# Cerebral Amyloid Angiopathy-related inflammation presenting with SIADH : a case report

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## Objective and materials

Cerebral amyloid angiopathy-related inflammation (CAA-ri) is a rare but potentially treatable immune-mediated variant of CAA. [1] We hereby report the case of a patient diagnosed with CAA-ri and presenting with acute confusion in the context of severe hyponatremia.

## Methods

A 76-year-old female patient presented to the emergency department for sudden onset of confusion. Upon neurological examination, only subtle aphasic features were evident. No seizures were reported. Electroencephalographic registration revealed generalized slow wave activity, with no epileptiform discharges. Urgent computed tomography with angiographic and perfusion studies were conducted and excluded any acute cerebrovascular event or mass lesion. Initial laboratory investigations revealed severe hyponatremia, with a serum sodium level of 120 mmol/L. It is noteworthy that venlafaxine was initiated to treat a mood disorder three days prior to clinical presentation. Based on these clinical data, a provisional diagnosis of inappropriate anti-diuretic hormone secretion syndrome (SIADH) was made. Additional diagnostic work-up was planned.

## Results

Magnetic resonance imaging (MRI) revealed multiple, bilateral lobar microbleeds and multiple foci of cortical superficial siderosis on susceptibility-weighted sequences (SWI). Fluid attenuated inversion recovery (FLAIR) sequences demonstrated multifocal and asymmetric white matter hyperintensities in cortico-subcortical and periventricular regions in combination with a diffuse leptomeningeal contrast enhancement along multiple cortical sulci bilaterally on T1 sequences (Figure). Cerebrospinal fluid (CSF) analysis revealed clear fluid with mild hyper-proteinorrachia. Search for neurotropic bacteria and viruses was negative, thus excluding infectious etiologies. Autoantibody screening ruled out an autoimmune or paraneoplastic etiology. CSF neurodegeneration markers exhibited an Alzheimer's disease-like profile. Considering the typical neuroradiological findings and the absence of alternative etiologies, a diagnosis of probable CAA-ri was made according to the diagnostic criteria by Auriel et al. [2]. The patient was treated for hyponatremia with hypertonic solution, and serum levels went back to normal range within 48 hours.

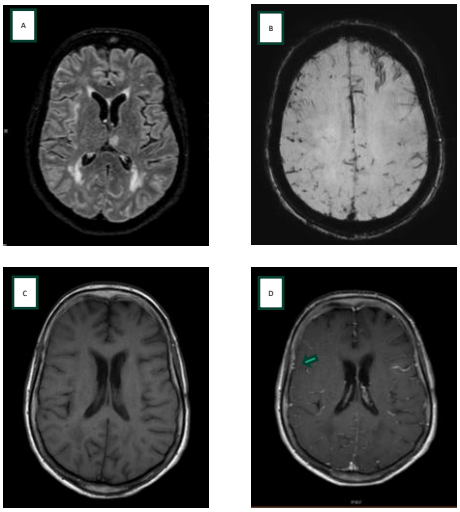


Figure 1: MRI images at baseline. Panel A: axial FLAIR sequence showing white matter hyperintensities in posterior periventricular and subcortical regions and in left thalamus; Panel B: axial SWI showing left frontal cortical siderosis (arrow) and multiple lobar haemorrhages Panel C: T1 pre contrast, D: T1 post contrast showing multiple sulcal leptomeningeal enhancement

Given the progressive clinical improvement, no immunosuppressive treatment was administered. At one month follow-up the patient was asymptomatic, and the MRI showed resolving leptomeningeal enhancement.

## Discussion and conclusions

Although isolated, our report raises the possibility of a potential link between CAA-ri and SIADH, a syndrome known to be associated with immune-mediated conditions. However, we note that the recent initiation of venlafaxine may have exacerbated hypernatremia. Furthermore, the diagnosis was based on brain MRI features and exclusion of alternative etiologies, but no pathological confirmation was available. These limitations notwithstanding, this case highlight how severe hyponatremia in the context of SIADH may represent an atypical and yet poorly characterized initial presentation of CAA-ri. Further studies will be required to validate this association.

**Bibliography**:
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[2] E. Auriel, A. Charidimou, M. E. Gurol et al., - Validation of clinicoradiological criteria for the diagnosis of cerebral amyloid angiopathyâ€"related inflammation - JAMA Neurology - 2016 - 73 - 197-202