Cerebral amyloid angiopathy related inflammation

Cerebral amyloid angiopathy-related inflammation (CAA-ri), a rare and treatable variant of cerebral amyloid angiopathy, lacks specific imaging and clinical features, and requires invasive brain biopsy to confirm the diagnosis.

Kusakabe et al., report the case of a patient with nonconvulsive status epilepticus (NCSE) caused by CAA-ri in the right occipital lobe.

A 78-year-old man with a history of hypertension and rheumatoid arthritis was admitted to our hospital following an episode of seizures. CT scan showed a low-attenuating subcortical lesion in the right occipital lobe. MRI revealed the lesion as hypointense on T1-weighted imaging (WI) and hyperintense on T2-WI, showing no enhancement on T1-WI contrast-enhanced with gadolinium. In addition, T2*-weighted gradient-recalled echo (T2*-GRE) and susceptibility-weighted imaging (SWI) revealed extensive cortical microbleeds. Biopsy to determine the exact diagnosis revealed histological findings of reactive changes and perivascular inflammatory infiltration associated with amyloid deposition in vessel walls. These findings were consistent with CAA-ri. Corticosteroid therapy with dexamethasone was initiated for a short period as a diagnostic and therapeutic maneuver, resulting in marked reductions in the lesion.

CAA is generally associated with intracerebral hemorrhage, dementia, and small cerebral infarctions in the elderly population, but in a small proportion of cases is related to inflammatory responses to vascular deposits of $A\beta$, as so-called CAA-ri.

CAA-ri should be considered among the differential diagnoses for causes of unprovoked seizure onset in elderly individuals, when associated with petechial hemorrhages on T2*-GRE and SWI sequences on MRI 1 .

1)

Kusakabe K, Inoue A, Matsumoto S, Kurata M, Kitazawa R, Watanabe H, Kunieda T. Cerebral amyloid angiopathy-related inflammation with epilepsy mimicking a presentation of brain tumor: A case report and review of the literature. Int J Surg Case Rep. 2018 May 29;48:95-100. doi: 10.1016/j.ijscr.2018.05.016. [Epub ahead of print] PubMed PMID: 29883923.

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