Cerebral amyloid angiopathy related inflammation presenting with rapid cognitive decline and isolated leptomeningeal effusion

Background

Cerebral amyloid angiopathy–related inflammation (CAA-RI) is a disease characterized by an inflammatory response to cerebral vascular deposits of amyloid-β (Aβ) which may present with cognitive decline, focal neurological deficits, epilepsy and headache. According to current diagnostic criteria, neuroimaging findings in CAA-ri are characterized by hemorrhagic markers of CAA and asymmetric white matter hyperintensities (WMH) due to vasogenic oedema.

We here present a case of CAA-RI with isolated sulcal hyperintensity without parenchymal vasogenic edema.

Case presentation

A 69-year-old man was admitted to the Neurology service for a 4-week history of rapidly progressive cognitive decline. Three years earlier, he had been diagnosed with temporal lobe epilepsy and treated with levetiracetam 500 mg twice daily with benefit. Brain MRI was normal. Figure 1 A-B

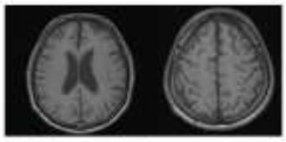

Fig. A-B: Brain MRI performed three years earlier for temporal lobe epilepsy showing no abnormalities.

At admission, neurological examination revealed disorientation in space, severe memory deficits, ideomotor apraxia, and impulsive behavior. MMSE score was 16/30. Blood tests and CSF analysis ruled out metabolic or infectious causes, although CSF showed pleocytosis and increased protein level Brain FLAIR-MRI showed extensive sulcal hyperintensities (SH) and leptomeningeal enhancement (LE) over both hemispheres without parenchymal inflammatory lesions (Figure 1 C_D). Steroid therapy led to clinical improvement and a diagnosis of non-infectious inflammatory encephalitis was made.

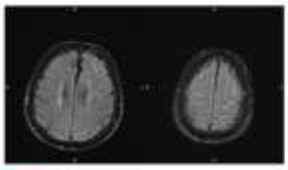

Fig. C-D: Brain MRI performed at the time of admission showed leptomeningeal enhancement (C) and sulcal hyperintensities (D).

Follow-up

After 5 months, the patient was re-admitted due to confusion, disorientation, and visual hallucinations.

MRI showed increase of previously known SH, diffuse LE, multiple small subcortical DWI hyperintense lesions and multiple microbleeds on susceptibility-weighted imaging (Figure 1 F), leading to a diagnosis of probable-CAA.

CSF analysis showed a A+T+N+ profile. Early phase amyloid-PET showed bilateral frontal and temporo-parietal hypoperfusion, while the late phase demonstrated Aβ deposition. The patient received a second cycle of IV methylprednisolone, without clear benefit.

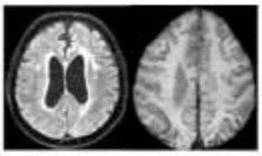

Fig. E-F axial FLAIR) showed a Fazekas score 2 bilateral and SWI showed multiple cortical microbleeds.

2021 transient speech disturbance	2024 rapidly progressive cognitive decline	2025 confusion, disorientation, visual hallucinations
no abnormalities	SH and LE over both hemispheres without parenchymal inflammatory lesions	Increased SH, LE and MB leading to a diagnosis of probable-CAA
temporal lobe epilepsy	pleocytosis, increased protein	A+T+N+
	steroid therapy led to clinical improvement	Second cycle of steroid therapy

Conclusion

SH and LE associated to CAA can be an early manifestation of CAA-RI even in the absence of parenchymal inflammatory lesions. This condition may be the cause of misdiagnosis since the current criteria for CAA-RI required detection of parenchymal vasogenic oedema. Validation and inclusion of isolated SH and LE as imaging findings in CAA-RI may increase diagnostic sensitivity in atypical cases.

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