Isolated CNS Whipple disease with a variant of oculofacial–skeletal myorhythmia (OFSM)

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Figure 1 Axial fluid-attenuated inversion recovery MRI

Axial fluid-attenuated inversion recovery image obtained at the level of the third ventricle discloses hyperintense lesions, bilateral and relatively symmetric, in thalami, lentiform nuclei, and caudate nuclei, without appreciable mass effect.

A 62-year-old woman presented with insidious onset of depressive mood and progressive difficulties with daily activities. She had no diarrhea, abdominal cramps, arthralgia, weight loss, or palpable lymphadenopathy. Physical examination revealed fever, delirium, bilateral upper motor neuron signs, and myorhythmic movements¹ (see the video on the *Neurology* Web site [www.neurology.org]). Supranuclear ophthalmoplegia and cranial nerve involvement were absent. CSF

Figure 2 Biopsy specimen

Right thalamus biopsy specimen stained by periodic acid-Schiff (PAS) showing PAS-positive bacilli inside histiocytes in the space of Virchow (arrows).

showed a predominantly lymphocytic pleocytosis (85 cells/mm³). MRI disclosed subcortical lesions (figure 1). Stereotactic biopsy (figure 2) confirmed CNS Whipple disease. Ceftriaxone (4 g/day for 28 days), sulfamethoxazole-trimethoprim (320/1,600 mg/day for 1 year),² and monthly gamma-globulin (2 g/kg) were started. After 3 months the oculofacial–skeletal myorhythmia (OFSM) variant disappeared, the CSF normalized, and she was afebrile and responded to verbal commands.

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