Teaching NeuroImages: Autoimmune glial fibrillary acidic protein meningoencephalomyelitis

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Neurology® 2019;93:e2076-e2077. doi:10.1212/WNL.0000000000008562

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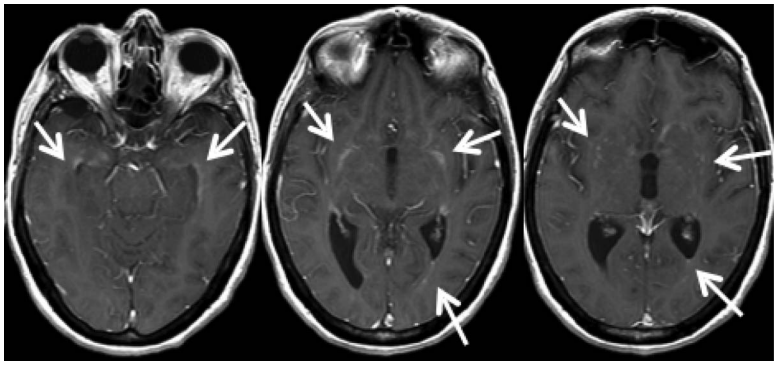
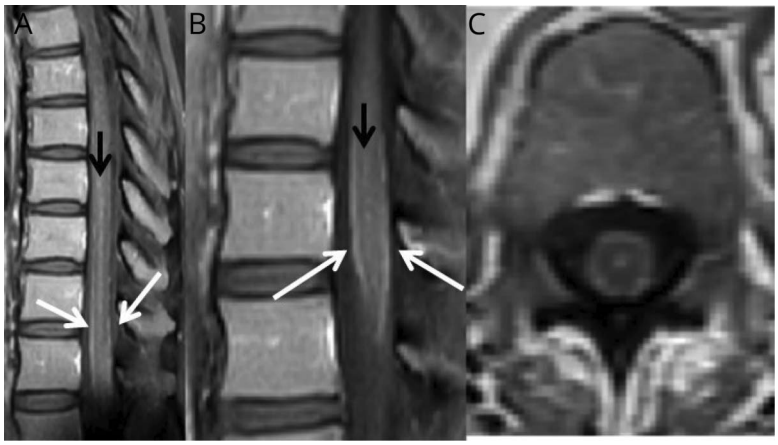
Figure 1 Axial T1 postcontrast brain MRI



Leptomeningeal, linear/stippled parenchymal, and ependymal postgadolinium enhancement (arrows).

Figure 2 Thoracic sagittal T1 postcontrast MRI



Leptomeningeal (white arrows) and central canal enhancement (black arrows) throughout the cord (A) and conus medullaris (B). Axial T1 postcontrast image (C) with peripheral leptomeningeal enhancement and central dot of central spinal canal enhancement.

A 73-year-old woman with myelodysplastic syndrome presented with 2 months of decline in cognition and mobility, accompanied by headache and weight loss. Examination revealed a somnolent woman with symmetric cogwheel rigidity, bradykinesia, arm myoclonus, and hyperreflexia. Brain MRI showed leptomeningeal, linear/stippled parenchymal, and ependymal enhancement (figure 1). Spine MRI demonstrated nonenhancing cervicothoracic cord signal change and circumferential enhancement of conus medullaris with central canal enhancement

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(figure 2). CSF was inflammatory with glial fibrillary acidic protein (GFAP)–immunoglobulin G (IgG). The patient responded promptly to IV methylprednisolone. GFAP-IgG identifies a severe but highly corticosteroid-responsive autoimmune meningoencephalomyelitis with distinctive MRI findings.[1,2]

Study funding

No targeted funding reported.

Disclosure

The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

Appendix Authors

Name	Location	Role	Contribution
Paula K.J. Lee, MD	Tufts Medical Center, Boston, MA	Author	Drafted the manuscript for intellectual content, primary clinical care of the patient

Appendix *(continued)*

Name	Location	Role	Contribution
Juan E. Small, MD	Lahey Hospital & Medical Center, Burlington, MA	Author	Major role in acquisition of imaging data, interpreted the data
Michal Vytopil, MD	Lahey Hospital & Medical Center, Burlington, MA	Author	Revised and supervised the manuscript for intellectual content, clinical care of the patient

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2. Flanagan EP, Hinson SR, Lennon VA, et al. Glial fibrillary acidic protein immunoglobulin G as biomarker of autoimmune astrocytopathy: analysis of 102 patients. Ann Neurol 2017;81:298–309.

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