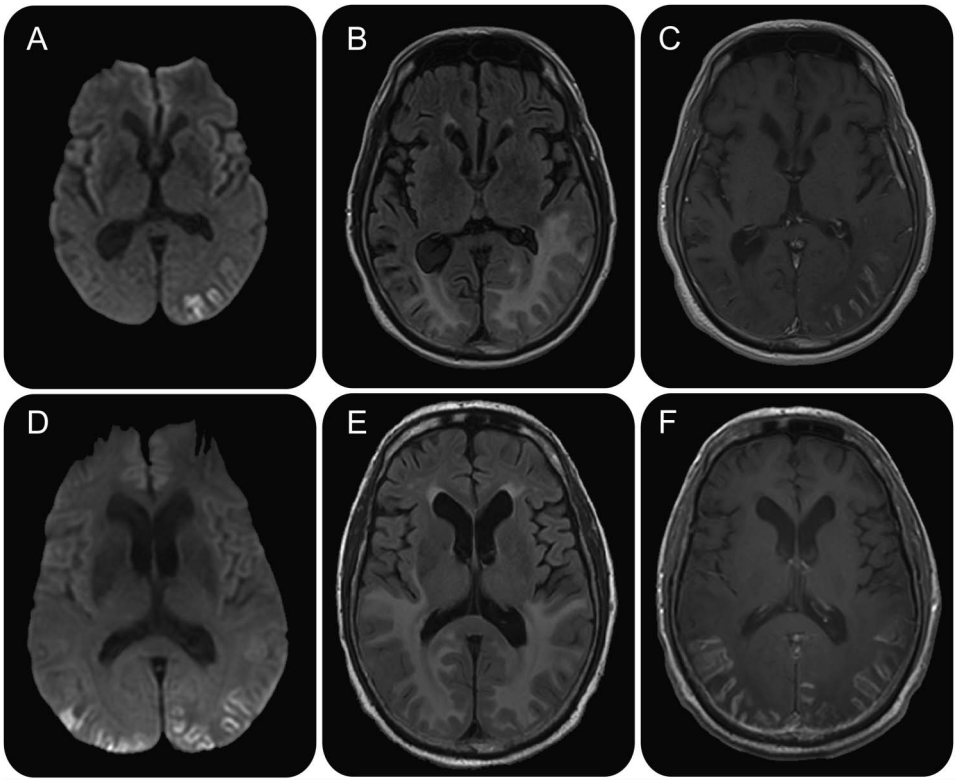
Meningioangiomatosis

A rare presentation with progressive cortical blindness

Figure 1 MRI brain



Initial MRI: restricted diffusion (A) with T2 hyperintensities (B) that enhanced with gadolinium (C) in the occipito-parietal lobes. MRI 18 months later: anterior extension of the above abnormalities (D–F).

A 58-year-old man presented with a 6-month history of headaches, progressive left homonymous hemianopia, and right-sided paresthesias. MRI revealed bilateral parietal and occipital enhancing lesions (figure 1, A–C) suggestive of cerebritis or posterior reversible encephalopathy syndrome due to history of hypertension. Worsening vision prompted neuroimaging 18 months later, showing contrast-enhancing cortical lesions in the posterior frontal region; fluid-attenuated inversion recovery abnormalities in the white matter (figure 1, D–F) suggested vasogenic edema and mass effect. Occipital lobe biopsy revealed cortical mesenchymal capillary proliferation typical of meningioangiomatosis (figure 2), a rare benign progressive disorder characterized by cortical vascular proliferation with associated perivascular mesenchymal cell proliferation.[1] Seizures are the presenting feature in 85% of sporadic cases; there have been no reports with permanent focal dysfunction[2] as in this instance.

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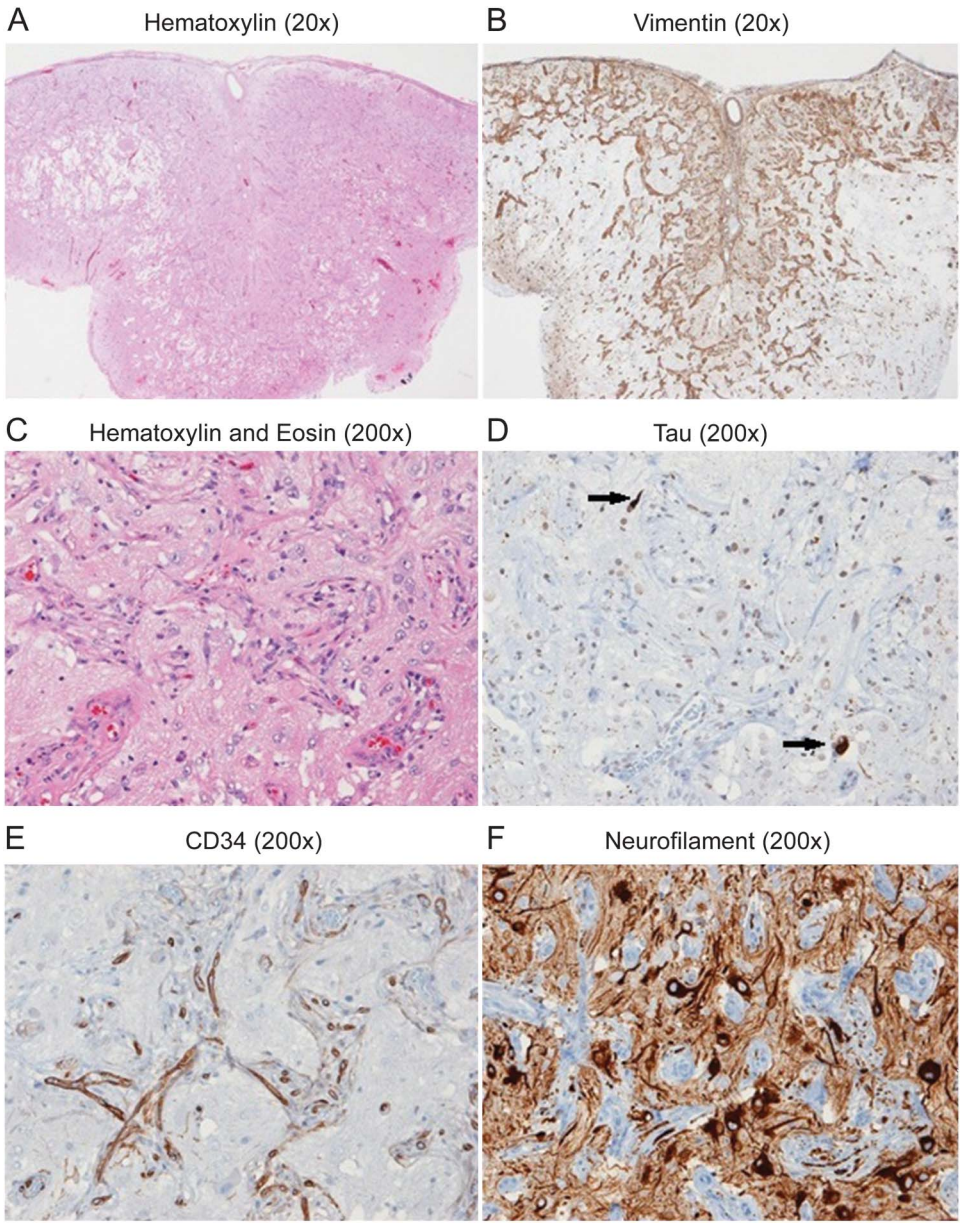
Author contributions: Akshay Shah, MD: primary author, case concept and design. Daniel Korya, MD: coauthor, critical revision of the manuscript for intellectual content. Brandon T. Larsen, MD, PhD: coauthor, critical revision of the manuscript for intellectual content. Myrka Torres, MD: critical revision of the manuscript for important intellectual content and supervision. Kendra Drake, MD: critical revision of the manuscript for important intellectual content and supervision. John La Wall, MD: critical revision of the manuscript for important intellectual content and supervision.

Figure 2 Brain biopsy



Plaque-like capillary proliferation encroaching the cortex (A) ensheathed by mesenchymal cells (C). Immunohistochemical staining highlighted mesenchymal proliferation (B), granulovacuolar degeneration (D) with neurofibrillary tangles (arrows), proliferating capillaries (E), and entrapped neurons (F).

Study funding: No targeted funding reported.

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

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Neurology 2013;81;511-512
DOI 10.1212/WNL.0b013e31829d8714