CLIPPERS: a recognizable image of a treatable disease

CLIPPERS: uma imagem reconhecível de uma doença tratável

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An 80-year-old man with a history of stroke presented with eyelid ptosis of three years' duration. Magnetic resonance imaging (MRI) of the brain revealed findings (Figure 1) consistent with chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (CLIPPERS). As limitations precluded anatomopathological examination and the imaging pattern was characteristic enough to support treatment without biopsy, the decision was made to prescribe long-term corticosteroids. At the two-year follow-up, MRI findings had regressed (Figure 2) and the patient had improved clinically, meeting criteria for treatment response¹. Biopsy is indicated when symptoms and MRI findings are unresponsive to treatment². The differential diagnosis should include CNS lymphoma, neurosyphilis, and vasculitides³⁴.



Figure 1. Contrast-enhanced brain MRI showing brainstem hyperintensity (white arrows) and disseminated perivascular areas of nodular enhancement, consistent with idiopathic inflammatory rhombencephalitis. There is evidence of cerebral and cerebellar volume reduction (Figure B).



Figure 2. Contrast-enhanced brain MRI at the two-year follow-up. There is significant reduction of abnormal hyperintensity and contrast uptake in the brainstem (arrowhead) compared with the previous scan.

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References

- Taieb G, Wacongne A, Renard D, Figarella-Branger D, Castelnovo G, Labauge P. A new case of chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids with initial normal magnetic resonance imaging. Brain; 2011.134(pt 8):182-183. https://doi.org/10.1093/brain/awq390
- Taieb G, Allou T, Labauge P. Therapeutic approaches in CLIPPERS. Curr Treat Options Neurol. 2017 May;19(5):17. https://doi.org/10.1007/s11940-017-0455-4
- 3. Taieb G, Duflos C, Renard D, Audoin B, Kaphan E, Pelletier J, et al. Long-term outcomes of CLIPPERS (chronic lymphocytic

inflammation with pontine perivascular enhancement responsive to steroids) in a consecutive series of 12 patients. Arch Neurol. 2012 Jul;69(7):847-55. https://doi.org/10.1001/archneurol.2012.122

 Dudesek A, Rimmele F, Tesar S, Kolbaske S, Rommer PS, Benecke R, et al. CLIPPERS: chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids: Review of an increasingly recognized entity within the spectrum of inflammatory central nervous system disorders. Clin Exp Immunol. 2014 Mar;175(3):385-96. https://doi.org/10.1111/cei.12204