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CAA-RELATED INFLAMMATION: AN ELUSIVE DIAGNOSIS

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Cerebral amyloid angiopathy (CAA) is characterised by deposition of β -amyloid in the vessel walls of leptomeningeal and small cortical arteries. It commonly presents as intracerebral lobar haemorrhage and is associated with dementia. However, rarely it can present as acute inflammation.

We describe the cases of three women who presented acutely with seizures, focal neurological deficits and cognitive dysfunction. T2-weighted MRI revealed white matter hyper-intensity suggestive of inflammation. Gradient echo found multiple microhaemorrhages within the inflamed brain parenchyma in two patients, but was absent initially in one patient. Follow-up imaging on this patient revealed new microbleeds. A presumptive diagnosis of CAA-related inflammation was made. After the initiation of high-dose steroids, rapid dramatic clinical and radiological improvement was seen in all three patients.

CAA is a complex condition lacking formal standardised criteria. Our experience highlights the importance of considering cerebral amyloid angiopathy-related inflammation, a rare subtype of CAA, in patients with parenchymal inflammation on MRI even in the absence of intracerebral haemorrhage. The MRI-based gradient echo sequence is especially useful here. Steroid treatment is highly effective, but long term immunosuppression may be required with recurrent inflammation.