IMAGEM EM NEUROLOGIA/IMAGE IN NEUROLOGY

Neuro-Behçet's Disease Mimicking Adult-Type Diffuse Glioma

Doença de Neuro-Behçet a mimetizar Glioma Difuso Tipo-Adulto

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A 40-year-old woman was admitted with bilateral tonic seizure and a 1-month history of difficulties planning and executing tasks. Past medical and family history were unremarkable. On admission, left palmar grasp reflex and left-sided mild hemiparesis were found. General physical examination, including the skin and mucosa, was normal. Routine electroencephalogram revealed right frontal paroxysmal activity. Bilateral, diffuse and asymmetrical, temporoparietal periventricular white-matter lesions, with right anterior frontal cortico-subcortical extension, were detected as hyperintensities at T2-FLAIR and hypointensities at T1-weighted brain magnetic resonance imaging (MRI) (**Figs. 1A, B**). There was no lesion enhancement and no abnormalities at diffusion-weighted imaging. MRI spectroscopy showed decreased N-acetylaspartate levels, increased coline and presence of lactates, considered suggestive of glioma. Cerebrospinal fluid had mild monocytic pleocytosis of 7/mm^3, normal protein levels and negative oligoclonal bands. A stereotactic-guided biopsy of right frontal lesion was performed, and histopathological examination was suggestive of adult-type diffuse glioma.

Temozolomide and dexamethasone were started, with clinical and radiological response. Three years later, she presented with subacute drowsiness, right internuclear ophthalmoplegia and gait ataxia. There was no evidence of disease progression on computed tomography

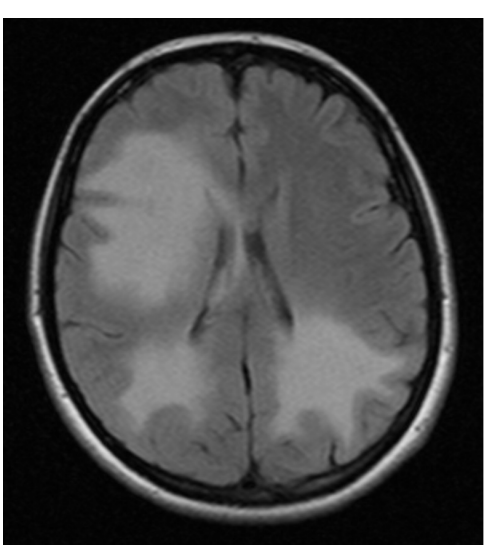

Figure 1A. Brain MRI showed bilateral, diffuse temporoparietal white-matter lesions, with right frontal cortico-subcortical extension, detected as hyperintensities at T2-FLAIR images.

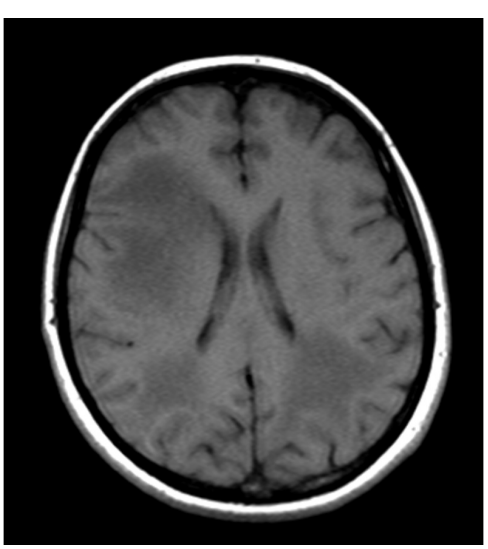

Figure 1B. Brain MRI showed bilateral, diffuse temporoparietal white-matter lesions, with right frontal cortico-subcortical extension, detected as hypointensities at T1-weighted images.

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(CT). Dexamethasone 10 mg id was restarted, with resolution of symptoms. Three months later, there was a new relapse characterized by left hemi-hypoesthesia and gait imbalance. Brain MRI showed a tumefactive hyperintense T2 lesion at right thalamus extending to the superior aspect of midbrain and ipsilateral cerebral peduncle (**Fig. 2A**). Given the patient's clinical atypical course for an infiltrative glioma, the previous neuropathological examination was reviewed and deeper sections were performed revealing perivascular and intraparenchymal mononuclear inflammatory cell infiltrate. This finding together with the presence of reactive astrocytes favored the hypothesis of an inflammatory disease. Two weeks later, the patient developed recurrent oral and genital mucocutaneus ulcerations, resulting in a diagnosis of neuro-Behçet's disease (NBD). She was treated with IV rituximab 1000 mg every 6 months with total resolution of the mesodiencephalic lesion (**Fig. 2B**) and complete clinical remission after a three-year follow-up period.

This patient fulfills the International Criteria for Behçet's disease1 and the consensus for NBD.[2] The remarkable diagnostic delay can be attributed to several atypical features. Firstly, epileptic seizures and cognitive symptoms are rare manifestations of NBD, compatible with the low prevalence of cortical involvement observed in brain MRI.[2] Secondly, the absence of systemic symptoms in the first three years of disease further complicated the diagnosis. Finally, the uncommon pseudotumoral MRI presentation with a cortico-subcortical distribution of lesions and lack of enhancement, and the MR spectroscopy considered suggestive of glioma. Typical parenchymal NBD is characterized by hyperintense lesions on T2-weighted imaging, which are enhancing and have a predilection for the upper brainstem and basal ganglia.[3]

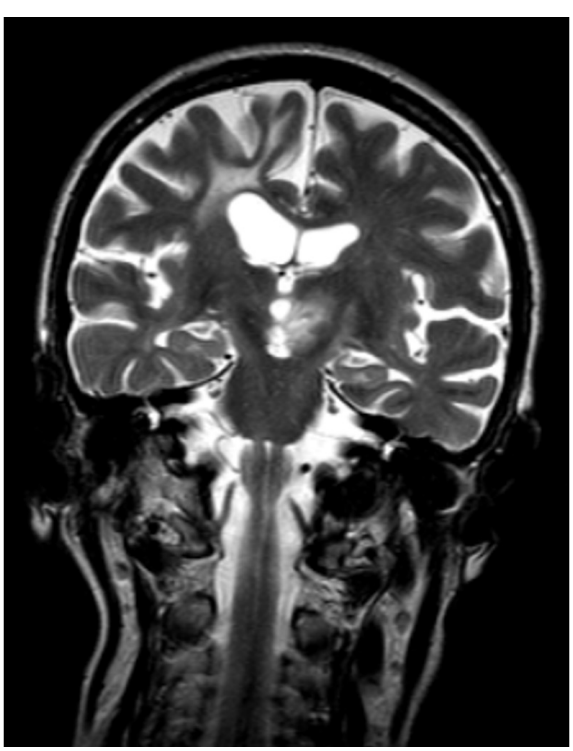

Figure 2A. T2 brain MRI showed a new tumefactive lesion at right thalamus extending to midbrain.

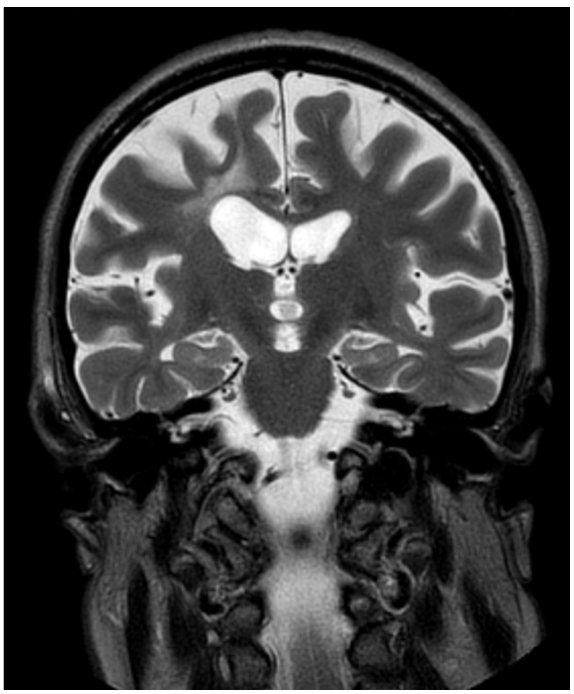

Figure 2B. Treatment with rituximab 1000mg 6/6 months was started with resolution of the mesodiencephalic lesion (T2-weighted images).

To the best of our knowledge, this is the second published case describing NBD mimicking the pattern of diffuse glioma.[4] It also highlights the potential significant clinical and radiological response to rituximab, in refractory NBD.

Few cases describing neurologic pseudotumoral presentation in NBD have been reported.[2] NBD should be considered in the differential diagnosis of brain lesions with neoplastic features, even after performing a brain biopsy. ■

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OR and SB: Conception, critical review and final approval.

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