cases are attributed to impaired absorption of CSF due to increased dural venous sinus pressure or a defect in the arachnoid villi. The possibility of dural venous sinus thrombosis was ruled out in this case by MR examination that showed the venous sinuses to be widely patent. As suggested by the MR findings in this patient, low hemoglobin levels may result in compensatory changes in cerebral blood volume, leading to increased intracranial pressure. This case serves as a reminder that an idiopathic intracranial hypertension–like picture may occur as a consequence of severe anemia and is reversible upon correction of the underlying hematologic disorder. In addition, this case underscores the fact that idiopathic intracranial hypertension may presage the onset of systemic lupus erythematosus.

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References

1. Taylor F. The practice of medicine. London: J & A Churchill; 1880:262.
2. Gowers W. Optic neuritis in chlorosis. BMJ 1881;1:796.
3. Lubeck M. Papilledema caused by iron-deficiency anemia. Trans Am Acad Ophthalmol Otol 1959;63:306–309.
4. Schwaber J, Blumberg A. Papilledema associated with blood loss anemia. Ann Intern Med 1961;55:1004–1007.
5. Corbett JJ, Digre K. Idiopathic intracranial hypertension: an answer to, "the chicken or the egg?" Neurology 2002;58:5–6.

Neuro *Images*

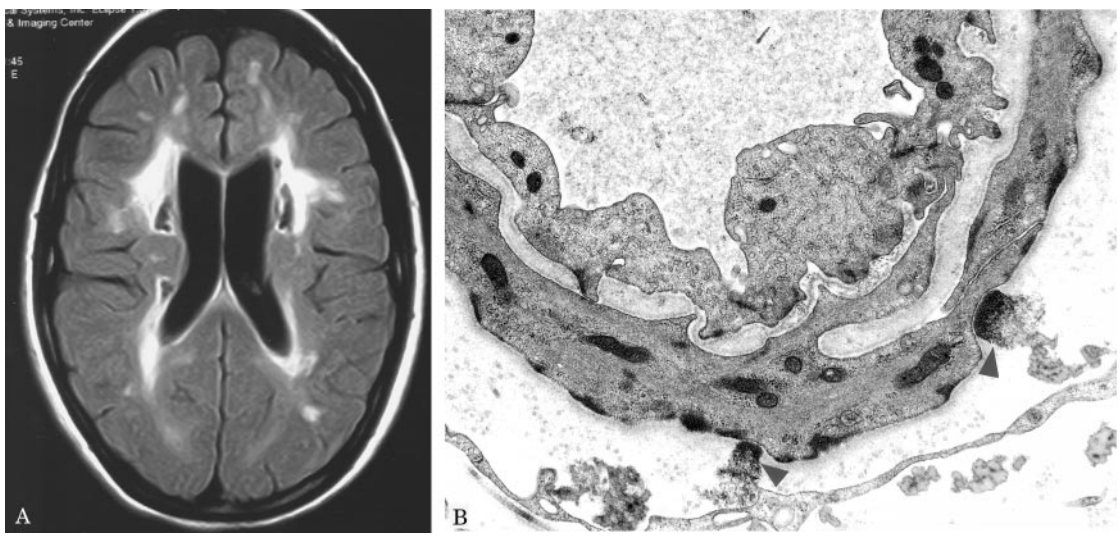

Figure. (A) FLAIR MR image showing extensive white matter signal hyperintensities in temporopolar regions and cystic changes. (B) Electron microscopy of skin biopsy shows electron dense granular deposits (arrows) in the basal lamina surrounding vascular smooth muscle cells.

Skin biopsy findings in CADASIL

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A 48-year-old woman was seen on neurology consultation at Baystate Medical Center for gait instability and abnormal CT scan of the brain. She had a medical history of stroke 5 years prior consisting of left-sided weakness with no residual symptoms. Her history also included seizures in the remote past, migraine without aura, and depression. Neurologic examination was notable for evidence of cognitive impairment, dysarthric speech, and brisk reflexes. MRI of the brain was obtained. There was diffuse white matter abnormality consisting of hyperintensity on T_2 and fluid-attenuated inversion-recovery (FLAIR) imaging and cystic changes (figure A). The findings were suggestive of cerebral autosomal-dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL).[1] A skin biopsy showed the characteristic granular deposits in the basal lamina of a small blood vessel on electron microscopy (figure B).[2] She was later found to have a *Notch3* gene mutation.

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1. Davous P. CADASIL: a review with proposed diagnostic criteria. European Journal of Neurology 1998;5:219-233.
2. Mayer M, Straube A, Bruening R, et al. Muscle and skin biopsies are a sensitive diagnostic tool in the diagnosis of CADASIL. J Neurol 1999; 246:526-532.

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