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Case Report

Cerebral Abscesses in Behcet's Disease: A Case Report

Behçet Hastalığında Serebral Abseler: Olgu Sunumu

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ABSTRACT

Behçet's disease is a multisystem relapsing inflammatory disorder of unknown cause. Neurological involvement is one of the most serious causes of long-term morbidity and mortality in Behçet's disease. Cerebral abscess is very rare in literature. A 45-yr-old man with Behçet's disease manifesting focal epileptic seizure and multiple cerebral abscesses is reported in the case. He was diagnosed with cerebral abscess and was treated with antibiotics but no improvement occurred. Excisional biopsy was performed and the lesions were consistent with abscess. The clinical state of the patient gradually improved. The patient had no further complications. The etiology, and clinical and magnetic resonance imaging findings are discussed.

KEYWORDS: NeuroBehcet disease, Behcet's disease, Cerebral abscess

ÖZ

Behçet Hastalığı nedeni bilinmeyen relapslarla seyreden multisistemik inflamatuvar bir hastalıktır. Nörolojik tutulum uzun dönem mortalite ve morbiditenin en ciddi nedenlerinden bir tanesidir. Serebral abse ise literatürde oldukça nadir görülmektedir. Fokal epileptik nöbetle gelen multipl serebral abseleri olan 45 yaşında bir erkek Behçet hastası sunuldu. Serebral abse tanısı alan hastaya antibiyotik tedavisi başlandı ancak bir düzelme görülmedi. Daha sonra eksizyonel biyopsi yapıldı ve alınan lezyonlar abse ile uyumluydu. Sonrasında hastanın klinik durumu giderek düzeldi. Hasta halen herhangi bir komplikasyonu olmaksızın yaşamını sürdürmektedir. Etiyoloji, klinik ve manyetik rezonans görüntüleme bulguları tartışıldı.

ANAHTAR SÖZCÜKLER: NöroBehçet hastalığı, Behçet hastalığı, Serebral abse

INTRODUCTION

Behçet's disease (BD) is a systemic inflammatory disease commonly characterized by oral and genital ulcerations, with skin and eye involvement, and meningoencephalitis. BD is a rare condition, seen more commonly in Turkey, as well as in Middle Eastern, Mediterranean and Far Eastern countries. Its cause is still unknown, but vasculitis is the major pathological feature (6). A brain abscess is a rare, serious, and life-threatening complication in BD. We present a BD case with cerebral abscess that presented with focal epilepsy.

CASE REPORT

A 45-year-old man was diagnosed as having BD four years ago. The day before admission, he had experienced focal tonic and clonic seizure, suddenly radiating from his left lower extremity to the left upper extremity. On examination, cranial nerves were intact, and mild muscular weakness was found on the left side. His deep tendon reflexes were hyperactive, muscles were hypertonic, and plantar reflex response was dorsiflexion on the left side. Body temperature, blood pressure, white blood cell count, and sedimentation were normal. No cell was observed in the analysis of cerebrospinal fluid (CSF), and the other biochemical findings of CSF were within the normal range. Microbiological examination of blood and CSF was normal in terms of bacteriological (tuberculosis, brucella, borrelia eg), viral (cytomegalovirus, herpes virus typeI-II, Ebstein-Barr virus, cytomegalovirus, human immunodeficiency virus eg), fungal (cysticercosis eg), and parasitic (toxoplasmosis, tenia solium) agents. Malignancy such as lymphomas or brain metastases was not detected. Conventional MR images showed a hypointense lesion (4 cm in diameter) with ring-like contrast enhancement on T1-weighted images in the right parietal region (Figure 1A, B). In addition, a smaller lesion than the first one was detected, which was similar feature to the first one. Perilesional edema was also observed. Diffusion-weighted images showed a bright cavity and a markedly decreased apparent diffusion coefficient (ADC) with perilesional edema. ADC map showed hypointensity, representing restricted diffusion in the corresponding region (Figure 2 A, B).

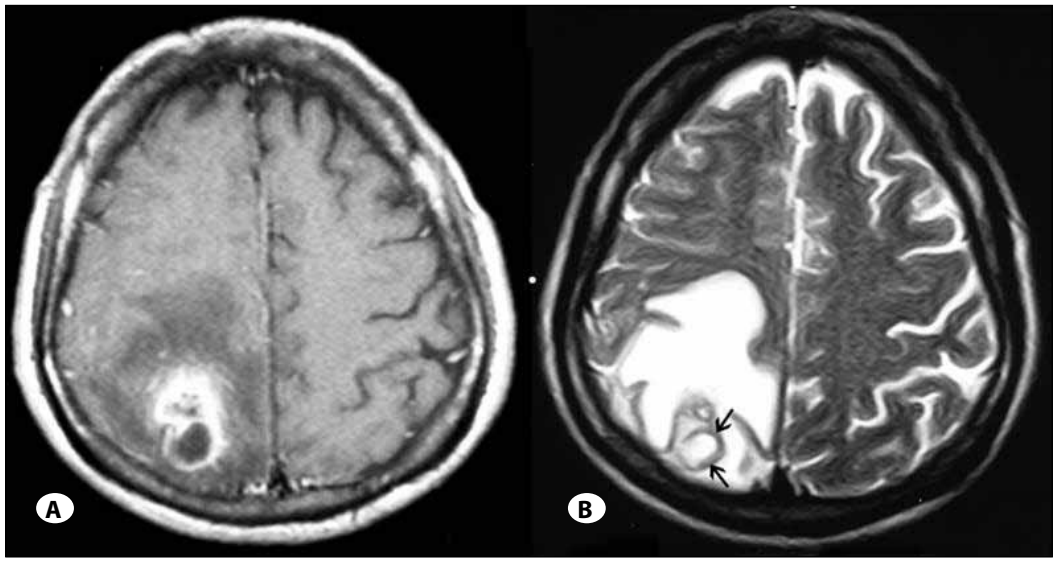

Figure 1: MR images show cerebral abscess in the patient with Behçet's disease.
A) Contrast-enhanced axial T1-weighted image shows ring contrast-enhancing lesion in the parietal region with perilesional hypointense edema,
B) Axial T2-weighted image shows hyperintesity in the centre of the lesion and surrounding oedema.

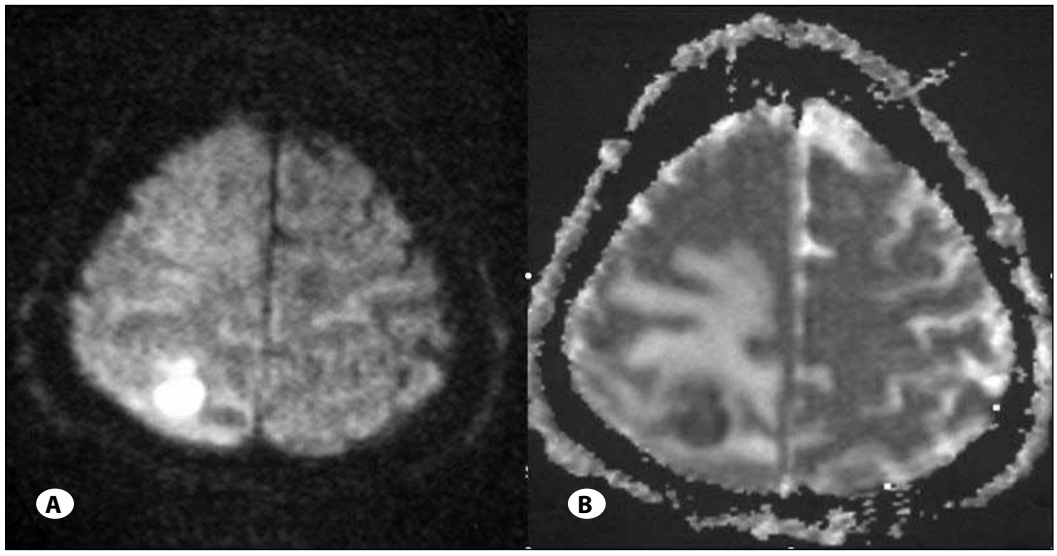

Figure 2: Diffusion-weighted MR images show cerebral abscess in the patient with Behçet's disease.
A) Diffusion-weighted image reveals increased signal in right parietal region surrounded by hypointensity that corresponds to edema,
B) ADC map reveals hypointensity, representing restricted diffusion in the centre of the lesion.

Ornidazole (1000mg/day) and ceftriaxone (2 gr/ day) were given to the patient diagnosed with intracerebral abscess, although no fever, stiff-neck, and infection symptoms were present. After a 26 to 42 days of antibiotic treatment, some improvement in his general condition and muscle strength were observed. No change in the size of intracerebral abscess was seen on subsequent cranial MRI. A lesion removed during the cranial operation was consistent with abscess macroscopically.

As a result of microbiological and histopathologic analysis, the lesions were consistent with sterile abscesses. The clinical state of the patient gradually improved. The patient had no further complications.

DISCUSSION

BD was first described by Hulusi Behçet in 1937. It is characterized by the presence of a diagnostic triad of oral and genital aphtous lesions, ulcers and relapsing uveitis (6).

CNS involvement is encountered at a rate between 5% and 30%, but cerebral abscess formation is rarely seen in the medical literature (5,6,8). CNS manifestations of BD are classified in two groups;

1- Parenchymal CNS involvement (Neuro–Behçet's Disease). These include hemispheric, brain stem, spinal and meningoencephalitic presentations.

2- Non-parenchymal CNS involvement (Neuro–vascular Behçet's Disease). Dural sinus thrombosis, arterial occlusion and arterial aneurysms (2).

Epileptic seizures and epilepsy are reported as manifestations of Neuro-Behçet's disease (NBD) at a rate of 2–5% in large series. Partial seizures are a presenting feature of NBD in one report. Generalized seizures are the predominant type. The relatively low prevalence of epilepsy in NBD is compatible with the low prevalence of cortical involvement seen on cranial MRI (1). MR imaging is more sensitive in revealing

these lesions than CT. Non-hemorrhagic lesions are the most typical, and they are usually hypointense on T1-weighted images and hyperintense on T2-weighted images. Brain abscesses show ring-shaped enhancement after contrast media administration and marked peri-lesional edema and exhibit restricted water diffusion on MR imaging. In addition, they are hyperintense on DWI and hypo-, iso-, or hyperintense on ADC map. Homogeneous, heterogeneous, linear, circular, and ring patterns have been described (4,7,8). Radiological imaging techniques provide early detection of neurological diseases, but they do not always provide an adequate and reliable diagnosis. With the help of stereotactic biopsy techniques, it is possible to access brain lesions safely and with high precision (3). In our patient, the findings on conventional MR imaging were similar to those reported in literature, and biopsy was essential in order to diagnose and to treat these lesions.

Although various CNS manifestations occur in BD, only four cerebral abscess cases concerning BD have been identified in the literature (6). Ho et al. noticed a cerebral abscess in a patient with BD, who suffered from generalized tonic clonic seizure and left-side-hemiparesis four years after the diagnosis, and reported that it was probably the result of brainstem encephalitis and meningoencephalitis (6). The neuropathology of parenchymal NBD in the acute phase involves meningoencephalitis with an intense inflammatory infiltration including polymorphs, eosinophils, lymphocytes, and macrophages, with areas of necrosis and apoptotic neuronal loss (1). Kaneko et al. showed the formation of cerebral abscess in the autopsy of a patient with BD. They reported that the formation of abscess had a close connection with softening, demyelization and cavitations. The infiltration of lymphocyte, histiocyte and leukocytes was found in the area surrounding the focus. They also reported that the strong and acute inflammation mainly contains lymphocytic and leukocytic infiltration in the perivascular area and causes necrosis in parenchyma (9).

In addition, there are various hypotheses regarding the reason of abscess in BD, including bacterial, viral, and fungal infections (8). In the case, microbiological examination of blood and CSF was normal in admission and on follow-up. No microbial agent had grown in the microbiologic examination of the removed material, and the histopathology was consistent with sterile abscess. Whether the reason for the formation of abscess is a microorganism that did not grow because of the antibiotics is questionable. However, as the history, clinical picture, and laboratory findings of an infection were absent, it is suggested that a softening, demyelization and cavity formation at the area of the lesions may cause cerebral abscess because of a vasculitic process and perivascular infiltration of leukocytes. A stereotactic biopsy may be performed to elucidate the etiopathogenesis.

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