

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

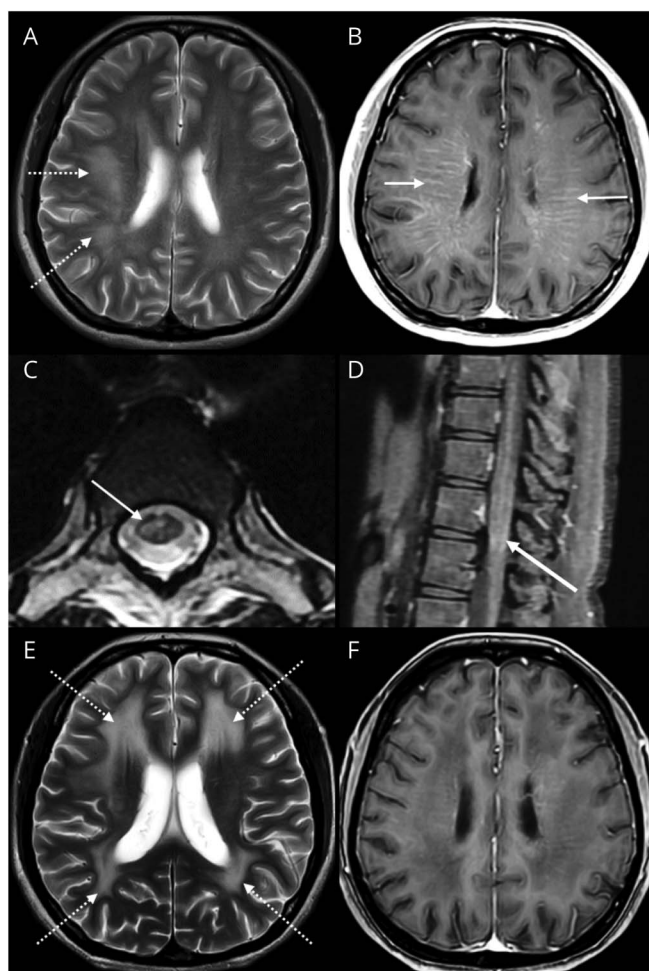
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Figure Baseline and Interval MRI Features in Glial Fibrillary Acidic Protein Astrocytopathy After Immunosuppressive Treatment



(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

Study Funding

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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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Appendix (continued)

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Adeline S.L. Ng, MD	Department of Neurology, National Neuroscience Institute, Singapore	Revision of manuscript for intellectual content

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