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Teaching NeuroImages: Treatment-resistant rapidly progressive amyloid β–related angiitis

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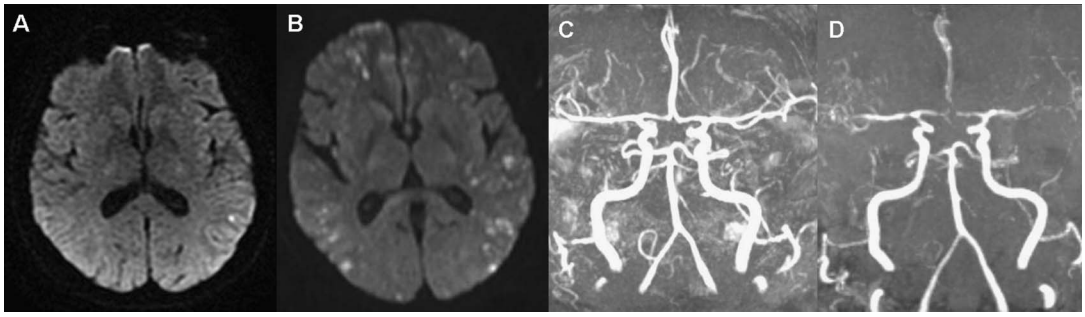
Figure 1 MRI and magnetic resonance angiography

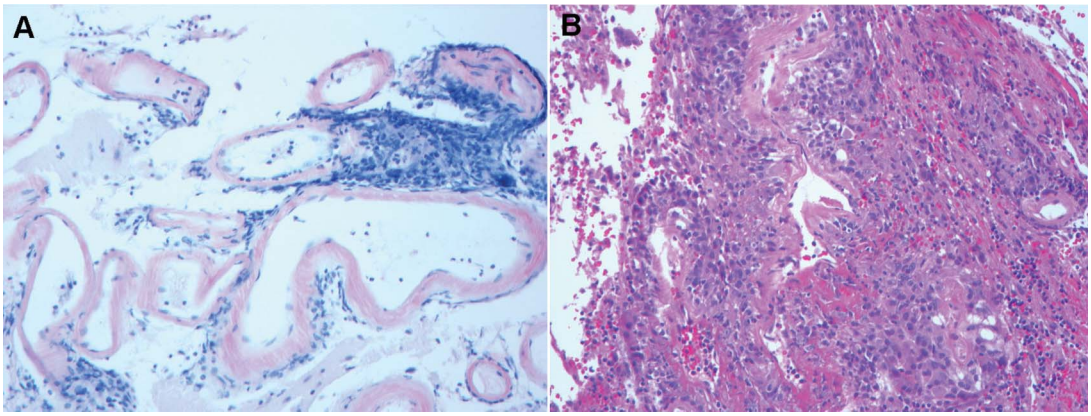


(A) Punctate restricted diffusion with normal intracranial vessels (C). (B) Increased burden of restricted diffusion with multifocal vessel attenuation (D).

A 76-year-old woman presented with 1 month of progressive aphasia, headache, and subsequent right hemiparesis. Initial brain MRI showed a punctate infarct (figure 1, A and C). Susceptibility-weighted imaging was unremarkable. A repeat study 16 days later demonstrated bihemispheric infarcts with multifocal attenuation of intracranial vessels on magnetic resonance angiography (figure 1, B and D). CSF showed a lymphocytic pleocytosis (101 leukocytes/μL) and elevated protein (480 mg/dL). Brain biopsy showed granulomatous angiitis with amyloid deposition and fibrinoid necrosis surrounded by inflammatory cells (figure 2). She rapidly deteriorated on immunosuppression with high-dose IV steroids and has not improved despite a combination of oral steroids and monthly cyclophosphamide.

Primary angiitis of the CNS is often suspected when multiple areas of ischemia on MRI are present along with multifocal vascular irregularity on angiography, without systemic involvement.[1] Amyloid β–related angiitis is thought to represent a distinct entity with amyloid

Figure 2 Meningeal (A) and frontal lobe (B) biopsies



Congo red–positive staining material consistent with amyloid in vessel walls surrounded by inflammatory cells and loose granulomas.

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β antibodies and more rapid progression compared to other CNS vasculitides.[2]

AUTHOR CONTRIBUTIONS

Meko Porter, Christopher R. Newey, and Gabor Toth contributed equally to the writing of the case and formatting the images.

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DISCLOSURE

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

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