CEREBRAL AMYLOID ANGIOPATHY-RELATED INFLAMMATION (CAA-ri) PRESENTING AS ACUTE CONFUSION IN AN OLDER PERSON

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INTRODUCTION

We are presenting a case of an elderly lady who was admitted with acute confusion which turned out to be a very rare cause of delirium.

CASE SUMMARY

- A 77-year-old female presented to our hyper-acute Stroke Unit with sudden onset agitation and dysarthria. A similar presentation had occurred in the past.
- MRI showed swelling and hyperintensity of the left parietal lobe extending to the occipital lobe, with the signal change extending to the cortical boundary. Numerous micro-haemorrhages were seen throughout the cerebrum in a lobar distribution particularly around the left parietal swelling.
- She was diagnosed with Inflammatory Cerebral Amyloid Angiopathy (CAA-ri) a year earlier and treated with IV methyl-prednisolone. On this occasion also the delirium recurred and a diagnosis of reversible encephalopathy was made. She was again treated with IV steroid successfully with significant radiological improvement on follow-up MRI.

DISCUSSION

- CAA-ri is a distinct subset of CAA^{1,2} with a presentation of acute or sub-acute cognitive decline rather than haemorrhage.
- Diagnostic tests include neuroimaging, CSF studies and brain biopsy. CSF may show higher level of autologous anti-Amyloid β antibodies than non-inflammatory form of CAA according to a small study³.
- MRI features include potentially reversible leukoencephalopathy with patchy or confluent white matter hyper-intensities and multiple micro-haemorrhages seen on gradient echo on T2 weighted MRI sequences.
- Treatment for CAA-ri is not standardized, however glucocorticoid or cytotoxic agent e.g. cyclophosphamide is often used as an initial agent.
- Clinical and radiological improvement is often seen after immunosuppression. Duration of treatment is not well-established and needs to be individualized.

CONCLUSION

It is important for clinicians to consider CAA-ri as a secondary cause of confusion in older patients presenting with stroke-like symptoms.

T2 MRI

GRADIENT ECHO

AFTER TREATMENT

1. Greenberg SM, Vonsattel JP. Diagnosis of cerebral amyloid angiopathy. Sensitivity and specificity of cortical biopsy. Stroke 1997; 28:1418.

2. Kinnecom C, Lev MH, Wendell L, et al. Course of cerebral amyloid angiopathy-related inflammation. Neurology 2007; 68:1411.

3. Piazza F, Greenberg SM, Savioardo M, et al. Anti-amyloid β autoantibodies in cerebral amyloid angiopathy-related inflammation: implications for amyloid-modifying therapies. Ann Neurol. 2013 Apr;73(4):449-58.