Cerebral Abscesses in Behcet’s Disease: A Case Report

Behçet Hastalığında Serebral Abseler: Olgu Sunumu

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 type-I-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).
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 aphthous 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 leukocyte 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.

REFERENCES