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Reversible stenosis of large cerebral arteries in a patient with combined Sjögren's syndrome and neuromyelitis optica spectrum disorder.

Ii Y, Shindo A, Sasaki R, Naito Y, Tanaka K, Kuzuhara S.

Department of Neurology, Mie University Graduate School of Medicine, 2-174, Edobashi, Tsu, Mie 514-8507, Japan.
ii-y@clin.medic.mie-u.ac.jp

We report a 49-year-old woman with neuromyelitis optica (NMO) spectrum disorder coexisting with Sjögren's syndrome (SS). She presented with acute brainstem symptoms and transverse myelitis. Brain MRI showed focal high signal intensity lesions in the hypothalamus and the pontine tegmentum on T2-weighted and FLAIR images. MRA revealed stenotic changes of the bilateral middle cerebral artery (MCA), posterior cerebral arteries (PCA) and basilar artery (BA). Spinal MRI revealed hyperintense lesions within the cord extending from the T4 to the T6 level on the T2-weighted image. The patient fulfilled the clinical criteria of primary SS. In addition, anti-AQP4 antibody which is highly specific for NMO was detected in the serum at the acute phase. The patient excellently responded to IVIg while methylprednisolon pulse therapy was not effective. Follow-up MRA displayed complete resolution of the stenosis of the MCA, PCA and BA.

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