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# Chronic Inflammatory Demyelinating Polyneuropathy (CIDP): An Uncommon Manifestation of Systemic Lupus Erythematosus (SLE)

Authors' Contribution:  
Study Design A  
Data Collection B  
Statistical Analysis C  
Data Interpretation D  
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**Conflict of interest:** None declared

**Patient:** Female, 40  
**Final Diagnosis:** Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)  
**Symptoms:** Gait disorder  
**Medication:** —  
**Clinical Procedure:** —  
**Specialty:** Rheumatology





**Objective:** Rare disease  
**Background:** Chronic inflammatory demyelinating polyneuropathy (CIDP) is an uncommon manifestation of systemic lupus erythematosus (SLE). We report a case of SLE presenting as CIDP and discuss the diagnosis, management, and prognosis of CIDP.

**Case Report:** A 40-year-old woman with a past medical history of SLE treated with hydroxychloroquine presented with bilateral, progressive, ascending, sensory and motor neuropathy. Physical examination showed weakness and reduced temperature of all extremities, reduced pinprick and vibration sense of the distal extremities, loss of reflexes, and walking with a wide-based unsteady gait. Laboratory investigations showed positive antinuclear antibodies (ANA), anti-(smooth muscle (SM) antibody, anti-RNP antibody, anti-SSA antibody, anti-ds-DNA antibody, and an erythrocyte sedimentation rate (ESR) of 75 mm/hr, low C4, leukopenia, and anemia. Electromyography (EMG) confirmed the diagnosis of CIDP. The patient's neuropathy and muscle weakness improved on treatment with intravenous immunoglobulin (IVIG) and high-dose steroids.

**Conclusions:** The early clinical diagnosis of CIDP, supported by serological autoantibody profiles associated with SLE, can predict a good response to steroids. Most patients with CIDP are treated successfully with steroids if the diagnosis is made early. IVIG, plasmapheresis, or immunosuppressive therapy should be considered if there is no response to steroids.

**MeSH Keywords:** Immunoglobulins • Lupus Vasculitis, Central Nervous System • Methylprednisolone • Polyradiculoneuropathy, Chronic Inflammatory Demyelinating • Prednisone

**Full-text PDF:** <https://www.amjcaserep.com/abstract/index/idArt/903541>

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## Background

Chronic inflammatory demyelinating polyneuropathy (CIDP) is an uncommon manifestation of systemic lupus erythematosus (SLE). To our knowledge, few cases of CIDP and SLE have been previously reported in the literature [1].

CIDP is a debilitating clinical condition that is characterized by symmetrical polyneuropathy with histologic findings of demyelination and occasionally remyelination, and is believed to be an acquired autoimmune disorder that targets myelin [1]. CIDP continues to progress, or may relapse, for more than eight weeks, a clinical finding that differentiates it from acute inflammatory demyelinating polyneuropathy which is a monophasic sub-acute illness that reaches its nadir within three to four weeks. CIDP is characterized by muscular weakness with or without sensory loss in the extremities and can have a chronic progressive course with remission and repeated relapses [2].

The diagnosis of CIDP is more likely when the patient has a predominance of sensory symptoms over motor symptoms. Although the cause of CIDP is unknown, there is evidence to support an autoimmune etiology with multiple immunological triggers [3,4]. Both the cellular and humoral components of the immune system appear to be involved in the pathogenesis of CIDP and its variants [3,4]. An estimated 10–20% of SLE patients show peripheral nervous system involvement and patients present with sensorimotor polyneuropathies, with less common syndromes including mononeuritis multiplex or asymmetric polyneuropathy and acute or chronic demyelinating polyneuropathy [5]. Multiple factors, including early diagnosis of CIDP and presence of multiple antibodies associated with SLE, predict a good response to intravenous immunoglobulin (IVIG).

## Case Report

A 40-year-old African American woman with a past medical history of SLE, diagnosed at the age of 40 years, and treated with hydroxychloroquine, presented with a three-month history of slowly progressive tingling sensation and weakness in both her lower and upper extremities, and difficulty in walking. She initially presented with fatigue, fever, myalgia and arthralgia at the time of her diagnosis of SLE diagnosis. She had no known complications of SLE and no significant past medical history at the time of presentation. Review of her systems showed worsening fatigue, myalgia, headache, and numbness and some weakness of upper and lower extremities. The sensory and motor symptoms progressed in an ascending fashion resulting in impaired balance without bowel or bladder involvement.

On her current admission to hospital, the differential diagnosis of her symptoms was broad and included idiopathic

inflammatory myopathy, CIDP, subacute combined degeneration of spinal cord, cervical myelopathy, SLE neuropathy, thyroid myopathy, amyotrophic lateral sclerosis, multiple sclerosis, Eaton-Lambert syndrome, and paraneoplastic syndrome associated with an unknown primary malignancy.

Physical examination showed a grading for motor strength of 4/5, decreased temperature of the limb extremities, reduced pinprick and vibration sense and absent reflexes of both the upper and lower extremities, and an unsteady gait. Her sensory symptoms were more prominent when compared to her motor weakness. Her positive and negative autoimmune panel, determined by indirect immunofluorescence, is listed in Table 1.

Other laboratory investigations showed a normal complete metabolic panel, cerebrospinal fluid analysis, thyroid profile, brain magnetic resonance imaging (MRI), an erythrocyte sedimentation rate (ESR) of 75 mm/hr, low C4 complement, leukopenia, and anemia. No paraproteins were identified on serum electrophoresis. Electromyography (EMG) showed axonal demyelinating polyradiculoneuropathy, abnormal peroneal distal latency with very low amplitude and disappearance of F waves consistent with CIDP. The patient was treated with intravenous immunoglobulin (IVIG) 2 gm/kg daily for five days and prednisone 60 mg daily for a total of seven days, while continuing hydroxychloroquine.

Following treatment with IVIG and prednisone, the patient showed marked clinical improvement and regained her ability to walk with minor assistance. She was discharged home on a tapering dose of prednisone dose with follow-up in the rheumatology clinic.

## Discussion

Neurological and psychiatric symptoms are reported to occur in between 10–80% of patients with the diagnosis of systemic lupus erythematosus (SLE) during their illness [6,7]. The range of neurological manifestations of SLE includes central nervous system encephalitis, hemiparesis, brain infarcts, seizures, schizophrenia, atypical trigeminal neuralgia, acute demyelinating polyneuropathy, and chronic inflammatory demyelinating polyneuropathy (CIDP) [8].

In 1948 Sedgwick and Von Hagen first described the clinical manifestation of a CIDP-like illness in patients with SLE [9]. Retchland and coworkers stated that CIDP can have an unusual presentation and can precede systemic symptoms of SLE [10]. About 50% of the patients with a concomitant diagnosis of SLE and CIDP can achieve a good clinical response to IVIG, and the remainder may have a minimal response. Certain characteristics, including early CIDP diagnosis, the involvement of all four extremities, hyporeflexia or areflexia, slowed peripheral motor

**Table 1.** Panel of autoimmune serology serological markers tested.

Serum markers and antibodies	Patient result	Normal range
ANA	Positive	Negative
ANA titer	1: 1280	<1: 80
ANA pattern	Speckled	
C4 complement	14.3 mg/dl	18.0–55.0 mg/dl
C3 complement	90.5 mg/dl	79.0–152.0 mg/dl
Anti-ds DNA antibodies	Positive	Negative
Anti-ds DNA antibodies titer	1: 44 IU	<1: 25 IU
Anti SSA antibodies	Positive	Negative
Anti SSA antibodies titer	>1: 25 IU	<1: 20 IU
Anti SM antibodies	Positive	Negative
Anti SM antibodies titer	>1: 25 IU	<1: 20 IU
Anti SSB antibodies	Negative	Negative
Anti RNP antibodies	Negative	Negative
Anti SCL 70	Negative	Negative
Anti-histone antibody	Negative	Negative

ANA – antinuclear antibodies; SSA, SSB – Sjögren syndrome A and B; SM – smooth muscle; RNP – ribonucleoprotein; SCL – scleroderma.

nerve conduction velocity, SLE involvement of critical internal organs, and the presence of multiple auto-antibodies associated with SLE, are of a good response to IVIG [1]. Patients with SLE and CIDP may present with recurrent episodes of Guillain-Barré syndrome-like symptoms, mononeuritis multiplex, or symmetric polyradiculopathy, over a period of weeks to months.

The proposed pathological mechanisms for the clinical symptoms of CIDP in SLE patients include abnormalities in blood vessels that supply the epineurium, resulting in nerve fiber loss, inflammation that involves the interstitial and causes separation of nerve fibers, and reduction in the amount of myelin [11,12]. CIDP is now believed to be an autoimmune mediated process involving cellular and humoral response that involve T-cells, activated macrophages, cytokines, anti-myelin antibodies, and costimulatory inflammatory molecules, resulting in loss of myelin and axonal loss [11,12].

The neuropathy of CIDP is usually polyradicular, symmetrical, and involves both proximal and distal muscles. [13]. Electromyography (EMG) testing in CIDP may help to determine the extent of sensory and motor deficits and categorize the demyelinating process (prolonged terminal latency, slowing of nerve conduction velocity, dispersion and conduction block) and axonal form of neuropathy (marginal slowing of nerve conduction, small compound muscle or sensory action potential, and denervation on EMG) [14]. Hereditary forms of

demyelination are suggested by features of uniform demyelination, whereas variable demyelination of nerve segments of the same nerve favor acquired demyelination [14]. Demyelination requires slowing of conduction velocities, according to the criteria by American Academy of Neurology (AAN) [15]. Most CIDP patients do not meet these AAN criteria, as conduction slowing can be absent when not enough fibers are affected when demyelination is proximal and not affected by distal stimulation, or when there is severe secondary axonal degeneration [16]. A study by Haq and colleagues showed that there was only 42% sensitivity of the electrodiagnostic criteria proposed by the AAN in CIDP patients who had sural nerve biopsies [17].

The presence of anticardiolipin antibodies in CIDP may also reflect myelin damage [18]. For patients with sensorimotor neuropathy, there is a good response to steroid pulse or IVIG treatment, despite age, concurrent illness, and positive anticardiolipin antibodies [18]. According to Sindern and colleagues, CIDP is also reported to occur episodically in other autoimmune diseases, including myasthenia gravis, acute glomerulonephritis, Hashimoto's thyroiditis, and multiple sclerosis [19]. Very rare associations have been reported with diseases that include hemophagocytic lymphohistiocytosis, which has an estimated prevalence of 0.9–4.6% in patients with SLE [20].

CIDP can occur before, after, or simultaneously with the onset of SLE. As shown in this case report, early identification of

peripheral limb weakness, areflexia or hyporeflexia, and slowed nerve conduction is crucial before treatment is commenced with steroids, IVIG, or plasmapheresis, all of which may be life-saving. Apart from treatment with steroids and IVIG, other immunosuppressive agents including azathioprine, mycophenolate, rituximab and cyclophosphamide may be considered [21,22]. Inflammatory neuropathies respond to therapy with glucocorticoids in moderate to higher doses, such as prednisone 30–60 mg/day, but not all patients improve on this treatment. Therapy with glucocorticoids, IVIG, or plasmapheresis may be indicated in CIDP. Evidence of axonal damage on EMG, or vasculitis on nerve biopsy, are indications for initiating more potent immunosuppressive therapy, such as cyclophosphamide. In 2005, Vina and colleagues suggested that more aggressive treatment should be used when more severe systemic manifestation of SLE were in order to suppress the immune-mediated processes involved in the pathogenesis of CIDP [1].

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## Conclusions

CIDP is an acquired autoimmune peripheral neuropathy that can be an unusual association seen in patients with SLE. As this case report has shown, early clinical diagnosis of CIDP, supported by serological autoantibody profiles associated with SLE, can predict a good response to steroids. Most patients with CIDP are treated successfully with steroids if the diagnosis is made early. IVIG, plasmapheresis, or immunosuppressive therapy should be considered if there is no response to steroids.

## Conflict of interest

None.