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Cerebral Amyloid Angiopathy as an Etiology for Cortical Superficial Siderosis: An Unproven Hypothesis

We read with great interest a recent article by Inoue et al[1] on the diagnostic significance of cortical superficial siderosis (cSS) for Alzheimer disease in patients with cognitive impairment. The article focused on presymptomatic cases of cSS diagnosed by MR imaging.

The major finding of the article was that cSS was associated with a lobar location of microbleeds (MBs) and may be an initial radiologic finding of cerebral amyloid angiopathy (CAA) in patients with cognitive impairment. Lobar location includes both the cortical gray matter and the subcortical white matter. The imaging manifestations of hemosiderin deposition from cSS and MBs from CAA can be indistinguishable on gradient-echo T2-weighted images,[2] especially when MBs are seen on the surface of the cerebral cortex. There is even a suggestion that CAA can be an underlying cause of cSS.[3] In the current study, there was 72% correspondence between the location of cSS and that of MBs. The definition of cSS was only based on the shape of the signal abnormality on SWI (ie, linear). Because there are currently no widely recognized criteria to distinguish hemosiderin deposition from MBs on imaging, it may be helpful to show interobserver variability in the assignment of individual lesions to ensure agreement on the nature of the hypointensity seen on T2-weighted MR imaging.

Although its detection has increased with the advances in MR imaging technology, cSS is still a rare disease.[4] The most accepted hypothesis for its etiology has been chronic iron deposition in neuronal tissues associated with CSF.[5] Chronic bleeding into the subarachnoid space of the brain releases erythrocytes into the CSF. The chronic bleeding source can be a result of past brain surgery or CNS trauma.[5] Less common bleeding sources include CSF cavity lesions, tumors, vascular malformations, and so forth.[4] The authors stated that cSS related to previous symptomatic subarachnoid hemorrhage, traumatic subdural hematoma, or intracranial surgery was not included, but they did not provide information on the number of patients excluded. According to the literature, the source of bleeding was never found in as many as half of all described cases.[4] Even for the 12 patients included in the analysis, it is possible that they still had an occult source of bleeding. Consequently, the relationship between cSS and CAA may be either overestimated or underestimated in the studied cohort, depending on how many patients were excluded due to a known bleeding source.

In conclusion, the pathogenesis of cSS from CAA is still an unproven hypothesis. An unidentified bleeding source may account for cSS in the studied cohort instead of CAA.

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