Images in Neurology

Cortical Vein Thrombosis, Tortuous Venous Vasculature, and Microhemorrhages in Neurosarcoidosis

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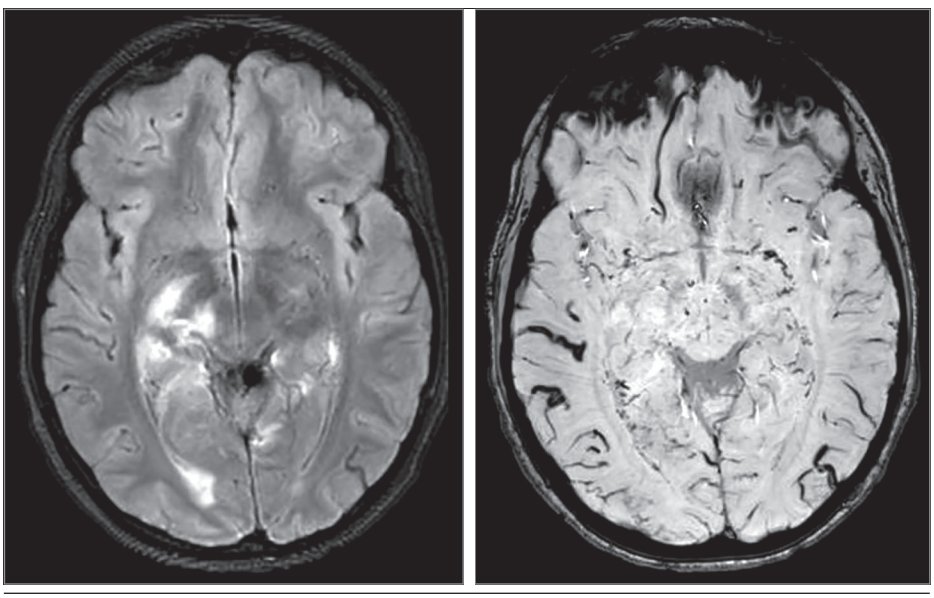
A previously healthy 13-year-old boy presented with an episode of left-sided facial droop and dysarthria lasting 90 minutes. His initial brain magnetic resonance imaging (MRI) demonstrated a nonenhancing fluid-attenuated inversion recovery hyperintensity in the corpus callosum and right cerebellar peduncle. Small tortuous veins were noted near the peduncular lesion, and punctate foci of susceptibility were noted in the area of the caudate, thalamus, and cerebellum. These were interpreted as a developmental venous anomaly and nonspecific prior injury, respectively. Serum anti–myelin oligodendrocyte glycoprotein and anti–aquaporin-4 antibodies tests were negative, and analysis of cerebrospinal fluid was normal except for a mild lymphocytic pleocytosis (7 leukocytes/mm^3 with 1 red blood cell count/mm^3) with a negative infectious evaluation. He was monitored closely every 3 months for clinical or radiographic progression. His MRI 8 months after presentation was notable for new and evolving T2 hyperintensities, with linear enhancement along the prior cerebellar lesion, and unchanged focal venous tortuosity and microhemorrhages. Despite several atypical features, this was interpreted as radiographic progression of presumed multiple sclerosis (MS), and he started rituximab infusions.

However, several months after rituximab initiation, the patient had a second episode of transient left-sided face and left arm tingling and dysarthria, along with development of daily migraine-type headaches. His brain MRI at this time (**Figure 1**) demonstrated interval development of new multifocal fluid-attenuated inversion recovery hyperintensities with nodular and linear enhancement patterns, a diffuse tortuous venous vasculature, new microhemorrhages, and a cortical vein thrombosis. Magnetic resonance angiography and spinal imaging results were normal. There was no diffusion restriction or cranial nerve enhancement. Hypercoagulable evaluation was negative, and anticoagulation was deferred in favor of monitoring. Given the clinical and radiographic features atypical of MS, he underwent brain biopsy. Hematoxylin-eosin–stained sections from the biopsy showed extensive non-necrotizing granulomatous inflammation of medium-sized blood vessels and leptomeninges, as well as the brain parenchyma (**Figure 2**).

The patient had negative subsequent serum angiotensin-converting enzyme, antineutrophil cytoplasmic antibodies, computed tomography of the chest, and infectious staining results, and his pathology was consistent with a diagnosis of granulomatous vasculitis secondary to isolated neurosarcoidosis. He started receiving a high-dose oral steroid taper, oral methotrexate, and infliximab infusions, with no further transient neurologic deficits and a decrease in his headaches. An MRI 3 months after initiation of this treatment regimen showed improvement in the size of previous fluid-attenuated inversion recovery lesions with no new lesions and resolution of enhancement and prior cortical vein thrombosis, although a second asymptomatic cortical vein thrombosis had developed.

The differentiation between MS and isolated neurosarcoidosis may be challenging because of potential overlap of clinical and radiographic features. The short-term and transient nature of the pa-

Figure 1. Brain Magnetic Resonance Imaging (MRI) 11 Months After Initial Presentation

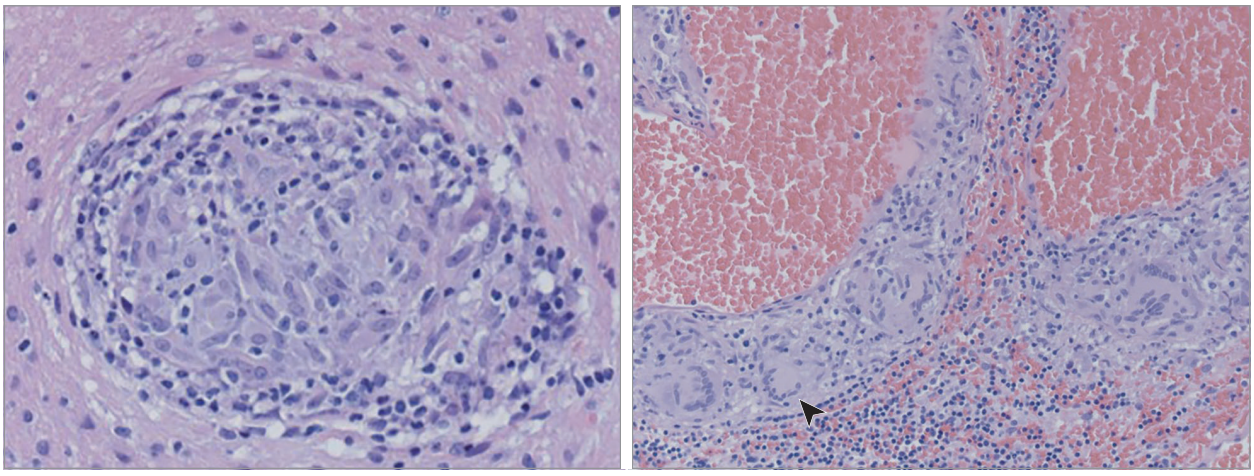


MRI demonstrated multifocal fluid-attenuated inversion recovery (FLAIR) hyperintensities with associated nodular and linear enhancement (not shown) (A) and susceptibility-weighted imaging (SWI) demonstrated diffuse prominent tortuous venous vasculature with scattered microhemorrhages (B).

Figure 2. Hematoxylin-Eosin Staining of Biopsy From Cerebellar Parenchyma

A Well-formed granuloma

B Granulomas within and surrounding cerebral blood vessels



Photomicrographs of a well-formed granuloma with associated epithelioid macrophages, surrounded by T lymphocytes (original magnification ×40) (A), and granulomas present within and surrounding the walls of cerebral blood vessels (original magnification ×20), with associated multinucleated giant cells (arrowhead; B). There was no necrosis and special stains ruled out mycobacterial infection.

tient's neurologic deficits was more consistent with a transient ischemic attack as might be seen in vasculitis, compared with demyelination. Radiographically, neurosarcoidosis may show tortuous or enlarged medullary veins, microhemorrhages, or venous thrombosis, all atypical for MS.[1] These were early radiographic identifiers of this patient's disease process and demonstrate the utility of susceptibility-weighted and vascular imaging in evaluation of suspected demyelinating disease. Other distinguishing characteristics that may be seen include leptomeningeal, dural, or pituitary/hypophyseal involvement, linear perivascular enhancement, mass-like lesions, pseudotumor, hydrocephalus, or enhancement persisting for more than 3 months.[2,3]

ARTICLE INFORMATION

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