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CASE REPORT: HEMIPARETIC PRESENTATION OF MULTIFOCAL CIDP

Neurology

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ABSTRACT

Background: Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP) is a rare immune-mediated neuropathy that predominantly affects motor and sensory nerves. It can present with symmetric or asymmetric limb involvement, with multifocal patterns being less common. Early diagnosis and treatment are essential to prevent long-term disability. **Case Report:** A 32-year-old male presented with a two-year history of progressive, asymmetric weakness and sensory deficits in the right upper and lower limbs, with distal muscles more severely affected than proximal. Clinical examination revealed muscle wasting, decreased strength, sensory loss, and areflexia in the affected areas. Investigations, including nerve conduction studies, showed features of demyelination with secondary axonal degeneration. A nerve biopsy confirmed axonal neuropathy with onion bulb formation. The patient was diagnosed with multifocal CIDP and initiated on immunosuppressive therapy. **Conclusion:** This case underscores the importance of recognizing atypical presentations of CIDP, such as this case, which had a hemiparetic presentation. A detailed clinical and diagnostic evaluation is necessary for timely immunosuppressive treatment, which is crucial to halt disease progression and improve patient outcomes.

KEYWORDS

CIDP, MADSAM, Multifocal CIDP, Hemiparetic CIDP.

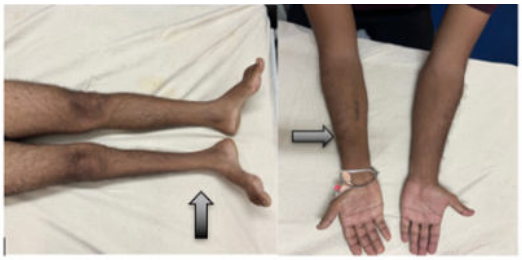
INTRODUCTION:

Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP) is a rare Autoimmune Neuropathy that affects motor and sensory nerves. Multifocal presentations are less common and pose diagnostic challenges[1]. This report details a case of multifocal CIDP with asymmetric limb involvement.

Case Details:

A 32-year-old gentleman presented with a two-year history of insidious onset and gradually progressive weakness in the right upper and lower limbs, predominantly distal. The weakness was associated with sensory deficits, muscle thinning, and areflexia. There was no history suggestive of cranial nerve involvement, bowel or bladder dysfunction, cerebellar symptoms, cognitive issues, or autonomic disturbances.

Clinical examination revealed muscle wasting and decreased strength in the right upper and lower limbs, decreased pain and temperature sensation, and a right high-steppage gait. Reflexes were diminished in the affected limbs. The left upper and lower limbs examination was unremarkable.



Pictures showing wasting of the Right Lower and upper limbs

Investigations:

Initial laboratory tests, including Complete Blood Picture, Renal and Liver Function Tests, and Viral markers, were normal. Auto Immune workup was negative. CT and MRI brain imaging showed no abnormalities. Nerve Conduction Studies demonstrated decreased Conduction Velocities, Conduction Blocks, and Reduced Compound Muscle Action Potential (CMAP) in the right median, ulnar, radial, peroneal, and tibial nerves, with normal findings on the left. Sensory NCS revealed absent Sensory Nerve Action Potential (SNAP) in the right median, ulnar, and sural nerves. CSF analysis revealed elevated protein (64 mg/dL) with normal glucose and Cell Count. High-resolution ultrasound showed loss of fascicular pattern in the Right Median, Ulnar, and Common Peroneal nerves, and Hyperechoic Calf muscles suggestive of Fatty Infiltration. Nerve biopsy findings demonstrated Axonal Neuropathy, along with Onion Bulb formation, indicative of Demyelination.

Diagnosis: Based on clinical presentation, nerve conduction studies, and biopsy findings, the patient was diagnosed with multifocal CIDP.

Management: The patient was started on IV Corticosteroids, IV Immunoglubulins followed by maintenance immunosuppression

DISCUSSION:

In the present case report, we describe a patient with motor weakness in the Right Upper and Lower extremities a hemiparetic presentation caused by MADSAM. MADSAM is a chronic acquired demyelinating neuropathy [2]. Symptoms of MADSAM include asymmetric weakness in the distribution of individual peripheral nerves combined with sensory deficit. MADSAM is caused by autoimmune mediated inflammation and has an onset age in the early 50s [3]. It is challenging for clinicians to recognize MADSAM hence, its diagnosis is often delayed for years. Because MADSAM usually shows an excellent response to IVIg or corticosteroid treatment, early treatment tends to result in better outcomes [4].

CONCLUSION:

This case highlights the importance of recognizing atypical presentations of CIDP, including Asymmetric or Multifocal involvement. A comprehensive diagnostic approach, incorporating clinical, electrophysiological, and pathological findings, is crucial for accurate diagnosis and early intervention to optimize patient outcomes.

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