

## 437 - COGNITIVE, PATHOLOGICAL, GENETIC AND NEURO-RADIOLOGICAL CORRELATES OF CEREBRAL AMYLOID ANGIOPATHY

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**Background:** Cerebral Amyloid Angiopathy related inflammatory process (CAA-ri), a rare condition caused by an inflammatory reaction occurring within essential cerebral blood vessels against beta-amyloid deposits, leads to subclinical cognitive decline. Often misdiagnosed as dementia, this process can be treated through aggressive immunosuppression, thereby reversing much of the cognitive impairment.

**Case Report:** We report a 69 year old female who came to the clinic for a second opinion and had received a previous diagnosis of Alzheimer's Dementia (AD) from an outside hospital two years prior. She presented with her husband who provided some key aspects of the history. The husband reported two years of worsening of memory, while the patient denied her symptoms. Per husband and patient, she was able to perform activities of daily living (ADLs), including bathing, dressing and toileting, but had difficulties with many instrumental ADLs (IADLs). The clinical course was somewhat fluctuating with progressive cognitive symptoms and significant word-finding difficulties. Patient had been started on Donepezil 5 mg daily by her primary provider.

**Results:** On examination, the patient did exhibit significant word-finding difficulties and scored 12/30 on the Montreal Cognitive Assessment (MoCA), indicating moderate cognitive impairment. The Patient was as well confused and disoriented to time and place. Neurological examination was otherwise unremarkable. Magnetic Resonance Imaging (MRI) studies were ordered and showed patchy and diffuse T2/FLAIR hyper intensities and particularly concentrated in the posterior cerebral artery and inferior division of the middle cerebral artery. These findings were consistent with cerebral amyloid angiopathy related inflammation (CAA-ri). Besides susceptibility weighted image (SWI) was showing multiple widely distributed microhemorrhages typical for CAA.

To address the acute inflammatory reaction the patient was hospitalized and received high dose, 3 day course of intravenous steroids, followed by an oral steroid taper. The treatment had to be monitored due to an unrelated hypertensive emergency and WPW syndrome (both newly diagnosed and treated emergently) thus the Patient was hospitalized for a 3 days and discharged on oral steroids taper in improved condition.

Additionally, imaging showed that the patient's hippocampal volumes were within normal range so this particular imaging biomarker didn't support the diagnosis of AD. CSF biomarkers analysis didn't support the diagnosis of AD either since had p-Tau levels were found to be within normal limits. Patient was found to be homozygous for the APOE e4 gene. Follow-up evaluation (including a repeat MRI study) was performed 2 months later showed clinical recovery and near complete resolution of diffuse hyperintensities, suggesting inflammation had resolved. Both the patient and the husband reported significant improvement in orientation and other aspects of cognition including working memory. The Patient scored 26/30 on MoCA.

**Discussion:** Cerebral amyloid angiopathy (CAA) has been commonly associated with brain hemorrhages in the elderly, but the inflammatory subtype CAA-I occur much less frequently and may be often misdiagnosed as a cancerous process (Ronsin et al. 2016). In a recent systematic review by Caldas A et al. 2015, of the 155 patients with documented CAA-I, almost half displayed some form of cognitive impairment and 86% received corticosteroids. Nearly half of the cases improved following treatment.

**Conclusion:** We present a case of a patient previously diagnosed with AD, upon further investigation, likely CAA-I, treated aggressively with intravenous steroids to good effect. Although rare, CAA-I is a

reversible disorder that may be masked by a dementia or/and delirious process and should be considered in patients showing relatively rapid and fluctuating cognitive decline.

References:

- 1) Ronsin A et al. Pseudotumoral presentation of cerebral amyloid angiopathy-related inflammation. *Neurology*. 2016 Mar 8;86(10):912-9
- 2) Caldas A et al. Cerebral Amyloid Angiopathy Associated with Inflammation: Report of 3 Cases and Systematic Review. *Journal of Stroke and Cerebrovascular Diseases*, Vol. 24, No. 9 (September), 2015: pp 2039-2048