LETTERS TO THE EDITOR

Whipple’s disease: Multiple systemic and neurological relapses. Reply

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., 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. Neurological manifestations, as this author has already mentioned, are extremely variable, and include stroke, convulsions, progressive cognitive decline, sleep disorders, supranuclear gaze palsy, communicating hydrocephalus, and oculomotoric 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. 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 an electron microscope. 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, 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.

References


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