MSR MALAYSIAN SOCIETY OF RHEUMATOLOGY
23RD MSR - SSR 2025 Workshop in Rheumatology
Singapore Society of Rheumatology

CNS lymphocytic vasculitis presented as tumefactive demyelination in a patient with Systemic Lupus Erythematosus: a diagnostic challenge

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Introduction

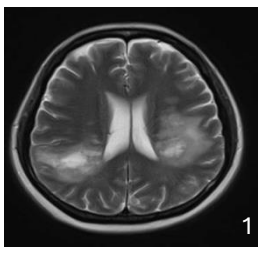
Lymphocytic vasculitis is a rare cause of CNS vasculitis in SLE and can be masked by multiple conditions like CNS tuberculosis or malignancy. Here we present a case of CNS vasculitis with biopsy-confirmed lymphocytic vasculitis.

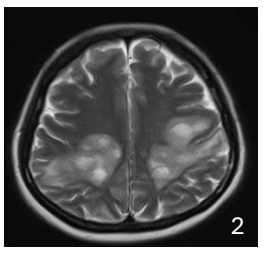
Case report

- Madam CB, a 47-year-old with known case of SLE, developed right facial numbness. MRI showed a focal lesion in the left pons. She was treated with pulsed methylprednisolone, and started on mycophenolate mofetil. Her symptoms improved, and she remained stable for a year.
- She later presented with bilateral limb weakness without altered sensorium, hallucinations, or seizures. MRI showed multiple patchy hyperintensities with perilesional oedema, suggestive of cerebral vasculitis. Despite IV methylprednisolone and cyclophosphamide, her condition worsened, requiring intubation.
- Empirical treatment for TB meningitis and cerebral toxoplasmosis was started due to positive Quantiferon Gold and Toxoplasma IgG. Brain biopsy confirmed lymphocytic vasculitis with glial microinfarcts. She underwent 5 plasma exchange cycles, 2 IVIG cycles, and 1 rituximab infusion. TB and toxoplasmosis treatments were discontinued.
- Following tracheostomy for prolonged ventilation, she made a slow but excellent recovery, regained limb strength, and could follow instructions during outpatient follow-up.

Discussion

- CNS lymphocytic vasculitis is a rare manifestation of NPSLE, occurring in less than 10% of post-mortem studies.(1)
- A mass-like lesion on neuroimaging in patients with SLE should require differential diagnosis among CNS vasculitis, brain infarcts due to secondary antiphospholipid syndrome (APS), cardioemboli, atherosclerosis, brain tumors, and infectious conditions.(2)
- Although it is rarely performed, brain biopsy remains the gold standard to diagnose such cases, as demonstrated in our case.
- Several studies have reported that cyclophosphamide and rituximab are effective as the induction therapy in NPSLE.(3)





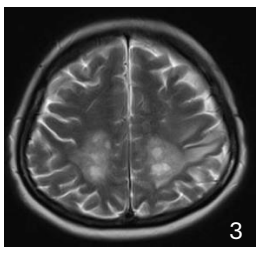


Figure 1,2,3; ***Multiple patchy lesions in the bilateral cerebral hemispheres with perilesional edema and rim enhancement are seen.***

Conclusion

- CNS lymphocytic vasculitis is a rare manifestation of NPSLE, therefore, the diagnosis can be challenging since there are other diseases which could present similarly such as oligodendrogliomas, infection or primary CNS demyelination or vasculitis.

References

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