A Wolf in Sheep's Clothing: A Mysterious Case of Seizures

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Patient Presentation

A 22M with a history of quiescent juvenile arthritis presented to ED in his northern community with arthralgias. He was incidentally found to have mild normocytic anemia and leukopenia, with a slightly elevated creatinine. He was discharged with a plan for further investigations including an autoimmune workup.

Two weeks later, he represented with **3 generalized tonic-clonic seizures.**

Initial Management

CT Head showed **ill-defined hypodense lesions in the left frontal lobe, right insula, and left cerebellar hemisphere**. The differential included **metastatic disease and disseminated infection**. MRI brain recommended for further assessment.

He was loaded with Keppra and transferred to tertiary care for ongoing management.

History and Physical Examination

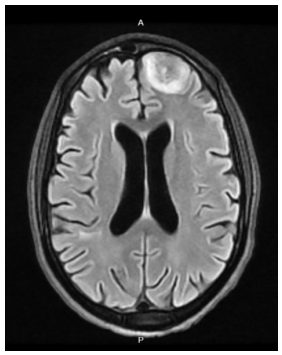
Assessment in ED revealed the following symptoms over the preceding months:

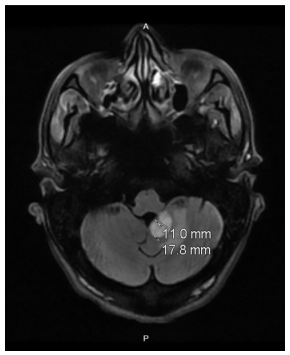
- Intermittent abdominal and joint pain
- 15 lbs weight loss, anorexia, and occasional night sweats
- Hyperpigmented facial rash
- Alopecia (see Image 1)
- Hyperpigmented lesions on the palmar aspects of his hands (see Image 2)

Image 1. Hand lesions

Image 2. Alopecia

Further Investigations





MRI Brain (T2 FLAIR images above): Multiple hemorrhagic lesions. **Etiology unclear, cannot exclude underlying mass lesion or septic infarct.**

Laboratory findings:
WBC 4.7, Hb 83 (MCV 95.5), Plat 285
Electrolytes N, **Cr 148**
BCx x2 negative. LP negative.

CRP 37.5, ESR 60
ANA 1:2560 (speckled), dsDNA >667
C3 0.46, C4 0.05
ENA +, Anti-Sm > 694, RNP>664, SSA >1,375
Cardiolipins +, B2 glycoprotein, and LA negative
RF 29, CCP <5, ANCA negative
TTE/TEE negative for thrombus/IE

Diagnosis

Final Dx: Systemic lupus erythematosus with neuropsychiatric involvement (vasculitis), bicytopenia, dermatologic manifestations, and arthritis. Renal biopsy later confirmed grade 2-3 lupus nephritis.

Patient was started on hydroxychloroquine, steroids, and Rituximab.

Discussion

Neuropsychiatric SLE (NPSLE) describes the CNS manifestations of lupus.

- Heterogeneous group including psychiatric presentations, seizures, and strokes. The ACR has defined 19 distinct NPSLE syndromes.[1]
- Prevalence varies significantly; some studies estimate NPSLE occurs in **40-50% of patients with ranges from 12-95% described.**[4]
- NPSLE is a major source of morbidity and mortality in SLE.[2]
- Pathophysiology is complex; suspected to involve microvasculopathy and neurotoxicity due to cytokines, autoantibodies, and immune complexes.[2]
- Diagnosis is based on clinical assessment, serologies, and imaging (MRI preferred).[4]

Management is challenging:

- Foundation is symptomatic management, especially in mild cases (ie. Antipsychotics, antiepileptics).[1]
- Immunosuppression with corticosteroids, azathioprine, cyclophosphamide, and mycophenolate is often required. IVIG and rituximab have been used in cases unresponsive to steroids or cytotoxic therapies.[3]

LEARNING OBJECTIVES

1) Recognize CNS manifestations of SLE, including vasculitis and seizure
2) Review the pathophysiology, epidemiology, and management of neuropsychiatric SLE

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CONFLICTS OF INTEREST

Co-Author	Conflict disclosures
Isabel Shore	None
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Delvina Hasimja	None