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CASE REPORT

ATYPICAL CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY WITH RELAPSING SENSORY-PREDOMINANT SENSORIMOTOR NEUROPATHY

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Abstract

Introduction: Chronic inflammatory demyelinating polyneuropathy (CIDP) is a rare, acquired autoimmune neuropathy with progressive symptoms. Intravenous immunoglobulin (IVIg) and corticosteroids are effective first-line therapies; however, many patients experience residual symptoms or relapse within the first year.

Case Presentation: A 39-year-old male with an initial diagnosis of carpal tunnel syndrome presented with a 13-month history of progressive sensory disturbances, evolving from lower limb paresthesia with gait instability. EMG confirmed demyelinating and axonal sensorimotor polyneuropathy consistent with CIDP. He was hospitalized and responded well to intravenous methylprednisolone and IVIg, but relapsed nine months later after a respiratory infection, presenting with distal sensory symptoms and preserved strength. Cerebrospinal fluid analysis showed albuminocytologic dissociation, while autoimmune nodopathy markers were negative. Retreatment with IVIg and methylprednisolone led to clinical improvement, with only mild exertion-related symptoms at two-month follow-up.

Discussion: This case highlights a rare sensory-predominant variant of CIDP with relapse triggered by infection. Relapses often require renewed immunotherapy, with corticosteroids and IVIg as first-line treatments; long-term management may include maintenance IVIg and gradual steroid tapering.

Conclusions: Early recognition, appropriate immunotherapy, and long-term monitoring are essential to sustain remission and prevent disability in sensory-predominant CIDP.

Keywords: Chronic inflammatory demyelinating polyneuropathy; electrodiagnostic studies; immunotherapy; relapse; sensory-predominant

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Introduction

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) is a rare, acquired autoimmune neuropathy characterized by progressive symptoms lasting at least 8 weeks.[1] It involves demyelination of both motor and sensory nerves within the peripheral nervous system and nerve roots, leading to conduction block, reduced conduction velocity, muscle weakness, and sensory

impairment. Although uncommon, CIDP is the most frequent autoimmune disorder of the peripheral nervous system, with a global prevalence estimated between 0.67 and 10.3 per 100,000 individuals. Its pathogenesis reflects a combination of cellular and humoral immune mechanisms: T-cell activation, macrophage-mediated demyelination, and deposition of immunoglobulin G and M (IgG and IgM, respectively) on Schwann cells contribute to myelin injury and secondary axonal degeneration.[2,3]

While typical CIDP accounts for 50-60% of cases, sensory-predominant CIDP is much less common, representing only 5-35%.[1,4,5] It is characterized by gait ataxia, impaired proprioception, and altered vibration or cutaneous sensation with little or no weakness. Diagnosis requires symptoms persisting beyond 8 weeks, supported by electrodiagnostic evidence of demyelination in sensory nerves, with or without motor conduction abnormalities. The 2021 European Academy of Neurology/Peripheral Nerve Society (EAN/PNS) guidelines emphasize the role of supportive findings, such as treatment response, cerebrospinal fluid (CSF) albuminocytologic dissociation, or nerve imaging abnormalities. Antibody testing is also recommended to exclude autoimmune nodopathy, which differ in prognosis and treatment response.[1,4]

Intravenous immunoglobulin (IVIg) and corticosteroids are effective first-line therapies; however, many patients experience residual symptoms or relapse, often within the first year of treatment[2,6] Relapse in sensory-predominant CIDP is particularly rare and poses additional diagnostic and therapeutic challenges. To our knowledge, most reports describe sensory CIDP as stable or slowly progressive disease[7], with relapses usually reported in the typical, motor-involved variant. This limited documentation underscores the uniqueness of the present case and highlights its value in expanding understanding of the clinical course and management of relapsing sensory-predominant CIDP.

Case Presentation

A 39-year-old male presented with a 13-month history of progressive sensory disturbances. Eight months earlier, in September 2023, he experienced numbness and tingling in the fingertips. Electrodiagnostic studies revealed bilateral ulnar sensory axonal neuropathy, and he was diagnosed with left-sided carpal tunnel syndrome (CTS, grade 1), treated with oral methylcobalamin, methylprednisolone, and omeprazole.

By February 2024, he developed new-onset paresthesia over the dorsum of both feet that gradually progressed proximally to the calves, leading to gait instability and a sensation of heaviness in the lower extremities. However, he did not seek medical care, assuming these symptoms were related to his prior CTS diagnosis. Symptoms later extended to the forearms and worsened by June 2024, leading to difficulty in walking and stair climbing. Repeat electromyography (EMG) demonstrated demyelinating and axonal sensorimotor polyneuropathy affecting both upper and lower limbs, consistent with chronic inflammatory demyelinating polyneuropathy (CIDP). He

was hospitalized and received a 5-day course of intravenous methylprednisolone and intravenous immunoglobulin (IVIg), with marked symptomatic improvement and restoration of daily function.

In March 2025, following an upper respiratory infection, he experienced a relapse with recurrent sensory symptoms (INCAT disability score: 4). EMG confirmed ongoing demyelinating polyneuropathy, although with improved compound motor action potential (CMAP) amplitudes compared to previous studies. Serum testing for CIDP-associated antibodies was negative for Contactin-1 IgG and Neurofascin-155 IgG4. Cerebrospinal fluid (CSF) demonstrated albuminocytologic dissociation (protein 56 mg/dL; 3 cells/µL, 100% mononuclear; CSF/serum glucose ratio 65%).

The patient was readmitted in May 2025 due to progressive symptoms. He was treated with intravenous methylprednisolone (4 × 250 mg, tapered) and a 5-day course of IVIg (5% 2.5 g/, 10 vials daily), resulting in gradual symptom improvement. Mild erythema and pruritus developed during corticosteroid therapy but resolved spontaneously (INCAT disability score: 2). He was discharged with oral methylcobalamin and was planned for immunosuppressive maintenance therapy.

At the two-month follow-up in July 2025, the patient reported only minimal sensory symptoms occurring during fatigue (INCAT disability score: 1).

Discussion

Sensory-predominant CIDP is an atypical variant that often leads to diagnostic delays, as it can resemble other neuropathies such as autoimmune nodopathy or chronic immune sensory polyradiculopathy (CISP). CISP is confined to the sensory roots and presents with large-fiber sensory loss, gait ataxia, frequent falls, and preserved strength, with normal nerve conduction studies and EMG. In contrast, autoimmune nodopathy is characterized by antibodies against nodal or paranodal proteins (e.g., neurofascin-155, Contactin-1, Caspr1) and typically manifests with asymmetric sensory or sensorimotor involvement, severe ataxia, tremor, poor response to IVIg, and distinctive MRI findings of root or plexus enlargement. Clinical red flags that help differentiate sensory-predominant CIDP from these conditions include comorbid diabetes mellitus, vitamin B12 deficiency, prior exposure to neurotoxic chemotherapy, or systemic symptoms suggestive of immune-mediated pathology. In the present case, electrophysiological studies revealed sensorimotor changes, underscoring subclinical motor involvement despite the predominance of sensory symptoms, which is consistent with prior observations that even sensory-predominant CIDP may exhibit subtle motor abnormalities on nerve conduction studies.[11,12]

Relapses are generally less common in sensory-predominant CIDP compared with typical CIDP, in which relapses affect approximately 30-50% of patients, often triggered by infections or other immune challenges.[8] However, the recurrent episode in this case indicate that even variants considered relatively indolent

may follow a fluctuating course similar to typical CIDP. Clinically, relapses may mimic other causes of acute or subacute sensory neuropathy, emphasizing the importance of thorough differential diagnosis. Electrodiagnostic confirmation, cerebrospinal fluid analysis, and antibody testing are recommended in complex or atypical presentations, as outlined in the 2021 EAN/PNS guideline.[4]

Corticosteroids and IVIg remain effective first-line therapies. Induction IVIg is typically administered at 2 g/kg over 2-5 consecutive days, with recommended maintenance dosing of 1 g/kg every 3 weeks based on clinical response.[4,13] Corticosteroids can be administered either as intravenous methylprednisolone, 500 mg/day for 4 consecutive days repeated every 4 weeks, or as oral prednisolone, starting at 60 mg/day for 4 weeks, followed by gradual tapering over 6–12 months. Alternate-day regimens may be considered to minimize long-term side effects.[4] Sensory-predominant CIDP patients often respond favorably to immunotherapy[7,9,11], with some evidence suggesting superior steroid responsiveness compared to typical CIDP. Early treatment initiation and milder baseline impairment are associated with improved outcomes, whereas higher baseline disability scores, such as Inflammatory Neuropathy Cause and Treatment (INCAT), correlate with increased relapse risk.[8,14] Regular neurological assessment, functional scoring, and follow-up electrophysiological studies, combined with patient education on early relapse recognition, are critical to guide timely therapy and prevent permanent deficits.

This case contributes to the limited literature on relapsing sensory-predominant CIDP and underscores its clinical relevance. Recognition of this variant is crucial to ensure timely initiation of immunotherapy and prevent cumulative disability. Future research should focus on identifying prognostic factors for relapse and optimizing long-term management, including individualized immunotherapy schedules and monitoring strategies, to improve outcomes and expand understanding of disease variability within the CIDP spectrum.

Conclusion

Sensory-predominant CIDP is an atypical variant with relapses occurring far less frequently than in typical CIDP. Relapse can lead to worsening sensory deficits and functional impairment. Early initiation of therapy has been associated with better recovery and lower residual disability, highlighting the importance for prompt diagnosis, appropriate immunotherapy, and comprehensive long-term management to sustain remission and optimize functional outcomes.

Conflict of Interest

The authors declared no conflict of interest.

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