CASE REPORT

# Sensory Chronic Inflammatory Demyelinating Polyneuropathy—A Variant of Chronic Inflammatory Demyelinating Polyneuropathy: A Rare Case Report

Sanjeev K Saggar[1], Richa G Thaman[2], Kawalinder K Girgla[3]

## Abstract

**Background:** Chronic inflammatory demyelinating polyneuropathy (CIDP) is progressive, sometimes relapsing disorder of peripheral nerves. It usually develops over months or years. Abrupt onset resembling Guillain-Barre syndrome (GBS) can occur and symptoms can persist. Pure sensory form of CIDP is rare and visualized in just 5% patients of CIDP.

**Case description:** A 44-year-old female presented to emergency department with a 12-month history of progressively worsening numbness in hands, feet, and extreme weakness. She had decreased pin prick and touch sensation in a stocking-glove distribution. Patient's history and nerve conduction study were indicative of sensory variant of CIDP.

**Discussion:** In patients who exhibit pure sensory form of CIDP, clinically sensory deficit predominantly manifests. Moreover, there is decrease in amplitude of sensory nerve action potential (SNAP) in case of sensory nerve, i.e., sural nerve, median sensory nerve, etc. In a given case, nerve conduction studies showed more than 50% decrease in SNAP. A significant decrease in amplitude of SNAP with no motor nerve conduction abnormality is the requirement for the sensory variant of CIDP, which is a characteristic feature seen in the given case.

**Keywords:** Chronic inflammatory demyelinating polyneuropathy (CIDP), Compound muscle action potential (CMAP), Guillain-Barre syndrome (GBS), Nerve conduction studies (NCS), Sensory nerve action potential (SNAP).

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## Introduction

Chronic inflammatory demyelinating polyneuropathy (CIDP) is progressive, sometimes relapsing disorder of peripheral nerves. It is a steroid-dependent demyelinating sensorimotor neuropathy which primarily affects the limbs. It usually develops, over months or years. Usually, onset of neurological deficit is critical for diagnosis of CIDP and it generally exceeds 8 weeks.[1] This disease can strike at any age; however, its prevalence increases between 40 and 60 years of age. Abrupt onset resembling Guillain-Barre syndrome (GBS) can occur and symptoms can persist.[2] Relapsing variety and chronic progressive variety are visualized in CIDP. About 60% of patients have chronic progressive course while one-third have relapsing remitting type. Generally, the lower limbs are more affected than the upper limbs. Weakness is a characteristic feature visualized in proximal and distal muscles which could be due to the root and peripheral nerve involvement. In it, distal paresthesia is accompanied by weakness at the time of presentation.

More than 80% of patients presented with mixed sensory motor neuropathy which is relatively symmetrical. Generally, predominantly motor or sensory form is encountered rarely. Very few cases of sensory polyneuropathy have been identified as an entity within the spectrum of CIDP.[3,4] The pure sensory form of CIDP is rare and is seen in just 5% patients of CIDP. Sensory form manifests with entirely sensory symptoms and signs. However, disease course and response to steroids is similar to conventional sensory motor type of CIDP.[3–5] We report here the details of the case which presented as pure sensory variant of CIDP. Also, the details of the patient regarding clinical and electrophysiological features are being presented in this report.

[1–3]Department of Physiology, SGRDIMSAR, Amritsar, Punjab, India

**Corresponding Author:** Richa G Thaman, Department of Physiology, SGRDIMSAR, Amritsar, Punjab, India, Phone: +91 9815963778, e-mail: richaghaythaman@yahoo.co.in

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## Case Description

A 44-year-old female patient presented to emergency department with a 12 month history of progressively worsening numbness in her hands, feet, and extreme weakness. She had impaired balance which caused infrequent falls. Patient did not smoke, drink alcohol, used drugs or dietary supplements. However, patient had noncompressive myelopathy with headache. Neurological examination revealed decreased pin prick and touch sensation in a stocking-glove distribution. The patient exhibited wide-based stance and gait, walk was unsteady, tandem gait was performed with difficulty, and Romberg's sign was positive with closed eyes. Magnetic resonance imaging of the brain and the spine did not show any acute changes. During the first day of admission, nerve conduction studies were conducted on the patient whereby median, ulnar, tibial, peroneal, and sural nerves were studied. These studies showed acute sensory neuropathy affecting the lower limb and the upper limb.

**Table 1:** Nerve conduction study report of patient

| *Sensory nerve* | *Latency (ms)* | *Amplitude (μv)* | *Velocity (m/s)* |
|---|---|---|---|
| Right median sensory nerve | | Nonstimuable | |
| Left median sensory nerve | 1.25 | 7.3* | 37.85 |
| Right ulnar sensory nerve | 1.17 | 5.4* | 39.83 |
| Left ulnar sensory nerve | 1.42 | 5.2* | 39.31 |
| Right sural nerve | 2.04 | 8.5* | 43.92 |
| Left sural nerve | 2.17 | 8.9* | 40.69 |

*Abnormally reduced value

Patient's history and nerve conduction studies were strongly indicative of sensory variant of CIDP.

Nerve conduction studies revealed that the right median sensory nerve was nonstimulable while the left median sensory nerve showed decrease in SNAP amplitude. It was less than 50% of lower limit of normal sensory amplitude. Similarly, the ulnar sensory nerve bilaterally showed decrease in SNAP amplitude, with mild decrease of nerve conduction velocity. However, no conduction block was visualized (Table 1).

In case of sural nerve, SNAP amplitude was reduced bilaterally. It was less than 50% of lower limit of normal sensory amplitude. Cerebral spinal fluid (CSF) analysis was done within 3 days of initial presentation. It revealed an elevated protein level of 89 mg/dL against normal value (45–60 mg/dL).

Patient was treated with intravenous immunoglobulin (IVIG) therapy for the treatment of CIDP by a neurologist. Patient showed gradual improvement with IVIG therapy and was discharged after hospital stay of 10 days.

## Discussion

This case report describes a patient of CIDP. It is a pure sensory form of CIDP. It is a rare case because pure sensory variant is found only in 5% cases of CIDP.[5] Patient did not exhibit any history of diabetes, hematological, or neoplastic disorder. Patient had symptoms of acute numbness, pain in extremities especially lower extremities. The given case exhibited (i) pure sensory deficits; (ii) a significant decline in amplitude of sensory nerve action potential (SNAP) and in sural nerve, median, and ulnar sensory nerve; (iii) there was complete absence of any motor nerve conduction abnormality as far as CMAP, latency, nerve conduction velocity were concerned in the motor nerves; (iv) the protein level was elevated without cells in CSF. This finding is a characteristic feature in 85% cases of CIDP as noted from other studies.[6]

Majority patients with sensory symptoms are likely to develop subclinical motor conduction abnormalities and become compatible with CIDP.[7,8] Immune-mediated sensory polyradiculoneuropathy is recognized as a specific type of CIDP.[9]

This case can be described as chronic sensory demyelinating neuropathy. It exhibited features of sensory variant of CIDP as sensory neuropathy showed chronic progression over a period of 12 months. Patient exhibited polyneuropathy, pain sensation, diminished pin prick, and touch sensation in stocking-glove distribution. Motor weakness was absent in reported case. Moreover, electrophysiological findings and elevated protein level substantiate the findings as a sensory variant of CIDP.[1]

## Conclusion

Pure sensory variant is a rare presentation in CIDP and can be tricky to diagnose due to variation in clinical presentation. Nerve conduction study is an important hallmark which helps in diagnosis of sensory variant of CIDP as affected neurophysiological variables, i.e., SNAP and NCV are put in light in clear cut terms. Proper diagnosis of CIDP with its variant helps to manage the patient in a better manner.

## References


1. McCombe PA, Pollard JD, McLeod JG. Chronic inflammatory demyelinating polyradiculoneuropathy. A clinical and electrophysiological study of 92 cases. Brain 1987;110(Pt 6): 1617–1630. DOI: 10.1093/brain/110.6.1617.
2. Mori K, Hattori N, Sugiura M, et al. Chronic inflammatory demyelinating polyneuropathy presenting with features of GBS. Neurology 2002;58(6):979–982. DOI: 10.1212/wnl.58.6.979.
3. Oh SJ, Joy JL, Kuruoglu R. "Chronic sensory demyelinating neuropathy": chronic inflammatory demyelinating polyneuropathy presenting as a pure sensory neuropathy. J Neurol Neurosurg Psychiatry 1992;5(8):677–680. DOI: 10.1136/jnnp.55.8.677.
4. Sinnreich M, Klein CJ, Daube JR, et al. Chronic immune sensory polyradiculopathy: a possibly treatable sensory ataxia. Neurology 2004;63(9):1662–1669. DOI: 10.1212/01.WNL.0000142507.12763.58.
5. Mishra UK, Kalita J. Clinical Neurophysiology. 3rd ed. New Delhi, India. Elsevier;2014. p. 219.
6. Busby M, Donaghy M. Chronic dysimmune neuropathy. A subclassification based upon the clinical features of 102 patients. J Neurol 2003;250(6):714–724. DOI: 10.1007/s00415-003-1068-2.
7. van Dijk GW, Notermans NC, Franssen H, et al. Development of weakness in patients with chronic inflammatory demyelinating polyneuropathy and only sensory symptoms at presentation: a long-term follow-up study. J Neurol 1999;246(12):1134–1139. DOI: 10.1007/s004150050531.
8. Rajabally YA, Wong SL. Chronic inflammatory pure sensory polyradiculoneuropathy: a rare CIDP variant with unusual electrophysiology. J Clin Neuromuscul Dis 2012;13(3):149–152. DOI: 10.1097/CND.0b013e31822484fb.
9. Ayrignac X, Viala K, Koutlidis RM, et al. Sensory chronic inflammatory demyelinating polyneuropathy: an under-recognized entity? Muscle Nerve 2013;48(5):727–732. DOI: 10.1002/mus.23821.