Central nervous system involvement in a case of Whipple disease

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Introduction

Whipple disease (WD) is a rare infectious disease caused by the gram-positive bacteria *Tropheryma whipplei*. In its typical form, it is characterized by low-grade fever, polyarthralgia and chronic diarrhea, but it may also present as a complex multisystemic disorder. In particular, between 20 and 40% of patients may develop neurological manifestations in the course of the disease.

Presentation and history

A 46-year-old man came to our observation for the subacute onset of agitation, walking difficulties and involuntary movements. He had previously been discharged from a gastroenterology department, where he had been admitted for diarrhea and weight loss. In this setting, a duodenal biopsy had been performed, showing evidence of WD. He was admitted in our Unit for further evaluation.

General and Neurological Examination

At the time of admission, his gait was ataxic; eye movement examination showed bilateral blepharoptosis, vertical supranuclear ophthalmoplegia, severe horizontal ophtalmoparesis, and rotatory nystagmus. He was dysarthric and hypophonic. His muscles were hypotonic and hypotrophic; his voluntary movements were dysmetric. **A peculiar pattern of involuntary movements could be observed: repetitive, jerky lateralized contractions of the mimic and masticatory muscles, that spread to the ipsilateral platysma.**

His cognition was also affected; he was partially oriented, easily distractable and showed deficits in his memory and executive abilities.

Magnetic resonance imaging of the brain was unremarkable, except for a faint T2 and FLAIR hyperintensity of the corticospinal tracts (Figure 1).

Electromyography and nerve conduction studies showed myopathic changes. A lumbar puncture was performed; polymerase chain reaction performed on blood and on cerebrospinal fluid was positive for *Tropheryma whipplei* DNA, thus confirming the diagnosis of Neuro-Whipple.

Conclusion

Neurological involvement is not a constant in WD, but its occurrence may produce clinical pictures that are striking for their severity and uniqueness. Particularly, the involuntary movements that were observed in this patient are consistent with **oculomasticatory myorhythmia**, a movement disorder that is pathognomonic of WD. This case emphasizes the importance of suspecting WD when confronted with complex neurological syndromes.

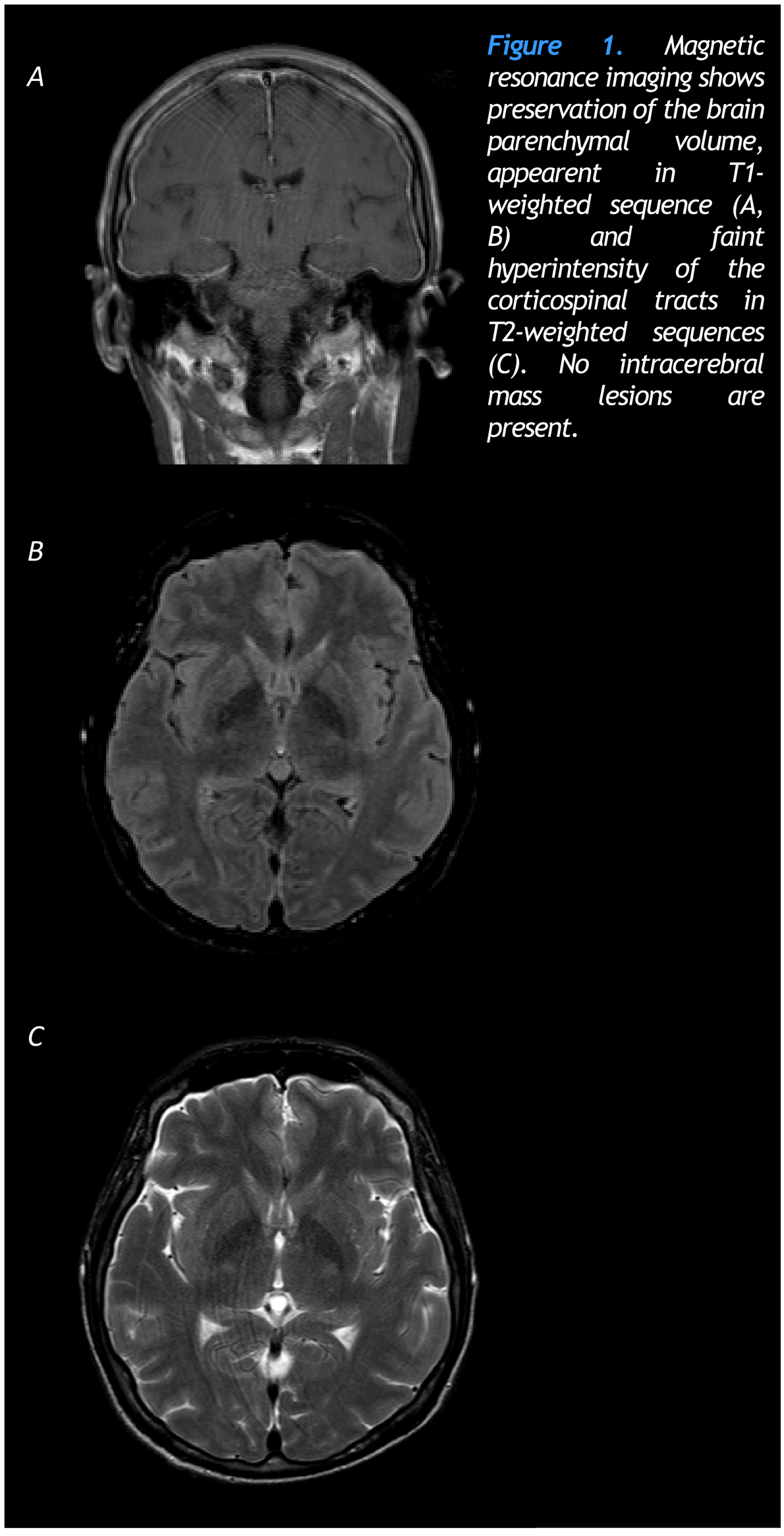

Figure 1. *Magnetic resonance imaging shows preservation of the brain parenchymal volume, appearent in T1-weighted sequence (A, B) and faint hyperintensity of the corticospinal tracts in T2-weighted sequences (C). No intracerebral mass lesions are present.*