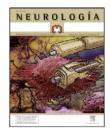


NEUROLOGÍA



www.elsevier.es/neurologia

LETTERS TO THE EDITOR

Whipple's disease: Multiple systemic and neurological relapses. Reply $^{\diamond}$

Enfermedad de Whipple: Múltiples recaídas sistémicas y neurológicas. Réplica

Dear Editor:

It was with great interest that we read the study recently published by Domínguez et al.¹ in which they describe a clinical case of Whipple's disease (WD) and also present the different systemic and neurological manifestations of that entity.

Our objective, focusing on isolated central nervous system involvement, is to emphasise the relapsing—recurring course that may characterise Whipple's disease with neurological symptoms. Recognition of this characteristic inherent to WD is essential when WD is clinically suspected in complex cases with no accompanying systemic manifestations (polyarthralgia, diarrhoea, or others).

Post-mortem studies have shown that the central nervous system is affected in up to 50% of patients diagnosed with WD, although only 10%–20% of cases are estimated to show clinical manifestations.^{2–4} Neurological manifestations, as this author has already mentioned,¹ are extremely variable, and include stroke, convulsions, progressive cognitive decline, sleep disorders, supranuclear gaze palsy, communicating hydrocephalus, and oculomasticatory myorhythmia.

The ample variability of the neurological signs and symptoms with which WD may present creates extremely complex diagnostic challenges. Four years ago, we published case reports on 2 patients with isolated CNS involvement and atypical presentations.⁵ In both patients, presentation was hyperacute; the first case resembled viral encephalitis and the second resembled a stroke. Only the disease progression with multiple neurological relapses led us to suspect WD, and the diagnosis was confirmed histologically by using

* Please cite this article as: Labiano-Fontcuberta A, Benito-León J. Enfermedad de Whipple: Múltiples recaídas sistémicas y neurológicas. Réplica. Neurología. 2012;27:376. an electron microscope.⁵ These examples illustrate the importance of suspecting WD in all cases of neurological disorders that present with a relapsing-recurring pattern.

Early diagnosis of WD is particularly important, since the disease can be fatal if treatment is not administered. As the authors rightly state,¹ proper treatment is still a matter for debate, since the optimal type of antibiotics and treatment time have not yet been established. In our experience, combining several different antibiotic regimens over an extended period of time can effectively prevent relapses.^{5,6}

References

- 1. Domínguez RO, Müller C, Davolos I, MacKeith P, Arias E, Taratuto AL. Enfermedad de Whipple Múltiples recaídas sistémicas y neurológicas. Neurologia. 2011, doi:10.1016/j.nrl.2011.04.009.
- Gerard A, Sarrot-Reynauld F, Liozon E, Cathebras P, Besson G, Robin C, et al. Neurologic presentation of Whipple disease: report of 12 cases and review of the literature. Medicine (Baltimore). 2002;81:443–57.
- Fenollar F, Puéchal X, Raoult D. Whipple's disease. N Engl J Med. 2007;356:55–66.
- 4. Louis ED. Whipple disease. Curr Neurol Neurosci Rep. 2003;3:470-5.
- 5. Benito-León J, Arpa J, Louis de, Herrera I, De La Loma A. Isolated CNS Whipple disease: acute onset and relapsing-remitting course. Scand J Infect Dis. 2007;39:623–5.
- Benito-León J, Sedano LF, Louis ED. Isolated central nervous system Whipple's disease causing reversible frontotemporal-like dementia. Clin Neurol Neurosurg. 2008;110: 747–9.
- A. Labiano-Fontcuberta^{a,b}, J. Benito-León^{a,b,c,*}

^a Servicio de Neurología, Hospital Universitario 12 de Octubre, Madrid, Spain

^b Departamento de Medicina, Facultad de Medicina,

- Universidad Complutense, Madrid, Spain
- ^c Centro de Investigación Biomédica en Red sobre
- Enfermedades Neurodegenerativas, Madrid, Spain

* Corresponding author.

E-mail address: jbenitol@meditex.es (J. Benito-León).