Diagnosis and Management of Neuromyelitis Optica

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Abstract

Typical NMO is characterized by simultaneous or sequential acute transverse myelitis and optic neuritis. Spinal cord lesions extending over 3 or more vertebral segments and normal brain imaging are the typical MRI findings in NMO. In typical cases with positive NMO antibody the diagnosis is easy but in seronegative and atypical cases with different clinical manifestations and MRI features the diagnosis will be challenging. Brainstem syndromes such as ataxia, intractable vomiting and cranial nerve palsies are less common initial manifestations of NMO. Shorter spinal cord lesions in MRI can be found very early during relapse or in residual atrophic stages. Some of the lesions may appear hypointense on corresponding T1-weighted images which reflects severe inflammation with necrosis. T1-weighted hypointensities may help in differentiating NMO lesions from MS in spinal cord. Brain MRI is initially normal in most patients with NMO but serial imaging may depict lesions in up to 85% of the patients at later stages. Brain lesions are usually nonspecific but may fulfill the Barkhof criteria for dissemination in space. Acute, large, edematous callosal lesions with a heterogeneous intensity occasionally develop in patients with NMO (marbled pattern). In contrast, in MS, callosal lesions are small, non-edematous, and the intensity was homogeneous in the acute phase and located at the lower margin of the corpus callosum. Multiple patchy enhancements with blurred margins (cloudlike enhancement) have been reported to be typical for NMO. Pencil-like ependymal enhancement is another type of enhancement around the anterior horns of lateral ventricles. Extensive hemispheric lesions, periependymal lesions surrounding the aqueduct, the third and fourth ventricles and brainstem lesions should raise the diagnosis of NMO. Actually NMO should be considered in any atypical lesion for MS in appropriate clinical settings. Other auto antibodies such as anti MOG have extended the spectrum of this entity and should be evaluated in special cases who are seronegative for anti-aquaporin 4. Treatment of NMO consists of acute relapse management with steroids and/or plasma exchange and prevention of exacerbations with cytoxic agents or Rituximab.

Keywords: Neuromyelitis Optica, Inflammation, Spinal cord

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