muscles of the left lower extremity. There was normal bilateral reflex in the sole skin, and deep tendon reflexes were normoactive in all extremities. Cranial examinations were normal. He described a severe pain sensation in the skin over the superficial peroneal nerve innervation during cutaneous sensory examination of his left foot with a soft cotton. The vibration sense reduced lower limbs by diapozon. Brain magnetic resonans imaging (MRI) revealed hyperintense lesions in both sides of the pons, left crus cerebri in the midbrain, and right side of the thalamus extending through the internal capsule on T2-FLAIR sequences (Figure 1a, b, c). After injection of gadolinium, these lesions showed contrast enhancement on T1-weighted images (Figure 1d, e, f). CSF analysis could not be performed because the patient did not give consent. The following laboratory examinations were normal: complete blood count, blood sedimentation rate, blood glucose level, glycated hemoglobin, serum electrolytes, urea, creatinine, serum protein content, cholesterol, triglycerides, vitamin B12, vasculitis markers, and liver and muscle enzymes.

Nerve conduction studies (NCSs) revealed an elongation of the distal motor latency of the left tibial nerve on the abductor hallucis muscle (4.10 ms on left side versus 3.05 ms on right side), slowing of the motor nerve conduction velocity (37.6 m/s on left side vs. 42.2 m/s on right side), and elongation of minimum F wave latency (56.9 ms on left side versus 50.4 ms on right side). Although the latency and conduction velocity were similar, the sensory nerve action potential amplitude of the superficial peroneal nerve was lower in the left foot than in the right foot (6.5 μV versus 20.9 μV, respectively). The other peripheral NCSs were completely normal. Electromyography conducted with a concentric argent needle electrode revealed spontaneous denervation potentials on the left abductor hallucis muscle. During maximal contraction, neurogenic motor unit potential changes characterized with decreased frequency and polyphasic pattern were observed on the left gastrocnemius and abductor hallucis muscles. The clinical and electrophysiological findings of our patient

Cite this article as: Kotan D, Alemdar M, Alaçam Köksal S. A Patient with Neuro-Behçet's Syndrome Presenting with Peripheral Nerve Involvement. Arch Neuropsychiatry 2017; 54:376-378.

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Received/Geliş Tarihi: 09.05.2016 **Accepted/Kabul Tarihi:** 06.06.2016

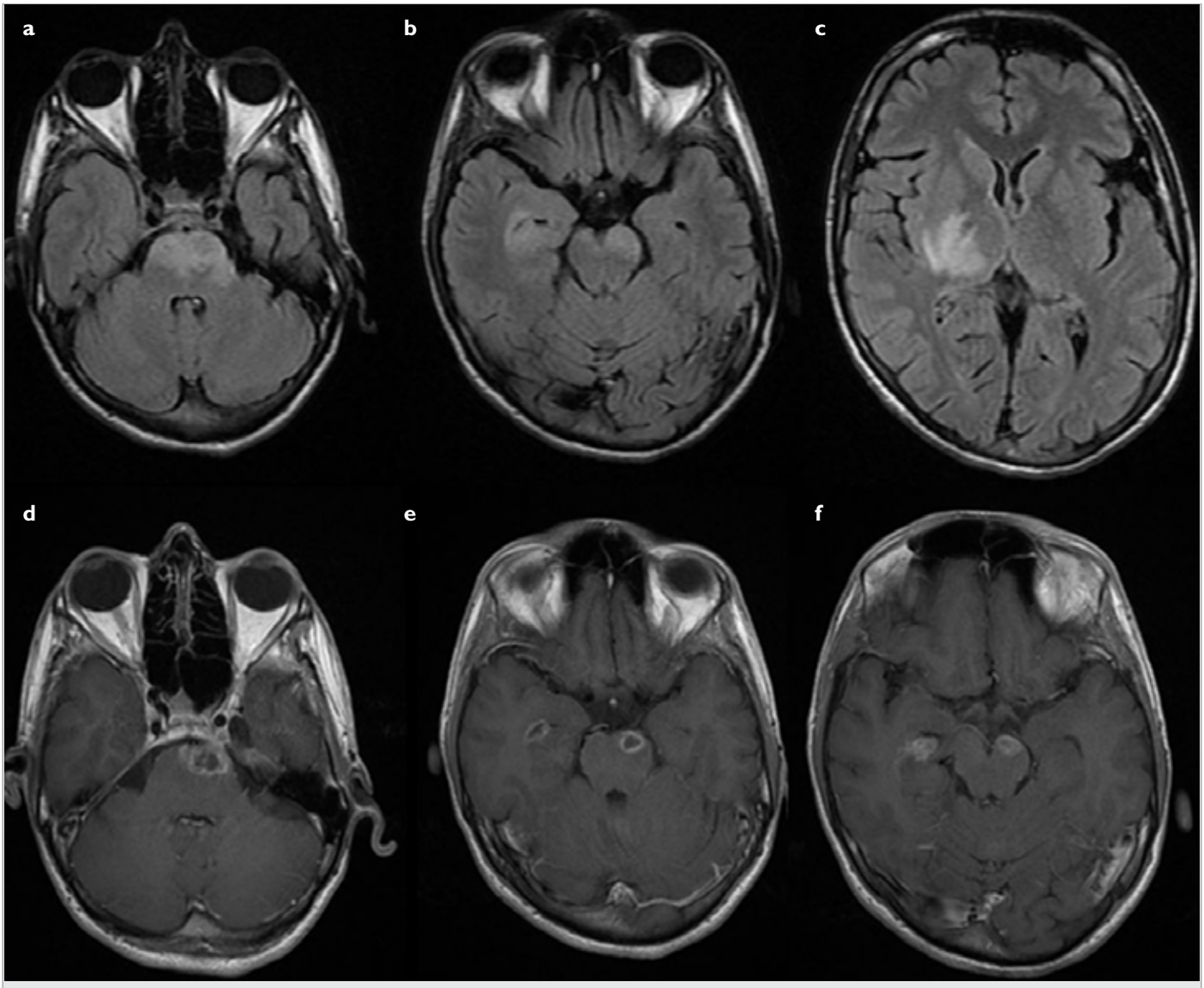

Figure 1. a-f. Brain MRI revealed hyperintense lesions on both sides of the pons, left crus cerebri in the midbrain, and right side of the thalamus on T2-FLAIR sequences (a, b, c). After injection of gadolinium, these lesions showed contrast enhancement on T1-weighted images (d, e, f)

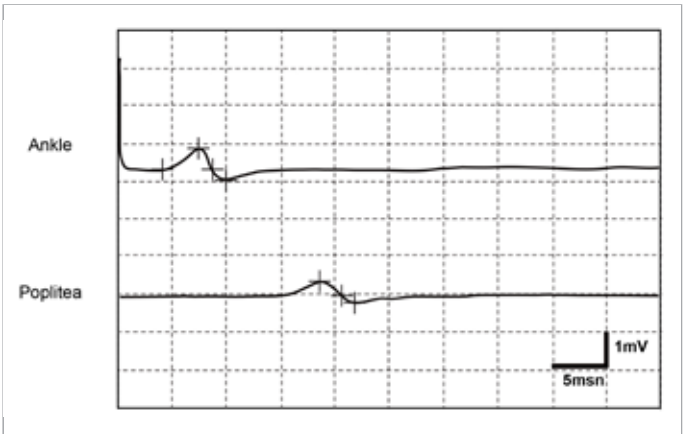

Figure 2. The trace obtained during the motor nerve conduction study of the left tibial nerve

were concordant with probable vasculitic neuropathy. Because of the serious neuropathic complaints and allodynia, he clearly ignored other problems related to his neurologic deficits. We administered intravenous pulse methylprednisolone (1 g/day for 10 days) treatment and new planned on continuing infliximab for the management of BD. Moreover, we added 600 mg/day of pregabalin and tramadol/paracetamol combination for the neuropathic pain. The maintenance treatment with pregabalin tablet resulted in alleviation of his complaints.

Neuro-Behçet's syndrome is one of the more serious manifestations of BD, which is a relapsing inflammatory multisystem disease. Although NBS is relatively uncommon, being potentially treatable, neurologists need to consider it in the differential diagnosis of inflammatory, infective, or demyelinating disorders (6). The reported frequency of peripheral nerve disorders varied in different case series of NBS (3%-14%) (1,2,3). Sensorimotor polyneuropathy is the most frequent type of involvement. However, Guillain-Barré syndrome, mononeuritis multiplex, and autonomic neuropathy could also occur (1,2,3,4,5). To the best of our knowledge, there is no report in the literature that presented the case of a patient with BD with the signs and symptoms owing to isolated unilateral tibial nerve and superficial peroneal nerve involvements as in our patient. The cases of patients suffering from cutaneous allodynia are rarely seen in NBS.

The symptoms secondary to disturbances of sensory fibers could negatively affect the quality of life of patients with NBS. As the affected cutaneous region was well-demarcated with its sensory innervation area and NCSs confirmed its axonal involvement, allodynia was regarded as a result of secondary central sensitization triggered by the disturbance of superficial peroneal nerve in this case, rather than a central pain secondary to thalamic involvement. As the addition of symptomatic agents to the pharmacological treatments of the patients could alleviate the neuropathic pain and increase their quality of life, clinicians should consider the possibility of peripheral nerve involvement when evaluating patients with BD. Pain, allodynia, and other noxious sensory complaints coexisting with motor signs in the same extremity should be redolent of peripheral sensory nervous system involvement, and NCSs must be performed in suspected cases.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - D.K.; Design - D.K.; Supervision - D.K., M.A.; Resource - D.K.; Materials - D.K., M.A.; Data Collection and/or Processing - D.K., S.A.K.; Analysis and/or Interpretation - D.K., M.A.; Literature Search - D.K.; Writing - D.K.; Critical Reviews - D.K.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

Hasta Onamı: Yazılı hasta onamı bu çalışmaya katılan hastadan alınmıştır.

Hakem Değerlendirmesi: Dış Bağımsız.

Yazar Katkıları: Fikir - D.K.; Tasarım - D.K.; Denetleme - D.K., M.A.; Kaynaklar - D.K.; Malzemeler - D.K., M.A.; Veri Toplanması ve/veya İşlemesi - D.K., M.A.; Analiz ve/veya Yorum - D.K., M.A.; Literatür Taraması - D.K.; Yazıyı Yazan - D.K.; Eleştirel İnceleme - D.K.

Çıkar Çatışması: Yazarlar çıkar çatışması bildirmemişlerdir.

Finansal Destek: Yazarlar bu çalışma için finansal destek almadıklarını beyan etmişlerdir.

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