Chronic Inflammatory Demyelinating Polyradiculoneuropathy as the First Manifestation of Systemic Lupus Erythematosus in an Elderly Male

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Abstract:

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a rare neurological manifestation of systemic lupus erythematosus (SLE), particularly uncommon as an initial presentation. A 61-year-old man presented with progressive symmetric paraparesis, sensory ataxia, and areflexia. Neurophysiology showed demyelinating sensorimotor polyneuropathy; cerebrospinal fluid (CSF) demonstrated albuminocytologic dissociation. He improved with corticosteroids but relapsed after two months with quadriparesis and severe sensory ataxia. Repeating studies supported CIDP. Subsequent workup revealed features consistent with SLE. He responded to intravenous immunoglobulin (IVIG) and tapering oral steroids, regaining functional ambulation. CIDP can be an initial manifestation of SLE even in elderly males. Early recognition and immunomodulatory therapy are critical for recovery.

Key words: Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP), Systemic Lupus Erythematosus (SLE), Neuro-Lupus, CNS Involvement in SLE.

Introduction

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease with diverse neuropsychiatric involvement.[1] Peripheral neuropathy occurs in approximately 10%–20% of SLE patients; however, chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is rare, estimated in about 0.2% of cases.[2] Distinguishing CIDP from acute demyelinating neuropathies such as Guillain-Barré syndrome (GBS) is crucial, as CIDP evolves over eight weeks or more and typically requires sustained immunomodulation. The pathogenesis of SLE-associated CIDP likely involves humoral and cellular immune mechanisms,[2] including immune-complex mediated microvascular injury and autoantibody-mediated demyelination. We report a case of CIDP as the first manifestation of SLE in a 61-year-old male, highlighting diagnostic challenges and therapeutic response.

Case Report

A 61-year-old man with hypertension, chronic smoking, and alcohol use presented with one month of progressive, symmetric weakness of the lower limbs, sensory disturbances, moderate back pain, and headache.

Examination revealed:

- **Lower limbs:** Proximal power 4–, distal power 4+, areflexia, graded sensory loss, absent joint position and vibration sense, positive Romberg's sign.
- **Upper limbs:** Reduced grip strength and areflexia.
- **Higher mental functions and cranial nerves:** Normal.

Nerve conduction studies (NCS) demonstrated bilateral symmetric sensorimotor demyelinating polyneuropathy, more pronounced in the lower limbs. Cerebrospinal fluid (CSF) analysis showed elevated opening pressure with albuminocytologic dissociation (protein 100, cells 2) and oligoclonal bands. Magnetic resonance imaging (MRI) of the brain and spine were unremarkable. Initial autoimmune panel, including anti-nuclear antibodies (ANA), and serum protein electrophoresis, were negative. He received intravenous corticosteroids followed by tapering oral steroids, with improvement over 3–4 weeks. His motor strength improved, reflexes returned, and ataxia resolved.

After two months of remission, he developed an abrupt headache, back pain, and rapidly progressive symmetric paraparesis with ataxia over three days. Examination showed greater severity in quadriparesis compared to prior tests: power 2/5 in lower limbs, 4/5 in upper limbs, generalised areflexia, loss of proprioception, and sensory ataxia. Repeat CSF analysis showed albuminocytologic dissociation, and NCS reconfirmed demyelinating sensorimotor polyneuropathy, predominantly in lower limbs. A diagnosis of CIDP was made. Subsequent evaluation identified features consistent with SLE. He was treated with intravenous immunoglobulin (IVIG), with marked improvement in motor power, independent ambulation and discharged on tapering oral corticosteroids.

Discussion

CIDP is an uncommon but important peripheral nervous system manifestation of SLE. In contrast to GBS, CIDP evolves over at least eight weeks and often relapses, requiring ongoing immunomodulatory therapy. The clinical phenotype typically includes symmetric sensorimotor deficits, proprioceptive loss, and areflexia affecting both upper and lower limbs.[3] Diagnostic hallmarks include demyelinating features on NCS (slowed conduction velocities, conduction block, prolonged F-waves) and albuminocytologic dissociation on CSF analysis. MRI is often unremarkable. While autoimmune serologies in SLE-associated CIDP may show ANA, anti-double-stranded DNA (anti-dsDNA), anti-Smith (anti-Sm), anti-SSA/Ro, and low complement levels, serological activity can be variable, and initial tests may be negative early in the disease.[4]

Proposed mechanisms include immune-complex deposition, microvascular injury resembling vasculitis, and autoantibody-mediated demyelination at the nodes of Ranvier. Anti-ganglioside antibodies have been reported in a subset of SLE patients with neuropathy, but associations with CIDP are inconsistent. Treatment typically involves high-dose corticosteroids,[5] IVIG, or plasmapheresis, alone or in combination. In SLE-associated cases, early recognition and prompt immunotherapy are associated with favourable outcomes; some patients respond well to steroids alone, while others require IVIG, particularly with systemic activity or relapse.

Conclusion

CIDP may be the first manifestation of SLE, even in older males. Neurophysiology and CSF studies are pivotal for diagnosis when imaging is unrevealing. Relapse after steroid response should prompt reassessment for underlying systemic autoimmunity. IVIG can be effective in steroid-responsive but relapsing disease, supporting combined immunomodulatory strategies.

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