CASE REPORT

An Atypical Case of Myasthenia Gravis: Purely Bulbar in a Young Male

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INTRODUCTION

Myasthenia Gravis (MG), an uncommon autoimmune syndrome caused by the failure of neuromuscular transmission, results from binding of autoantibodies to proteins that are involved in signaling at the neuromuscular junction (1). Acquired myasthenia gravis (MG) is an uncommon disorder (occurring in 200–400 cases per million people) (2).

Its symptoms are caused by a characteristic muscle weakness that worsens after repeated use. In most cases of MG, the initial sign is ocular weakness of any sort. The next most common sign is bulbar weakness (5) . In about two-thirds of patients, the extrinsic ocular muscles (EOMs) present with the initial symptoms. These symptoms usually progress to other bulbar muscles and limb muscles, resulting in generalized MG (gMG). In about 10% of MG patients, symptoms remain limited to the EOM, and this condition is termed ocular MG (oMG)(3,4). However in cases where weakening of other striated skeletal muscles occurs, this is referred to as generalized MG. We present a rare case of early onset Myasthenia Gravis affecting a young male with only and purely bulbar involvement.

CASE REPORT

A 22 year old male presented in the Ear Nose Throat out-patient department of Holy Family Hospital complaining of dyphagia and dysarthria and no significant past medical history. On examination, orophyrangeal edema was suspected, for which Coamoxiclav (Augmentin ® 625mg) and Citrizin (Sedil ® 10mg) Serratiopeptidase (denzin ® DS) were prescribed.

On the first week of follow-up, the symptoms of dyphagia and dysarthria had worsened and there was evidence of facial muscle weakening. The patient was referred to the neurology department. The patient had no significant past medical or surgical history. He denied smoking, IV drug abuse, alcohol abuse, and was sexually inactive. The family history was unremarkable as well. There was no history of fever, cough, cold, or recent upper respiratory tract infection. He had no known drug allergies and his current medications consisted of those prescribed for orophyrangel edema.

On physical examination, the patient was alert, awake and oriented, and his blood pressure, heart rate, respiratory rate, SpO2, and temperature were within normal limits. On neurological examination the patient showed signs of bulbar weakness, however, there were no other signs and symptoms to direct a diagnosis. There was no external ocular weakness, motor weakness, muscle wasting, sensory deficits, or sphincter dysfunction. He was diagnosed with a rare descending form of Gullian barre syndrome and an NCS was ordered to confirm the diagnosis. The NCS was negative for Gullian Barre syndrome. However, decremental response was seen in the proximal muscles at 3 Hz, suggesting Myasthenia Gravis (MG). To confirm MG, an acetylcholine receptor antibody titer was ordered and found to be 60nmol/L (where a measurement of less than 0.25nmol/L is negative). Despite this significant elevation, no signs of generalized myasthenia were detectable.

The patient was treated with acetylcholin-

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esterase (AchE) inhibitor, Pyridostigmine (Amygra ® 360 mg per day) and steroids, Prednisolone (Delta Cortef ®) over a period of 4 weeks. The patient was unable to talk, chew, swallow, or demonstrate proper facial expressions. However, there were no signs of ptosis, diplopia, difficulty seeing, double vision, dizziness, unsteady gait, fatigue, or any difficulty maintaining balance. The patient's condition did not improve much with Acetylcholinesterase (AchE) inhibitors and steroids. However, he was able to ingest liquids, consume a soft diet, and speak a few sentences after medication.

The patient's complete blood count was normal with no eosinophilia. His electrolytes were normal as was his thyroid function test, liver function test, coagulation profile, pulmonary function test. His chest CT scan with contrast showed no evidence of thymoma. At follow-up visits, the patient remained otherwise healthy with no evident development of other myasthenic signs except bulbar weakness. This case is atypical since bulbar onset MG does not usually persist with complete absence of other myasthenic signs and symptoms (of the generalized form), especially if it is an early-onset MG.

DISCUSSION

Myasthenia Gravis is a rare neurological disorder involving neuromuscular junctions and most commonly affects young females or older men6. Those having history of some autoimmune disorder are at higher risk for MG (7).

The most common initial presentation of MG is ocular weakness (5,8). Ocular weakness most commonly manifests itself as ptosis in 90% of cases. However an accommodative/vergence insufficiency or a concomitant or noncomitant oculomotor paresis may also be seen (9). Generalized MG eventually develops in 50% of those who manifest an ocular sign, whereas others only retain the ocular weakness and are thus considered as cases of ocular MG (3,4). The second most common presentation of MG is bulbar weakness. However, bulbar weakness usually progresses to generalized myasthenia gravis (3,4,5). Fatigue is the hallmark of any kind of myasthenia (10). Although fatigability of peripheral skeletal muscle is the hallmark of the disease, it can be absent in the bulbar forms. Isolated bulbar presentation is common in late-onset MG and may be confused with diseases of the oropharynx and other neurological conditions (11,12,13). MG that develops after the age of 50 is said to be late onset MG (14).

In the case of early onset MG, isolated bulbar presentation and its persistence are not commonly seen. In the present case report, the patient was a 22 year old male with an unremarkable past medical or family history, and as such, was unlikely to develop MG. Before a diagnosis of MG was made, orophyrangeal edema and descending gullian barre syndrome were also suspected. Diagnosis of MG was confirmed on the basis of a laboratory workup, which showed a significant elevation of the for acetylcholine receptor antibody. However, there were no signs and symptoms of fatigue, nor were there ocular signs and symptoms. The most widely accepted classification of MG is based onthe Myasthenia Gravis Foundation of America's Clinical Classification (15):

Class I: Any eye muscle weakness, possible ptosis, no other evidence of muscle weakness elsewhere.

Class II: Eye muscle weakness of any severity, mild weakness of other muscles

- Class IIa: Predominantly limb or axial muscles
- Class IIb: Predominantly bulbar and/or respiratory muscles

Class III: Eye muscle weakness of any severity, moderate weakness of other muscles

- Class IIIa: Predominantly limb or axial muscles
- Class IIIb: Predominantly bulbar and/or respiratory muscles

Class IV: Eye muscle weakness of any severity, severe weakness of other muscles

- Class IVa: Predominantly limb or axial muscles
- Class IVb: Predominantly bulbar and/or respiratory muscles (Can also include feeding tube without intubation)

Class V: Intubation needed to maintain airway.

In all classes, eye muscle weakness is standard along with varying weakness of limb, axial, bulbar or respiratory muscles. In the presented case study, only bulbar weakness not responsive to standard medications for MG was present. Although the bulbar form of MG is usually seen in cases of late-onset myasthenia, the presented case study provides an interesting example of bulbar weakness in a case of early onset MG.

CONCLUSION

It is evident from the case report that early onset MG can present in strictly bulbar form as well, which has not been reported elsewhere up until now. Therefore, in the case of any bulbar weakness with no signs and symptoms of fatigue, limb weakness, and ocular involvement, MG should not be ruled out. The prognosis for this patient does

not seem hopeful given his non- responsiveness to therapy. Further study is required to find out the cause of therapeutic failure in this case. The continued absence of ocular weakness, fatigue, and generalized symptoms in his six month follow-up visit is astonishing, as far as early onset myasthenia is concerned. Further follow-up of three years or more is indicated for this patient since previous reports show increased risk of developing the generalized form of the disorder during this time period.

ACKNOWLEDGEMENTS

We wish to express our sincere gratitude to Sajal Akhtar, a 4th year student at the Rawalpindi Medical College, for her devoted cooperation in completion of this project. We are also very thankful to Dr. Mohammad Bilal for all of his guidance and support in making this project possible.

CONSENT

Written informed consent was obtained from the patient for publication of this case report.

CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest

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