Commentary

Cerebral amyloid angiopathy (CAA) is defined as the deposition of ß-amyloid in the walls of cortical and leptomeningeal blood vessels. The most important clinical presentation of CAA is lobar intracerebral hemorrhage (ICH), and cerebral microbleeds (MBs) are a common magnetic resonance imaging feature of CAA. ICH can be often multiple.[1,2]

Identifying CAA as a cause of ICH is challenging. CAA as a cause of ICH often goes undiagnosed and is frequently overlooked.[1] CAA can be clinically suspected in normotensive patients with headache, vomiting, seizures, decreased of consciousness, dementia, recurrent TIA and/or ischemic stroke and/or hemorrhagic stroke. We can diagnose CAA by autopsy and immunohistochemical

methods as a definitive diagnosis of this disease. It is done after death or intravitally using brain biopsy or after neurosurgical brain resection if other disease is suspected.[3]

The diagnosis of CAA can be done using neuroimaging methods such as a CT brain scans, MR imaging with T2-weighted spin-echo sequences, gradient-echo (GRE) sequences (T2*-weighted gradient-echo MR), the susceptibility weighted imaging (SWI), positron emission tomography (PET) imaging with the ß-amyloid-binding compound Pittsburgh Compound B (PiB). MR spectroscopy can also be helpful.[1,2,4] Using neuroimaging methods we can observe often lobar multiple and recurrent hemorrhages, ischemic lesions, leukoaraiosis, and MBs.

The definite diagnosis of CAA can be made postmortem or after biopsy, but having newest neuroimaging methods we can more often make diagnosis of "probable CAA."[3,5]

A set of clinical and neuropathological criteria has been proposed by the Boston group to diagnose the CAA as a definitive, probable with pathology, probable without pathology, and possible.[6] Recent developments of brain imaging techniques more often allows to diagnose the "probable CAA" and "possible CAA" using classic and modified Boston criteria for CAA-related hemorrhage.[2]

I would like to emphasize the role of superficial siderosis (SS) in the diagnosis of CAA. Recently, SS has been described as a potential magnetic resonance marker of CAA.[1,7] CAA may also be associated with evidence of hemosiderin deposition similar to that seen in SS. Superficial cortical hemosiderosis and subarachnoid hemosiderosis are potentially useful new MR imaging criteria to facilitate the noninvasive diagnosis of CAA.[1] SS of the central nervous system results from hemosiderin deposition in subpial layers of the brain. The hemosidrin deposition is a consequence of recurrent and persistent bleeding into the subarachnoid space.[8] Commonly performed investigations during the work-up of SS include MR imaging of the brain and spine, CT myelography, MR angiography, and cerebrospinal angiography. Brain MR imaging is the investigation of choice for the diagnosis of SS. MR imaging has shown that the hemosiderin deposition around the brain is the cause of characteristic hypointensity seen on T2-weighted MR imaging. Gradient-echo T2*-weighted MR images have even a higher sensitivity for hemosiderin deposition.[8]

We should be very cautions with patients suspected for CAA. It is a devastating brain disease with various clinical manifestations.

References

1. Linn J, Herms J, Dichgans M, Brückmann H, Fesl G, Freilinger T, *et al.* Subarachnoid hemosiderosis and superficial cortical hemosiderosis in cerebral amyloid angiopathy. AJNR Am J Neuroradiol 2008;29:184-6.
2. Linn J, Halpin A, Demaerel P, Ruhland J, Giese AD, Dichgans M, *et al.* Prevalence of superficial siderosis in patients with cerebral amyloid angiopathy. Neurology 2010;74:1346-50.
3. Greenberg SM, Vonsattel JPG. Diagnosis of cerebral amyloid angiopathy: Sensitivity and specificity of cortical biopsy. Stroke 1997;28:1418-22.
4. Nandigam RN, Viswanathan A, Delgado P, Skehan ME, Smith EE, Rosand J, *et al.* MR imaging detection of cerebral microbleeds: Effect of susceptibility-weighted imaging, section thickness, and field strength. AJNR Am J Neuroradiol 2009;30:338-43.
5. Vonsattel JP, Myers RH, Hedley-Whyte ET, Ropper AH, Bird ED, Richardson EP. Cerebral amyloid angiopathy without and with cerebral hemorrhages: A comparative histological study. Ann Neurol 1991;30:637-49.
6. Knudsen KA, Rosand J, Karluk D, Greenberg SM. Clinical diagnosis of cerebral amyloid angiopathy: Validation of the Boston criteria. Neurology 2001;56:537-9.
7. Vernooij MW, Ikram MA, Hofmann A, Krestin GP, Breteler MM, van der Lugt A. Superficial siderosis in the general population. Neurology 2009;73:202-5.
8. Kumar N. Neuroimaging in superficial siderosis: An in-depth look. AJNR Am J Neuroradiol 2010;31:5-14.

Tadeusz Mendel

Second Department of Neurology, Institute of Psychiatry and Neurology, Sobieskiego Str. 9, 02-957 Warsaw, Poland,

Address for Correspondence:
Dr. Tadeusz Mendel, Second Department of Neurology, Institute of Psychiatry and Neurology, Sobieskiego Str. 9, 02-957 Warsaw, Poland, E-mail: mendel@ipin.edu.pl

Access this article online	
Quick Response Code:	Website: www.ruralneuropractice.com