

Cerebral amyloid angiopathy

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DESCRIPTION

A 72-year-old man presented with headache, vomiting and diminution of vision. There was no history of smoking, diabetes mellitus, hypertension or use of aspirin or recreational drugs. The general physical examination, pulse, blood pressure, chest and cardiovascular examination were normal. On central nervous system examination, the visual acuity was reduced to perception of light at 6 m and pupils were 3 mm and reacting to light. Tests for cerebellar functions showed dysdiadochokinesia and impaired tandem walking. Blood investigations were normal and the axial MRI of the head disclosed a right occipital haematoma (figure 1) and other changes of probable cerebral amyloid angiopathy (figures 2 and 3).

Cerebral amyloid angiopathy (CAA) accounts for 15% of all non-traumatic intracerebral haemorrhage in persons greater than 60 years.^{1 2} In people above 70 years, it accounts for 50% of all lobar haematomas. CAA and CAA-related intracranial haemorrhage are also common in elderly individuals with Alzheimer's disease and Down syndrome. Histopathologically, the deposition of β -amyloid material occurs in the walls of medium and small arteries mainly in the parietal and occipital lobes.^{1 2} The amyloid deposit in the media can be extensive as to cause necrosis of the vessel wall, microaneurysm formation and lobar haemorrhage.

The Boston Criteria were proposed in 1990 to standardise the definition of CAA.² They comprise of clinical, imaging and pathological criteria. According to these criteria, probable CAA is diagnosed in patients older than 55 years with appropriate clinical history and MRI findings demonstrating multiple haemorrhages limited to lobar, cortical, corticosubcortical region of varying sizes and ages (cerebellar haemorrhages allowed) with no other explanation.

Patients of CAA may present with neuroimaging evidence of small subclinical bleed called microbleeds. Gradient-echo (GRE) MRI sequences can demonstrate small haemorrhagic lesions which are not seen on T1-weighted and T2-weighted images.^{1 2} GRE sequences are broadly divided into coherent and incoherent or spoiled sequences. For detecting microbleeds, T2 GRE images and SWI (susceptibility weighted imaging) are used. SWI are high-resolution, three-dimensional, fully velocity compensated GRE sequences and are currently the gold standard for the evaluation of haemorrhagic lesions. Cerebral microbleeds are not pathognomonic of CAA and can also be detected in 5–23% of older individuals in population-based studies.^{3 4} In the Rotterdam cohort, microbleeds were more prevalent among those on antiplatelet therapy.⁴ Another study has suggested that microbleeds arise from amyloid-based pathology.⁵ Microbleeds also occur in hypertensive microangiopathy in the basal ganglia and thalamus and infective endocarditis generally in anatomic locations different from CAA.⁶ In patients of CAA with primary lobar haematoma, the

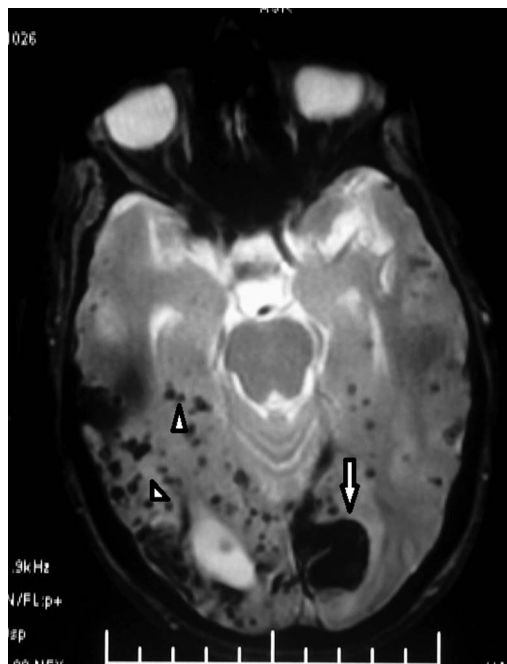


Figure 1 Axial MRI—gradient-echo technique showing multiple microbleeds (white arrowheads) in the cerebral hemispheres and a large left occipital haematoma (arrow)—cerebral amyloid angiopathy.

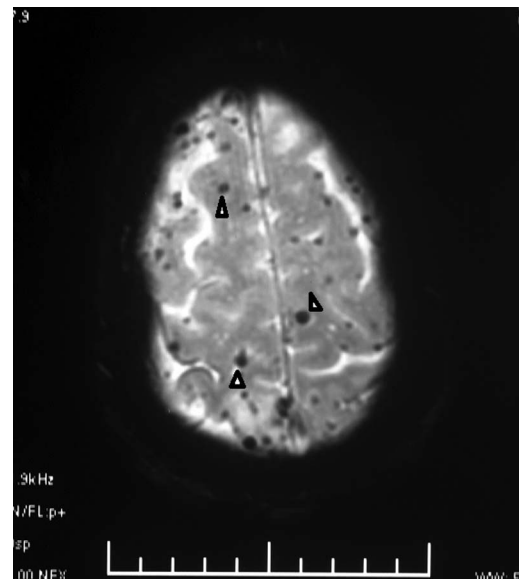


Figure 2 Axial MRI—gradient-echo image showing extensive superficial microbleeds (arrowheads) in both the cerebral hemispheres.



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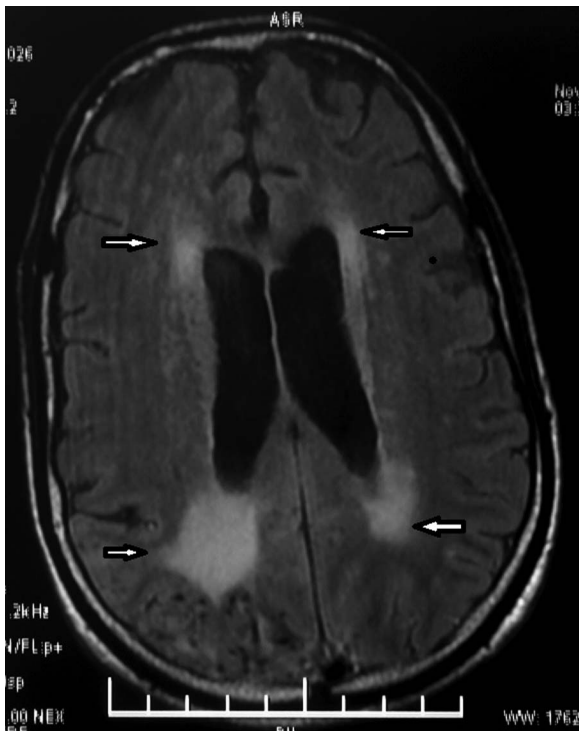


Figure 3 T2-weighted fluid attenuation inversion recovery axial MRI—periventricular white matter intensities signifying small vessel angiopathic changes (arrows).

number of microbleeds may have some prognostic implications as a number above six microbleeds increases the 3-year cumulative risk of recurrent haemorrhage by 51%, causes greater cognitive impairment, loss of function and death.⁷

Learning points

- ▶ Above 70 years of age, cerebral amyloid angiopathy accounts for 50% of all lobar haematomas.
- ▶ T2-weighted gradient-echo MRI sequences are the gold standard of demonstrating cerebral microbleeds.
- ▶ Cerebral microbleeds also occur in patients on antiplatelet and antithrombotic therapy and in cases of infective endocarditis and hypertensive angiopathy.

Contributors NS was involved in preparing the manuscript, gathering and editing the images. SV was involved in editing the images and the manuscript.

Competing interests None.

Patient consent Obtained.

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