Abstracts

Articles appearing in the May 2019 issue

GABAA receptor autoimmunity: A multicenter experience

**Objective** We sought to validate methods for detection and confirmation of GABA\(_A\) receptor (R)-IgG and clinically characterize seropositive cases.

**Methods** Archived serum and CSF specimens (185 total) suspected to harbor GABA\(_A\)R-IgG were evaluated by indirect immunofluorescence assay (IFA). Twenty-six specimens from 19 patients appeared suspicious for GABA\(_A\)R-IgG positivity by IFA, based on prior reports and comparison with commercial GABA\(_A\)R antibody staining. Aliquots of those specimens were tested at the University of Oxford, United Kingdom, and Euroimmun, Lubeck, Germany, for GABA\(_A\)R-IgG by cell-based assays (CBAs) using HEK293-indicator cells transfected with plasmids encoding different GABA\(_A\)R subunits.

**Results** Eight specimens (of 26 tested; 4 serums, 4 CSFs) from 5 patients were confirmed by CBA to be GABA\(_A\)R-IgG positive. Patient IgGs were always reactive with \(\alpha_1\beta_3\) GABA\(_A\)R subunits. One more patient was identified clinically after this validation study. Median age for the 6 patients at serologic diagnosis was 44 years (range, 1–71 years), and 4 of them were male. Among the 4 for whom clinical information was available (2 treated by the authors), all had encephalitis and antiepileptic drug refractory seizures. Three out of 4 patients treated with a combination of immunotherapies had good outcomes. The fourth, recognized to have an autoimmune cause late in the clinical course, had severe permanent neurologic sequelae and brain atrophy.

**Conclusions** Though not as common as NMDA-R encephalitis, GABA\(_A\)R encephalitis generally has a characteristic clinical-radiologic presentation and is treatable, making accurate laboratory diagnosis critical.

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Quantitative 7T MRI does not detect occult brain damage in neuromyelitis optica

**Objective** To investigate and compare occult damages in aquaporin-4 (AQP4)-rich periependymal regions in patients with neuromyelitis optica spectrum disorder (NMOSD) vs healthy controls (HCs) and patients with MS applying quantitative T1 mapping at 7 Tesla (T) in a cross-sectional study.

**Methods** Eleven patients with NMOSD (median Expanded Disability Status Scale [EDSS] score 3.5, disease duration 9.3 years, age 43.7 years, and 11 female) seropositive for anti-AQP4 antibodies, 7 patients with MS (median EDSS score 1.5, disease duration 3.6, age 30.2 years, and 4 female), and 10 HCs underwent 7T MRI. The imaging protocol included T2*-weighted (w) imaging and an MP2RAGE sequence yielding 3D T1w images and quantitative T1 maps. We semiautomatically marked the lesion-free periependymal area around the cerebral aqueduct and the lateral, third, and fourth ventricles to finally measure and compare the T1 relaxation time within these areas.

**Results** We did not observe any differences in the T1 relaxation time between patients with NMOSD and HCs (all \(p > 0.05\)). Contrarily, the T1 relaxation time was longer in patients with MS vs patients with NMOSD (lateral ventricle \(p = 0.026\), third ventricle \(p = 0.173\), fourth ventricle \(p = 0.016\), and cerebral aqueduct \(p = 0.048\)) and vs HCs (third ventricle \(p = 0.027\), fourth ventricle \(p = 0.013\), lateral ventricle \(p = 0.043\), and cerebral aqueduct \(p = 0.005\)).

**Conclusion** Unlike in MS, we did not observe subtle T1 changes in lesion-free periependymal regions in NMOSD, which supports the hypothesis of a rather focal than diffuse brain pathology in NMOSD.

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