LONGITUDDINALLY EXTENSIVE TRANSVERSE MYELITIS IN WEGENER’S GRANULOMATOSIS: IS IT RELATED TO NEUROMYELITIS OPTICA?

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Introduction
Both the central and the peripheral nervous system are involved in Wegener’s granulomatosis. The most frequent manifestation is peripheral neuropathy followed by cranial nerve involvement. Transverse myelitis is poorly described in literature. Herein reported is a two-case series of Wegener’s granulomatosis presenting with longitudinally extensive transverse myelitis lesions.

Case reports
Case 1: a 60-year-old man, previously blind since his childhood due to a bilateral corneal lesion, presented with a slowly-progressive lower-limb symmetrical paresthesia followed by urinary and fecal retention. Patient had previously been submitted to a nasal septoplasty to treat recurrent episodes of sinusitis. In a five-month period, he was readmitted with vertigo, gait disturbance with a left tendency to fall, dysphonia, grade 4 lower-limb weakness and sensitive level at T10. Thoracic spinal resonance imaging depicted a longitudinally extensive lesion intertwined with an atrophic tissue. Cerebrospinal fluid analysis was normal. Laboratory work-up showed a positive p-ANCA test and ruled out other diseases. Serum anti-aquaporin 4 antibody was negative. He was then submitted to a five-day course of methylprednisone with partial improvement of symptoms and azathioprine was started at 150 mg/day. He persisted with a mild spastic-ataxic gait and an EDSS score at 3.0. Case 2: a 60-year-old man presented with a history of bouts of neurological symptoms spared in a six-month period. In 2001, he presented with fatigue, arthralgia and an acute onset of left hemiparesis which completely resolved in 24 hours. In 2002, he developed paraparesis associated with urinary retention which lasted six months and was insufficiently investigated and treated with oral prednisone. In 2003, he complained of fatigue and arthritis and he was started on azathioprine after a rheumatologic consult. In 2004, painful erythematous skin changes and arthritis simultaneously occurred. A c-ANCA test was positive (1:40). Chest computed tomography showed bilateral posterior and basal pulmonary segments opacifications. In 2005, he developed global hyperreflexia and pyramidal signs which resulted in a spastic-ataxic gait. In 2010, only three days after an H1N1 vaccination, he presented with right hemiparesis. Spinal cord magnetic resonance imaging showed no active lesions, but an atrophic tissue from C4 to C7 levels.

Conclusion
In this series of two patients with Wegener’s granulomatosis, longitudinally extensive transverse myelitis were observed. Patients also had criteria of neuromyelitis optica spectrum disorder. Is it only a spurious association or does it have a causal link?

References