Improvement of neurovascular coupling but not vasoreactivity after steroid treatment in a patient with Cerebral Amyloid Angiopathy-related Inflammation

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Background

Cerebral amyloid angiopathy (CAA) is characterized by β_2-amyloid deposition in leptomeningeal and cortical vessels. CAA-related inflammation (CAA-ri) is a rare, typically reversible inflammatory subtype of CAA, presenting with acute onset of cognitive and behavioral disturbances, seizures, and headaches. It has typical MRI features such as asymmetric vasogenic edema and microbleeds. Neurosonological data on CAA-ri are lacking.

Case report

A 73-year-old man was admitted with generalized seizures, severe cognitive impairment, psychomotor agitation and apraxia. Cognitive testing was not feasible. Brain CT revealed a right posterior hypodense lesion. MRI showed extensive vasogenic edema in the right parieto- temporo-occipital regions, mildly diffused contralaterally, leptomeningeal enhancement, and multiple cortical microbleeds (Figure 1 a and b).

CSF analysis showed only mild protein elevation (0.80 mg/ml) and amyloid deposition without p-tau increaAmyloid PET was negative, and ApoE genotyping was E3/E4.

A diagnosis of probable CAA-ri was made. High-dose ev methylprednisolone (1 g/day for 5 days) followed by oral prednisone (50 mg/day) led to rapid clinical improvement.

Neurosonological Investigation

Transcranial color-coded duplex sonography (TCCD) performed during the acute phase revealed increased vascular resistance in major cerebral arteries and significantly impaired vasoreactivity. Breath-holding index (BHI) assessment of the middle cerebral arteries (MCAs) confirmed reduced reactivity.

Visual stimulation failed to elicit vascular response in the right posterior cerebral artery (PCA) and showed only a mild increase (7%) in mean flow velocity (MFV) on the left side. At one-month follow-up, left in response to visual stimulation. Vasoreactivity alteration persisted.

Follow up

After three months, the patient's neurological examination was normal with residual mild cognitive impairment (MoCA core: 20/30). MRI showed complete resolution of the edema. (Figire 2)

Repeated TCCD demonstrated an improvement of the neurovascular coupling, with FV increases of 20% in the right PCA and 10% in the

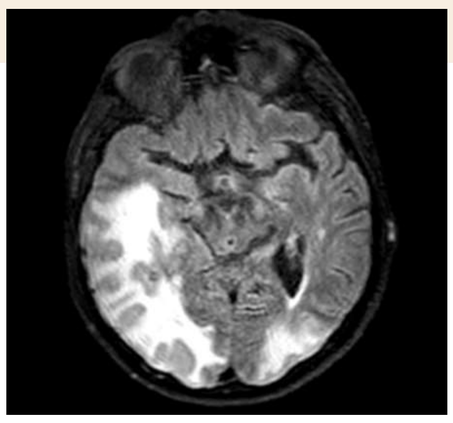

Fig 1a

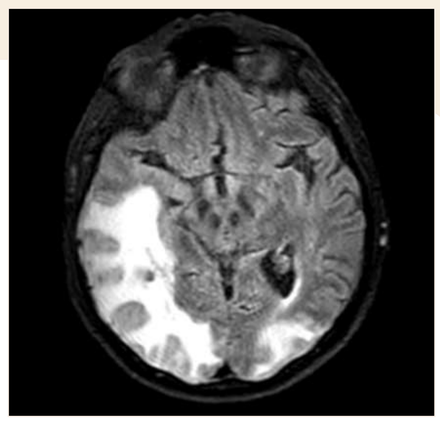

Fig 1b

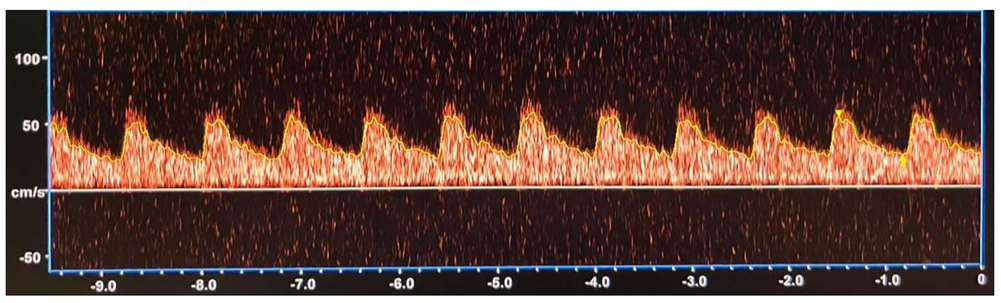

Fig. 3 a

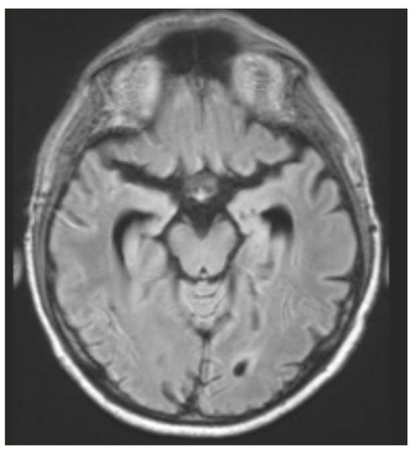

Fig.2a

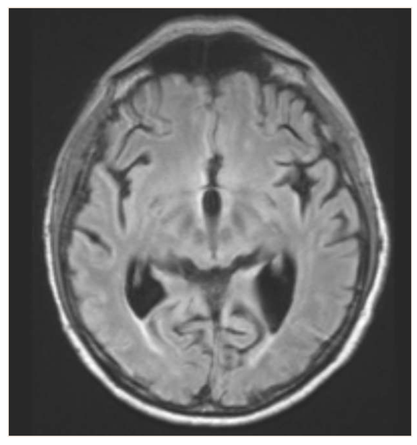

Fig.2b

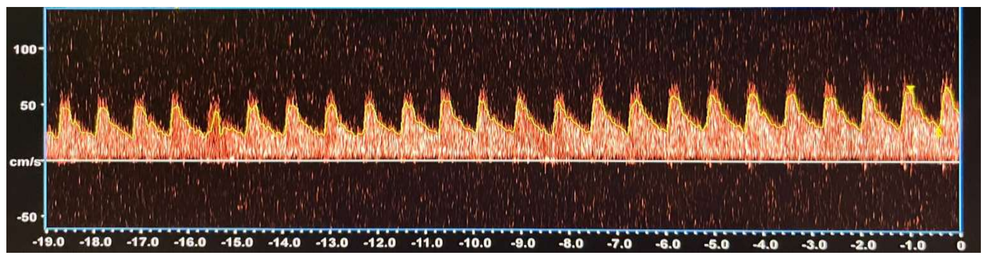

Fig. 3 b

Fig. 1a -1b MRI AX FLAIR images before starting steroid treatment with evidence of extended vasogenic edema in the posterior regions. Fig. 2a-2b resolution of the oedema three months after treatment start.

Fig. 3a-3b neurovascular coupling (15 s of direct light stimulus) in the right PCA before and after steroids.

Conclusion

This case illustrates the utility of TCCD in detecting functional small vessel abnormalities in CAA-ri. Inflammatory involvement of the neurovascular unit may contribute to impaired autoregulation and metabolic coupling, which appears to improve after steroid therapy. TCCD ay serve as a useful, non-invasive adjunct in monitoring disease progression and therapeutic response in CAA-ri.

1. Staszewski J, Dèsbiec A, Skrobowska E, Stepiea„ A. Cerebral Vasoreactivity Changes Over Time in Patients With Different Clinical Manifestations of Cerebral Small Vessel Disease. Front Aging Neurosci.2021 Oct 20;