Spinal Cord Involvement in CNS Whipple Disease: An Ongoing Experience in the Magnetic Resonance Imaging Era

In their case report Kremer et al (1) presented findings in a patient with Whipple disease (WD) that involved the brain, optic chiasm, posterior fossa, and spinal cord. They underscored the rarity of spinal cord involvement by calling the case described by Clarke et al (2) a unique reported case of myelopathy secondary to WD. Actually, in the 62-year-old woman reported on by Clarke et al, myelopathy was a unique presentation and MR imaging abnormalities (ie, high signal intensity on T2-weighted images, minimal contrast enhancement, and cord enlargement) were confined to the cord and medulla. Such an isolated spinal cord and medullary lesion suggested that a neoplasm was one of the diagnostic possibilities, and cord biopsy was performed. The finding of large numbers of foamy macrophages containing bacilliform structures that had positive results at periodic acid-Schiff staining made the correct diagnosis and treatment possible. Findings at jejunal biopsy were normal, but results of the polymerase chain reaction (PCR) for Whipple agent (*Tropheryma whippelii*) were positive.

We would like to emphasize that, while a myelopathy associated with WD is a multisystem disease or one that is confined to the CNS but involves several compartments, it is not a substantial diagnostic problem. An isolated spinal cord lesion in a patient without other system involvement is most likely to be misdiagnosed or diagnosed late. We recently described the case of a 65-year-old woman who had a severe myelopathy and an expansive spinal cord lesion that was highly suggestive of an intrinsic neoplasm on MR images (3). (To our knowledge, this is the second reported case in which CNS WD had only a spinal presentation.) Biopsy was not performed; the disease had a remitting-relapsing course with corticosteroid treatment, and cerebral lesions developed after only 3 years. While histologic and PCR findings in jejunal biopsy samples were negative, PCR analysis of the peripheral blood finally revealed the DNA of *T. whippelii*; the clinical and imaging improvement with specific treatment was dramatic and long-lasting. (At present, it persists at 22-mo follow-up.) MR images show that spinal cord involvement in CNS WD is either synchronous or early and possibly isolated, and they show either signal intensity abnormality or a tumorlike lesion. A high index of suspicion should be maintained for this challenging condition, although it is rare. Cord biopsy may not be necessary, even in the case of an isolated lesion that resembles a spinal cord tumor, since noninvasive molecular biologic techniques sometimes may make diagnosis possible.

Anna Messori, M.D.,
Ugo Salvolini, M.D.
University of Ancona
Ancona, Italy

References