Teaching NeuroImages:

Cerebral amyloid angiopathy–related inflammation presenting with isolated leptomeningitis

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Figure 1 Brain MRI

T1-weighted images postgadolinium (A) demonstrate leptomeningeal enhancement of the bilateral temporal, parietal, and occipital lobes (arrowheads), with associated nonsuppression of sulcal CSF (arrows) on fluid-attenuated inversion recovery (B). No microhemorrhages or areas of restricted diffusion were seen on gradient echo or diffusion-weighted images (not shown).

A 69-year-old woman presented with 1 month of rapid cognitive decline preceded by 1 year of very mild memory changes. Examination showed impaired attention, logopenic aphasia, anosognosia, simultagnosia, graphesthesia, and apraxia. Neuroimaging demonstrated leptomeningeal enhancement without significant parenchymal lesions or cerebral microbleeds (figure 1). A lymphocytic pleocytosis (15 nucleated cells/ μ L) was noted on CSF analysis. Extensive diagnostic testing was inconclusive, including flow cytometry and cytology, conventional angiography, body CT, and PET. Cerebral amyloid angiopathy–related inflammation (CAA-RI) was confirmed on brain biopsy (figure 2). CAA-RI rarely presents with isolated leptomeningeal enhancement and can be a challenging diagnosis.^{1,2}

AUTHOR CONTRIBUTIONS

Dr. Kang: design and conceptualization, obtaining images, drafting, revision and final submission of manuscript. Dr. Bucelli: design and conceptualization, revision of manuscript. Dr. Ferguson: histopathologic analysis, obtaining images, revision of manuscript. Dr. Corbo: histopathologic analysis, obtaining images, revision of manuscript. Dr. Kim: design and conceptualization, revision of manuscript.

Dr. Day: design and conceptualization, obtaining images, revision of manuscript.

STUDY FUNDING

No targeted funding reported.

DISCLOSURE

P. Kang receives research funding from the Washington University NIH CTSA Grant #UL1 TR000448. R. Bucelli receives a recurring annual financial gift from a patient's family for Parsonage-Turner syndrome research. C. Ferguson, J. Corbo, and A. Kim report no disclosures relevant to the manuscript. G. Day is involved in research supported by an in-kind gift of radiopharmaceuticals from Avid Radiopharmaceuticals, and holds stocks (>\$10,000) in ANI Pharmaceuticals (a generic pharmaceutical company). He is a recipient of an American Brain Foundation Clinical Research Training Fellowship. Go to Neurology.org for full disclosures.

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(A) Parenchymal gliosis and perivascular inflammation without overt angiitis. (B) β -Amyloid immunostain demonstrates amyloid angiopathy (arrows) and plaque (arrowhead). (C) CD3 and CD68 immunostains show no definite angioinvasion by immune cells.

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Neurology 2017;89:e66-e67

DOI 10.1212/WNL.0000000000004218

This information is current as of August 7, 2017

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