23RD MSR - SSR 2025 Workshop in Rheumatology

# When Recovery Stalls: Refractory Vasculitic Neuropathy in Systemic Lupus Erythematosus with Emerging Autonomic Dysfunction

**Izzatul Nadiah Zolkiply[1],**
**[1]Department of Medicine, Hospital Pakar Sultanah Fatimah Muar,Johor, Malaysia**

## Introduction

Peripheral nervous system (PNS) involvement in systemic lupus erythematosus (SLE) is under-recognized and diagnostically challenging. Vasculitic neuropathy, particularly mononeuritis multiplex, is among the most specific but least reversible manifestations. This case highlights an unusual clinical trajectory: despite full NIH-standard immunosuppression, neurological deficits persisted, and autonomic symptoms emerged late, raising concern for neurodominant disease progression.

## Case Summary

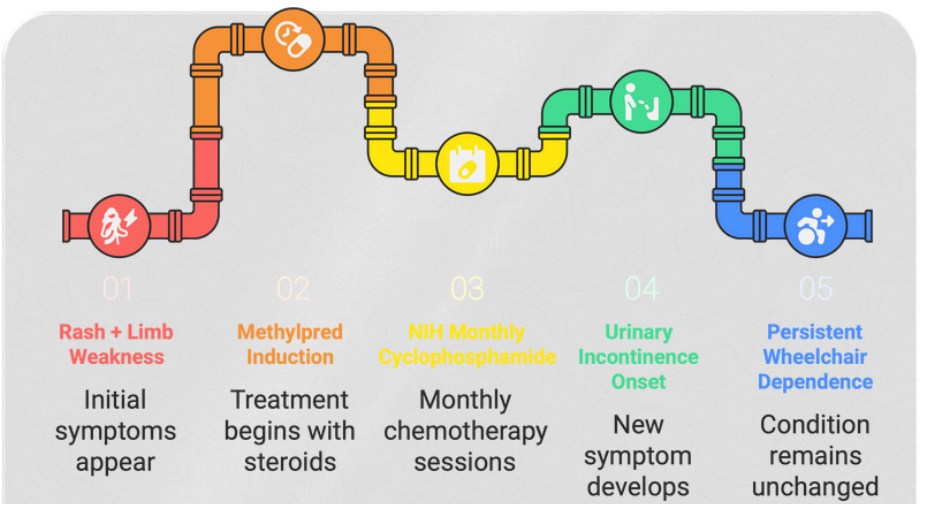
**Patient Profile** 68-year-old woman, Diabetes melitus
**Initial Presentation:**
- Palpable purpura (UL/LL/trunk), right hand weakness, progressive lower limb numbness

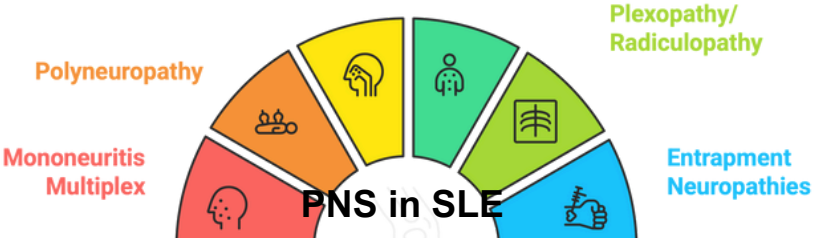
**Investigations:**
- ANA 1:80 speckled, low C3/C4, Coombs-positive hemolytic anemia, ANCA negative, APLS negative
- Skin biopsy: leukocytoclastic vasculitis
- NCS: Mononeuritis multiplex (asymmetrical axonal neuropathy); bilateral median entrapment

**Treatment:**
- IV methylprednisolone ×3 days → oral steroids
- Followed by 6 monthly pulses of IV cyclophosphamide (NIH regimen)

**Outcome:**
- Cutaneous resolution ✅
- Persistent paraparesis and sensory deficits ❌
- New-onset urinary incontinence after 5th CYC pulse → autonomic dysfunction vs spinal cord vasculitis (MRI pending)

## Clinical timeline




## Discussion

🧠 **Non-Responsive Neuropathy**
Lack of improvement after 6 NIH CYC pulses suggests either irreversible axonal damage or suboptimal immune control—despite systemic remission.

🧩 **Entrapment Mimicry vs Vasculitis**
Vasculitic neuropathy may mimic entrapment syndromes, especially in diabetics or elderly.
The asymmetric involvement of median, ulnar, and sural nerves in this case is more consistent with mononeuritis multiplex than compressive mononeuropathies.

🚽 **Delayed Autonomic Signs**
Urinary incontinence post-5th CYC is atypical.
Differentials include:
• Autonomic neuropathy (neurogenic bladder)
• Spinal vasculitis (occult myelitis; imaging pending)

⚠️ **Phenotypic Uncertainty**
Low-titre ANA and a seronegative profile raise key questions:Is this evolving neurodominant SLE, or early UCTD with vasculitic overlap?

💉 **Therapeutic Reflection**
• Should rituximab be considered earlier?

## Conclusion

Despite resolution of cutaneous vasculitis, this case showed no neurological recovery and emergence of autonomic symptoms after standard NIH cyclophosphamide induction.
This trajectory suggests a neurodominant SLE variant, potentially under-recognized in current classification and management pathways.
It calls for early escalation, repeat neuro-axis imaging, and rethinking of therapeutic timing in vasculitic neuro-lupus.

💡 **Learning Points**

🔹 Skin may heals, nerves may not.
Cutaneous improvement doesn't predict neurological recovery in vasculitic SLE.
🔹 Don't wait to escalate.
Persistent deficits warrant earlier transition to biologics or combination therapy.
🔹 Autonomic red flags matter.
Late-onset incontinence should prompt urgent evaluation for spinal or autonomic involvement.

## Referrence

1.Peripheral nervous system involvement in systemic lupus erythematosus: a review of the evidence — Clinical and Experimental Rheumatology, 2019, by Bortoluzzi et al.