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

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INTRODUCTION

Systemic Lupus Erythematosus:

- SLE is a clinically heterogeneous autoimmune disease more common in females affecting up to 150 per 100,000[3]
- Transverse myelitis is 1000x more prevalent in SLE compared to the general population[1] **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 (+)
ANCA: <1.20
ANA: +1:2560 titer (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

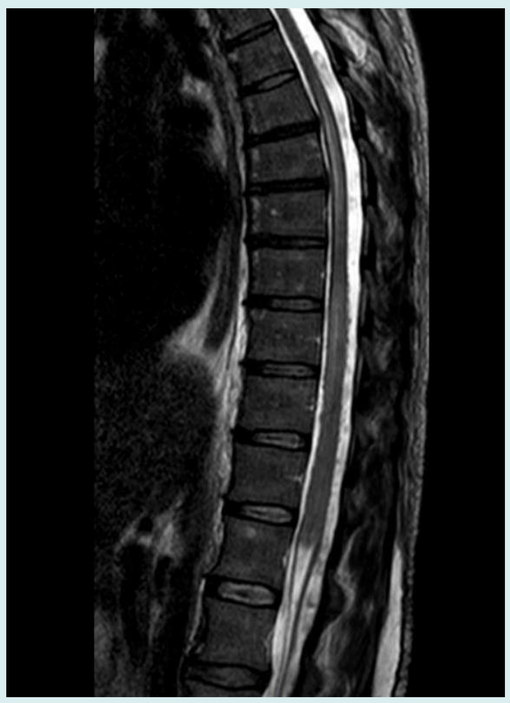

Figure 1: MRI thoracic cord - central cord T2 hyperintensity from around T7 to conus, notably restricted to gray matter.

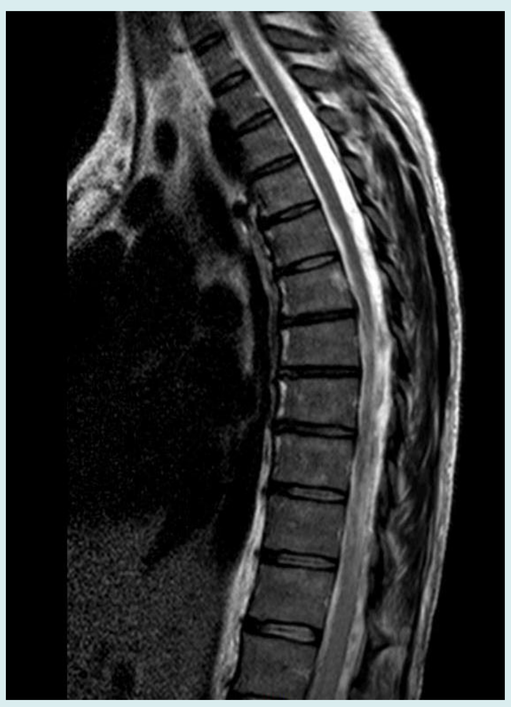

Figure 2: MRI thoracic cord 6 days later - persistent T2 hyperintensity in central cord, although more diffuse cord signal abnormality inferior to T9.

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 prognoses[2]
- **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

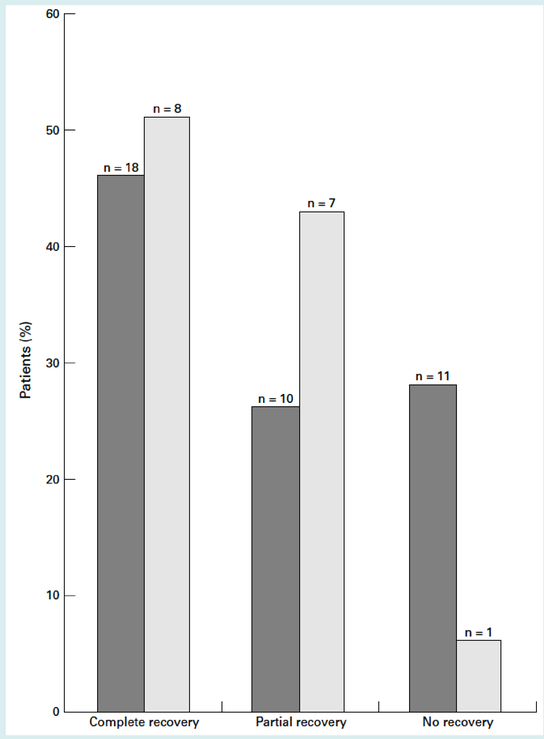

Figure 3: Comparison of MRI results with outcome in 55 patients w/ TM and SLE - abnormal results shown in black, normal results shown in white[1].

REFERENCES

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4. Lukjanowicz M, Brzosko M. Myelitis in the course of systemic lupus erythematosus. Pol Arch Med Wewn. 2009;119(1-2):67-72.