Role of diffusion-weighted MRI in Neuro-Behçet’s disease

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ABSTRACT

We report the findings on serial diffusion-weighted MRI in a 29-year-old male with neuro-Behçet’s disease. Initial T2-weighted and fluid-attenuated inversion recovery images showed a hyperintense lesion in the brain stem. The lesion showed slight hyperintensity on diffusion-weighted images with no evidence of diffusion restriction on apparent diffusion coefficient maps. A follow up study after 7 months showed complete resolution of the brain stem lesion. Our findings indicate that diffusion-weighted imaging is a useful tool to differentiate acute exacerbation of neuro-Behçet’s disease from acute infarction, and therefore it helps in selecting the appropriate therapy.

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Behçet’s disease (BD) is a multi-system inflammatory disorder of unknown origin. Involvement of the central nervous system (CNS) occurs in up to 49% of cases of BD. The most commonly affected sites in the CNS are the mesodiencephalic junction, the pontobulbar region, the hypothalamic-thalamic region and the basal ganglia. Vasculitis and hypercoagulability are characteristics of BD. Diffusion-weighted imaging (DWI) is a newly developed technique that has proved to be useful in discrimination between the cytotoxic edema that is seen in infarction and the vasogenic edema that is associated with neuro-Behçet’s lesions. The aim of this report is to demonstrate the importance of DWI in distinguishing an acute exacerbation of neuro-Behçet’s disease from an acute infarction.

Case Report. A 29-year-old man was diagnosed 11 years ago to have Behçet’s disease on the basis of recurrent oral and genital ulcers, arthralgia, eye involvement and a positive pathergy test. Recently, he presented with double vision, slurred speech, difficulty in swallowing, and weakness in the left side of the body for a period of a few days. He had left sixth and seventh cranial nerves palsy, dysarthria, and left sided-weakness. There were no signs of meningeal irritation or skin lesions. The full blood count, the coagulation profile, and the cerebrospinal fluid analysis were normal. The initial T2-weighted images (T2WI) revealed a hyperintense lesion in the brain stem, especially within the pons, involving the right thalamus. The lesion showed slight hyperintensity on DWI and apparent diffusion coefficient (ADC) maps (Figure 1). He was started on systemic steroids and showed significant improvement over time with resolution of the neurological manifestations except for a minimal residual left-sided weakness. Seven months later, T2WI showed complete resolution of the previously noted abnormality in the brain stem (Figure 2).
Discussion. Behçet’s disease, named after Turkish dermatologist Hulusi Behçet in 1937, is a systemic vasculitis of unknown etiology. The onset is typical in young adults with recurrent oral and genital ulceration, uveitis, skin manifestations, arthritis, neurological involvement, and a tendency for thrombosis. The disease has a worldwide distribution but is prevalent in Japan, the Middle East, and some Mediterranean countries. The etiological factors remain obscure, but viral agents, immunological factors, genetic causes, bacterial factors and fibrinolytic defects have been implicated. Currently, the most widely used diagnostic criteria of BD is the International Study Group’s classification, which requires recurrent oral ulcerations plus 2 of the following in order to establish a definite diagnosis: recurrent genital ulcerations, skin or eye lesions, or a positive pathergy test.

Neurological involvement has been reported in 4-49% of patients, however the neurological manifestations of BD can be the presenting symptoms in approximately 5% of cases. The neurological presentation and clinical course of BD are variable. Some patients have an acute presentation followed by a relapsing remitting, or secondary progressive course. Others may have an insidious onset with a primary progressive CNS dysfunction. A small group has no neurological complaints but shows abnormal neuroimaging findings. This group is said to have “silent neurological involvement.” Two main patterns of CNS involvement occur: parenchymal and non-parenchymal. The neuropathology of the parenchymal form is that of multifocal necrotizing lesions with marked inflammatory cell reactions within the vascular wall and with perivascular mononuclear
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cell infiltration, which is consistent with vasculitis involving both arterial and venous systems. 4,5

There have been several reports describing the distribution and appearance of CNS lesions in BD on conventional MRI. The common parenchymal locations are the mesodiencephalic junction, the pontobulbar region, the hypothalamic-thalamic region and the basal ganglia. 4 During the acute/subacute phase, the lesions show hyperintensity on T2WI. These lesions tend to resolve or decrease in size in the chronic phase. 4 Non-parenchymal lesions include cerebral venous thrombosis as well as the extremely rare finding of enhanced thick meninges. 8

There are 2 types of brain edema that are indistinguishable by the conventional MRI while distinction is possible with application of DWI. The cytotoxic edema is characterized by abnormal uptake of water by the cellular elements of the brain resulting in cell swelling, while the vasogenic edema is caused by increased permeability of the blood-brain barrier with fluid accumulation outside the cells in the extracellular spaces. The cytotoxic edema tends to have decreased ADC due to restriction of water motion within the swollen cells and therefore demonstrates remarkable hyperintensity on DWI. The vasogenic edema on the other hand tends to have increased ADC due to free water motion in the extracellular space and therefore shows either no increase in signal intensity on DWI or a mild increase in signal intensity due to the T2-shine through effect. This shows the role of DWI in evaluation of neuro-Behçet’s disease since discrimination between a cytotoxic edema caused by acute infarction and a vasogenic edema seen in acute exacerbation of BD has important clinical implications because these 2 conditions are managed differently. 6,9

Only 3 previous reports in the English literature have described DWI findings in neuro-BD. 1,5,6 Our case showed a finding that is consistent and in accordance with and supportive to the previously documented radiological features of acute neuro-BD.

In conclusion, DWI plays a very important role in the management of patients with BD because it can distinguish between infarction and acute exacerbation of BD.

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References