Contrasting disease patterns in seropositive and seronegative neuromyelitis optica

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Neuromyelitis optica (NMO) is a severely disabling inflammatory disorder of the central nervous system (CNS) of putative autoimmune aetiology that predominantly affects the optic nerves and spinal cord .NMO is associated with serum antibodies to aquaporin-4, the most abundant water channel in the CNS in up to 80% of cases . These antibodies (termed NMO-IgG or AQP4-Ab) are thought to be directly involved in the pathogenesis of the condition .The clinical spectrum of NMO as defined by Wingerchuk et al. (2007) comprises cases of simultaneous optic neuritis (ON) and myelitis, cases of myelitis and ON, in which the two index events do not develop simultaneously but successively, and limited or inaugural forms such as single or recurrent events of longitudinally extensive myelitis (LETM) or recurrent ON. More rarely, patients may present with brain stem encephalitis . Similar to other autoimmune neurological diseases such as myasthenia gravis, a subset of patients exists who are seronegative. There are indications that seropositive and seronegative patients might differ with regard to clinical presentation or prognosis .

In a large multicenter study these results were seen:

The female to male ratio was significantly higher among seropositive patients compared to seronegative patients.

A monophasic course was more common among seronegative patients and, accordingly, also more frequent in patients in whom the disease started with simultaneous myelitis and ON compared to those in whom the disease started with either myelitis or ON . No significant difference regarding the annual myelitis relapse, the annual ON relapse rate, or the ON to myelitis ratio was found between seropositive and seronegative patients.

Importantly, the annualized EDSS progression index did not differ significantly between seropositive and seronegative patients.

Signs of co-existing autoimmunity were significantly more frequent in the seropositive group .

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