Case Report

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# Myasthenia Gravis and Chronic Inflammatory Demyelinating Polyradiculoneuropathy in the Same Patient: A Clinical Rarity

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**Abstract**

We report a 23 year old female who concomitantly developed Chronic inflammatory polyradiculoneuropathy (CIDP) with Myasthenia gravis (MG). She initially presented with slowly progressing proximal and distal weakness in all four limbs with areflexia and elevated CSF protein. Electrophysiological studies confirmed the diagnosis of CIDP and started on corticosteroids and immunodulators. However, two months into treatment, her condition deterioted and she presented to emergency department with bilateral ptosis, breathing difficulty, dysphagia and no improvement in motor power. High titres of AchR antibodies, presence of ophthalmoplegia along with neostigmine test confirmed the concomitant diagnosis of MG with CIDP. Steroids along with acetylcholinesterase inhibitors were used to treat MG. Coexistence of two autoimmune disorders in the same patient can be a diagnostic labyrinth which can delay diagnosis and treatment.

**Keywords:** Myasthenia Gravis; Chronic Inflammatory Demyelinating.

## Introduction

CIDP and MG are both autoimmune neuromuscular disorders which have similar presenting features. While CIDP patients typically present with chronic progressive, stepwise progressive or relapsing weakness, MG is characterized by muscle weakness that fluctuates, worsening with exertion, and improving with rest. [1, 2] The probability of CIDP and MG occurring in the same patient is extremely rare. In this report, we describe the case of a 23 year old female who was initially diagnosed only as CIDP but later confirmed as a case of concomitant CIDP and MG.

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## Case Report

A 23 year old female presented to our hospital with complaints of slowly but gradually progressing bilateral symmetrical proximal and distal weakness of both upper and lower limbs since 1 month. She had numbness of both hands and feet. She had difficulty in swallowing, mainly solid food since last 3 months. She was conscious and responding to commands without any breathing difficulty. Her muscle strength in distal and proximal lower and upper limb was 4/5 and 3/5 respectively. Muscle tone was reduced and bulk was normal. Deep tendon reflexes were absent while superficial reflexes were preserved. CSF examination revealed elevated protein (100mg/dl) with normal cell count. Electrophysiological studies showed F- wave latency prolongation in all four limbs.

A diagnosis of CIDP was made provisionally. She was started on tablet prednisolone 50 mg once daily and mycophenolate mofetil 250 mg twice daily. However, she again came to the emergency department. She presented with further deterioration of the motor and sensory symptoms along with

breathing difficulty, bilateral ptosis, hoarseness of voice and generalized areflexia. CSF (protein-245mg/dl) and electrodiagnostic tests were again performed which validated the previous results.

An overlapping diagnosis of Myasthenia Gravis was suspected and Neostigmine challenge test was done in the emergency. Neostigmine 1.5 mg was injected intramuscularly which was positive with improved vocalization and decreased breathing difficulty. She also had almost immediate increased strength in all four limbs. She was then shifted to intensive care unit for ventilatory management as the symptoms relapsed after 50 minutes. Elective tracheostomy was done to prevent any lung infection in the ICU after 5 days. Acetylcholine receptor (AchR) antibody titre was elevated to 30.48 nmol/l(Normal-0.25-0.4 nmol/l) which confirmed our diagnosis.

As there was no evidence of cholinergic crisis, she was then started on Injection Methylprednisolone 1g IV once daily for three days and tab. Pyridostigmine 60 mg thrice daily. Methylprednisolone was replaced with prednisolone 50 mg orally once daily after three days. Patient responded drastically with improvement in ptosis and power of all limbs within a few days. She was weaned off ventilator support and shifted to the ward. Her motor power improved to 4/5 in all four limbs at discharge.

## Discussion

Chronic inflammatory demyelinating polyneuropathy (CIDP) is an acquired immune mediated chronic inflammatory disease of the peripheral nervous system (PNS) characterized by hyporeflexia or areflexia and progressive or relapsing motor or sensory dysfunction developing over weeks. [3] The pathogenesis of CIDP is not completely clear and a number of humoral and cell-mediated mechanisms are implicated. [4]

Myasthenia gravis (MG), on the other hand, is an autoimmune disorder characterized by fluctuating muscle weakness, worsening with exertion, and improving with rest. Extrinsic ocular muscles involvement is seen in about two-thirds of cases as the initial presentation which later progresses to involve other bulbar muscles and limb musculature [5].

The coexistence of polyneuropathy with Myasthenia gravis is quite uncommonly reported in literature [6-11].

Our patient had clinical, laboratory and electrodiagnostic features of both CIDP and Myasthenia gravis which initially was diagnosed as only demyelinating polyneuropathy. Diagnosis of CIDP is made based on clinical and electrophysiological data along with supportive criteria such as elevated CSF protein, demyelination on nerve biopsy and improvement following immuomodulatory treatment [9]. Our patient showed progressive motor weakness with numbness in all limbs with raised CSF protein and F wave latency on nerve conduction studies which made us initially diagnose her as a case of demyelinating polyneuropathy. However, lack of improvement in symptoms on steroid therapy raised our suspicions regarding a coexisting pathology. Corticosteroids have proven their efficacy in the treatment of MG, but the symptoms of MG may worsen during the initial phase of steroid therapy [12]. We assume the worsening of symptoms during the initial phase of steroid therapy in our case might have been due to the above stated effect also.

Involvement of ophthalmoplegia further strengthened our belief. Ophthalmoplegia being rare in CIDP, the possibility of MG must be given due consideration when this sign is present in a patient with CIDP [7]. AchR antibody is the most common autoantibody out of many which attacks the nicotinic acetylcholine receptor [13].

The diagnosis was of myasthenia gravis in our case was clinched by evidence of a positive neostigmine test, and positive anti acetylcholine receptor (AchR) antibodies. She also had CIDP as proved by prolonged conduction velocity, prolonged F latency and elevated CSF protein consistent with CIDP.

This may not be a chance association. Both CIDP and MG having a common immunopathogenesis has been suggested. Cytokines produced by Th1 cells (IL-2 and IFN-g) are inflammatory mediators involved in various autoimmune processes whereas those produced by Th2 cells (IL-4, IL-5, IL-6 and IL-13) mediate in antibody production, anti-inflammatory cascade and resolution of inflammatory and autoimmune processes. Th1/Th2 balance is critical in many pathological conditions of immunomediated disorders, including coexistence of CIDP and MG although the balance remains contentious [14,15,16].

## Conclusion

Although rare, a patient who presents with features of CIDP may also have another autoimmune disorder like MG. Looking out for possible signs of another autoimmune pathology can help prevent delay in its diagnosis and treatment.

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