

The Diagnostic Odyssey in Myasthenia Gravis

Patient #3: Delayed diagnosis leading to disease exacerbation in man with late-onset MG 57-year-old-male

Case contributor and commentary:

Yohei Harada, MD and Donald Sanders, MD
Duke University
Durham, North Carolina

Age 56-57

Initial symptoms:

- » “Numbness” of the right side of his face with drooping of the right eyelid
- » Slurred speech and difficulty chewing; had to hold his jaw closed to eat
- » Frequent choking episodes and nasal regurgitation of liquids
- » Lost 30 pounds over 3-4 months
- » In the last 1-2 months in his job as a carpenter, had difficulty holding a hammer, drilling above his head and carrying heavy items
- » Could use stairs and get out of chairs without difficulty

PCP workup:

- » Considered Bell’s palsy as the cause of facial weakness
- » Ordered AChR binding antibody test



Outcomes and Management:

- » Prescribed 5 days of prednisone, with no appreciable response
- » AChR antibodies were present

5 months later

Patient was referred for neuromuscular evaluation and presented with the following symptoms:

- » Five-month history of intermittent droopy eyelids
- » Difficulty chewing solid foods
- » Slurred speech
- » Weakness in arms
- » Intermittent double vision with vertical displacement when staring at objects
- » Worsening symptoms in the evening

Diagnostic evaluation

Neurological exam:

Cranial nerves

- » Bilateral ptosis, moderately severe on the right, mild on the left
- » No diplopia
- » Moderately weak eyelid closure and cheek puff

Deep tendon reflexes

- » Normal

Motor exam

- » Moderately severe weakness of neck flexion and shoulder abduction bilaterally
- » No muscle atrophy

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Electrodiagnostic testing:

- » Repetitive nerve stimulation study of nasalis and trapezius muscles showed a decremental response, which confirmed the presence of abnormal neuromuscular transmission as seen in myasthenia gravis

Management based on MG diagnosis

- » Admitted to the hospital and received 3 courses of plasma exchange, which resolved all symptoms except for persistent bilateral mild ptosis and mild eye closure weakness
- » Chest CT did not reveal thymoma
- » Started prednisone 20mg daily followed by mycophenolate mofetil 1000mg twice per day



Follow-up evaluations

- » Examination 2 months after discharge showed further improvement of symptoms except for residual mild eyelid closure weakness

Commentary: The peak onset of MG in males occurs in their 60s.¹ Prompt therapy is required in patients with MG as they may develop life-threatening weakness.

In this patient, a 5-month delay in diagnosis resulted in severe bulbar symptoms requiring emergent plasma exchange.

Key learning points

- » Delay in diagnosis can lead to life-threatening weakness
- » Severe bulbar involvement requires emergent therapy such as plasma exchange

References

1. Sanders DB, Raja SM, Guptill JT, Hobson-Webb LD, Juel VC, Massey JM. The Duke myasthenia gravis clinic registry: I. Description and demographics. Muscle and Nerve. 2021;63(2):209-216. doi:10.1002/mus.27120