Case Report / *Olgu Sunumu*

Ege Journal of Medicine / *Ege Tıp Dergisi* 2025; 64 (4): 716-720

A case of myasthenia gravis emerging after prior diagnosis of CIDP: Clinical challenges in overlapping autoimmune disorders

Önceden CIDP tanısı almış bir hastada ortaya çıkan miyastenia gravis: Örtüşen otoimmün hastalıklarda klinik zorluklar

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ABSTRACT

Chronic inflammatory demyelinating polyneuropathy (CIDP) and myasthenia gravis (MG) are distinct autoimmune neuromuscular disorders that rarely coexist. We present a 52-year-old man with stable CIDP who developed progressive lower limb weakness and facial involvement. Electrophysiological studies revealed neuromuscular junction dysfunction, and elevated acetylcholine receptor antibodies confirmed MG. Treatment with pyridostigmine, intravenous immunoglobulin, and rituximab resulted in clinical stabilization. This case shows the importance of considering overlapping autoimmune disorders in CIDP patients with new or atypical symptoms. Early recognition using electrophysiological and serological tests is important for accurate diagnosis and effective management of coexisting CIDP and MG.

Keywords: CIDP, myasthenia gravis, diagnostic challenge, immunosuppression, rituximab.

Öz

Kronik inflamatuar demiyelinizan polinöropati (CIDP) ve miyastenia gravis (MG), nadiren birlikte görülen iki farklı otoimmün nöromüsküler hastalıktır. Stabil CIDP'si olan 52 yaşındaki erkek hasta, ilerleyici alt ekstremite ve fasyal güçsüzlükle başvurdu. Elektrofizyolojik incelemelerde nöromüsküler kavşak disfonksiyonu saptandı ve asetilkolin reseptör antikorlarının yüksek olması MG tanısını doğruladı. Piridostigmin, intravenöz immünglobulin ve rituksimab tedavisiyle klinik stabilite sağlandı. Bu olgu, CIDP'li hastalarda yeni veya atipik semptomlarda eşlik eden otoimmün hastalıkların düşünülmesinin önemini vurgulamaktadır. Elektrofizyolojik ve serolojik testlerle erken tanı, eşzamanlı CIDP ve MG yönetiminde etkilidir.

Anahtar Sözcükler: *CIDP, myasthenia gravis, tanı güçlüğü, immunsüpresyon, rituximab*

INTRODUCTION

Chronic inflammatory demyelinating polyneuropathy (CIDP) and myasthenia gravis (MG) are distinct autoimmune disorders affecting the peripheral nerves and the neuromuscular junction, respectively (1–3). Although rare, co-occurrence of CIDP and MG has been documented, with the conditions presenting simultaneously or sequentially in the same patient (4-16).

Despite some overlapping clinical features, fluctuating symptoms involving bulbar or ocular muscles are more indicative of MG, whereas a progressive, motor-predominant neuropathy with sensory involvement is characteristic of CIDP. We present the case of an adult patient with a previous diagnosis of CIDP who developed new-onset progressive lower extremity weakness during the remission phase. Further evaluation confirmed a diagnosis of MG, and treatment with rituximab led to clinical improvement.

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Application date: 11.07.2025 Accepted: 30.09.2025

CASE REPORT

A 52-year-old man presented to our clinic with a six-month history of progressively worsening weakness in his lower limbs. His past medical history was notable for a diagnosis of CIDP made four years earlier, at which time he had experienced numbness, paresthesia, and weakness in the lower extremities. Electromyography (EMG) confirmed a demyelinating polyneuropathy and cerebrospinal fluid (CSF) analysis showed elevated protein levels. He was initially treated with intravenous immunoglobulin (IVIG), corticosteroids, and azathioprine. Over time, his symptoms improved significantly with INCAT score 2 to 1. However, due to side effects from corticosteroids and azathioprine, his long- term treatment was adjusted to monthly IVIG infusions at a reduced dose of 0.5 mg/kg. His condition remained stable until the recent onset of progressive weakness. Neurological examination revealed marked proximal weakness in the lower extremities (thigh flexion 3/5, extension 4/5), mild weakness of the orbicularis oculi muscles bilaterally, absent deep tendon reflexes, and reduced vibration sensation in the lower limbs.

Routine hemogram and serum biochemistry tests were unremarkable. Given the patient's clinical progression, a repeat EMG study was conducted. Nerve conduction studies—including median and ulnar motor-sensory, posterior tibial, fibular motor, and sural nerves—revealed a sensory-motor neuropathy predominantly affecting the lower extremities. Due to the presence of proximal weakness, needle EMG was performed on both distal and proximal muscles of the upper and lower limbs, including the tibialis anterior, vastus medialis, first dorsal interosseous, and biceps brachii muscles; all findings were within normal limits. Repetitive nerve stimulation (RNS) testing at low frequencies (1–3–5 Hz) applied to the nasalis muscle demonstrated a decremental response, with a 15–20% reduction between the first and fourth responses. Single-fiber EMG of the orbicularis oculi muscle revealed increased jitter values (131–139–195 μs). Serological analysis showed a significantly elevated acetylcholine receptor antibody (AchR-Ab) level (>8 mg/dL; normal <0.5 mg/dL), confirming the diagnosis of MG. Thoracic and abdominal computed tomography (CT) scans revealed no evidence of malignancy or thymic pathology. Additional immunological workups, including vasculitis markers, antiganglioside antibodies, and paraneoplastic neurological antibodies, were negative.

The patient was subsequently started on pyridostigmine three times daily, along with intravenous immunoglobulin (IVIG) at a dose of 1 mg/kg administered over two days. Although the myasthenia gravis foundation association (MGFA) score remained at 2 from baseline, the patient's weakness gradually improved. Long-term immunosuppressive therapy continued with rituximab. During follow-up, he remained clinically stable without the emergence of new symptoms

DISCUSSION

We present a case of a patient previously diagnosed with CIDP, who developed new symptoms four years later, ultimately leading to a diagnosis of MG. Before the onset of these symptoms, the patient's CIDP had remained in remission. MG may be easily overlooked when it arises concurrently with or after CIDP. However, in patients with CIDP who develop ophthalmoplegia (7), fluctuating symptoms, or a poor response to treatment (12), the possibility of coexisting MG should be considered. Cranial nerve involvement is uncommon in CIDP, though facial palsy, diplopia, and ptosis due to ophthalmoplegia have been reported in 3–10% of cases (1,17). Symmetrical proximal and distal muscle weakness is typical of CIDP. In our case, although the patient did not report fluctuating weakness, the pattern had changed; he exhibited prominent proximal weakness in the lower extremities and mild bilateral facial weakness, which he had not previously experienced. Initially, these findings were attributed to the possible reactivation of CIDP. However, follow-up EMG did not show motor conduction blocks, as seen in previous studies, nor any evidence of axonal loss on needle EMG. It is well-recognized that IVIG and immunosuppressive agents can reverse conduction blocks. Additionally, needle EMG findings suggestive of axonal damage are often associated with disease progression in CIDP (18). However, a repetitive nerve stimulation test revealed a decremental response in cranial muscles, supporting the diagnosis of MG in our case.

Several cases have been reported in the literature describing the coexistence of MG and presumed autoimmune neuropathies, such as CIDP (4-16). In most of these reports, patients

with MG demonstrated seropositivity, particularly the presence of AchR-Ab, as was observed in our patient. Notably, only one reported case described the development of MG following CIDP in the absence of MG-related antibodies. In that case, RNS was non-diagnostic, while single-fiber electromyography (SFEMG) confirmed the diagnosis of MG (15). Conversely, cases have also documented the development of CIDP following MG (8,10,15,16). These rare overlaps suggest a possible shared immunopathogenic mechanism. Pathological studies strongly support the view that T cell–mediated immune mechanisms are central to the pathogenesis of chronic inflammatory demyelinating polyneuropathy (CIDP). This process appears to be antigen-driven and restricted by the major histocompatibility complex (19). Autoantibodies targeting diverse antigens have also been identified in a subset of patients, though their prevalence and specificity vary widely. The rapid improvement observed after plasmapheresis further underscores the role of humoral factors, suggesting that circulating immune components contribute to functional impairment (20). A similar antigen-driven T cell response is well recognized in MG. In this condition, regulatory T cells and $CD4^+$ T cells bind to AChR epitopes, activating B cells and thereby promoting the production of pathogenic antibodies (21). Infectious triggers or other immune-activating events may lead to the generation of cross-reactive antibodies targeting both acetylcholine receptors at the neuromuscular junction and peripheral nerve myelin (22). Additionally, thymic abnormalities—both benign and malignant—have been associated with autoimmune neuromuscular disorders. Thymic pathologies are considered to cause an aberrant autoimmune response. The thymus is a primary source of this regulatory CD4+CD25+ T-cell subset, which participates in the active suppression of potentially autoreactive T cells in the periphery. CD4+CD25+ Treg cells have been described in a variety of experimental systems to provide protection from T cell–mediated autoimmune disorders (23). In rare instances, thymoma has been linked with the sequential or concurrent development of MG and CIDP (16,24). In our case, no specific trigger or underlying pathology such as thymoma or infection was identified, similar to most previously reported cases (Table-1).

Due to adverse effects associated with corticosteroids and azathioprine, rituximab was selected as the preferred treatment option for our patient. Rituximab is a chimeric monoclonal antibody targeting the CD20 antigen on B cells, leading to rapid peripheral B-cell depletion following administration. While its efficacy is well established in muscle-specific kinase (MuSK)-positive myasthenia gravis, recent evidence also supports its potential benefit in AChR-positive myasthenia, mainly when initiated during the early stages of the disease (<2 years) (25,26). Additionally, rituximab has shown effectiveness in patients with CIDP, especially when associated with other autoimmune diseases. It has been reported to enhance clinical outcomes and reduce the need for first-line therapies (27,28). In our case, treatment with rituximab resulted in significant symptom improvement and stabilization of both autoimmune conditions—myasthenia gravis and CIDP.

Table-1. Reported Cases of MG and CIDP in Literature: Co-occurring or Sequential Presentations

References	Co-occurring or Sequential Presentations	MG related Ab positivity	Etiology
Weinreb H. et al. 1986 (4)	co-occurring; ocular and neuropathic symptoms	Not known	Not known
Inatus A. et al. 1992 (5)	sequential; the first weakness and paresthesia due to neuropathy and later appearing of bulbar symptoms	AchR-Ab +	Thymoma
Patwa H.S. et al. 1996 (6)	co-occurring; not known	Not known	Not known
Sempere AP. et al.1996 (7)	sequential; the first weakness and paresthesia due to neuropathy and later appearing of ocular, bulbar symptoms and weakness	AchR-Ab +	Not known
Kimura K. et al. 1998 (8)	sequential; the first ocular symptoms and later weakness due to neuropathy	AchR-Ab +	Not known

Tam D. A. et al.1999 (9)	co-occurring; recurrent extremity weakness	AchR-Ab +	Not known
Shankar V. et al. 1999 (10)	sequential; the first ocular symptoms and later weakness due to neuropathy	AchR-Ab +	Not known
Magot A. et al. 2002 (11)	co-occurring; ocular symptoms with mastication difficulty and extremity weakness	AchR-Ab +	Not known
Mori Masahiro. et al. 2006 (12)	co-occurring; ocular symptoms with extremity weakness	AchR-Ab +	CMV infection
Quan W. et al. 2018 (13)	co-occurring; progressive weakness with bulbar symptoms	AchR-Ab +	Not known
Sumit D. et al. 2018 (14)	co-occurring; progressive weakness with ocular and bulbar symptoms	AchR-Ab +	Not known
Jiang N. Et al. 2020 (15)	sequential; the first weakness and paresthesia due to neuropathy and later appearing of bulbar symptoms	Seronegative	Not known
	sequential; the first bulbar and extremity weakness and then neuropathic symptoms	AchR-Ab +	

CONCLUSION

Although rare, MG and CIDP may coexist or develop sequentially in the same patient. In individuals with a prior diagnosis of CIDP, the emergence of new ocular or bulbar symptoms should prompt consideration of superimposed MG. Electrophysiological studies are essential to differentiate whether these symptoms reflect the progression of the underlying neuropathy or indicate an additional neuromuscular junction disorder. The presence of AchR-Ab can further support the diagnosis. Rituximab may be a suitable treatment option, particularly in cases where conventional therapies are poorly tolerated.

Conflict of interest: None of the authors have conflict of interest to declare.

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