

Cerebral Amyloid Angiopathy Related inflammation

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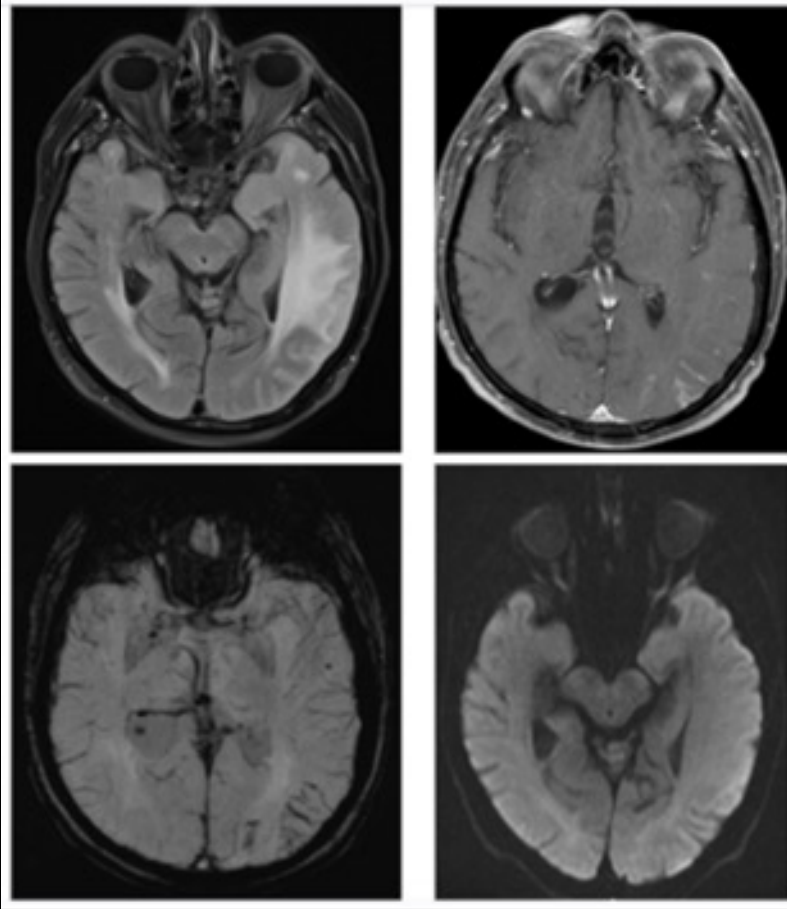


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Clinical Presentation

- A 78 year old male presented to ED with generalized tonic-clonic seizure.

Imaging Discussion



MRI brain imaging reveals extensive T2/FLAIR white matter hyperintensities most severe in left temporoccipital lobes with extension into cortex. There was associated leptomeningeal hemosiderin staining and leptomeningeal enhancement in left temporoccipital lobes as well as mild mass effect. There was facilitated diffusion associated with affected areas. There were scattered punctate foci of hemosiderin staining in peripheral distribution in the left temporal lobe and right occipital lobe.

Management and Outcome

- Patient underwent left temporal mini-craniotomy for biopsy of meninges and gray and white matter. Pathology revealed Beta4-amyloid immunostain positivity in frequent leptomeningeal and superficial cortical vessel walls, some of which appeared to be only partially intact. It also highlighted frequent diffuse cortical plaques. PAS stain is positive in the thickened vessel walls.
- In summary, the morphologic and immunohistochemical findings supported the diagnosis of cerebral amyloid angiopathy with inflammatory reaction (also referred to as the inflammatory form of CAA or CAA-related inflammation).
- Follow up MR brain imaging showed significant improvement after 4 mg/day steroid therapy.

Take Home Points

- Cerebral amyloid angiopathy related inflammation is an uncommon presentation of the disease which mimics a variety of other diseases such as encephalitis, demyelination and tumor. Key imaging findings include chronic subarachnoid hemorrhages in peripheral and leptomeningeal distribution as well as leptomeningeal enhancement. Immunosuppression to improve inflammation is the treatment of choice.

References

- Chao CP, Kotsenas AL, Broderick DF. Cerebral amyloid angiopathy: CT and MR imaging findings. *Radiographics*. 2006 Sep-Oct;26(5):1517-31. doi: 10.1148/rg.265055090. PMID: 16973779.