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**Case Report**

# Successful management of chronic inflammatory demyelinating polyradiculoneuropathy: a rare case report

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## ABSTRACT

Acute immune-mediated demyelinating polyradiculoneuropathy is a rare neuropathic disorder characterized by significant motor involvement. The remitting and relapsing form of this disease is known as chronic inflammatory demyelinating polyradiculoneuropathy (CIDP). This case report presents the clinical features, diagnostic process, and successful management of a 37-year-old female patient with CIDP. The patient visited the emergency department with complaints of fever, numbness, and progressive weakness in all limbs. There was a similar previous history of weakness in limbs. The patient was treated with administration of high-dose methylprednisolone which led to resolution of symptoms. This case shows the importance of thorough clinical evaluation and prior history taking.

**Keywords:** CIDP, Polyradiculoneuropathy, Demyelinating, Neuropathy, Quadriplegia, Case report

## INTRODUCTION

Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP) is a rare disorder of the peripheral nervous system characterized by demyelination and impaired nerve conduction, resulting in prominent motor deficits.[1] This autoimmune disorder attacks own body tissues called myelin sheaths, which are fatty coverings on the nerves in the brain and the spinal cord.[2] This leads to polyneuropathy of the nerve roots and peripheral nerve inflammation leading to sensory impairment and motor-dominant weakness.[2,3] Incidence and prevalence rates are poorly defined, it is found based on a meta-analysis study that prevalence of CIDP ranges from 0.7 to 10.3 per 100,000 people.[4] The incidence rate is seen to be greater in men compared to women and in elderly population of >50 years old.[5] Most cases are idiopathic, and this condition can follow a chronic, relapsing, and remitting course, posing diagnostic and therapeutic challenges.[6-8] Prompt recognition and treatment are crucial to prevent long-term disability and improve patient outcomes.[9,10] CIDP typically presents with ascending muscle weakness, sensory deficits, and occasionally cranial nerve involvement.[6,11] In this case report, we discuss the clinical presentation, management, and outcome of CIDP in a 37-year-old female,

emphasizing the importance of thorough clinical evaluation and the role of past medical history in achieving an accurate diagnosis and effective treatment.

## CASE REPORT

A 37-year-old female presented to Rajindra Hospital, Patiala, with complaints of fever, numbness, and progressive weakness in all limbs. The weakness began in the left lower limb, progressed to the left upper limb, and eventually affected the right side. The disease also involved bilateral facial muscles but spared bowel and bladder functions. The patient had experienced a similar episode 18 months earlier. On examination, the patient exhibited flaccid, a-reflexic quadriplegia with significant sensory loss in all four limbs.

Additionally, there was bilateral lower motor neuron type of 7th cranial nerve palsy. Following are the diagnostic evaluations to rule out alternative diagnoses causing nerve damage. Nerve conduction/Electrophysiological studies show increased latent period and decreased velocity without any prominent change in the amplitude of graph, probably due to a demyelinating pathology. MRI showing Early degenerative disease of lumbar spine with mild diffuse bulges and neural compromise. Management included treatment for fever and administration of high-dose methylprednisolone, which led to a resolution of symptoms.

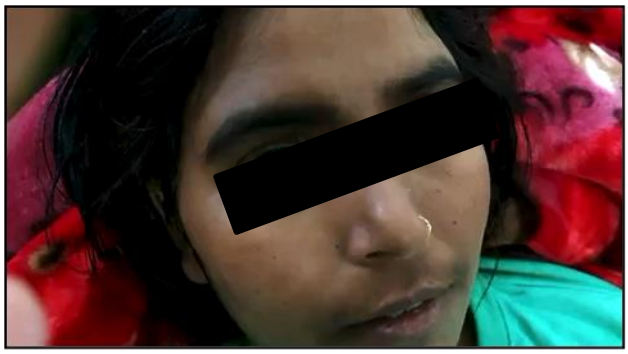

**Figure 1: Clinical features showing bilateral LMN facial nerve palsy.**

**Figure 2: Decreased power at wrist joint causing wrist drops.**

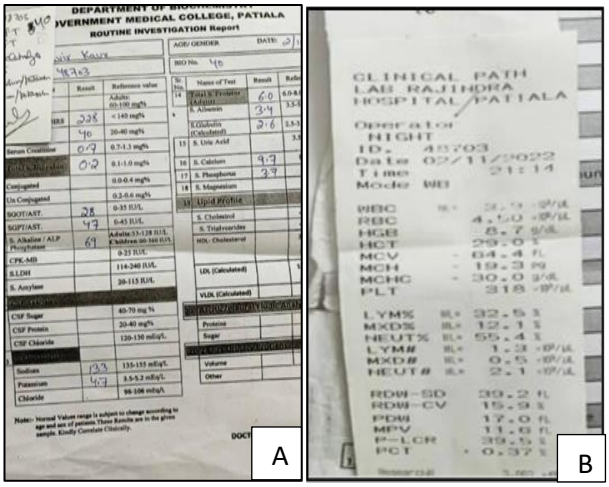


**Figure 3: (A) Complete blood picture. (B) Biochemical investigation.**

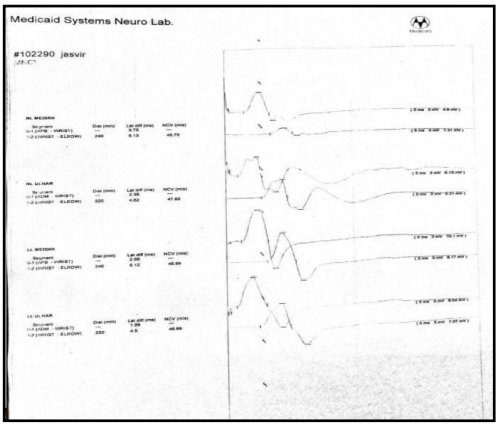


**Figure 4: Nerve conduction/electrophysiological studies.**

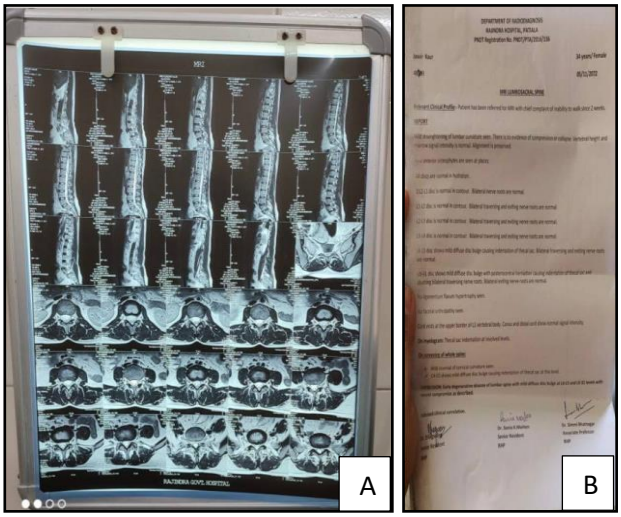


**Figure 5: (A) Radiological investigation-MRI of lumbar spine (film). (B) Radiological investigation-MRI of lumbar spine (report).**

## DISCUSSION

This case highlights the clinical presentation and management of CIDP, a rare yet treatable condition. The patient's history of a similar episode 18 months prior, along with the acute onset of fever, numbness, and progressive weakness, was pivotal in diagnosing CIDP. The involvement of both upper and lower limbs, coupled with facial paralysis and sensory loss, is consistent with typical CIDP presentations. The sparing of bowel and bladder functions helps differentiate CIDP from other neuropathies. According to the diagnostic criteria for CDIP by European federation of neurological societies (EFNS) in cooperation with the peripheral nerve society (PNS) established in 2021, any case of recurrent or worsening symptoms polyradiculoneuropathy should be suspected of CDIP.[12,13] In line with these guidelines, nerve conduction test as well as MRI examinations must be supplemented to rule out diagnosis.[14]

The administration of high-dose methylprednisolone was effective in resolving the patient's symptoms, emphasizing the importance of timely intervention. The response to corticosteroids supports the immune-mediated nature of CIDP, involving inflammation and demyelination. The primary goal of therapy in CDIP is improving strength and functional ability in the patient literature has shown that immunomodulating treatments such as corticosteroids, plasma exchange, and intravenous (IV) immunoglobulin have all been successful.[15] The choice of therapy seems, to have varied depending on several factors such as cost, time to administer, availability, adverse event profile and disease severity.[16]

The recurrence of similar symptoms 18 months earlier indicates the chronic, relapsing nature of CIDP, highlighting the need for ongoing monitoring and potentially long-term immunomodulatory therapy to prevent future exacerbations. Corticosteroids have worked in this scenario since its given in high dose and produces a quick immune response, however low maintenance dose needs to be administered to sustain remission.[8] Long-term steroid therapy is well known to be associated with potential adverse events and thus seeks novel interventions for CDIP.[17] Several studies including animal models recently have increased understanding of cellular mechanics for CDIP and development of targeted therapies which not just provide symptomatic relief but also mechanism driven.[18]

## CONCLUSION

This case underscores the importance of comprehensive clinical evaluation and consideration of past medical history in diagnosing CIDP. Early and appropriate treatment can lead to significant improvement in symptoms and prevent long-term disability. CIDP, though potentially life-threatening, is a reversible condition with timely intervention. Further research is needed to explore optimal treatment strategies and long-term outcomes in CIDP patients, particularly those presenting with acute exacerbations similar to this case.

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## REFERENCES

1. Chronic inflammatory demyelinating polyradiculoneuropathy. Available at: www.hopkinsmedicine.org. Accessed on 17th March 2024.
2. Chronic inflammatory demyelinating polyradiculoneuropathy: practice essentials, background, pathophysiology. Medicine. 2024. https://emedicine.medscape.com/article. Accessed on 17 March 2024
3. Chronic inflammatory demyelinating polyneuropathy-NORD (National Organization for Rare Disorders). 2015. Available at: https://rarediseases.org/rare-diseases/chronic. Accessed on 12th March 2024.
4. Broers Merel C, Bunschoten C, Nieboer D, Lingsma Hester F, Jacobs Bart C. Incidence and prevalence of chronic inflammatory demyelinating polyradiculoneuropathy: a systematic review and meta-analysis. Neuroepidemiol. 2019;52(3-4):161-72.
5. Broers MC, Marcel de Wilde, Lingsma HF, van, Verhamme K, Jacobs BC. Epidemiology of chronic inflammatory demyelinating polyradiculoneuropathy in The Netherlands. J Peripheral Nerv Sys. 2022;27(3):182-88.
6. Gogia B, Rocha Cabrero F, Rai PK. Chronic Inflammatory Demyelinating Polyradiculoneuropathy. PubMed. 2022. Available at: https://www.ncbi.nlm.nih.gov/books.
7. Mathey EK, Park SB, Hughes RAC. Chronic inflammatory demyelinating polyradiculoneuropathy: from pathology to phenotype. J Neurol, Neurosurg Psych. 2015;86(9):973-85.
8. Dalakas MC. Advances in the diagnosis, pathogenesis and treatment of CIDP. Natu Rev Neurol. 2011;7(9):507-17.
9. Quint P, Schroeter CB, Kohle F. Preventing long-term disability in CIDP: the role of timely diagnosis and treatment monitoring in a multicentre CIDP cohort. J Neurol. 2024;271(9):5930-43.
10. Van Doorn IN, Eftimov F, Wieske L, van Schaik IN, Verhamme C. Challenges in the early diagnosis and treatment of chronic inflammatory demyelinating polyradiculoneuropathy in adults: current perspectives. Therap Clin Risk Manag. 2024;20:111-26.
11. Van den Bergh PYK, Rajabally YA. Chronic inflammatory demyelinating polyradiculoneuropathy. La Presse Médicale. 2013;42(6):203-15.

12. Van den Bergh PYK, Doorn PA, Hadden RDM. European academy of neurology/peripheral nerve Society guideline on diagnosis and treatment of chronic inflammatory demyelinating polyradiculoneuropathy: Report of a joint task force-second revision. European J Neurol. 2021;28(11):3556-83.
13. Dziadkowiak E, Waliszewska-Prosół M, Nowakowska-Kotas M, Budrewicz S, Koszewicz Z, Koszewicz M. Pathophysiology of the different clinical phenotypes of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP). Int J Molec Sci. 2021;23(1):179.
14. Bunschoten C, Jacobs BC, Van den Bergh PYK, Cornblath DR, van Doorn PA. Progress in diagnosis and treatment of chronic inflammatory demyelinating polyradiculoneuropathy. The Lancet Neurol. 2019;18(8):784-94.
15. Lopate G, Pestronk A, Al-Lozi M. Treatment of chronic inflammatory demyelinating polyneuropathy with high-dose intermittent intravenous methylprednisolone. Archives Neurol. 2005;62(2):249.
16. Ropper AH. Current treatments for CIDP. Neurol. 2003;60(3):16-22.
17. Nobile-Orazio E, Cocito D, Jann S. Intravenous immunoglobulin versus intravenous methylprednisolone for chronic inflammatory demyelinating polyradiculoneuropathy: a randomised controlled trial. The Lancet Neurol. 2012;11(6):493-502.
18. Brun S, de Sèze J, Muller S. CIDP: current treatments and identification of targets for future specific therapeutic intervention. Immuno. 2022;2(1):118-31.

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