

NEUROLOGICAL INVOLVEMENT IN SYSTEMIC LUPUS ERYTHEMATOSUS: UNVEILING AN ENIGMA



Main Line Health®

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Introduction

- Systemic lupus erythematosus (SLE) is an autoimmune disease that causes diffuse inflammation and damage to the tissues in the body including the nervous system. Neuropsychiatric events may precede, occur concomitantly with, or follow the diagnosis of SLE. (1)
- Given the wide range of symptoms ranging from headaches, confusion, seizures and strokes; it can make diagnosing neuropsychiatric systemic lupus erythematosus (NP-SLE) difficult.

Case Presentation

- 63-year-old female with past medical history of polymyalgia rheumatica presented with subacute encephalopathy and weakness.
- Home medications: amlodipine, levothyroxine, ciprofloxacin and metronidazole for recent diverticulitis episode.
- Vitals were normal. Physical exam revealed a disheveled appearance, dry mucous membranes, flat affect but oriented x3
- Laboratory findings: serum sodium 121, TSH 2.94, WBC 2.6 with absolute lymphocytes 0.23 and normal bands, Hemoglobin 11.4, PLT 127, ALP 176, AST 328, ALT 175. Urinalysis, toxicology screen, CT head, CXR, and MRI brain were unremarkable
- Encephalopathy thought to be secondary to hyponatremia, however she showed no improvement after correction of the hyponatremia so workup for her encephalopathy broadened.
- Bone marrow biopsy and flow cytometry were unrevealing. SPEP and IFE were drawn showing a faint lambda monoclonal light chain band. Subsequent bone marrow biopsy revealed no evidence of plasma cell disorder and flow cytometry was unremarkable.
- Serological work-up revealed a positive ANA of greater than 1:640 homogenous pattern, dsDNA Ab 469, positive SSA, and a positive Histone Ab. Further testing revealed an elevated ESR, ferritin, positive Coombs test, and hypocomplementemia.
- Lumbar puncture revealed pleocytosis of 10, elevated protein, IgG index, oligoclonal bands.
- NP-SLE was diagnosed and she received high-dose steroids and hydroxychloroquine. She was later given cyclophosphamide then transitioned to mycophenolate mofetil with resolution of her pancytopenia, transaminitis, and encephalopathy.

Discussion

- The pathogenesis of (NP-SLE) is not fully understood.
- One theory is that autoantibodies cause direct neuronal damage by triggering reactive cytokines and chemokines. Specific antibodies thought to be linked with the development of NP-SLE are: antiphospholipid Ab, anti-ribosomal P Ab, and N-methyl-D-aspartate receptor (NMDAR) Ab. (2)
- Since diseases can present with atypical symptoms, maintaining a broad differential diagnosis ensures potential diagnoses are not overlooked and avoids delayed workup and management.

References

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2. Schwartz N, Stock AD, Putterman C. Neuropsychiatric lupus: new mechanistic insights and future treatment directions. *Nat Rev Rheumatol*. 2019 Mar;15(3):137-152. doi: 10.1038/s41584-018-0156-8. PMID: 30659245; PMCID: PMC8023338.
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