

Mystery Case: Cerebral amyloid angiopathy–related inflammation

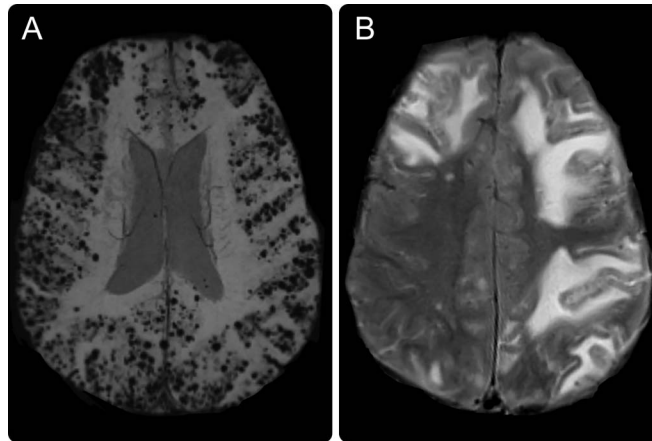


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Figure Cerebral amyloid angiopathy–related inflammation



MRI demonstrates innumerable lobar microhemorrhages on susceptibility-weighted imaging sequence (A) and asymmetric subcortical T2 hyperintensities on fluid-attenuated inversion recovery sequence (B).

A 59-year-old man was arrested for unruly public behavior. He was subsequently found unresponsive with bilateral upper extremity extensor posturing and lower extremity triple flexion when stimulated. Over the next 48 hours, he awoke and became belligerent. MRI revealed innumerable cortical microhemorrhages on susceptibility-weighted imaging (figure, A) and asymmetric subcortical T2 hyperintensities (figure, B) consistent with cerebral amyloid angiopathy–related inflammation (CAA-ri).¹ With corticosteroid treatment, he rapidly became calm and conversant, returning to near his premorbid baseline with only mild residual confusion within 1 week. CAA-ri is an uncommon, possibly autoimmune² variant of CAA characterized by cognitive changes, seizures, headaches, and often, responsiveness to immunosuppressive therapy.¹

AUTHOR CONTRIBUTIONS

Dr. Berkowitz conceived of the manuscript, drafted the initial manuscript, revised the manuscript, created the figure, and revised the accompanying PowerPoint. Dr. Baker revised the manuscript. Dr. Miller revised the manuscript and developed the accompanying PowerPoint. Dr. Greenberg drafted the initial manuscript and revised the manuscript.

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DISCLOSURE

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1. Kinnecom C, Lev MH, Wendell L, et al. Course of cerebral amyloid angiopathy–related inflammation. *Neurology* 2007;68:1411–1416.
2. Piazza F, Greenberg SM, Savoirdo M, et al. Anti-amyloid β autoantibodies in cerebral amyloid angiopathy-related inflammation: implications for amyloid-modifying therapies. *Ann Neurol* 2013;73:449–458.

MYSTERY CASE RESPONSES

The Mystery Case series was initiated by the *Neurology*[®] Resident & Fellow Section to develop the clinical reasoning skills of trainees. Residency programs, medical student preceptors, and individuals were invited to use this Mystery Case as an educational tool.

From the Department of Neurology, Brigham and Women's Hospital (A.L.B., J.M.B., J.J.M.), and the Department of Neurology, Massachusetts General Hospital (J.M.B., J.J.M., S.M.G.), Harvard Medical School, Boston.

Responses were solicited through a group e-mail sent to the American Academy of Neurology Consortium of Neurology Residents and Fellows and through social media.

All the responses we received came from individuals rather than groups. Sixty percent of respondents correctly stated that the lobar microhemorrhages seen on the patient's susceptibility-weighted imaging sequence and the subcortical hyperintensities on the

T2 sequence are most consistent with CAA-ri. A total of 47% of respondents correctly identified corticosteroid therapy as the treatment of choice for this condition.

This case highlights the importance of MRI in diagnosing this rare, but often treatable, variant of CAA.

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This Week's *Neurology*[®] Podcast



Mystery Case: Cerebral amyloid angiopathy-related inflammation (See p. 1678)

This podcast begins and closes with Dr. Robert Gross, Editor-in-Chief, briefly discussing highlighted articles from the October 28, 2014, issue of *Neurology*. In the second segment, Dr. Prachi Mehndiratta talks with Dr. Aaron Berkowitz about his paper on cerebral amyloid angiopathy-related inflammation. Dr. Adam Numis then reads the e-Pearl of the week about nasociliary reflex. In the next part of the podcast, Dr. Chenjie Xia focuses her interview with Dr. Karen Roos on neurocysticercosis, its common manifestations, investigations and treatment.

Disclosures can be found at Neurology.org.

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