Case report

A Rare CaseReport on Chronic Inflammatory DemyelinatingPolyradiculoneuropathy

ABSTRACT:

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) CIDP is a rare autoimmune disorder of the peripheral nervous system that attacks myelin sheath around the peripheral nerves. It is characterized by muscle weakness and sensory deficits, numbness that can lead to significant neurological disability. The diagnosis is based on a combination of clinical examination findings, electrodiagnostic studies, and other supportive evidence. Recognizing CIDP and distinguishing it from other chronic polyneuropathies is important because many patients with CIDP are highly responsive to treatment with immunosuppressive or immunomodulatory therapies. This case report summarizes the variants of CIDP, diagnosis and current treatment strategies.

KEYWORDS: Chronic inflammatory demyelinating polyradiculoneuropathy, neuroinflammatory, autoimmune, Guillain-Barré Syndrome, immunosupperessive, immunomodulatory.

INTRODUCTION:

Chronic inflammatory demyelinating polyneuropathy (CIDP) is a neurological disorder.CIDP is a rare autoimmune disorder of the peripheral nervous system that attacks myelin sheath around the peripheral nerves. It is characterized by muscle weakness and sensory deficits, numbness that can lead to significant neurological disability.CIDP is an uncommon immune-mediated neuropathy that primarily affects men and is linked to aging especially at birth. The exact reason for CIDP is not clearly known. CIDP is usually classified asProgressive, Recurrent, Monophasic[(1)]. Worldwide, there are between 0.8 and 10.3 cases of CIDP for every 100,000 individuals, according to estimates [(2).]

The most common symptom is muscle weakness that gets worse over at least eight weeks.The typical features of CIDP are Slowly progressing, symmetric, proximal, distal paresis and sensory dysfunction. The duration of the symptoms is more than two months, and

they may progress or relapse. It typically affects the muscles of Hips and thighs, shoulders and upper arms, hands and feet equally on both sides of your body:Other symptoms of CIDP may include atrophy, paraesthesia, clumsiness, Loss of mobility, Loss or weakening of deep tendonreflexes.These symptoms may change in severity over time. They may come on slowly or rapidly and sometimes come and go over time.Early diagnosis and treatment are key to recovery.

Although the underlying pathophysiology of CIDP is not well understood, humoral and cellular autoimmunity are known to be involved. Certain CIDP cases are linked to recognized antibodies that specifically target different forms of neurofascin, a protein found in the neuronal cytoskeleton. When a patient's presentation points to a neuroinflammatory condition, it's important to think about whether GBS or CIDP (or another condition) best explains the weakness pattern. About 50% of patients exhibit the "typical" CIDP phenotype, which is defined by symmetric and primarily motor manifestations. Presently reclassified as "CIDP variants," "atypical" CIDP includes multiple well-characterized entities (multifocal, focal, distal, motor, or sensory CIDP). Guillain-Barré Syndrome (GBS), another immune-mediated peripheral neuropathy with an acute onset, is closely linked to CIDP. GBS usually last for one or two weeks, on the other hand CIDP have a slower natural history and last longer term of more than 8 weeks . GBS, once treated people recover quickly and CIDP shows re-occurrence. In GBS, the patient can typically pinpoint the precise moment when symptoms begin to appear, but in CIDP, this is not the case. Typically, prominent sensory indicators that favour CIDP include ataxia and impaired sensation. Like in this instance, proximal limb weakness favours CIDP, whereas GBS typically results in length-dependent "ascending paralysis" that first affects the strength of distal muscles [(3)]. In rare cases, GBS leads to CIDP. There’s no test to diagnosis CIDP.The diagnosis is based on a combination of Physical and clinical examination findings, electrodiagnostic studies, neurological studies and other supportive evidence. Early treatment is key option. It can help prevent. As it's an autoimmune disorder, healthcare providers use medicines that suppress the immune response to treat CIDP. Treatment includes: Corticosteroids, Intravenous immunoglobulin (IVIG), Plasma exchange (PE), are first line therapy and also treatment Immunotherapy, Stem cell transplant[(1)].

Case report:

A 45 year old male patient who has a history of HTN since 3 years and GBS since 1 year came to hospital with complaints of weakness in B/L lower limb since 2 months , initially started as tingling and numbness in lower limb , patient was apparently asymptomatic 2 months ago, then noticed slippage of chappals and unable to hold chappals, then developed proximal weakness of lower limbs since 20 days, history of weakness of upper limbs in the form of unable to mix food since 1 month, unable to lift the arm above shoulder level, static since 15 days not progressed, history of B/L lower limb oedema since 1 month insidious onset gradually progressed upto ankle associated with skin pigmentation, history of facial puffiness since 1 month.

On examination the patient is conscious and coherent, Blood Pressure: 140/90 mmhg, Pulse Rate: 86/min, Cardio Vascular Sounds: S1S2+, Respiratory System: BAE + clear, Central Nervous System: power Upper Limb, Lower Limb 3/5(Right and Left), Tone: Upper Limb decreased, Lower Limb Voluntary resistance +, Reflexes BTKAP (BICEPS, TRICEPS, KNEE, ANKLE, PLANTAR) negative, hand grip 40%. Clinical serology report shows Anti-Nuclear Antibody IgM Antibodies detectable. Electromyography shows severe motor axonal neuropathy involving both upper and lower limbs. Laboratory findings show that decreased levels of haemoglobin (9.1) increased WBC (14.97k cells $/\mu$), Cerebrospinal Fluid examination shows: colour- blood hinged with plenty of RBCs, increased protein count (70mg/dl) and borderline sugar content(80mg/dl), skin pigmentation and thickening over dorsum of both foot and hand. Treatment given to the patient was methyl prednisolone 1g, injection IvIg 400mg per day, injection vitamin B_{12} 1 amp, injection Multi-vitamin 1 ampule , tablet amoxicillin and potassium clavulanate, tablet paracetamol, tablet pantoprazole, injection human albumin 20%, tablet pregabalin, and advised for physiotherapy. IvIg was given for 5 days methylprednisolone is given for 10 days which partially improved the symptoms of the patient. Methyl prednisolone was stopped and wysolone 40mg (oral Prednisolone) was prescribed.His symptoms improved, the motor powertest results were normalized.

DISCUSSION:

Chronic inflammatory demyelinating polyradiculopathy (CIDP) is a chronic and disabling neuropathy with a postulated immune pathogenesis [(4)].CIDP is an idiopathic condition in which a primarily T cell-mediated immune response is directed against myelin components of peripheral nerves. For CIDP, corticosteroids, intravenous immunoglobulins (IVIG), and plasma exchange (PLEX) are the first-line treatments available [(6)]. Since corticosteroids have long-term side effects, they are used as bridge therapies, the cornerstones of treatment are plasmapheresis and serial IVIG. Their efficacy is comparable. The cost effectiveness and the long-term adverse effects need to be considered. And we can also use maintenance therapy like steroid sparing immunosuppressive agents [(7)]. In our case patient was treated with ivig and corticosteroids by using the combined therapy the treatment was effective and successful and the patient recovered from the symptoms.

REFERENCES:

1. Querol, L et al., Novel Immunological and Therapeutic Insights in Guillain-Barré Syndrome and CIDP. *Neurotherapeutics* **18**, 2222–2235 (2021). https://doi.org/10.1007/s13311-021-01117-3
2. Hans-Peter Hartung et al., The Role of the Complement System in Chronic Inflammatory Demyelinating Polyneuropathy: Implications for Complement-Targeted Therapies, Neurotherapeutics, Volume 19, Issue 3,2022, Pages 864-873, ISSN 1878-7479.

3. Saad TC et al., Chronic inflammatory demyelinating polyneuropathy: a rare cause of falls. BMJ Case Rep. 2019 Dec 2;12(12): e231676. doi: 10.1136/bcr-2019-231676. PMID: 31796460; PMCID: PMC7001711.
4. Nobile-Orazio E. Chronic inflammatory demyelinating polyradiculoneuropathy: where we are, where we should go. J PeripherNervSyst 2014; 19:2–13.
5. Nobile-Orazio E et al., Comparing treatment options for chronic inflammatory neuropathies and choosing the right treatment plan. Expert Rev Neurother 2017; 17:755–765.
6. Dimachkie MM, Barohn RJ. Chronic inflammatory demyelinating polyneuropathy. Curr Treat Options Neurol. 2013 Jun;15(3):350-66.
7. Gogia B, Rocha Cabrero F, Khan Suheb MZ, et al. Chronic Inflammatory Demyelinating Polyradiculoneuropathy. [Updated 2023 Jun 1]. In: StatPearls. Treasure Island (FL): StatPearls Publishing; 2024 Jan.