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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 $GABA_AR$-IgG were evaluated by indirect immunofluorescence assay (IFA). Twenty-six specimens from 19 patients appeared suspicious for $GABA_AR$–IgG positivity by IFA, based on prior reports and comparison with commercial $GABA_AR$ antibody staining. Aliquots of those specimens were tested at the University of Oxford, United Kingdom, and Euroimmun, Lubeck, Germany, for $GABA_AR$-IgG by cell-based assays (CBAs) using HEK293-indicator cells transfected with plasmids encoding different $GABA_AR$ subunits.

Results Eight specimens (of 26 tested; 4 serums, 4 CSFs) from 5 patients were confirmed by CBA to be $GABA_AR$-IgG positive. Patient IgGs were always reactive with α1β3 $GABA_AR$ 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, $GABA_AR$ 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 $p = 0.056$, 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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F. Luessi, S. Engel, A. Spreer, S. Bittner, F. Zipp. 2018;5:e481. doi.org/10.1212/NXI.0000000000000481

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J.O Dalmau. 2019;6:e564. doi.org/10.1212/NXI.0000000000000564

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