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WHEN CIDP BEHAVES LIKE GBS: A CASE OF RECURRENT ACUTE-ONSET DEMYELINATING NEUROPATHY

Neurology

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KEYWORDS

BACKGROUND:
Chronic inflammatory demyelinating polyneuropathy (CIDP) is an immune-mediated neuropathy characterized by progressive or relapsing sensorimotor deficits. While typically chronic, some patients exhibit acute or relapsing-onset variants, which may clinically mimic acute inflammatory demyelinating polyneuropathy (AIDP). We report a case of recurrent acute-onset quadriparesis over several years, ultimately managed with corticosteroids and long-term immunosuppression.

Case Presentation:
A 45-year-old male presented with acute difficulty in walking over four days, followed by progressive bilateral upper limb weakness and numbness. Symptoms began with difficulty rising from a squatting position and numbness in both lower limbs. He subsequently developed bilateral hand weakness and sensory changes.

He had a history of five similar episodes over several years, each presenting with acute ascending quadriparesis. Previous episodes responded to intravenous immunoglobulin (IVIG) or plasma exchange (PLEX), with full recovery. He was diagnosed with CIDP and advised immunosuppressive therapy (Azathioprine), but discontinued it after a short period.

Investigations:
Routine hematological and biochemical investigations were within normal limits, including complete blood count, renal and liver function, thyroid profile, HbA1c, serum vitamins, inflammatory markers, and iron studies. Serum and urine protein electrophoresis showed no monoclonal bands. Screening for porphyria, syphilis, tuberculosis, and cryptococcal infection was negative.

Cerebrospinal Fluid (CSF) Analysisrevealed:
- Elevated protein level with normal glucose
- Acellular
- Negative India ink stain

Neurological Examination:
Mental Status: Alert, MMSE 30/30
Cranial Nerves: Bilateral facial weakness, absent jaw jerk; other cranial nerves normal

Motor Examination:
Flaccid quadriparesis, with greater involvement of lower limbs
Upper limbs: Power approximately 4-/5 proximally, hand grip \~80%
Lower limbs: Power initially 2–3/5, improved to 4-/5 at discharge
Reflexes: Absent
Plantar: Mute
Sensory: Decreased pain and touch in hands and feet; impaired vibration and proprioception below the knees

Differential Diagnosis:
- Acute-onset CIDP (relapsing variant)
- Recurrent AIDP
- Vasculitic neuropathy
- Paraneoplastic neuropathy
- Metabolic, toxic, or infectious neuropathies (ruled out)

Treatment:
The patient received pulse intravenous methylprednisolone therapy for five days, resulting in improved strength, particularly in the lower limbs. Due to prior poor compliance with oral immunosuppression and multiple relapses, a long-term plan was made for monthly intravenous cyclophosphamide for six months. The first dose was administered with MESNA for uroprotection. Physiotherapy and outpatient follow-up were advised.

Outcome And Follow-up:
The patient demonstrated objective improvement in lower limb strength, enabling ambulation with one-person support. He was discharged with a structured immunosuppressive regimen and counseling regarding the importance of adherence. Regular follow-up was planned to monitor for side effects including infections, cytopenias, and infertility.

DISCUSSION:
This case highlights a relapsing, acute-onset variant of CIDP that closely resembles AIDP. In such cases, the diagnosis relies on clinical history, CSF findings, and response to immunotherapy. Due to frequent relapses and poor adherence to oral immunosuppressants, cyclophosphamide was chosen for long-term control. This case underscores the importance of treatment compliance and the role of individualized immunosuppressive strategies in managing refractory CIDP.