A Rare CaseReport on Clinical Insights of Chronic Inflammatory Demyelinating Polyradiculoneuropathy

ABSTRACT:

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) CIDP is a rare and heterogenous 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 evolving over more than 8 weeks. Raised protein concentrations in CSF and heterogeneous slowing of nerve conduction are typical of the condition. Understanding of its pathophysiology has recently improved, although its causes remain unclear. Diagnosis is sometimes challenging and can require use of neuro imaging and nerve biopsy.The diagnosis is also 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 corticosteriods , 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)]. “The pathogenesis of CIDP is linked to cellular immune mechanisms, as evidenced by the presence of inflammatory infiltrates in sural nerve biopsies, modifications in T cell subset frequencies and functions, altered expression of cytokines and other inflammatory mediators in the blood and CSF of CIDP patients, and the role of T cells in EAN (Experimental Autoimmune neuritis)” (4).“The spinal roots, proximal nerve trunks, and major plexuses are the primary locations of the inflammatory lesions in CIDP, though they can also spread throughout the PNS, according to a combination of autopsy, MRI, and ultrasound studies. However, the majority of biopsies are taken from the sural nerve because the proximal nerves and nerve roots are relatively inaccessible. Pathological changes in sural nerve biopsies cover a wide range of changes, including no abnormalities, oedema, demyelination, formation of onion bulbs, axonal degeneration, and perivascular or endoneurial inflammatory infiltrates of macrophages and T cells, despite the fact that this site is distant from the most prominent inflammatory activity” (5,6,7,8). “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), muscle 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/μ), 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. Thus by knowning the history , chief complaints, and physical and diagnostic examination it is confirmed that the patient is diagnoised as suffering from Chronic inflammatory demyelinating polyneuropathy (CIDP). Treatment given to the patient was methyl prednisolone 1g- corticosteriods indicated for immnunereactions, injectionIvIg 400mg per day - an immnunosupressant, injection vitamin B_{12} 1 amp,&Multi-vitamin 1 ampule- indicated for nervousness , tablet amoxicillin and potassium clavulanate - antibiotics, tablet paracetamol - to reduce pain and inflammamtion, tablet pantoprazole- antacid, injection human albumin 20%, tablet pregabalin- for neuropathic pain 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”[(9)]. “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”[(10)]. 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 [(11,12)]. In our case patient presented complaints of weakness in Both the lower limb since 2 months , initially started as tingling and numbness in lower limb , patient was apparently asymptomatic 2 months ago, then noticed that he is unable to wear and hold footwear, then developed proximal weakness of lower limbs since 20 days, history of weakness of upper limbs, unable to lift the arm above shoulder level, static since 15 days not progressed and also with lower limb oedema since 1 month insidious onset

gradually progressed upto ankle associated with skin pigmentation. On examination the patient had elevated BP and abnormal reflexes with hand grip 40%, Anti-Nuclear Antibody IgM Antibodies detectable. Electromyography shows severe motor axonal neuropathy involving both upper and lower limbs.Abnormal CSF was also proved that the patient was suffering from CIPD. The patient was initially treated with corticosteriods and then withIVIG by using the combined therapy the treatment was effective and successful and the patient recovered from the symptoms.

Conclusion:

The typical presentation of chronic inflammatory demyelinating polyneuropathy (CIDP), an uncommon but treatable auto-immune mediated chronic disorder of the peripheral nervous system, includes symmetrical weakness of the proximal and distal muscles, hyporeflexia or areflexia, and occasionally paraesthesia. Even with all of these difficulties, the diagnosis of CIDP is made using a combination of clinical criteria, electrodiagnostic characteristics, and, when in doubt, further laboratory and imaging tests.An elderly man with growing muscular weakening in both his upper and lower limbs is the subject of our case study. Corticosteroids and IVIg have shown to be safe and effective in treating CIDP. For the purpose of assessing the effectiveness of a treatment, objective outcome assessments are essential. For improved results, thorough assessment, prompt detection, and appropriate treatment are therefore suggested.

Consent

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

Ethical Approval:

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

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