ORIGINAL RESEARCH PAPER

Internal Medicine

A CASE OF CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPTH

KEY WORDS: Chronic Inflammatory Demyelinating Polyneuropathy, DADS, MADSAM.

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ABSTRACT

Chronic inflammatory demyelinating polyneuropathy (CIDP) is immune-mediated neuropathy defined by clinical progression for more than 2 months, and electrodiagnostic evidence of peripheral nerve demyelination. However, there are several clinical phenotypes, classified into "typical CIDP," and "atypical CIDP" such as "multifocal acquired demyelinating sensory and motor neuropathy (MADSAM)." Typical CIDP is a most common form, characterized by symmetric proximal and distal muscle weakness and motor-dominant manifestation. In typical CIDP, demyelination predominantly affects the distal nerve terminals and nerve roots, where the blood-nerve barrier is anatomically deficient. These features suggest antibody-mediated demyelination in typical CIDP. By contrast, MADSAM is characterized by multifocal demyelination in the nerve trunks, and such distribution of lesions results in multiple mononeuropathy or asymmetric polyneuropathy. In MADSAM, cellular immunity is likely to be involved in the breakdown of the blood-nerve barrier at the site of conduction block. Clinical features are probably determined by the distribution of demyelinative lesions and reflect the different immunopathogenesis of each CIDP subtype. CIDP can be successfully treated with immunoglobulin, glucocorticoids or plasma exchange. A second line treatment like Immunosuppressant agents and monoclonal antibodies may be proposed in case of no response, intolerance or inaccessibility to the three reference treatments.

INTRODUCTION

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) is an acquired, immune-mediated neuropathy affecting peripheral nerves and nerve roots, characterized by a relapsing-remitting or progressive course, glucocorticoid responsiveness, and electrodiagnostic or pathologic features of demyelination.

CASE STUDY

A 45 year old male resident of Nerul, Navi Mumbai, presented to the emergency department with complaints of:

Weakness in bilateral lower limb since 2 months . acute in onset,gradual in progression , noticed as slippage of chappals and eventually progressed to inability to walk in 2 months no significant past history.

General Examination:

Patient is conscious oriented to time place and person
Afebrile
Pulse-92/min
B.P- 130/80 mm-hg
Spo2 -98% on RA
Systemic Examination:
CVS – S1S2 normal. No murmur appreciated
RS – Bilateral Vesicular breath sounds heard.
P/A – Soft, NonTender. No organomegaly.
CNS – Patient is conscious and oriented to time place and person

Motor examination	Right	Rt	Lt
left			
Upper limb – proximal	4/5 4/5	4/5	4/5
Distal	4/5 4/5	4/5	4/5
Lower limb -proximal -	3/5 3/5	3/5	3/5
Distal	1/5 1/5	1/5	1/5

Sensory examination : loss of sensations below L1 level
Cranial nerves not involved.
CSF Examination
R/M – 1.Sugars – 186mmol/L CSF ADA - 20
2.Proteins – 224mmol/L
3.Cells – 7/cu.mm
CSF C/S – No Growth
Nerve Conduction Study :
s/o generalized sensory motor demyelinating +axonal polyradiculopathy

The patient was started on injection IVIG 2gm/kg over 5 divided doses for 5 days

DISCUSSION :

Typical CIDP (50-60%)-is a fairly symmetric sensorimotor polyneuropathy with proximal and distal motor involvement that exceeds sensory involvement. The presentation is usually one of gradually progressive symptoms over the course of several months or longer

Atypical Variants –

MADSAM (Lewis Sumner Syndrome)-Multifocal distribution of weakness and sensory deficits are seen. The electrophysiological hallmark of the disease is the presence of conduction block.

DADS (Distal Acquired Symmetrical Demyelinating Neuropathy) – symmetrical length dependent sensory or sensorimotor neuropathy (only distal limb involvement) often associated with an IgM paraprotein and markedly increased distal motor latencies.

Chronic Immune Sensory Polyradiculopathy-

Selective involvement of sensory nerve roots. Patients present with a sensory ataxic syndrome with normal motor and sensory conduction studies.

Chronic Inflammatory Lumbosacral Polyradiculopathy- lower

extremity variant presents with weakness and sensory involvement limited to legs with normal motor and sensory nerve conduction studies.

CANOMAD is a rare chronic ataxic neuropathy associated with ophthalmoplegia, IgM paraprotein, cold agglutinins and disialosyl (ganglioside) antibodies.

CONCLUSION :

Misdiagnosis is common and is reported in up to 50% of patients referred with a CIDP diagnosis, mainly in patients with an atypical presentation.

CSF is usually acellular with an elevated protein level, sometimes several times normal.

Electrodiagnostic: findings reveal variable degrees of conduction slowing, prolonged distal latencies, distal and temporal dispersion of CMAPs, and conduction block as the principal features.

Treatments:

IVIG - administered as 2.0 g/kg body weight given in divided doses over 2–5 days.

Plasma exchange -initiated at 2-3 treatments per week for 6 weeks; periodic re-treatment may also be required.
Corticosteroids - 60–80 mg prednisone PO daily for 1–2 months, followed by a gradual dose reduction of 10 mg per month as tolerated.

Patients who fail therapy with IVIg, PE, and glucocorticoids may benefit from treatment with immunosuppressive agents such as azathioprine, methotrexate, cyclosporine, and cyclophosphamide, either alone or as adjunctive therapy.

CIDP associated with anti-CNTN1 and NF155 antibodies is typically refractory to IVIg, but a few studies suggest a response to Rituximab

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