A 70 year old with confusion - another delirium? Cerebral vasculitis as a rare cause of encephalitis

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Introduction
We present a case of cerebral vasculitis as a rare cause of encephalitis in a patient with a history of autoimmune pathology.

The Case
Presentation: 70 year old female presented with 4 days of confusion, fever, drowsiness and headaches.

Background: Autoimmune haemolytic anaemia, splenectomy, AF, stroke, HTN and Statin induced myopathy. No offending medications.

Assessment: Confused, pyrexial, no focal neurology

Investigations: FBC/U&E’s/LFTs/Bone profile/Viral serologies normal. CRP 26. Septic screen negative. CT head old infarction. CSF WCC 25 (100% lymph) protein 1.7, glucose 3.9; negative gram stain and TB culture.

Treatment: Started on Acyclovir for viral encephalitis pending CSF PCR

Progress: Despite treatment GCS fluctuated from 8-15 and fevers continued. CSF viral PCR returned negative. MRI head showed subcortical oedema (figure 1). ANA titre 1/1280, dsDNA 61 and strongly positive anticardiolipin IgG

Diagnosis: Cerebral Vasculitis likely lupus related

Treatment: Pulse Methylprednisolone -> tapering prednisolone course

Outcome: GCS improved and fever settled in days; patient walked out of hospital. Follow up MRI (figure 2) showed vast improvement; repeat CSF showed: WCC 0 and protein 0.46.

Discussion
- Cerebral vasculitis is a rare cause of encephalopathy
- Presents with confusion, fever, systemically unwell patient +/- focal neurology
- MRI demonstrates a generalized inflammatory process and has become the most sensitive non-invasive test for the majority of neuroimmunological and infectious disorders.
- CSF shows lymphocytosis with raised protein; aseptic
- Viral PCR’s have a sensitivity of >90%
- Patient had Hx of autoimmune pathology and myositis
- SLE presents with neurological symptoms in <3%
- Presence of neurology confers a poor prognosis
- Treatment classically with induction with steroids or cyclophosphamide prior to maintenance agents.

References
RL DeBiasi; molecular methods for diagnosis of viral encephalitis, clinical microbiology review 2004 Oct; 17(4): 903-925
Kuker W; Cerebral vasculitis: imaging signs revisited, Neuroradiology 2007; 49; 471-479

Figure 1: subcortical oedema affecting the white matter, deep nuclei, brainstem and cerebellum with several foci of microhaemorrhages.

Figure 2: significant resolution post methylprednisolone.