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Diffusion-weighted MR imaging of neuro-Behçet's disease: a case report

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Abstract We present a serial study of diffusion-weighted imaging (DWI) in a patient with neuro-Behçet's disease. Initial T2-weighted magnetic resonance images showed a hyperintense lesion in the brain stem. The lesion was slightly hyperintense on DWI and the apparent diffusion coefficient (ADC) was slightly increased. Ten months later, DWI showed an improvement in the abnormal signal intensity and the region of increased ADC had increased in size, especially on the left side. DWI is useful for differentiating an acute exacerbation of neuro-Behçet's disease from acute infarction.

Keywords Behçet's disease · Brain Vasculitis · Magnetic resonance imaging · Diffusion weighted imaging · Apparent diffusion coefficient

Introduction

Behçet's disease (BD) is a systemic vasculitis of unknown origin with neurological involvement in between 4% and 49% of patients [1, 2, 3]. It was first described in 1937 and consists of a triad of recurrent aphthous ulcers of the oral and genital mucosa with relapsing uveitis [1, 2, 3, 4, 5, 6, 7]. The common neural parenchymal locations are the mesodiencephalic junction, pontobulbar region and hypothalamic-thalamic region [1, 2]. There are few reports on diffusion-weighted imaging (DWI) of neuro-BD [7, 8]. We report a patient with neuro-BD who underwent serial magnetic resonance imaging (MRI) with DWI.

Case report

A 24-year-old man was admitted to the emergency department with slurred speech and right arm and leg weakness that had gradually

increased over a 2 month period. The patient also had ulceration of the tongue and uveitis of the right eye, but no genital ulceration. Initial T2-weighted MR images showed a hyperintense lesion in the midbrain extending into the bilateral temporal lobes with prominent enlargement of the left cerebral peduncle (Fig. 1). DWI showed mild hyperintensity in the left cerebral peduncle. The apparent diffusion coefficient (ADC) was bilaterally increased slightly in the cerebral peduncle ($0.91 \times 10^{-3} \text{ mm}^2/\text{s}$ in the right and $0.98 \times 10^{-3} \text{ mm}^2/\text{s}$ in the left cerebral peduncle) compared with the normal-appearing white matter in the frontal lobes ($0.62\text{--}0.86 \times 10^{-3} \text{ mm}^2/\text{s}$). BD was suspected and the symptoms disappeared after steroid treatment.

Ten months later, the patient was admitted again complaining of severe right arm weakness and lower facial droop with slurred speech. At this time, the patient had genital and oral ulcers with uveitis; the diagnosis of BD was made clinically according to the criteria of the International Study Group for Behçet's Disease [6]. T2-weighted MR images showed a hyperintense lesion in the left side of the midbrain extending into the left temporal lobe and enlargement of the left cerebral peduncle, which was more prominent compared with the previous examination (Fig. 2). The left red nucleus was spared and compressed medially. Fluid-attenuated inversion recovery (FLAIR) images showed a hyperintense lesion

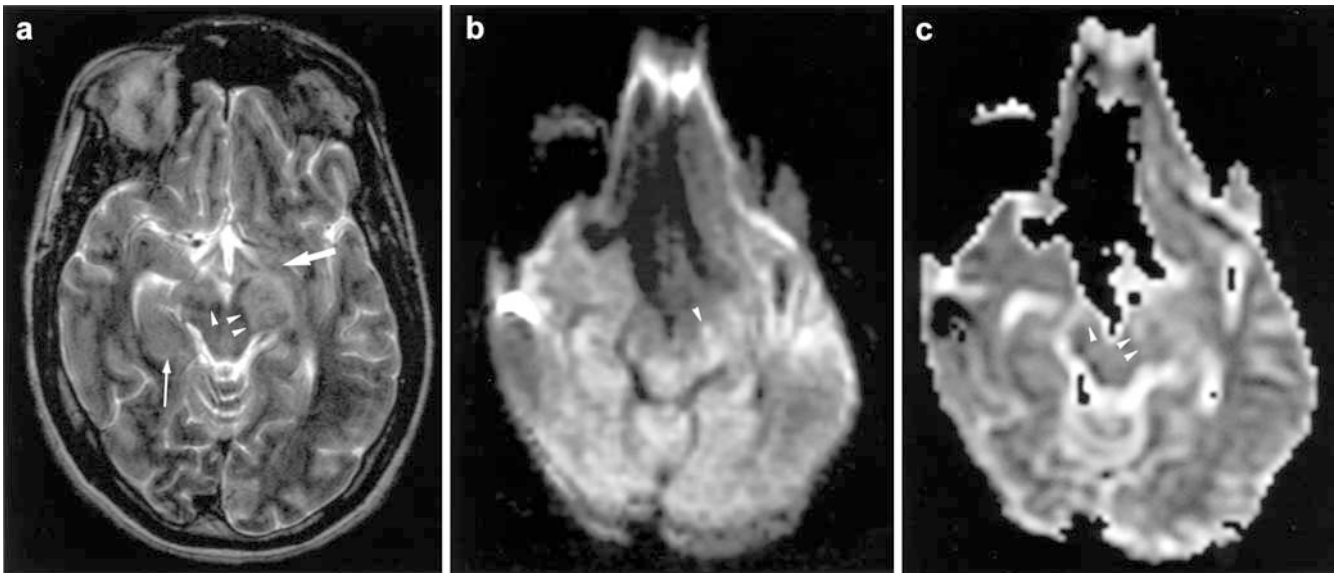


Fig. 1a–c The initial presentation. **a** Initial T2-weighted fast spin echo magnetic resonance (MR) image (TR/TE/NEX, 5400/105/2) shows a hyperintense lesion in the midbrain (*arrowheads*) extending bilaterally into the temporal lobes (*arrows*) with prominent enlargement of the left cerebral peduncle. **b** On the diffusion-weighted image (TR/TE/NEX, 10000/125/1; $b=1000$), the left cerebral peduncle (*arrowhead*) is slightly hyperintense. **c** The apparent diffusion coefficient (ADC) values of the lesions (*arrowheads*) are slightly increased ($0.91 \times 10^{-3} \text{ mm}^2/\text{s}$ in the right and $0.98 \times 10^{-3} \text{ mm}^2/\text{s}$ in the left cerebral peduncle), suggesting vasogenic edema

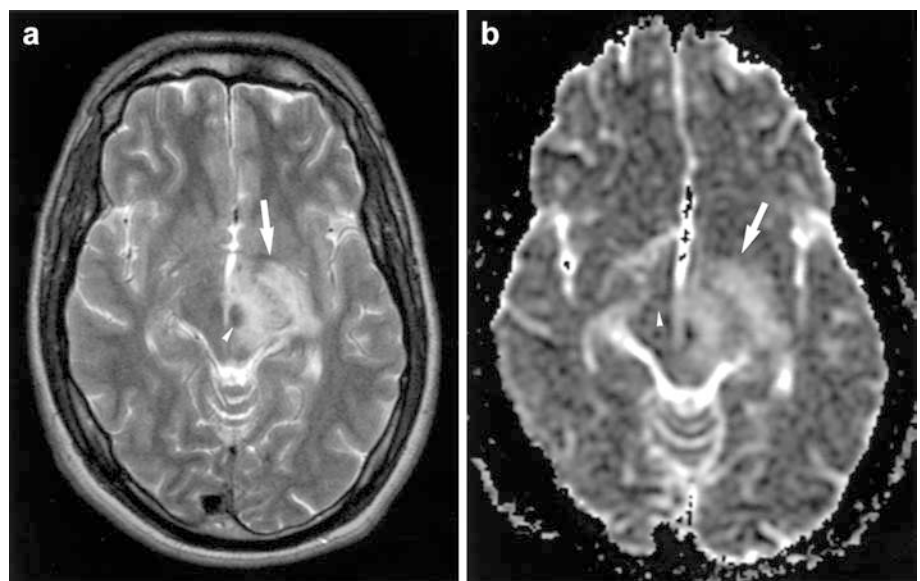
cerebral peduncle was slightly hyperintense on DWI, the signal intensity was decreased compared with the previous study. The region of increased ADC in the left side of the midbrain and temporal lobe had increased in size and was more prominent ($1.30 \times 10^{-3} \text{ mm}^2/\text{s}$) than in the previous study, while the ADC of the right cerebral peduncle was reduced ($0.82 \times 10^{-3} \text{ mm}^2/\text{s}$). Following administration of 0.1 mmol/kg of gadolinium diethylenetriaminepentaacetic acid, heterogeneous, intense enhancement was noted predominantly in the left side of the midbrain and thalamus. The patient was again treated with steroids and his symptoms subsequently resolved.

in the midbrain extending into the left temporal lobe and optic tract. The left side of the pons, the left thalamus and posterior limb of the left internal capsule were also involved. The lesion seen previously in the right cerebral peduncle and the right temporal lobe was no longer visualized. Although the lesion of the left

Discussion

BD is a systemic inflammatory disorder of unknown etiology. It was first described in 1937 and consists of a

Fig. 2a, b Ten months after the initial presentation. **a** T2-weighted fast spin echo MR image (TR/TE/NEX, 5400/98/2) shows a hyperintense lesion in the left midbrain extending into the left temporal lobe with prominent enlargement of the left cerebral peduncle (*arrow*). The hyperintense lesion seen initially in the right temporal lobe is not visualized. The left red nucleus is compressed medially but spared (*arrowhead*). **b** The region of increased ADC in the left side of the midbrain and left temporal lobe (*arrow*) is increased in size and is more prominent ($1.30 \times 10^{-3} \text{ mm}^2/\text{s}$), while the ADC of the right cerebral peduncle (*arrowhead*) is reduced ($0.82 \times 10^{-3} \text{ mm}^2/\text{s}$)



triad of recurrent aphthous ulcers of the oral and genital mucosa with relapsing uveitis [1, 2, 3, 4, 5, 6, 7]. Additional involvement has been reported including central nervous system, skin, kidney, lung, deep vein thrombosis and arterial occlusion [2, 5, 6]. The etiological factors remain obscure, but viral agents, immunological factors, genetic causes, bacterial factors and fibrinolytic defects have been implicated [1].

Neuro-BD causes inflammatory changes in the meninges and brain parenchyma with perivascular lymphocytic infiltrates involving veins, venules, capillaries and, less frequently, arteries. As the lesions become more chronic, gliosis, atrophy, and thickening and fibrosis of the meninges may ensue [1].

There have been several reports describing the conventional MRI appearances of the brain in BD [1, 2, 3, 7, 8]. The common parenchymal location is the mesodiencephalic junction, pontobulbar region, the hypothalamic-thalamic region, basal ganglia and telencephalon [1, 2]. During an acute/subacute phase, the lesions show hyperintensity on T2-weighted images and contrast enhancement on T1-weighted images. These lesions tend to resolve or decrease in size in the chronic phase [1, 2, 7, 8].

DWI is a relatively new technique and has proved useful in the detection of acute ischemic changes [7, 8, 9, 10, 11, 12]. Moreover, DWI can discriminate the two main types of brain edema: (1) cytotoxic edema, characterized by abnormal uptake of water by the cellular elements of the brain; and (2) vasogenic edema, caused by increased permeability of the blood-brain barrier [9, 10, 11, 12]. The lesion of cytotoxic edema tends to have decreased ADC and shows hyperintensity on DWI, while that of vasogenic edema tends to have increased ADC and shows isointensity to slight hyperintensity on DWI. Conventional MRI can not always distinguish between these types of edema [9, 10].

The DWI findings of neuro-BD have been mentioned in a few reports. Ohta et al. [7] reported two cases of acute/subacute BD and showed T2 hyperintense lesions in the brain stem. DWI showed an isointense lesion in one case and a slightly hyperintense lesion in the other. These signal abnormalities improved in about 1 month. Kang et al. [8] reported a patient who had T2 hyperintense lesions with slight hyperintensity on DWI and increased ADC. These signal abnormalities had resolved 1 year later. Our patient is the first case to show the recurrent aspect of neuro-BD on DWI, indicating that the lesion of neuro-BD seen on MRI is caused by vasogenic edema, probably due to vasculitis [13]. Moreover, DWI including ADC maps is important clinically to diagnose acute neuro-BD, because the lesion of acute neuro-BD should be treated by steroid, while the treatment of cytotoxic edema due to acute infarction may include thrombolysis or anticoagulation [7].

The differential diagnosis includes infarction, all other types of vasculitis such as systematic lupus erythematosus, multiple sclerosis, sarcoidosis and meningitis [1, 14, 15]. Ischemic lesions tend to show hyperintensity on T2-weighted images, but it can be difficult to discriminate cytotoxic edema from vasogenic edema on conventional MRI alone without chronological change [9, 10]. The MRI findings in neuro-BD are similar to those in systemic lupus erythematosus, but involvement of the brain stem is rare in the latter [1, 14]. The typical periventricular ovoid lesions in multiple sclerosis and meningeal lesions in sarcoidosis are not usual findings of BD [1, 15].

In conclusion, neuro-BD tends to show hyperintense lesions mainly in the brain stem due to vasogenic edema on T2-weighted images and DWI. DWI seems to be useful for distinguishing an acute exacerbation of BD from acute infarction.

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