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# Steroid-responsive edema in CAA-related inflammation

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Sirs: Cerebral amyloid angiopathy (CAA) forms a heterogeneous group within the cerebral small vessel diseases, characterized by congophilic amyloid vessel wall deposits [1]. Several studies have emphasized the importance of CAA in cerebral brain hemorrhage of non-traumatic and non-hypertensive origin, and to a lesser extent in ischemic stroke and leukoencephalopathy with cognitive impairment and cortical dysfunction [2]. CAA classically presents as a spontaneous intracerebral hemorrhage. We present a less common manifestation of CAA.

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An 80-year old woman was admitted with sudden right-sided hemiparesis, dysphasia and altered mental status. There was no preceding infection or vaccination. She had an epileptic seizure after which we started phenytoin. Vital signs were BP 130/70, pulse 87, temperature 37.1 °C. MRI revealed white matter abnormalities, exerting a slight mass effect (Fig. 1 A). No gadolinium enhancement was observed. Serum and cerebrospinal fluid studies were within normal limits. After exclusion of infection we started dexamethasone 4 mg/day and observed a clear recovery. The initial follow-up MRI was comparable with the previous MRI. Diffusion weighted images suggested that the white matter abnormalities represented vasogenic edema (Fig. 1 D). A T2*-weighted gradient echo sequence revealed multiple petechial hemorrhages or cerebral microbleeds (CMB, Fig. 1 B). In the absence of the classical vascular risk factors, we diagnosed CAA and continued dexamethasone in a gradually tapering scheme. The patient recovered, but relapsed after two months. She responded well to a temporary increase of dexamethasone. Four months later we could stop the dexamethasone. She had a mild residual dysphasia with a full recovery of cognition. Repeated MRI showed a clear reduction in brain edema (Fig. 1 C).

Following the Boston criteria [3], we classified our patient as 'probable CAA'. Post-mortem neuropathological examination is still considered the most reliable tool for diagnosing CAA. CAA can be identified in vivo by histological analyses upon evacuating a hematoma or by cortical biopsy. Because our patient responded well to steroid treatment, we did not perform a brain biopsy. The need for biopsy in the diagnosis of CAA is likely to decrease in the future, since Knudsen et al. reported that a reliable diagnosis of CAA can be reached from MR imaging and clinical findings alone [3].

In recent years, inflammatory changes associated with CAA have been increasingly recognized. A study with 42 patients diagnosed with CAA suggested that in these patients accumulation of cerebral vascular fibrillar Aβ promotes perivascular inflammation, according to localization and close relation of immune cells to Aβ-containing plaque deposits. The clinical presentation of these patients differs from most CAA patients. As in other CAA patients, multiple CMB and small infarctions in the cerebral cortex are seen, resulting in subacute cognitive decline and seizures. However, lobar hemorrhagic stroke is rare. These differences might have a genetic background, since the APOE ε4/ε4 genotype is much more frequently seen in patients who present with CAA-related inflammation [4]. Furthermore, these patients have a considerable chance of recovery with steroids [5, 6].

In our case, radiographic imaging showed extensive edema, which was also reported in similar CAA cases in the literature [7]. The corresponding reversible, asymmetric T2-hyperintense MRI changes are fairly specific and enable differentiation from CAA without associated inflammation and reversible posterior leukoencephalopathy [6]. CMB will further support a diagnosis of CAA and should be looked for using T2*-weighted (FLASH2D gradient-echo) MR sequences. This leads Kinnecom et al. to suggest that in the appropriate clinical setting CAA-related inflammation can be diagnosed without brain biopsy [6]. Most of these patients respond well to anti-inflammatory treatment [6]. With this case study, we wish to stress the importance of recognizing this less well-known

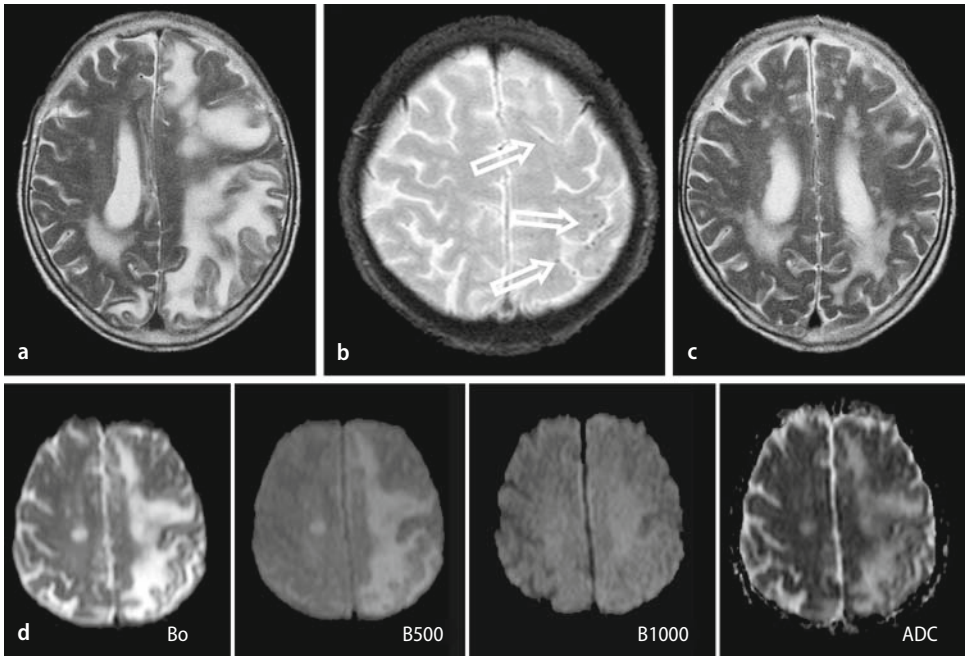


**Fig. 1** **A** Conventional T2-weighted spin-echo sequence (September 2006) showing diffuse, high signal intensity involving the white matter of the left hemisphere and to a much lesser extent the right parietal and occiptal lobes. **B** T2*-weighted (FLASH2D gradient-echo) sequence (September 2006) showing multiple foci of low signal intensity (arrows) in the cerebral cortex. **C** Conventional T2-weighted sequence (June 2007) showing clear resolution of edema. **D** Axial DWI images and corresponding ADC map (September 2006). DWI images show a rapid decrease in signal intensity present in the cerebral white matter (B0-B1000) corresponding to a highly increased diffusion coefficient (ADC). Imaging characteristics indicate presence of vasogenic edema

CAA-related inflammation in vivo, since it is likely to respond well to anti-inflammatory treatment.

**Conflict of interest** The authors declare no conflict of interest.

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