Amyloid spells

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Figure MRI and EEG findings

Brain T2-fast low-angle shot MRI showing superficial siderosis (B-arrowhead) with well-defined, homogeneous, hypointense, curvilinear cerebrocortical signal intensity, and multiple cortical microbleeds with gradient echo susceptibility foci (B-arrows).^{1,2} These findings were not visualized on T2-fluid-attenuated inversion recovery MRI (A), demonstrating the importance of blood-sensitive MRI sequences in the investigation of CAA.^{3,4} EEGs captured during TNFE (C and D) were normal. CAA = cerebral amyloid angiopathy; TNFE = transient focal neurologic episode.

A 66-year-old woman presented with a history of recurrent, transient neurologic symptoms including limb shaking, focal paresthesias, focal weakness, dysphasia, and unresponsiveness. The episodes showed no preference for time of day, lasted less than 6 minutes, and occurred less than monthly in small clusters, increasing in frequency slowly over 2 years. She lived independently, denied cognitive or other complaints, and had no history of smoking, hypertension, hyperlipidemia, coronary atherosclerosis or diabetes mellitus, or family history of stroke. Clinical examination was unremarkable. Cognition was grossly intact but not formally tested. Differential diagnosis included transient ischemic attacks (TIAs), seizures, and psychogenic nonepileptic seizures (PNESs). MRI of the brain was consistent with probable cerebral amyloid angiopathy (CAA) (figure) with cortical microbleeds and superficial siderosis.^{1,2} Two habitual transient focal neurologic episodes (TFNEs) were captured on video EEG (video 1).

TFNEs, also called amyloid spells, occur commonly in CAA as brief, recurrent, stereotypical episodes including both positive seizure-like and negative TIA-like phenomena.^{3,4} CAA is characterized by progressive deposition of amyloid- β in cortical and leptomeningeal vessel walls. Risk factors include aging, Alzheimer disease, and Apo ϵ 2 or Apo ϵ 4 carrier status, but not

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other stroke risk factors.¹⁻³ Apoε4 carriers also have increased risk of Alzheimer disease.^{1,2} No treatment is available for CAA or TFNE.² Absence of epileptiform activity in TFNE may lead to the diagnosis of PNES, but neither TFNE nor PNES responds to antiseizure medications.³ TFNE may also be misdiagnosed as TIA, but are predictive of elevated risk of symptomatic cortical hemorrhage, and antithrombotic medications should be used cautiously.²⁻⁴

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Disclosure

S. S. Cooperman reports no disclosures. A. K. Shah: performed video-EEG monitoring in his clinical practice at Detroit Medical Center and currently at Carilion Clinic, serves as an editorial board member of *Epilepsia*, serves on the advisory council for the ROW Foundation, has received funding from Lundbeck, and has participated in clinical trials sponsored by UCB, Eisai, and Sunovion. K. Rajamani: serves as site principal investigator for the ARCADIA study at Detroit Medical Center. Full disclosure form information provided by the authors is available with the full text of this article at Neurology.org/cp.

Appendix 1 Author contributions

Name	Location	Role	Contribution
Sharon S. Cooperman, MD, PhD	Detroit Medical Center, Detroit	Author	Analyzed and interpreted the data and drafted and revised the manuscript for intellectual content.
Aashit K. Shah, MD	Detroit Medical Center/Wayne State University, Department of Neurology, Comprehensive Epilepsy Program, Detroit	Author	Acquisition of data; analyzed and interpreted the data; and drafted and revised the manuscript for intellectual content.
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References

- Charidimou A, Martinez-Ramirez S, Shoamanesh A, et al. Cerebral Amyloid Angiopathy with and without hemorrhage: evidence for different disease phenotypes. *Neurology* 2015;84:1206–1212.
- Yamada M Cerebral amyloid angiopathy: emerging concepts. *J Stroke* 2015;17:17–30.
- Charidimou A, Peeters A, Fox Z, et al. Spectrum of transient focal neurological episodes in cerebral amyloid angiopathy: multicentre magnetic resonance imaging cohort study and meta-analysis. *Stroke* 2012;43:2324–2330.
- Nadarajan V, Perry RJ, Johnson J, Werring DJ. Transient ischaemic attacks: mimics and chameleons. *Pract Neurol* 2014;14:23–31.