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.

From the Neurogenetics Branch (Dr. Taylor), NINDS, National Institutes of Health, Bethesda, MD; Departments of Neurology (Drs. Galetta and Asbury) and Ophthalmology (Dr. Volpe), University of Pennsylvania Medical Center, and the Scheie Eye Institute (Drs. Galetta, Asbury, and Volpe), Philadelphia, PA.

Dr. Taylor is supported by a fellowship from the Howard Hughes Medical Institute.

Received February 22, 2002. Accepted in final form June 6, 2002.

Address correspondence and reprint requests to Dr. J. Paul Taylor, Building 10, Room 3B-11, 10 Center Dr., Bethesda, MD 20892.

Copyright © 2002 by AAN Enterprises, Inc.

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

Brian W. Smith, MD, Jean Henneberry, MD,
Timothy Connolly, BS, Springfield, MA

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.

Address correspondence and reprint requests to Dr. Brian W. Smith, Bay State Medical Center, Department of Neurology, 759 Chestnut St., Springfield, MA 01199-1001.

MRI of the brain was obtained. There was diffuse white matter abnormality consisting of hyperintensity on T₂ 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.

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.

Neurology®

Skin biopsy findings in CADASIL

Brian W. Smith, Jean Henneberry and Timothy Connolly

Neurology 2002;59;961

DOI 10.1212/WNL.59.6.961

This information is current as of September 24, 2002

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/59/6/961.full
References	This article cites 2 articles, 0 of which you can access for free at: http://n.neurology.org/content/59/6/961.full#ref-list-1
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): CADASIL http://n.neurology.org/cgi/collection/cadasil MRI http://n.neurology.org/cgi/collection/mri
Permissions & Licensing	Information about reproducing this article in parts (figures,tables) or in its entirety can be found online at: http://www.neurology.org/about/about_the_journal#permissions
Reprints	Information about ordering reprints can be found online: http://n.neurology.org/subscribers/advertise

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright . All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.