Cytotoxic Edema in Neuro-Behcet’s Disease?

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Abstract

We report a case of a 52-year-old woman with Behcet’s disease who presented with dysarthria and right-sided hemiparesis. T2-weighted and diffusion-weighted images (DWI) showed a hyperintense lesion in the left pons with a relatively decreased apparent diffusion coefficient (ADC). Imaging showed almost complete resolution of the lesion after treatment with prednisolone. The atypical DWI and ADC findings in this case may reflect cytotoxic edema due to excitotoxic brain injury. This case thus illustrates the radiological diversity of neuro-Behcet’s lesions.

Key words: neuro-Behcet’s disease, magnetic resonance imaging, diffusion-weighted imaging, apparent diffusion coefficient, cytotoxic edema

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Introduction

Behcet’s disease (BD) is a systemic recurrent vasculitis of unknown etiology. BD was first described in 1937 by a Turkish dermatologist, Hulusi Behcet, and manifests as a classical triad of recurrent oral and genital ulcers and relapsing uveitis (1). Central nervous system (CNS) involvement occurs in 4-49% of cases and these neurological entities are referred to as neuro-Behcet’s disease (NBD) (2, 3). Several reports have described the diffusion-weighted imaging (DWI) of NBD along with the apparent diffusion coefficient (ADC) values of the lesions; most reports state that the lesions have increased diffusivity with a relatively increased ADC value (4, 5). Recently a case of NBD with unusual radiological findings leading to diagnostic difficulties was reported (6). Here, we describe the second case of NBD with unusual radiological findings.

Case Report

We report a case of a 52-year-old woman who was diagnosed with BD 18 years previously. At that time, she had presented with the history of recurrent oral and genital ulcers and had uveitis at presentation; thus, her symptoms fulfilled the criteria for BD as proposed by the International Study Group (7). There was no history of hypertension, diabetes mellitus, coronary artery disease, or stroke, and her family history was irrelevant. Prednisolone (5 mg/day) and colchicine (0.5 mg/day) were prescribed to prevent BD relapse, and she received regular follow-up examinations in another hospital. When she recently sought medical help, she presented with dysarthria and right-sided hemiparesis of subacute onset. She also complained of headache and mild fever. After examination, she was referred to our hospital. We found that her vital signs were stable, she was conscious and oriented, but she had dysarthria and her tongue was deviated towards the right. She also had some dysphagia. In an examination of motor function, the patient showed right-sided hemiparesis, with manual muscle strength testing giving a result of 3/5 in both the upper and lower limbs. The tendon reflexes were brisk on the right side, pathological reflex (Hoffmann reflex) were present on the right side, and there was no Babinski sign. Sensory examination revealed no abnormalities, and there was no sign of meningeal irritation. Blood cell counts, chemistry, and coagulation profiles were within normal limits, except for an increased erythrocyte sedimentation rate. At the hospital where the patient originally sought treatment, cerebrospinal fluid (CSF) examination revealed pleocytosis with 323 cells; however, a

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repeat examination in our hospital after 2 weeks of prednisolone revealed normal cell counts and CSF protein and IL-6 levels. The following were unremarkable: antithrombin III levels, serum immunoglobulin levels, rheumatoid factor, double-stranded DNA and antinuclear antibody tests, antiphospholipid antibody assay, C3 and C4 complement assay, and HLA-B51 assay.

The patient was examined on a 1.5T MR unit (Siemens Medical Systems) with echoplanar imaging capability. Her T1-weighted MRI (T1WI) showed a low intensity lesion on the left side of the pons with ring enhancement after contrast administration; the T2-weighted MRI (T2WI) showed a high intensity, well-defined, round lesion in the same region (Fig. A, arrow). DWI also showed hyperintensity in the corresponding area (Fig. B, arrow). The ADC map revealed a low signal intensity and a relatively low ADC value in the lesion area \((0.67 \times 10^{-3} \text{ mm}^2/\text{sec})\) compared with the ADC value in the contralateral lesion-free pons \((0.92 \times 10^{-3} \text{ mm}^2/\text{sec})\) (Fig. C, arrow).

Based on the clinical picture and the prior diagnosis of BD, NBD was diagnosed. The patient was started on prednisolone (50 mg/day) and observed clinically. After initiation of the treatment, the patient showed rapid improvement: the headache and fever declined, and there was marked progressive improvement in right-sided hemiparesis and dysarthria. The prednisolone was slowly tapered off at the rate of 5 mg every two weeks. Five months later, the follow-up MRI showed almost complete resolution of the previously hyperintense lesion in the T2WI and DWI, with a small focus of high signal intensity remaining (Fig. D, arrow). The patient recovered with no lasting neurological deficits.

Discussion

NBD may present as acute focal or multifocal CNS dysfunction with a relapsing and remitting course. Several reports have described the conventional MRI appearance of the brain in NBD (2, 3, 5, 8). The commonest parenchymal location of involvement is brainstem, followed by basal ganglia, cerebral hemispheres and spinal cord. During the acute/
subacute phase, the lesions show hyperintensity on T2WI and contrast enhancement in T1WI. These lesions tend to resolve in the chronic phase.

Most reports of NBD note that the DWIs show iso- or hyperintensity with relatively high ADC values, as these lesions are thought to be caused by vasogenic edema, probably due to vasculitis (4, 5, 9, 10). Therefore such lesions, with high intensity signals on DWI and increased ADC values, are usually reversible. Contrary to the previous reports, the DWI lesion in the present case showed hyperintensity with a relatively decreased ADC value. Following the treatment, the patient improved clinically and had no residual neurological deficits. Furthermore, the lesion also showed considerable resolution in the follow-up imaging, although a small area of hyperintensity was still seen in the T2 MRI performed 5 months post-treatment.

The MRI lesion in the left side of the pons raised the differential diagnosis of infarction. However, the T1WI after contrast administration on the second day of presentation revealed a ring-enhanced lesion, an unusual finding for an acute/subacute ischemic lesion. The well-defined round shape of the lesion, as seen using T2WI and DWI, was also somewhat unusual for an ischemic lesion in the pontine base. These features seemed to exclude the possibility of acute infarction.

Heo et al reported a case of NBD with imaging findings resembling ours; in that case, stereotactic biopsy was performed with initial consideration being brain tumor, which was possible due to the cortical location of the one of the lesions (6). In our case as well, the ring-enhanced lesion with high DWI and low ADC value prompted the differential diagnosis of brain abscess or a mass lesion. But the typical location of the lesion in the pons, the absence of any systemic sign symptoms of infection, the marked clinical improvement of the symptoms with corticosteroid therapy, and the patient’s background of BD helped us make a prompt diagnosis and begin appropriate treatment.

High signal intensities on DWI and decreased ADC values, observed in patients with cerebral infarction, are usually interpreted as cytotoxic edema. These lesions are usually found to be irreversible, except for a few cases (11). It has been suggested that excitotoxic brain injury is one of the mechanisms of this type of DWI abnormality, which occurs due to increased extracellular glutamate in different neuropathological conditions and causes cytotoxic edema (12). Cytotoxic edema due to excitotoxic injury with less energy failure caused by excessive release of glutamate is associated with conditions such as seizures, viral encephalitis, multiple sclerosis, and toxic metabolic disease and occasionally is resolved on follow-up MRI. The high intensity lesions in DWI with decreased ADC values, which are reversible, have also been observed in some of these conditions (12-15). The DWI and ADC characteristics of cases like ours may reflect an excitotoxic brain injury mechanism that can lead to cytotoxic edema in NBD lesions. The neuropathological findings during the chronic stage of NBD consist of mainly longstanding inflammatory changes and gliosis (5). In the case presented here, the remaining small focus of high signal intensity seen in the T2WI after five months of treatment can be attributed to the presumed gliosis.

This case highlights the unusual neuroradiological features that can occur in NBD. These features should be kept in mind, especially when the location of the lesion precludes easy intervention, so that diagnosis and treatment can be performed in a timely manner.

References


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