Central Nervous System Involvement in Behçet's Syndrome

MONICA BALLESTA MORATALLA, Hospital Universitario La Fe, Avenida Campanar 21, Valencia, 46009 Spain. E-mail: monica_ballesta@hotmail.com

A 45-year-old man presented at our hospital with fever, left hemiparesia, dysarthria, drowsiness, facial paresia, and fever. His history was nonspecific. No meningeal signs were found. Physical examination revealed aphthous ulcers (tongue and inside of the cheek) and genital sores. Computed tomography showed no abnormality. Magnetic resonance imaging (MRI) showed hyperintense lesions on T2-weighted sequences, 2 in the pons, one in the left cerebral peduncule, and one in the occipital deep white matter (Figures 1 and 2), with vasogenic edema. Diffusion MRI revealed high signal intensity changes at the lesion site (Figure 3). Gadolinium-enhanced T1-weighted MRI showed a ring-enhancement of the lesions (Figure 4). He was treated with intravenous corticosteroid (methylprednisolone 1 g/day) and immunosuppression treatment (azathioprine). He achieved stable condition, without fever or neurological signs. Followup MR study showed stability of the lesions, with minor contrast enhancement.

Behçet's disease is a multisystem inflammatory chronic disorder, common in the eastern areas of the Mediterranean, the Middle East, and Japan[1]. It is more prevalent among men, and onset is usually in the third decade of life. It is an autoimmune disease[1] that results from damage to blood vessels. The most common symptoms are mouth and genital sores, inflammation of the eye, and skin problems. Central nervous system involvement is one of the most dangerous manifestations (5% to 10% of all patients)[2]. There are 2 patterns of involvement, parenchymal (in which the brain stem is the most common location) and nonparenchymal (associated with dural sinus thrombosis)[2,3]. There is no cure for Behçet's disease and the goal of treatment is to reduce discomfort and prevent serious complications.

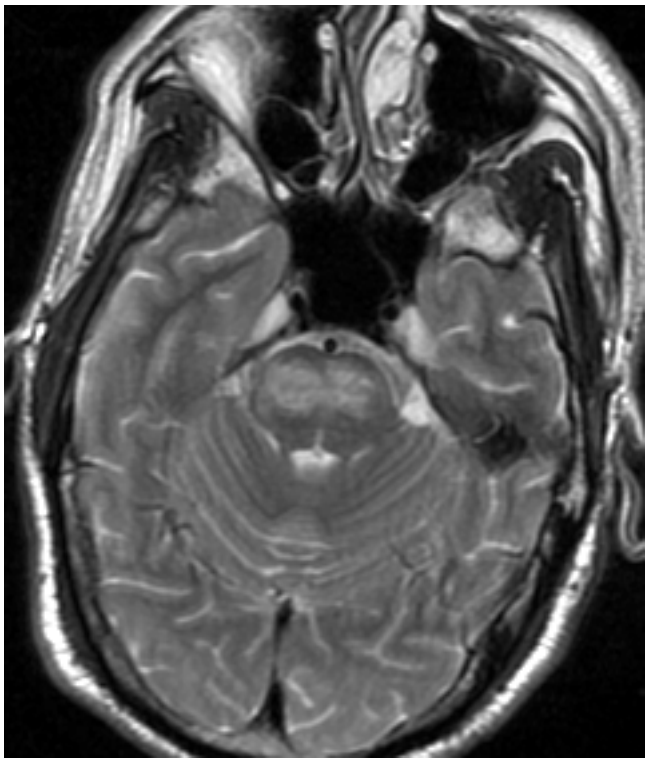

Figure 1. Axial T2 weighted MR image shows inhomogeneous high signal intensity abnormalities throughout the pons.

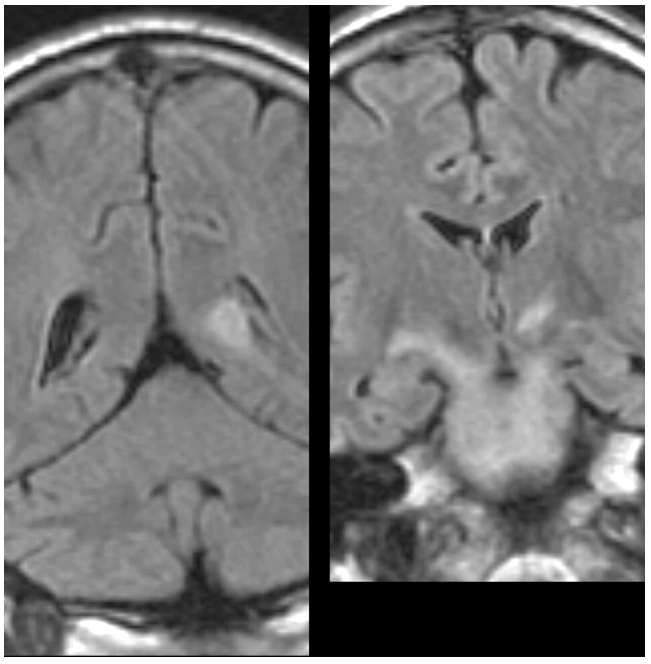

Figure 2. Coronal fluid-attenuated inversion-recovery MR sequence reveals hyperintense lesions in the pons and the left cerebral peduncule and within the deep left occipital white matter.

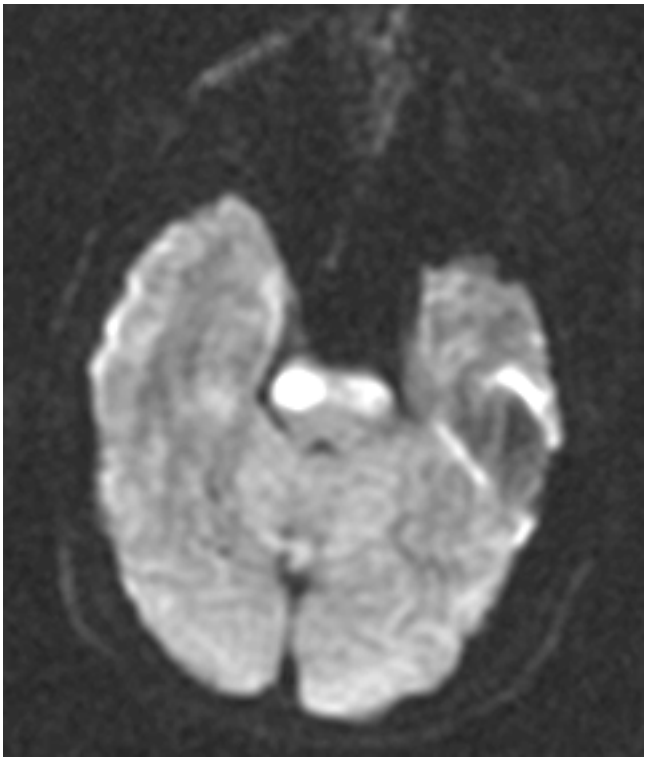

Figure 3. Diffusion-weighted image reveals high signal intensity in the pons compared with the temporal white matter, consistent with the presence of increased diffusion.

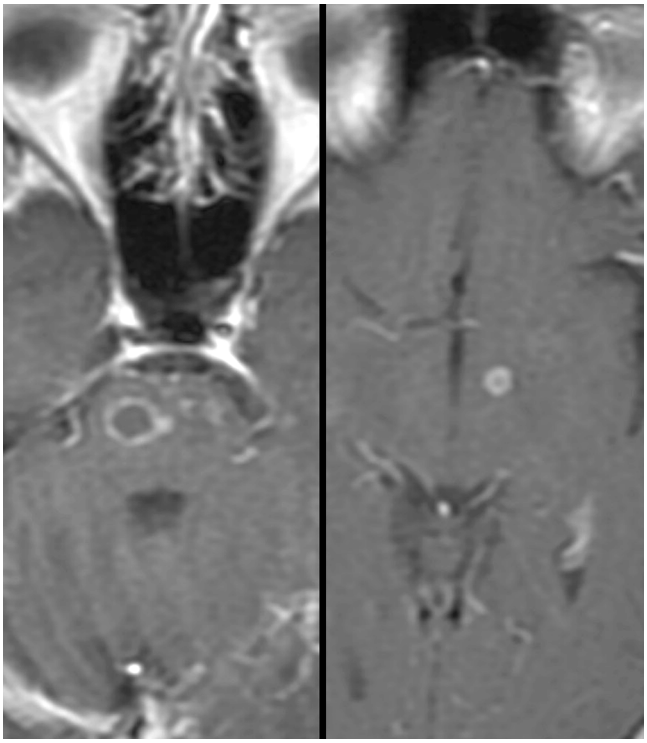

Figure 4. Contrast enhanced axial T1-weighted MR images show marked ring enhancement of the lesion located in the pons and in the left cerebral peduncule.

REFERENCES

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