Teaching NeuroImages: Linear Radial Periventricular Enhancement in Glial Fibrillary Acidic Protein Astrocytopathy

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Figure Baseline and Interval MRI Features in Glial Fibrillary Acidic Protein Astrocytopathy After Immunosuppressive Treatment

(A) Baseline T2-weighted deep white matter hyperintensities. (B) Baseline contrast-enhanced T1-weighted linear/radial perivascular enhancement. (C) Baseline T2-weighted dorsal cord focal hyperintensity. (D) Contrast-enhanced T1-weighted leptomeningeal enhancement over conus. (E) Interval T2-weighted progression/confluence of hyperintensities in periventricular and deep white matter with background involution. (F) Corresponding resolution of radial perivascular enhancement.

A 48-year-old woman presented with asymptomatic painless optic disc swelling, seizures, cognitive decline, and weakness over 3 months. She had an upper motor neuron pattern of grade 4/5 weakness in all 4 limbs. Brain MRI revealed striking linear radial perivascular enhancement in the deep white matter^{1,2} (figure, A and B); MRI spine revealed longitudinally extensive C3-to-conus cord T2 hyperintensity and T1 enhancement² (figure, C and D). CSF

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demonstrated glial fibrillary acidic protein (GFAP) pattern by indirect immunofluorescence with GFAP α specificity confirmed by cell-based assay. The patient improved clinically (figure, E and F) with steroid treatment. Linear/radial perivascular enhancement in the deep white matter prompts consideration of a corticosteroid-responsive meningoencephalomyelitis termed autoimmune GFAP astrocytopathy.¹

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Disclosure

A. McKeon serves as an editorial board member of *Neurology®: Neuroimmunology and Neuroinflammation*; received research support from Medimmune, Euroimmun, Grifols, and Alexion; and has patents pending for Septin 5, GFAP, PDE10A, Kelch-11, and MAP1B as markers of neurologic autoimmunity and paraneoplastic disorders. A. Zekeridou has a patent pending for PDE10A-IgG paraneoplastic autoimmunity. Y. Kong, O.S.Q. Koh, Y.R. Chiew, B. Purohit, C.F. Chin, and A.S.L. Ng report no disclosures. Go to Neurology.org/N for full disclosures.

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Yongyao Kong, MD	Department of Neurology, National Neuroscience Institute, Singapore	Writing of manuscript

Appendix (continued)

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Andrew McKeon, MD	Departments of Laboratory Medicine and Pathology and Neurology, Mayo Clinic College of Medicine, Rochester, MN	Writing of manuscript
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Anastasia Zekeridou, MD, PhD	Departments of Laboratory Medicine and Pathology and Neurology, Mayo Clinic College of Medicine, Rochester, MN	Writing of manuscript
Adeline S.L. Ng, MD	Department of Neurology, National Neuroscience Institute, Singapore	Revision of manuscript for intellectual content

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