Systemic lupus erythematosis with cerebral vasculitis: a diagnostic and therapeutic challenge

Shireen R. Chacko M.D. [1], Harpreet Kaur M.D. [1], Nidhi Mody D.O. [1], Peter Moussa M.D., M.Sc.[1], Shraddha Jatwani M.D. [1, 2]

[1]Department of Medicine, Albert Einstein Medical Center, PA

[2]Division of Rheumatology, Albert Einstein Medical Center, PA

Learning Objectives

- Neuropsychiatric systemic lupus erythematosus (NPSLE) is a diagnosis of exclusion.
- Mycophenolate mofetil (MMF) along with steroids, can be used as first line therapy.

Case

21-year-old African-American female with SLE presented with headache, intermittent fever, photophobia, fatigue, and reduced oral intake for 2 months. These symptoms worsened and she started experiencing with nausea and vomiting for 3 days prior to presentation.

Physical examination:

- Febrile (39.3 degrees Centigrade)
- Awake and oriented to time, place and person
- No focal neurologic deficits or nuchal rigidity
- Brudzinski's sign positive

Differential diagnosis:

Meningitis, NPSLE

Initial work-up and management on admission:

- **Brain CT Scan:** Mild cerebral atrophy; no space occupying lesion
- Started on empiric ceftriaxone, vancomycin and acyclovir.
- **Lumbar CSF:** lymphocytic pleocytosis (23 cells/mm^3; 92% lymphocytes); elevated protein (69.4 mg/dL) and normal glucose (77 mg/dL).
- **Culture and PCR panel** returned negative for bacterial and viral pathogens and antimicrobials were discontinued.

Further work-up during admission:

- **EEG:** Diffuse slowing suggestive of moderate diffuse encephalopathy
- **Brain MRI with contrast:** Linear enhancement of the cerebellar folia bilaterally; no infarcts
- **CT Angiography Brain:** Diffuse multifocal irregularities of the distal branches of the anterior, middle, and posterior cerebral arteries suggestive of vasculitis
- **Psychiatric evaluation:** Ruled out depression or eating disorder to explain her poor oral intake
- **Serology** was obtained (see table 1)

Serology	Value	Serology	Value
ANA	Positive (Speckled, >1:1280)	C3, C4	Normal
Anti-dsDNA	21 IU/ml	ESR	102 mm/hr
Anti-Smith	>8 AI	CRP	17 mg/L

Table 1. Serology (values reported are on admission)

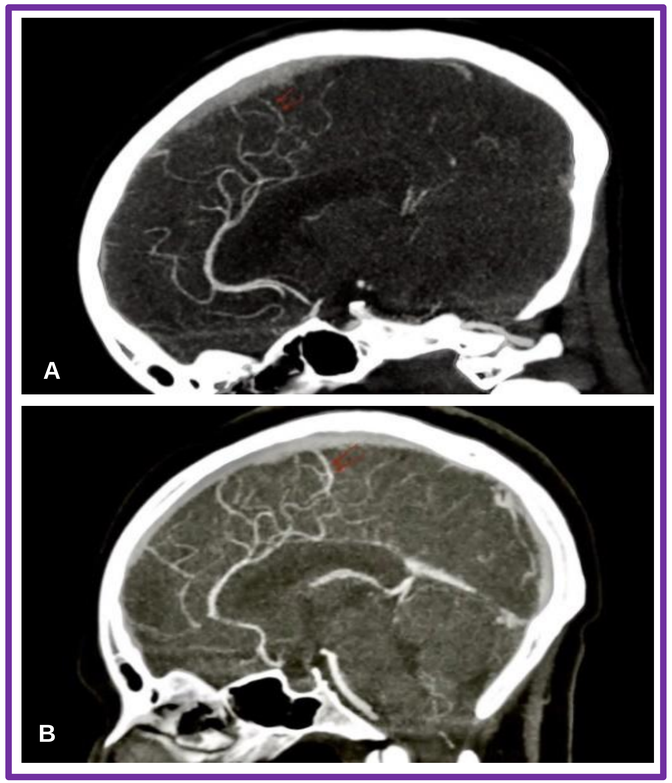

Figure 1. CT angiography at diagnosis (panel A) and at follow up 6 weeks later (Panel B) showing significant improvement in multifocal vascular irregularities.

Case Continued

Management during admission:

- Pulse dose Methylprednisone for three days, then
- Prednisone 60 mg daily, MMF 1500 mg twice daily and Hydroxychloroquine 400 mg daily
- At 6 weeks, there was significant symptomatic and radiologic improvement (Figure 1), although antibody levels remained high

Discussion

- NPSLE is a challenging clinical diagnosis due to its varied clinical presentations and remains a diagnosis of exclusion.
- The typical treatment for NPSLE consists of induction of remission with a steroid taper combined with an additional immune suppressant, typically cyclophosphamide (CYC). Hydroxychloroquine is also typically continued for treatment of underlying SLE.
- There a number of case reports and case series that have evidenced benefit in the use of MMF with steroids in both the induction and maintenance of remission NPSLE
- We favored the use of MMF over CYC for induction and maintenance therapy given the relatively lower incidence of adverse effects and better tolerability.

References

1.Hanly JG. Diagnosis and management of neuropsychiatric SLE. Nat Rev Rheumatol. 2014;10(6):338-347.

2.Gupta N, Ganpati A, Mandal S, et al. Mycophenolate mofetil and deflazacort combination in neuropsychiatric lupus: a decade of experience from a tertiary care teaching hospital in southern India. Clin Rheumatol. 2017;36(10):2273-2279.

3.Salvarani C, Brown RD Jr, Christianson TJ, et al. Mycophenolate mofetil in primary central nervous system vasculitis. Semin Arthritis Rheum. 2015;45(1):55-59.

4.Hu W, Liu Z, Chen H, et al. Mycophenolate mofetil vs cyclophosphamide therapy for patients with diffuse proliferative lupus nephritis. Chin Med J (Engl). 2002;115(5):705-709.

.