

Inflammatory CAA protocol

Diagnosis and Treatment of cerebral amyloid angiopathy–related inflammation (CAA-RI)

Prior to making any medical decisions, please view our [disclaimer](#).

Diagnosis

Definite CAA-RI : Tissue-based evidence (autopsy or brain biopsy) of CAA-related inflammation in the cerebral vasculature.

- CAA identified by Congo Red staining on pathological samples
- Evidence of perivascular or vascular inflammatory infiltrate associated with CAA in the blood vessels

Probable CAA-RI : Clinical and neuroimaging evidence strongly suggestive of CAA-related inflammation.

- Age 40 years or older
- At least one of the following clinical features: headache, mental status or behavioral change, focal neurological symptoms/signs, or seizures
- Pattern of hemorrhagic lesions consistent with probable CAA (multiple exclusively lobar-based hemorrhages, microbleeds, or superficial siderosis)
- MRI T2-weighted or FLAIR sequence showing unifocal or multifocal hyperintensities (not due to intracerebral hemorrhage) that are asymmetric and extend to the immediately subcortical white matter or sulci
- CSF with mildly elevated protein or presence of leukocytes may be seen in some cases but normal CSF does not rule out the diagnosis.

Treatment

- Five-day course of high-dose corticosteroids (500mg-1g / day x 5 days) as an inpatient or outpatient with rapid steroid taper. The treating clinician may also wish to consider the following if appropriate:
 - Ensure adequate calcium (1200-1500mg/day), and vitamin D (400-800 units/day). (Two-three caltrate tabs daily)
 - Yearly bone density measurements
 - Vitamin D level every 6 months. Supplement if below 30ng/ml
- Re-imaging and clinical follow-up approximately 3-4 weeks after initial treatment to monitor radiographic changes and clinical status. Radiographic changes may precede or occur in the absence of clinical change. In these cases, other reasons for clinical impairment (progression of underlying Alzheimer's or CAA pathology) should be sought.
- Consider repeat lumbar puncture on follow-up, especially in cases with baseline CSF abnormalities
- If there is no radiographic and/or clinical change, consider cyclophosphamide, 1-2 mg/kg/day PO for 2 weeks [consideration of brain biopsy to confirm diagnosis prior to initiating treatment is recommended]
- Other immunosuppressive agents may be further alternatives in high refractory cases, although clinical experience with these agents remain very limited

Authoring Information

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