Neuropathic itch as a feature of Neuromyelitis Optica spectrum disorders

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Background: Neuropathic itch, an uncommon symptom in neurology, has been recently described in patients with Neuromyelitis Optica (NMO) spectrum disorders (SD) [1]. Longitudinally extensive transverse myelitis (LETM) is a characteristic feature of NMO but it can also occur in other inflammatory or infectious diseases of the central nervous system (CNS). An IgG autoantibody specific for Aquaporin-4 (AQP4) is a reliable biomarker to distinguish NMOSD from other inflammatory diseases of the CNS [2].

Objective: To evaluate the frequency, the clinical characteristics and the neuroimaging correlates of neuropathic itch in patients with LETM with and without AQP4 autoantibodies.

Methods: Retrospective review of all patients with LETM who attended our clinic from January 2010 to September 2014. Serum samples from all patients were tested for AQP4-IgG by a cell-based assay.

Results: We identified 43 patients (28 females, 65.1%) with a first-ever episode of LETM, of whom 19 (44.1%) were AQP4 antibody (Ab) positive. Neuropathic itch was present at LETM onset, in 7/19 (36.8%) of the AQP4-Ab positive and in 3/22 (12.5%) of the AQP4-Ab negative patients (p=0.026). One AQP4-Ab negative patient with neuropathic itch experienced another LETM episode and optic neuritis during the follow-up period, thus fulfilling the NMO diagnostic criteria [3]. In all patients itch involved the dermatomes corresponding to the levels of the spinal lesions, which always involved the central grey matter of the spinal cord. In most patients (80%) it was the first clinical manifestation of LETM. In 7/10 patients, itch responded to intravenous steroids or plasmapheresis; 2 patients were successfully treated with gabapentin.

Conclusions: Neuropathic itch can herald or accompany LETM, being more frequent in AQP4-Ab positive patients. The role of AQP4 autoantibodies in neuropathic itch warrants further investigations.