HEMORRHAGE IN THE NORMOTENSIVE ELDERLY: THE FACES OF AMYLOID ANGIOPATHY

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Purpose

To describe both the typical and atypical features of Cerebral Amyloid Angiopathy (CAA) on MRI and to review the imaging protocol to reach a correct diagnosis.

Methods and Materials

Since 2001 we retrospectively reviewed the MRI findings of 200 patients with a probable or probable with supporting pathologic evidence Cerebral Amyloid Angiopathy based on the Boston Criteria.

Apart from the standard cranial protocol, T2*-weighted gradient-echo (T2* GRE) and more recently susceptibility-weighted imaging (SWI) were systematically performed. The use of contrast was limited to the cases with atypical MRI findings.

Results

The most frequent Symptoms at presentation were: progressive dementia (29%), sudden neurologic deficit due to CAA-related hemorrhages (28%), difficulty walking (14%) and rapid cognitive decline (10%, one of them finally diagnosed as Creutzfeldt-Jakob disease).

MR findings were: isolated cortical-subcortical microhemorrhages (21%); or associated with periventricular leukoencephalopathy (36%), with lobar macrohemorrhages (36%), or less frequently, with asymmetric lobar leukoencephalopathy (12%, two of them with patchy leptomeningeal contrast enhancement). Only one case of amyloidoma was reported.

Cortical-subcortical microhemorrhages were predominantly located supratentorially (80%), with no lobar preference; or both, supra and infratentorially (20%).

CAA is an important but underrecognized cause of cerebrovascular disorders that predominantly affects normotensive elderly patients (30%), and increases in both prevalence and severity with age, although many patients are asymptomatic.
CAA results from the impaired elimination and accumulation of soluble and insoluble 
#-amyloid peptide and its deposition within the media and adventitia of cerebral 
arterioles (and less frequently, veins), mainly in small and medium-sized arteries of the 
leptomeninges and superficial cortex.

It ranges in severity from asymptomatic amyloid deposition in otherwise normal cerebral 
vessels to complete replacement and breakdown of the cerebrovascular wall.

Vessels loose elasticity and become fragile, more easily damaged, predisposing the 
patient to repeated episodes of blood vessel leakage in and around the arteriole vessel 
wall, or frank hemorrhage. The amyloid deposition and fibrinoid degeneration can 
contribute to occlusion of small arteries and microaneurysm formation and cause small 
cortical and subcortical infarcts and hemorrhages. Some evidence suggests that CAA 
has a role in some anticoagulant- and thrombolytic-related hemorrhages.

CAA can be Sporadic or Hereditary (Dutch or Icelandic type); Idiopathic or Secondary 
(dialysis-related amyloidosis, …).

CAA is particulary common in association with Alzheimer disease (80%) and Down 
syndrome. Other associations are Kuru, Creutzfeldt-Jakob disease, plasmacytoma…

CAA may coexist with Hypertension.

Typical clinical presentations include sudden neurologic deficit related to acute 
Intracerebral Hemorrhage (ICH), TIA, seizures or dementia.

The BOSTON CRITERIA were developed to standarize the diagnosis of CAA. There are 
four diagnostic categories:

**Definite CAA** (postmortem).

**Probable CAA with supporting pathologic evidence** (after biopsy, generally of ICH).

**Probable CAA** (>60 years, with multiple cortical-subcortical hemorrhages with no other 
clinical/radiological causes).

**Possible CAA** (>60 years, with single cortical-subcortical hemorrhage or hemorrhage in 
an atypical location with no other clinical/radiological causes).

As histologic analysis is often not practical, Magnetic Resonance Imaging is an important 
tool for the diagnosis of these patients (some of them treated with anticoagulant therapy, 
because the use of anticoagulants may result in the enlargement of hemorrhages that 
remained asymptomatic (CASE 6) Fig. 7 on page 11), exclusion of other causes of 
acute cortical-subcortical hemorrhage and assessment of disease progression.
Cerebral amyloid deposition occurs in 3 morphologic varieties: CAA-related hemorrhages, Leukoencephalopathy and Amyloidoma (more rare).

**CAA-RELATED HEMORRHAGES:**

Typically affect the cerebral cortex and cortico-subcortical or lobar regions of the brain, and are absent in brain locations characteristic of hypertensive hemorrhages (putamen, thalamus and brainstem).

They can be large, > 5 mm, (CASE 1) Fig. 1 on page 5 cortical-subcortical microhemorrhages, < 5 mm, (often asymptomatic) or other types of hemorrhages (subarachnoid or subdural hemorrhages). They usually coexist in the same patient.

Conventional gradient-echo (T2* GRE) imaging is a well-established technique for detecting cerebral microhemorrhages, although it has been recently substituted by a new neuroimaging technique, susceptibility-weighted imaging (SWI). It is a technique based on a 3D, velocity-compensated, GE sequence that combines both magnitude information with phase information, based on tissue magnetic susceptibility differences to generate a unique contrast, being more detailed in identifying the microangiopathic changes seen in CAA and a more sensitive diagnostic tool (CASE 2) Fig. 2 on page 6.

**LEUKOENCEPHALOPATHY:**

- **SYMmetric periventricular** (CASE 3) Fig. 3 on page 7 related to chronic hypoperfusion of deep white matter caused by diffuse narrowing of penetrating cortical vessels resulting from #-amyloid deposition in the media and adventitia of their walls.

Sparing the U fibers, usually symmetric, no reversible and usually associated with atrophy.

Associated clinically with dementia.

- **ASymmetric lObAR** (CASE 4) Fig. 4 on page 8 secondary to # amyloid-induced vasculopathy, cerebral amyloid angiopathy related inflammation, due to an inflammatory response to #-amyloid in the walls of blood vessels, with perivascular and intramural inflammatory infiltrates.

Involving the U fibers, with mass effect, asymmetric, it may coexist with leptomeningeal or parenchymal enhancement (CASES 5, 6) Fig. 5 on page 9, Fig. 6 on page 10.

Unusual. Reversible although can recurr.

Associated clinically with a subacute cognitive decline, cognitive and behavioural changes, seizures or focal neurological deficits.
AMYLOIDOMA:

The least common form of amyloid deposition. Tumoral deposition of amyloid, with slow growth and a benign clinical pattern.

It has been described as solitary or multiple supratentorial white matter masses with little or no mass effect. It is not calcified but may be hiperintense in T1-WI and heterogeneous in T2-WI depending on the amount of amyloid deposits.

It shows variable enhancement degree and a characteristic medial extension of the mass up to the lateral ventricular ependyma, with thickening of lateral ventricular wall and fine, irregular, radiating lines at the edge of the tumor indicating the deposition of amyloid along the vessels, a finding that has been observed in pathology specimens (CASE 7) Fig. 8 on page 12.

CAA is untreatable at this time. Management depends on each individual case, paying attention to the reversal of anticoagulation and the prevention of complications.

Images for this section:
Fig. 1: CASE 1: Axial TSE-T2 (a), FLAIR (b) and SWI (c, d) MR images. A 80-year-old woman with rapid cognitive decline. Left cortical hematoma with associated intraventricular and left posterior parafalcaline subdural hemorrhages (arrows), and bilateral cortical microbleeds.
**Fig. 2:** CASE 2: Axial GRE (a, b) and SWI (c, d) MR images. A 78-year-old man with seizures. Multiple bilateral cortical deposits of hemosiderin sparing the basal ganglia (c, d), underestimated in the conventional axial GRE MR images (a, b).
Fig. 3: CASE 3: Axial SWI (a, b) and FLAIR (c, d) MR images. A 66-year-old man with seizures. Multiple bilateral cortical deposits of hemosiderin sparing the basal ganglia (a, b) and symmetric periventricular leukoencephalopathy sparing the U fibers (c, d).
Fig. 4: CASE 4: Axial FLAIR (a, b) and SWI (c, d) MR images. A 76-year-old woman with motor and cognitive decline. Asymmetric lobar leukoencephalopathy with involvement of U fibers and mass effect (a, b). Superficial siderosis from a previous subarachnoid hemorrhage and multiple bilateral cortical microhemorrhages sparing the basal ganglia (c, d).
Fig. 5: CASE 5: Axial GRE (a, b) and contrast-enhanced T1-weighted (c, d) MR images. A 80-year-old woman with seizure and oral commissure deviation. Superficial siderosis from a previous subarachnoid hemorrhage (arrows in a, b), cortical microhemorrhages (arrowheads in b) and left frontal lobe hematoma (circle in b). T1-weighted images with gadolinium show abnormal leptomeningeal enhancement in the right frontal lobe secondary to cerebral amyloid angiopathy related inflammation.
Fig. 6: CASE 6A: Axial GRE (a, b), FLAIR (c) and contrast-enhanced T1-weighted (d) MR images. A 84-year-old man with left hand paresis. Yuxtacortical parietal microbleeds (arrows in a, b), symmetric periventricular leukoencephalopathy and asymmetric lobar parietal leukoencephalopathy (c). T1-weighted images with gadolinium show associated abnormal leptomeningeal enhancement.
**Fig. 7:** CASE 6B: Axial GRE MR images. After anticoagulant treatment for a pulmonary embolism the patient from case 6 suffered a rebleeding in the left parietal lobe (arrow in a) and developed a new intracerebral haematoma extending to ventricles and subarachnoid space (arrows in d).
Fig. 8: CASE 8: Axial FLAIR (a, b), SWI (c, d), and contrast-enhanced T1-weighted (e, f) MR images. A 78-year-old man with progressive cognitive decline. There is a mass around left lateral ventricle and deep white matter of left frontal lobe with hemosiderin-amyloid deposit and associated vasogenic edema. The mass presents an inhomogeneous enhancement and a medial extension up to the lateral ventricle ependyma (arrows in e, f).
Conclusion

Accurate recognition of imaging findings is important in guiding clinical decision making in patients with CAA (particularly with anticoagulant therapy).

SWI (or T2* GRE imaging if SWI is not available) should be included in every MR cranial protocol in patients older than 60 years.

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