A Diagnostic Dilemma: Venturing into the Gray Zone with Lupus Myelitis

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INTRODUCTION

Systemic Lupus Erythematous:
- SLE is a clinically heterogeneous autoimmune disease more common in females affecting up to 150 per 100,0003
- Transverse myelitis is 1000x more prevalent in SLE compared to the general population1 AND can be the primary manifestation of lupus

CASE DESCRIPTION

- 45 y/o female with hypothyroidism initially presented to her PCP with a pruritic truncal rash, diffuse abdominal cramping and diarrhea → symptoms resolved
- Developed right perineal & thigh numbness a few days later
- Subsequently experienced rapid-onset, bilateral ascending paralysis with urinary retention → emergent hospitalization
- Physical exam: lower extremity areflexia, flaccid paralysis & sensory loss up to T8, saddle anesthesia, decreased rectal tone and urinary retention
- Pertinent labs:
  - UA 7/26: 5 RBCs, 3 WBCs, protein 100
  - ESR: 92
  - CRP: 15.2
  - Anti-SSA: >8.0 (+)
  - Anti-SSB: 3.2 (+)
  - ANA: +1:2560 litter (H)
  - C3: 82 (L)
  - C4: 15 (L)
  - Anti-Smith Ab: 0
  - Anti-DS-DNA Ab: negative
- MRI spine demonstrated extensive T2 hyperintensity predominantly in the gray matter of the spinal cord from T7 through conus suggestive of transverse myelitis
- LP was performed → CSF: 750 WBCs (78% PMNs), 31K RBCs, total protein 658 and glucose 40

HOSPITAL COURSE

- Extensive infectious & hypercoagulable workup were negative
- Completed 5 days of methylprednisolone, 2 rounds of plasmapheresis without neurologic improvement
- Consulted Rheumatology due to high ANA titer, +anti-SSA/SSB and mild hypocomplementemia concerning for SLE
- Repeat MRI with persistent T2 signal abnormality but visceral and spinal angiogram did not show evidence of infarct or vasculitis
- Developed nephrotic range proteinuria (6650 mg in 24hr) felt to be more consistent with lupus membranous nephropathy
- Ultimately treated for lupus myelitis with monthly cyclophosphamide and prolonged steroid taper to prevent further neurologic complications
- Discharged to inpatient rehab without recovery of neurologic deficits

DISCUSSION

- Common neurologic features in SLE: stroke, peripheral neuropathy, headache, seizures, cognitive dysfunction
- Lupus-associated myelitis is one of the most devastating consequences of SLE, may be the initial manifestation of lupus
- Risk of recurrence estimated at 21-55%4
- Recent cohort studies: 2 distinct subtypes of myelitis in SLE patients based on involvement of gray vs. white matter associated with differing prognoses2
- Gray matter myelitis = more acute presentation, flaccidity, hyporeflexia → higher rates of irreversible paraplegia despite intensive immunosuppression
- White matter myelitis = slower presentation, spasticity, hyperreflexia → overall better prognosis
- It is critical to recognize salient prodromal features (e.g. urinary retention and fever) associated with gray matter myelitis to expedite spinal cord imaging for earlier diagnosis and treatment in pursuit of better outcomes

REFERENCES


Figure 1: MRI thoracic cord - central cord T2 hyperintensity from around T7 to conus, notably restricted to gray matter.

Figure 2: MRI thoracic cord 6 days later - persistent T2 hyperintensity in central cord, although more diffuse cord signal abnormality inferior to T9.

Figure 3: Comparison of MRI results with outcome in 55 patients w/ TM and SLE - abnormal results shown in black, normal results shown in white1.