Intracranial high-resolution vessel wall imaging in CADASIL

Eric D. Goldstein, MD, Jennifer J. Majersik, MD, MS, and Scott McNally, MD, PhD

Neurology[®] 2020;94:1040-1041. doi:10.1212/WNL.00000000009588

Correspondence Dr. Goldstein eric.goldstein@hsc.utah.edu

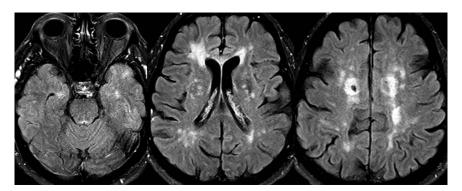
Figure 1 Intracranial vessel wall MRI of cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy



Top row showing T1 precontrast axial images compared with bottom row showing T1 postcontrast mural enhancement of the subcortical and leptomeningeal arteries. Note cortical venous mural enhancement in the bottom right pane. Arrows indicate intramural enhancement.

A 64-year-old man with cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) (heterozygous, c.544C>T; p.Arg182Cys) underwent an intracranial high-resolution black-blood protocoled vessel wall MRI (vwMRI). Intramural patchy gadolinium enhancement in the subcortical and leptomeningeal arteries and veins was noted, consistent with the histopathologic findings of CADASIL (figures 1 and 2).^{1,2} We hypothesize that vwMRI allowed for an in vivo view of the vasculopathy intrinsic to CADASIL. Pending investigation of larger cohorts, this imaging technique may provide a novel mechanism for understanding CADASIL progression and pathogenesis, as well as potentially serving as

Figure 2 Baseline white matter burden



Axial fluid-attenuated inversion recovery sequences show T2 subcortical white matter hyperintensities consistent with cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy. Images provided for comparison.

From the Departments of Neurology (E.D.G., J.J.M.) and Neuroradiology (S.M.), University of Utah, Salt Lake City.

Go to Neurology.org/N for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.

Copyright © 2020 American Academy of Neurology. Unauthorized reproduction of this article is prohibited.

a biomarker in future disease modification trials and aiding in the differential diagnosis for interpreting clinicians.

Study funding

No targeted funding reported.

Disclosure

The authors report no relevant disclosures. Go to Neurology. org/N for full disclosures.

Appendix Authors

Name	Location	Contribution
Eric D.	University of	Design and conceptualization,
Goldstein,	Utah, Salt Lake	drafted manuscript for intellectual
MD	City	content

Appendix (continued) Location Contribution Name Jennifer J. Drafting and revision University of Majersik, MD, Utah, Salt Lake of manuscript for MS City intellectual content Scott University of Drafting and revision McNally, MD, Utah, Salt Lake of manuscript for PhD City intellectual content

References

- Coupland K, Lendahl U, Karlström H. Role of NOTCH3 mutations in the cerebral small vessel disease cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy. Stroke 2018;49:2793–2800.
- Rafalowska J, Fidzianska A, Dziewulska D, Podlecka A, Szpak GM, Kwiecinski H. CADASIL or CADVaSIL? Neuropathology 2004;24:16–20.

The AAN is at Your Side

When you're in the office, the AAN is at your side. The AAN is your #1 resource to support you and your care team. Whether it's resources to help you and your staff provide the best care for your patients, ensure proper reimbursement, or maximize practice performance, the AAN is at your side. Access these resources today at *AAN.com/view/practiceresources*.

Visit the Neurology® Resident & Fellow Website

Click on Residents & Fellows tab at Neurology.org.

Now offering:

- + $\mathit{Neurology}^{\texttt{B}}$ Resident & Fellow Editorial team information
- "Search by subcategory" option
- E-pearl of the Week
- RSS Feeds
- Direct links to Continuum®, Career Planning, and AAN Resident & Fellow pages
- Recently published Resident & Fellow articles
- Podcast descriptions
- Blogs by Editors and Resident & Fellow team members
 - Find Neurology® Residents & Fellows Section on Facebook: facebook.com/AANResidentsAndFellows
 - y Follow *Neurology*[®] on Twitter: @GreenJournal #NeurologyRF
 - (O) Find Neurology® Residents & Fellows Section on Instagram: @aanbrain #NeurologyRF

Neurology.org/N

Copyright © 2020 American Academy of Neurology. Unauthorized reproduction of this article is prohibited.