Extending brainstem and capsule-thalamic lesions in a patient with parenchymal neuro-Behçet disease

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ABSTRACT

A 44-year-old female patient with a history of recurrent oral and genital ulcers presented with rapid gait disturbance, headaches, dysphonia, dysphagia, and diplopia evolving for 3 weeks. On examination, spastic tetraparesis, cerebellar ataxia, and nerves palsies were noted. Pseudofolliculitis of the lower limbs and active buccal ulcers were identified. Cerebral MRI demonstrated T2-Flair hyperintense capsulothalamic lesion with midbrain and latero-protuberantial extension. The international criteria for the diagnosis of definite Neuro-Behçet’s disease (NBD) were met in our patient. Evolution under cyclophosphamide and intravenous methylprednisolone was favorable. The radiological findings in NBD are broad and challenging. Clinicians should be particularly suspicious of NBD in brain MRI with extensive lesions, involving the brainstem.

KEYWORDS
Behçet syndrome, neuro-Behçet Disease, magnetic resonance imaging, diagnostic

A 44-year-old female patient with a four-year history of recurrent oral (more than three recurrences per year) and genital ulcers presented with rapid gait disturbance, headaches, dysphonia, dysphagia, and diplopia evolving for 3 weeks.

On examination, spastic tetraparesis, cerebellar ataxia, and left abducens nerve and vagus nerves palsies were noted. Pseudofolliculitis of the lower limbs and active buccal ulcers were identified. C-reactive protein level was elevated (176 mg l⁻¹). Cerebrospinal fluid examination showed mild lymphocytic pleocytosis (24 cells mm⁻³) with normal protein and glucose levels. Cerebral MRI demonstrated T2-FLAIR hyperintense capsulo-thalamic lesion with midbrain and latero-protuberantial extension. The international criteria for Behçet’s disease [1] and for the diagnosis of definite Neuro-Behçet’s disease (NBD) [2] were met in our patient. Evolution under cyclophosphamide and intravenous methylprednisolone was favorable. An improvement was observed in the patient’s swallowing and gait disturbances.

The radiological findings in NBD is broad and challenging [3]. Although the MRI findings in the present case were typical of NBD, they were also suggestive of tumor, particularly lymphoma. However, the absence of neoplastic cells and the long follow-up period argued against this hypothesis.

Clinicians should therefore be particularly suspicious of NBD in brain MRI with extensive lesions, involving the midbrain and pons [3]. Typically, lesions are hyperintense on T2 sequences and hypo or isointense on T1 sequences. This is particularly suggestive of the chronic phase of parenchymal NBD [4], the period during which brainstem atrophy is remarkable [4] and the area of enhancement is restricted [3], as shown in the present case.
Fig. 1. Axial T2-FLAIR brain MRI images demonstrating bilateral capsule-thalamic lesions (A), extending to the mesencephalic regions, mainly on the left (B), and to cerebellar peduncles (C). T1 weighted post-contrast sequence (D) revealing a heterogeneous left enhancement in the left mesencephalic region. T2-weighted sagittal (E) and coronal sections (F) demonstrating the extent of T2-weighted signal abnormality along the majority of the brainstem. The brain MRI was obtained 5 years from the disease onset.
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