Cerebral amyloid angiopathy-related inflammation: a rare disease that needs to be diagnosed

Ioan Paul Voicu,¹ Corrado Tagliati ,² Samanta Sciamanna,³ Giuseppe Lanni²

¹Department of Services, Radiology and MRI Unit, "G. Mazzini" Hospital, Teramo, Italy, ASL 4 Teramo, Teramo, Abruzzo, Italy

²Department of Services, Radiology Unit, "S. Liberatore" Hospital, Atri, Italy, ASL 4 Teramo, Teramo, Abruzzo, Italy

³Department of medicine, Neurology Unit, "G. Mazzini" Hospital, Teramo, Italy, ASL 4 Teramo, Teramo, Abruzzo, Italy

Correspondence to

Dr Corrado Tagliati;
corrado.tagliati@gmail.com

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DESCRIPTION

A 70-year-old man with persistent headache for 2 weeks presented to our emergency department due to sudden clinical worsening characterised by global aphasia, right lateral homonymous hemianopia and cognitive deterioration. A CT scan revealed left parietal and occipital hypodensity consistent with oedema. MRI fluid-attenuated inversion recovery (FLAIR) sequence ([figure 1](#)) confirmed subcortical oedema, associated with effacement of the overlying sulci, without associated diffusion restriction ([figure 2](#)) or contrast material enhancement. T2*-weighted sequence ([figure 3](#)) showed multiple small hypointense cortical and subcortical signal changes, consistent with microhaemorrhages. Based on MRI findings, a diagnosis of cerebral amyloid angiopathy-related inflammation (CAA-ri) was suggested. Therefore, oral steroid therapy was initiated and the patient fully recovered.¹

CAA-ri is a disease subtype of cerebral amyloid angiopathy characterised by rapidly progressive cognitive decline, seizures, headaches, T2-weighted hyperintense MRI lesions, evidence of pre-existing CAA on susceptibility-weighted MRI sequences and neuropathological evidence of CAA-associated vascular inflammation.^{2,3}

Some authors have suggested that clinical and radiological criteria may help diagnose a probable CAA-ri with good sensitivity and specificity, therefore avoiding cerebral biopsy.⁴ The diagnosis of

Figure 2 Apparent diffusion coefficient image showed hyperintense signal, suggesting vasogenic oedema.

Figure 3 T2*-weighted sequence showed multiple small hypointense cortical and subcortical signal changes, consistent with microhaemorrhages.

Figure 1 MRI fluid-attenuated inversion recovery sequence showed subcortical oedema and effacement of the overlying sulci.

probable CAA-ri requires the following six findings: (1) acute or subacute onset of symptoms; (2) 40 years of age or older; (3) presentation with a variable combination of acute or subacute onset of headache, decrease in consciousness or

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Images in...

behavioural change, focal neurological deficits and seizures; (4) MRI with patchy or confluent T2-weighted or FLAIR lesions, usually asymmetric, with or without mass effect and with or without leptomeningeal or parenchymal enhancement; (5) multiple lobar microhaemorrhages and/or recent or past lobar intracerebral haemorrhage; (6) absence of neoplastic, infectious or other cause.⁵

There is usually a favourable response to immunosuppressive therapy. However, some patients may have a fluctuating course or relentless progression. Therefore, CAA-ri diagnosis is very important in clinical practice as many patients respond to immunosuppressive therapy.

Patient's perspective

I am very grateful to the doctors who diagnosed this disease for being able to recover completely simply with corticosteroid therapy.

Learning points

- ▶ A diagnosis of probable cerebral amyloid angiopathy-related inflammation (CAA-ri) may be made on the basis of typical clinical and radiological findings without requiring a biopsy.
- ▶ CAA-ri diagnosis is very important in clinical practice as many patients respond to immunosuppressive therapy.

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ORCID iD

Corrado Tagliati <http://orcid.org/0000-0002-4630-1090>

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