Syringomyelia in Neuromyelitis Optica Seropositive for Aquaporin-4 Antibody

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A 51-year-old woman suffered quadriparesis, hypalgesia at C5 to Th6, and dyspnea. Spinal T1- and T2-weighted MRI revealed a swollen cervical cord with irregularly shaped low- and high-intensity areas, respectively, in the central syrinx (C1-C7). Dorsal syringomyelia was gadolinium-enhanced, suggesting active demyelination (Picture 1). Steroid medication achieved complete recovery; MRI lesions disappeared in one year. Four years later, she suffered left hemiparesis and hypesthesia at Th10-12. T2-weighted MRI showed longitudinally extensive high intensity (partly gadolinium-enhanced) at Th2-6, without syringomyelia (Picture 2). No brain lesions were detected. Visual-evoked potential showed abnormal waves. Serum anti-aquaporin-4 antibody was positive.

Syringomyelia accompanies 4.5% of multiple sclerosis cases in Switzerland (1), but it is rare in Japan. One syringomyelia in neuromyelitis optica case was reported (2), but the present case is the first with anti-aquaporin-4 seropositivity. Interestingly, the localization of syringomyelia was thoracic/lumbar (1) in multiple sclerosis and the cervical...
cord in neuromyelitis optica in the present case and that of O’Riordan et al (2). Anti-aquaporin-4 antibody may facilitate syrinx formation through enhanced blood-brain barrier permeability (3).

References


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