Superficial hemosiderosis of central nervous system (CNS) in cerebral amyloid angiopathy (CAA).

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Learning objectives

In this educational exhibit we want to underline the role of this microangiopathic disease as a potential pathomechanism for superficial hemosiderosis (SS) and to illustrate the related neuroimaging features in SS of the central nervous system as a new prospective MR diagnostic marker of CAA.

Background

CAA is an important cause of spontaneous cerebral hemorrhage and occurs more frequently in elderly normotensive subjects. It is characterized by the deposition of #-amyloid protein in the media and adventitia of small and medium-sized vessels of the cerebral cortex, subcortex, and leptomeninges. Both sporadic and hereditary forms may occur; the latter generally demonstrate autosomal dominant transmission, is rare, and occurs at a younger age than the sporadic form that is more common in the elderly and increases in both prevalence and severity with increasing age.

More common sporadic, age-related form of CAA remains an underrecognized cause of cerebrovascular disease, clinically as well as at imaging, in part because many patients are asymptomatic. It manifests at imaging as part or all of a spectrum of findings including acute or chronic cortical-subcortical cerebral hemorrhages, leukencephalopathy, and atrophy.

The clinical differentiation of CAA-related vs non-CAA-related symptomatology may be very difficult and unreliable, as there is significant overlap in diseases that result in acute neurologic deficits, TIA-like symptoms, and dementia.

As a tool to both improve and standardize the diagnosis of CAA, in the mid 1990s were developed the Boston criteria that specify four diagnostic categories: 1) definite CAA, 2) probable CAA with supporting pathologic evidence, 3) probable CAA, and 4) possible CAA, depending on a combination of clinical, imaging, and histologic data.

According to these widely used criteria the definite diagnosis still requires the histopathologic demonstration of vascular amyloid as a criterion standard and it can be considered "probable" in subjects 55 ys old or older with clinical suspicion and MRI evidence of multifocal parenchymal hemorrhagic foci with no other clinical or radiologic cause of hemorrhage.

Thus, further improvement of the sensitivity and specificity of imaging methods would be desirable to define additional diagnostic features especially useful in asymptomatic patients in order to limit the use of invasive diagnostic procedures.
With regard to that, several Authors report on patients with histologically proved CAA in which observed superficial cortical and/or subarachnoid hemosiderosis, underlining the role of the disease as an important cause of SS.

We describe two cases of occult supratentorial superficial cortical hemosiderosis with evidence of parenchymal micro and macrohemorrhages with a distribution suitable with CAA, ruling out other causes.

**Findings and procedure details**

Two consecutive male patients were admitted to our institution between 2009 and 2011 for undergoing brain MR examination.

MRI was performed at 1.5 Tesla MR scanner, including GRE T2w and Fluid-Attenuated Inversion Recovery (FLAIR) sequences.

The first patient 63 yo without vascular risk factors presented with partial seizures and progressive dementia whereas the second 84 yo, totally asymptomatic, underwent MR as a consequence of the occasional evidence of parenchymal bleedings during MR examination of the temporo-mandibular joints.

Both brain MR studies showed multifocal supratentorial hemosiderinic foci in the cortical-subcortical junction in both hemispheres (fig. 1) and linear gyriform low signal deposits located predominantly in the cortical superficial-subpial space, mostly over the cerebral convexities (fig. 2).

The second patient's scan furtherly showed two cortical-subcortical 2 cm subacute (fig. 3) and chronic (fig. 4) hematomas.

The first patient was readmitted 5 weeks after MR examination with an acute subarachnoid haemorrhage (SAH) as evidenced by acute headache and subarachnoid blood on CT over the right parietal area. (fig. 5)

In both patients there was no involvement of subtentorial and deep structures (fig. 6) and there was no other cause of bleeding both in the MR scans and in the clinical history.

**Images for this section:**
Fig. 1: Axial GRE T2-w brain MR images from patients 1(a) and 2(b). Both images depict multiple foci of signal loss in cortical-subcortical locations that are consistent with chronic microhemorrhages, a finding highly suggestive of CAA.

Fig. 2: Axial (a-c) and coronal (b) GRE T2-w brain MR images from patients 1(a-b) and 2(c): gyriform low signal linear deposits located bilaterally over the cerebral convexities corresponding to superficial cortical hemosiderosis. Coronal image also shows a few cortical microhemorrhages.
Fig. 3: Axial T2-w (a), FLAIR (b), GRE T2-w and Sagittal T1-w (d) brain MR images from patient 1. Scans show a small ovoid, left-sided frontal cortical-subcortical subacute macrohemorrhage. A symmetric periventricular leukoencephalopathy is also apparent on axial images.
Fig. 4: Axial GRE T2-w brain MR image from the same patients 1 at superior level shows a small area of encephalomalacia and hemosiderin from prior macrohemorrhage in the right frontal lobe.
Fig. 5: Axial nonenhanced CT scans at two levels from patient 1 five weeks after MR examination. Image show a subtle right-sided parietal sulcal hyperattenuation favoring minimal subarachnoid hemorrhage.
**Fig. 6:** Axial GRE T2-w brain MR images from patients 1(a) and 2(b) doesn't show in the infratentorial compartment hemosiderin deposition along the cerebellar folia and pons. SS in CAA is typically found in a supratentorial distribution over the cerebral convexities whereas the "classic" SS mainly affects brain stem and posterior fossa.
Conclusion

The most important clinical presentation of CAA is spontaneous intracerebral macrohemorrhages or microbleeds (ICH), but identifying CAA as the cause of ICH is challenging.

The Boston criteria for CAA-related hemorrhage can be used to attribute an ICH with increasing certainty to CAA by using clinical data, imaging signs and, if available, histopathologic findings. The definite diagnosis requires a full postmortem examination.

In recent years several case reports also found superficial siderosis in patients with CAA with the authors providing a thorough review of neuroimaging as well as detailing important underlying causes of this phenomenon and listing CAA as a potential pathomechanism for SS.

CAA cases with SS lack the typical clinical presentation associated with "classic" SS, which are progressive gait ataxia with cerebellar dysarthria and sensorineural hearing loss, namely cerebellar and brainstem signs, but patients often present with headache, seizures, and cognitive impairment, most probably due to the characteristic localization of SS in patients with CAA: while the "classic" SS mainly affects brain stem and posterior fossa, SS in CAA is typically found in a supratentorial distribution over the cerebral convexities and only exceptionally occurs in the infratentorial compartment.

Superficial siderosis occurs with higher prevalence in CAA being rare in non-CAA forms of intracerebral hemorrhages.

Yet SS has been reported as the only bleeding manifestation in some patients with CAA, especially if they present with transient clinical manifestations, e.g., seizures.

In our opinion, CAA should be thoroughly considered as a cause of SS, especially in older patients with isolated supratentorial SS and an atypical clinical presentation.

Considering supratentorial cortical superficial and/or subarachnoid hemosiderosis, in the presence of multiple foci of microhemorrhages in cortical-subcortical locations, as another hemorrhagic manifestation of CAA, can result in a diagnostic upgrading in patients with CAA not lowering the specificity of the Boston criteria for CAA-related hemorrhage.

Nevertheless the sensitivity and specificity of SS as an in vivo noninvasive diagnostic MR imaging marker of CAA have to be validated in large prospective studies, and longitudinal follow-up studies of patients with SS should be performed.
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References


