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Mononeuritis Multiplex as a Rare Initial Presentation of Systemic Lupus Erythematosus: A Case Report

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INTRODUCTION

- **Mononeuritis multiplex (MNM)** is a neurological disorder involving damage to multiple peripheral nerves in different parts of the body. It typically presents with symptoms like weakness, numbness, pain, or loss of function.
- Unlike polyneuropathy, which affects nerves symmetrically, mononeuritis multiplex follows an irregular and asymmetric pattern. It is often linked to underlying conditions such as diabetes, vasculitis, infections, or autoimmune diseases.

CASE REPORT

Patient Overview

A 47-year-old woman with a history of hypertension and a strong family history of autoimmune disease, including her sister's recent diagnosis of systemic lupus erythematosus (SLE).

Clinical Presentation

She presented with a three-month history of numbness, weakness, and electric shock-like pain in both feet. The symptoms gradually progressed to involve her hands, with asymmetrical weakness and sensory loss.

She also experienced intermittent joint pain, fever, and unintentional weight loss.

Neurological examination showed asymmetrical weakness, sensory deficits, and reduced reflexes in all four limbs.

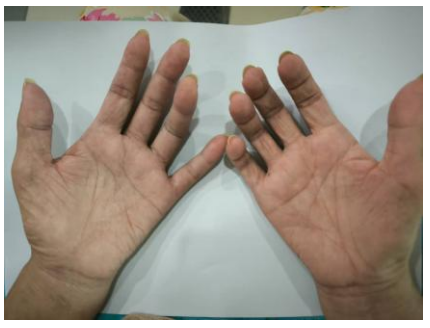


Figure 1 : Claw hands with wasting of bilateral hypothenar and left thenar region

Figure 2 : Showing positive rock sign over left hand where patient unable to make a fist.

Laboratory and Diagnostic Findings

Laboratory investigations revealed positive autoimmune markers, including ANA, anti-dsDNA, anti-Sm, anti-RNP, anti-Ro52, anti-Ro60, anti-SS-B, and anti-Centromere antibodies, along with low complement levels.

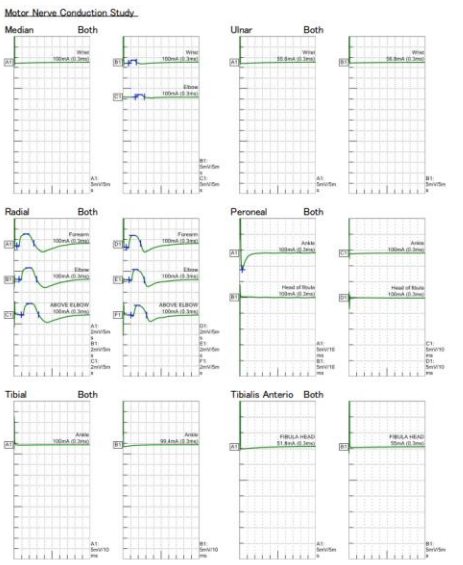
Her HbA1c was within normal limits, and infectious screening for Hepatitis B, Hepatitis C, and HIV was negative.

Electromyography and nerve conduction studies showed active denervation consistent with sensorimotor axonal polyneuropathy, indicating vasculitic neuropathy likely secondary to systemic lupus erythematosus or Sjogren's Syndrome.

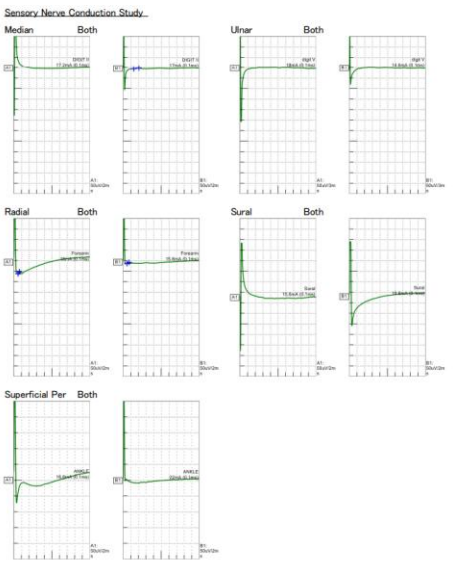
Urinalysis demonstrated 4+ proteinuria and hematuria, with a 24-hour urine protein measurement of 2.72 g/day. A subsequent renal biopsy confirmed Class II Lupus Nephritis.

Treatment & Outcome

She initially received a five-day course of intravenous methylprednisolone (1g daily); however, there was no significant neurological improvement. As a result, she was started on intravenous cyclophosphamide (500 mg per cycle), with a total of six cycles planned.



Graph 1 : Motor study
Right median and bilateral radial nerves show reduced CMAP, normal DML and CV. Left median, bilateral ulnar show absent CMAP. Bilateral peroneal and tibial show absent CMAP.



Graph 2: Sensory study
Right median and bilateral radial nerves show reduced SNAP, normal DSL and CV. Bilateral ulnar and left median show absent SNAP. Bilateral sural and superficial peroneal show absent SNAP.

DISCUSSION

- MNM occurs in an estimated 1% to 9% of SLE cases. Its presence is often associated with high disease activity and may signal a worse prognosis.
- The most common cause of MNM in SLE is small-vessel vasculitis, which leads to inflammation and ischemic injury to the blood vessels supplying peripheral nerves. This can result in nerve infarction, causing both sensory and motor dysfunction.
- MNM can either accompany other lupus features or appear independently.
- It most frequently affects the lower limbs, with symptoms such as: Sharp, burning, or deep aching pain (worse at night), Numbness, tingling, and muscle weakness. Foot drop, muscle atrophy, and a foot-slapping gait, especially due to tibialis anterior weakness
- Nerve conduction studies and EMG are key tools to assess the extent and type of nerve damage.
- In uncertain cases, nerve or muscle biopsy can help confirm vasculitis. MRI and CSF analysis may be useful in ruling out central or alternative causes of neuropathy.
- First-line treatment includes immunosuppressive therapy such as steroids and cyclophosphamide. Physical therapy supports recovery by preserving mobility and preventing joint contractures.
- Prognosis varies but depends on early recognition and aggressiveness of treatment. Some patients recover function, while others may experience permanent deficits.

CONCLUSION

- Mononeuritis multiplex can be a rare initial presentation of SLE.
- Early recognition aids prompt diagnosis and treatment.
- Timely management helps prevent complications and improves outcomes. Delays in diagnosis or treatment can increase the risk of long-term disability.

REFERENCES

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