

# Teaching NeuroImages: Steroid-responsive leukoencephalopathy in inflammatory cerebral amyloid angiopathy

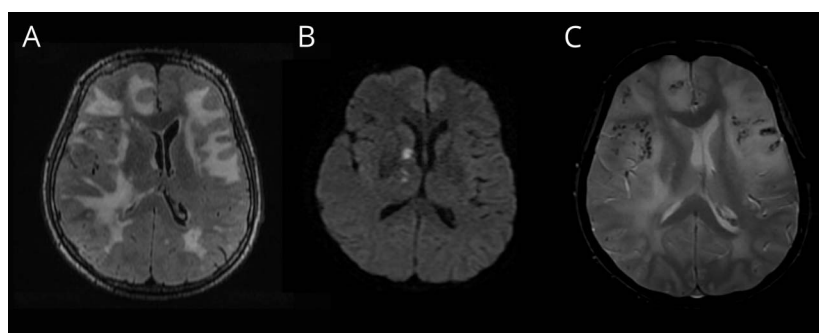
David Dongkyung Kim, MD, Teneille Gofton, MD, and G. Bryan Young, MD

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## Correspondence

Dr. Young  
Bryan.young@lhsc.on.ca

**Figure 1** Initial MRI



(A) T2-weighted confluent leukoencephalopathy extending to the subcortical white matter with mass effect. (B) Diffusion-weighted imaging 10 days later with associated changes on the apparent diffusion coefficient sequence (not pictured) was consistent with infarction. (C) Gradient echo sequence revealed microhemorrhages.

An obtunded previously healthy 60-year-old woman was intubated with a 2-week history of headaches and 1-month history of memory loss. MRI was performed (figure 1). Digital subtraction angiography of the head, varicella-zoster virus PCR, immunoglobulin G in the CSF, hepatitis B/C, CSF cryptococcal antigen, HIV, Lyme serology, and syphilis screen were negative. The initial differential diagnosis included CNS vasculitis and intracerebral lymphoma. She was diagnosed with inflammatory cerebral amyloid angiopathy based on clinico-radiologic criteria (which has a sensitivity of 82% and specificity of 97%),<sup>1</sup> with infarction suggesting amyloid- $\beta$ -related angiitis subtype. Within a few days of steroids, she was fully alert, and repeat MRI 5 months later showed improvement of leukoencephalopathy (figure 2). Montreal Cognitive Assessment scores were 23/30 2 weeks after initiation of therapy and 22/30 5 months later.

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## Author contributions

D.D. Kim: study concept and design, preparation of manuscript. T. Gofton, G.B. Young: study concept and design, supervision of manuscript.

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## Disclosure

The authors report no disclosures relevant to the manuscript. Go to [Neurology.org/N](https://Neurology.org/N) for full disclosures.

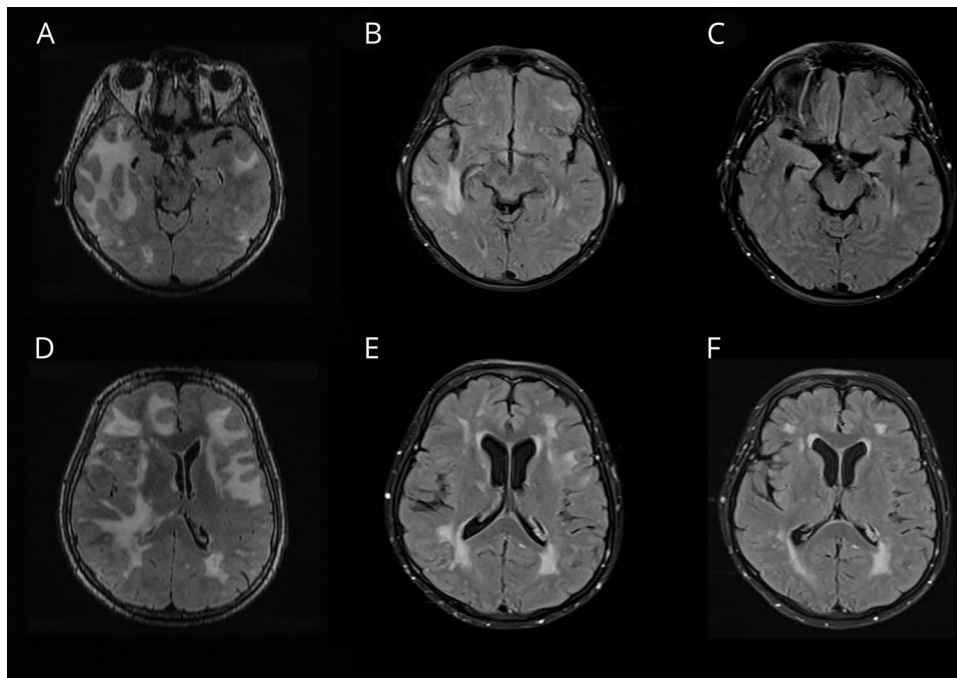
## Reference

1. Auriel E, Charidimou A, Gurol ME, et al. Validation of clinico-radiological criteria for the diagnosis of cerebral amyloid angiopathy-related inflammation. *JAMA Neurol* 2016;73:197–202.

From the Department of Clinical Neurological Sciences, Western University, London, Canada.

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**Figure 2** Radiologic improvement after steroids



Comparison between initial imaging (A, D), around 3 weeks after initiation of steroid therapy (B, E), and after 5 months of corticosteroid therapy (C, F) showed improvement of leukoencephalopathy.

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