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Systemic Lupus Erythematosus presenting with CNS vasculitis: A diagnostic challenge

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Background

Cerebral vasculitis with extensive intracranial haemorrhage is a rare NPSLE manifestation with <10% incidence in post-mortem studies and a 0.39% prevalence. It carries a high mortality risk. We report a case of spontaneous intracranial haemorrhage (ICH) attributed to cerebral vasculitis in a patient with active SLE.

Case Report

A 14-year-old girl was diagnosed with SLE in May 2024. She was initially treated with prednisolone and followed by five doses of intravenous Cyclophosphamide (IV CYC) for cutaneous vasculitis, but cutaneous activity persisted. At that time, she was on prednisolone 20mg daily and started on mycophenolate mofetil whilst awaiting rituximab approval.

In November 2024, she presented to the hospital with sudden loss of consciousness, right-sided weakness and slurred speech. On examination, active cutaneous lesions were noted on the face and upper limbs. Neurological examination showed right-sided weakness, facial asymmetry and dysarthria. Antiphospholipid antibody, hepatitis, HIV and VDRL were negative.

A CT brain scan showed an acute left basal ganglia haemorrhage with midline shift and mass effect. CT angiography and venography revealed no vascular malformations, while MRI findings suggested cerebral vasculitis. She was managed conservatively with steroids and received a sixth dose of IV CYC. Rehabilitation led to improved muscle strength, with no new skin lesions during follow-up. However, she later flared on prednisolone 12.5 mg daily and is now planned for rituximab due to steroid tapering difficulty.

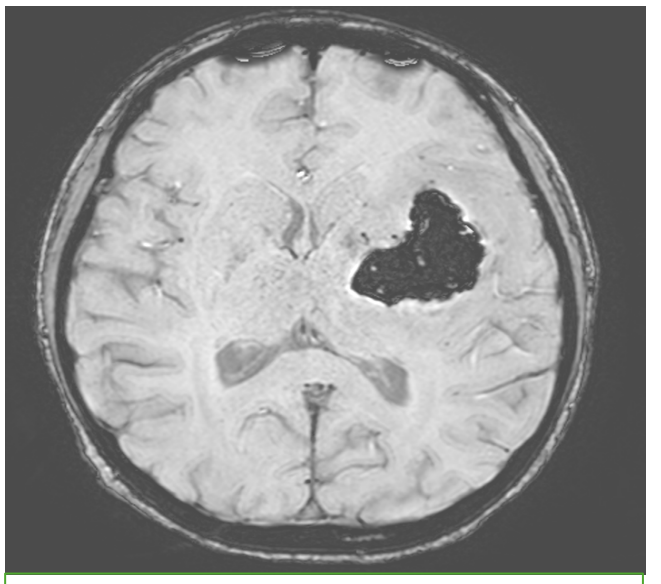

Fig 1: SWI axial view shows susceptibility or blooming artefact at the left basal ganglia suggestive of hemorrhage

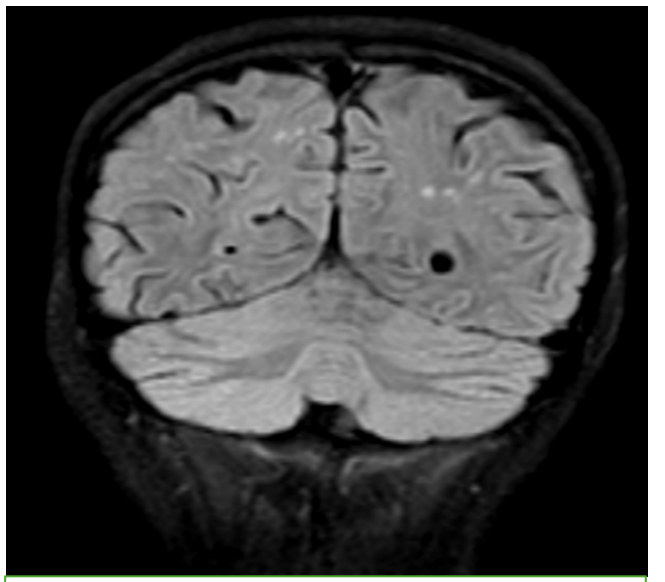

Fig 2: Flair Coronal view shows multiple foci subcortical hyperintensity at bifrontal and biparietal regions.

Conclusions

This case illustrates a rare but life-threatening manifestation of NPSLE, that is cerebral vasculitis leading to spontaneous ICH in a young patient with refractory cutaneous lupus. Early recognition, prompt initiation of immunosuppressive therapy, and early rehabilitation were critical components to her recovery. This case underscores the need for heightened vigilance and timely escalation of treatment in patients with active SLE, particularly when tapering steroids is difficult.

References

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