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An 82-year-old man with facial diplegia and bulbar involvement : case report

Hélène Salvadé

helene.salvade@hopitalvs.ch

Hôpital du Valais

Vadim Afanasiev

Hôpital du Valais

Vincent Alvarez

Hôpital du Valais

Case Report

Keywords: Case report, Facial diplegia, Myasthenia gravis, Guillain-Barré syndrome

Posted Date: November 12th, 2026

DOI: https://doi.org/10.21203/rs.3.rs-11378569/v1

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Additional Declarations: No competing interests reported.

Abstract

Introduction

Acute or subacute bilateral facial diplegia with bulbar involvement is an uncommon neurological presentation that encompasses a broad spectrum of potentially life-threatening disorders requiring prompt diagnosis and treatment. In cases with an acute or subacute course, the differential diagnosis can often be limited to two entities: Guillain–Barré syndrome (GBS) and myasthenia gravis (MG). Distinguishing between these conditions may be challenging, particularly when MG presents with atypical features such as predominant bulbar involvement, bilateral facial weakness, or an absence of obvious symptom fluctuation.

Case presentation

An 80-year-old man with a history of malignancies presented with rapidly progressive bilateral facial diplegia, tongue paresis, dysarthria, dysphagia, and ptosis. Hyporeflexia and a recent diarrheal episode initially suggested a GBS variant. Despite intravenous immunoglobulin therapy, the patient developed respiratory failure requiring mechanical ventilation. Plasma exchange resulted in rapid and dramatic improvement. Subsequent testing revealed strongly positive acetylcholine receptor and anti-titin antibodies, confirming seropositive MG.

Conclusion

Myasthenia gravis may initially present as a myasthenic crisis without clear diurnal fluctuation and with atypical manifestations, including bilateral facial weakness, isolated dropped head syndrome, isolated ocular symptoms without ptosis, distal limb weakness such as foot or hand drop, or limb-girdle weakness. In such atypical presentations, distinguishing myasthenia gravis from Guillain–Barré syndrome can be challenging. The temporal evolution of symptoms, electrophysiological findings, and response to treatment may therefore be crucial in establishing the correct diagnosis.

Introduction

Acute or subacute bilateral facial diplegia with bulbar involvement is an uncommon neurological presentation that encompasses a broad spectrum of potentially life-threatening disorders requiring prompt diagnosis and treatment. In cases with an acute or subacute course, the differential diagnosis can often be limited to two entities: Guillain–Barré syndrome (GBS) and myasthenia gravis (MG). Distinguishing between these conditions may be challenging, particularly when MG presents with atypical features such as predominant bulbar involvement, bilateral facial weakness, or an absence of obvious symptom fluctuation. Because confirmatory investigations, including autoantibody testing, often require several days before results become available, clinicians frequently need to make therapeutic

decisions based on clinical presentation and the most likely differential diagnoses. In this setting, the temporal evolution of symptoms, electrophysiological findings, and response to immunomodulatory therapies may provide valuable diagnostic clues. We report the case of an elderly patient presenting with rapidly progressive facial diplegia and bulbar dysfunction, in whom the clinical course and response to treatment were fundamental in establishing the diagnosis.

Case presentation

A man in his 80s presented with a progressive onset over several days of bilateral facial paresis, bilateral tongue paresis, dysarthria, and severe dysphagia without clear fluctuation. He was known for a prostatic adenocarcinoma with diffuse osseous and pelvic lymph node metastases (treated with radical prostatectomy and radiotherapy, with stable disease over the past five years under regular PSA monitoring), as well as a mucinous adenocarcinoma of the rectosigmoid junction causing stenosis but without metastases, recently diagnosed for which surgical management was planned.

Apart from the facial diplegia with bulbar involvement, the patient also reported asthenia. He mentioned an episode of diarrhea prior to the onset of symptoms and a recent insect bite in the mountains, followed by an erythematous rash on both hands, which had resolved upon admission. The remainder of the medical history was unremarkable except for an insect bite a few weeks ago.

Clinical examination revealed bilateral facial complete diplegia associated with bilateral tongue weakness, and swallowing difficulties with a preserved pharyngeal reflex. Bilateral ptosis was present without clear fatigability, and no oculomotor involvement was observed. Additionally, there was hyporeflexia in the upper limbs and areflexia in the lower limbs, without appendicular sensorimotor deficit or gait disturbance.

Given the history of an insect bite, an initial diagnosis of neuroborreliosis was quickly ruled out following negative serology results. A structural cause (including brain metastasis regarding clinical context) was also excluded by brain MRI. The nerve conduction study demonstrated a cupular decremental response of compound muscle action potentials upon repetitive nerve stimulation at 3 Hz of the left facial and right spinal accessory nerves recording orbicularis oculi and upper trapezius (Fig. 1). Immmunological tests were performed at admission but were not available for several days.

This patient presented with bilateral facial diplegia and rapidly progressive bulbar involvement in an oncological context with subacute onset and no clear fluctuation. Although several differential diagnoses may be considered in this clinical presentation, including lymphoma, amyotrophic lateral sclerosis, and sarcoidosis, the rapid progression of symptoms mainly pointed toward two principal differential diagnoses.

The first potential differential diagnosis was a variant of GBS. GBS classically manifests as flaccid limb paresis, sometimes associated with sensory disturbances and abolition of reflexes (1). However, several variants have been described, including the pharyngeal-cervical-brachial variant, characterized by

bilateral facial weakness and sometimes associated with paresthesias and paresis of the upper limbs, which may, however, be absent(2, 3). A presentation of GBS with predominant bulbar paresis has also been reported (4), which may correspond to our patient. The presence of hyporeflexia in all four limbs supported this differential diagnosis. The subacute onset over a few days was also consistent, as symptoms in GBS typically peak within 10 to 14 days. Furthermore, the occurrence of a diarrheal episode prior to symptom onset had suggested a post-infectious GBS. This condition classically presents with albuminocytologic dissociation in the cerebrospinal fluid, which was absent in our patient. However, this does not exclude the diagnosis, particularly given that lumbar puncture was performed less than one week after symptom onset(1).

Another major differential diagnosis to consider in the context of bulbar involvement is MG. This disease can be divided into two subtypes: oculomotor or generalized. In the generalized form, bilateral facial weakness is common. Predominant bulbar involvement, presenting with dysarthria, dysphonia, and dysphagia, may occur and represents the initial manifestation in approximately 15% of cases.(5) Although bulbar onset without ocular motor involvement has been described, it remains extremely rare (6). A characteristic feature of MG is symptom fluctuation over time, with fatigability and worsening toward the end of the day. In our patient, symptoms had been constant throughout the day and had progressively worsened over three days, which initially argued against this diagnosis. Nerve conduction studies had shown a cupular decremental response on repetitive nerve stimulation, supporting a neuromuscular junction disorder (Fig. 1). In contrast, in GBS, nerve conduction studies would typically reveal reduced nerve conduction velocities, prolonged distal latencies, and conduction blocks(1), which were absent in this patient in all four limbs.

Given these two leading differential diagnoses, and in light of negative infectious serologies and cerebrospinal fluid analysis showing neither protein elevation nor pleocytosis, intravenous immunoglobulin (IVIG) therapy was initiated. Despite several doses, the patient worsened with rapid respiratory distress with hypercapnic failure, requiring transfer to the intensive care unit (ICU) and endotracheal intubation. Plasma exchange (PLEX) therapy was initiated due to clinical deterioration, leading to a rapid and dramatic improvement after only three sessions.

PLEX is indicated for both major differential diagnoses, in case of rapidly progressive symptoms with respiratory distress. In the case of GBS plasmapheresis accelerates motor recovery (7), whereas in MG, transient and rapid improvement has been observed, allowing for clinical stabilization until initiation of long-term immunosuppressive therapy (8).

After several days and continued clinical improvement under plasmapheresis, the immunological tests performed at admission became available and revealed positive anti-titin and anti-acetylcholine receptor antibodies (AchR) (titer 78.00 nmol/l, n < 0.45nmol/L), confirming the diagnosis of MG. Anti-muscle-specific kinase (MuSK) antibodies were absent.

Regarding maintenance therapy, the patient was started on a tapering course of corticosteroids in combination with rituximab. During follow-up, he developed multiple complications related to

immunosuppression, including a respiratory tract infection and an aortic dissection, the latter possibly facilitated by corticosteroid-induced tissue fragility. In addition, pancytopenia secondary to myelodysplastic syndrome necessitated discontinuation of rituximab.

Ten months after the initial presentation, the patient experienced a relapse of myasthenia gravis, which was initially treated with intravenous immunoglobulin (IVIG). Owing to the lack of clinical response, therapeutic plasma exchange was subsequently performed. However, in view of the patient's progressive deterioration in general condition, multiple treatment-related complications, and concomitant local recurrence of the sigmoid malignancy, curative treatment was withdrawn, and the goals of care were transitioned to comfort-focused palliative management.

Discussion

Our patient showed a drastic and rapid clinical improvement after only three sessions of plasmapheresis over five days, which enabled extubation within 48 hours. This response suggested the presence of a pathogenic antibody, thereby favoring a diagnosis of MG. In contrast, in GBS, improvement in motor function would be expected but typically with a longer delay of approximately two weeks(7).

The absence of symptom fluctuation in this case was unusual. This case highlights the importance of non-prominent fluctuations in MG(9) and the absence of fluctuations in cases of rapidly deteriorating MG symptoms. Moreover, the dramatic and rapid clinical improvement following plasmapheresis and the characteristic electrophysiological findings supported the diagnosis of MG, which was ultimately confirmed by the presence of anti-acetylcholine receptor antibodies.

In this series of 290 patients with MG (10), 6.9% presented with unusual symptoms: bilateral facial weakness, isolated dropped head, isolated ocular findings without ptosis, distal limb weakness (e.g., foot and hand drop), and weakness of limb-girdle muscles. Anti-MuSK antibody positivity was statistically more prevalent among patients with unusual presentations, though only 20% tested positive for anti-MuSK antibodies (70% tested positive for anti-AchR antibodies).(10)

In this case, the chronological evolution of symptoms and the therapeutic response were crucial to guide the diagnostic process. The relationship with the coexisting gastrointestinal malignancy remains unclear.

Conclusion

Myasthenia gravis may present with an atypical clinical picture and even as an acute myasthenic crisis without clear fatigability or symptom fluctuation. In such cases, it may closely mimic Guillain–Barré syndrome. Careful assessment of symptom evolution, electrophysiological findings, and therapeutic response is essential to establish the diagnosis and guide appropriate management.

Declarations

Ethics approval and consent to participate

Ethical approval was not required for this case report in accordance with local institutional and national guidelines. Written informed consent to participate was obtained from the participant.

Consent for publication

The participant provided written informed consent for publication of the manuscript.

Availability of data and materials

Data sharing is not applicable to this article as no datasets were generated or analysed during the current study.

Competing interests

The authors declare no competing interests.

Funding

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Authors' contributions

Hélène Salvadé conceptualized the study and wrote the original draft. Vincent Alvarez and Vadim Afanasiev supervised the project and reviewed the manuscript. All authors approved the final version.

Acknowledgements

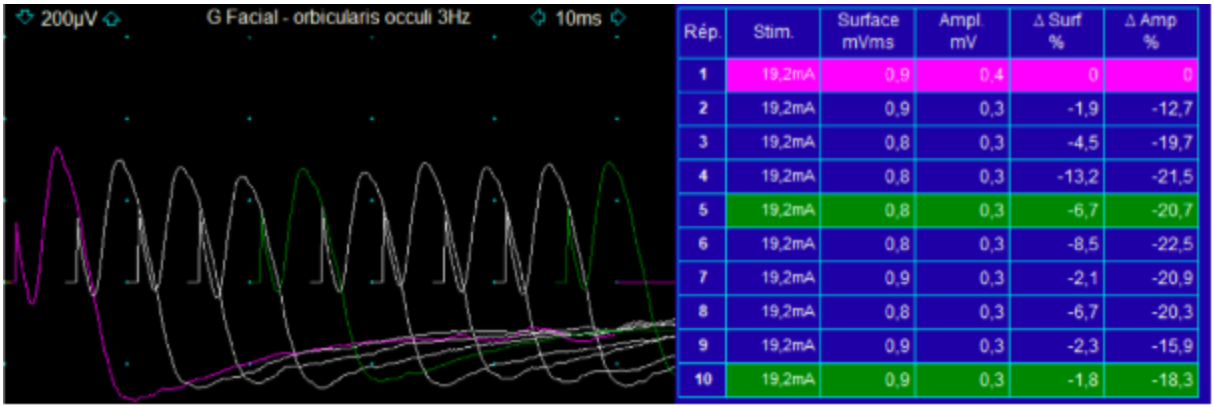
The authors have no acknowledgements to declare.

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Figures



Rép.	Stim.	Surface mV/ms	Ampl. mV	Δ Surf %	Δ Amp %
1	19,2mA	0,9	0,4	0	0
2	19,2mA	0,9	0,3	-1,9	-12,7
3	19,2mA	0,8	0,3	-4,5	-19,7
4	19,2mA	0,8	0,3	-13,2	-21,5
5	19,2mA	0,8	0,3	-6,7	-20,7
6	19,2mA	0,8	0,3	-8,5	-22,5
7	19,2mA	0,9	0,3	-2,1	-20,9
8	19,2mA	0,8	0,3	-6,7	-20,3
9	19,2mA	0,9	0,3	-2,3	-15,9
10	19,2mA	0,9	0,3	-1,8	-18,3

Figure 1

Repetitive nerve stimulation at 3 Hz of the left facial nerve recording orbicularis occuli

Repetitive nerve stimulation at 3 Hz of the left facial nerve recording orbicularis occuli demonstrates a decremental response of the compound muscle action potential amplitude, with a maximum decrement observed around the fifth stimulus, suggestive but not specific of myasthenia gravis.