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A RARE CASE OF AN ATYPICAL CASE OF MYASTHENIA GRAVIS: PURELY BULBAR INVOLVEMENT IN A YOUNG MALE

General Medicine

Dr Manisha Panchal — Associate professor, general medicine department PDU medical college, Rajkot

Dr Maharshi Prajapati — 3rd Yr Medicine Resident PDU medical college and hospital ,Rajkot ,Gujarat , India.

ABSTRACT

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. Symptoms are caused by a characteristic muscle weakness that worsens after repeated user. In most cases of MG, the initial sign is OCULAR weakness of any sort. The next most common sign is BULBAR weakness. In about two-thirds of patients, the extra ocular muscles (EOMS) present with the initial symptoms. These symptoms usually progress to other bulbar system, limb muscles etc resulting in to generalized MG Ocular MG (oMG) : in about 10%of MG patients symptoms remain limited to the EOM(1) 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

KEYWORDS

OMG Ocular Myasthenia Gravis, Bulbar Weakness ,myopyrollate Stimulation Test, Nasal Regurgitation, No Generalized Muscle Weakness

INTRODUCTION

Acquired myasthenia gravis (MG) is a relatively uncommon disorder, with prevalence rates that have increased to about 20 per 100,000 in the US population (2). This autoimmune disease is characterized by muscle weakness that fluctuates, worsening with exertion, and improving with rest. In about two-thirds of the patients, the involvement of extrinsic ocular muscles (EOMs) presents as the initial symptom, usually progressing to involve other bulbar muscles and limb musculature, resulting in generalized myasthenia gravis (g MG)

Classification of Myasthenia Gravis

Subtypes of MG are broadly classified as follows [3]:

1. Early-onset MG: age at onset <50 years. Thymic hyperplasia, usually females,
2. late-onset MG: age at onset >50 years. Thymic atrophy, mainly males,
3. Thymoma-associated MG (10%–15%)
4. MG with anti-MUSK antibodies,
5. ocular MG (oMG): symptoms only affecting extraocular muscles,
6. MG with no detectable AChR and muscle-specific tyrosine kinase (MuSK) antibodies.

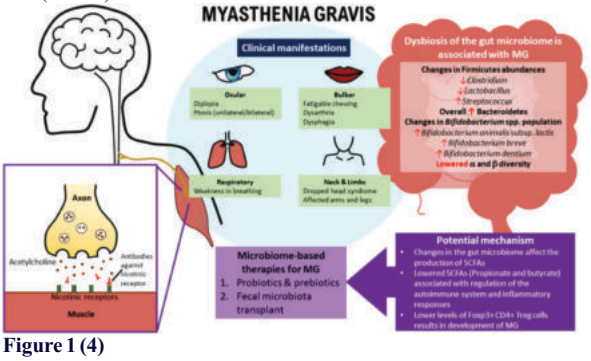

Figure 1 (4)

Case Report

A 30 year old male presented in the ENT out-patient department of civil Hospital Rajkot with 3 day history of acute onset c/o nasal regurgitation ,nasal twang in speech, difficulty in swallowing After clinical evaluation from ENT department Referred to physician with opinion s/o normal mobile vocal cord, normal barium swallow, gag reflex absent Patient was admitted in medicine ward with worsening of above symptoms.

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

On physical examination, the patient was alert, awake and oriented, his blood pressure, heart rate, respiratory rate, SpO2, and temperature were within normal limits. On neurological examination the patient showed signs of bulbar weakness, 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 After initial clinical evaluation with expert opinion provisional diagnosis of

1) Isolated 9th cranial nerve palsy was made Mri brain and csf examination ordered which was in normal limits Finally short course of steroids considering idiopathic lower cranial nerve neuritis initiated After 3 days of pulse therapy, patient discharge on tapering dose of steroids with somewhat clinical improvement

On follow up after 5 days, patient had flared up symptoms of nasal regurgitation, absent gag, nasal twang in voice.

Considering some differential of bulbar weakness. myopyrollate challenge test was given. Which was surprisingly positive with drastic improvement in symptoms (in seconds) Indicating us towards diagnosis of ? Myasthenia gravis, so for further evaluation Anti ACHR antibody test ordered

- Anti ACHR results positive with titre of anti ACHR a/b = 6.22 (normal range <0.5)
- The Patient was treated with Tablet
- Pyridostigmine (240 mg per day) and steroids,
- Prednisolone (omnacortil 40mg®) over a period of 4 weeks.
- There were no signs of ptosis, diplopia, difficulty seeing, double vision, dizziness, unsteady gait, fatigue, or any difficulty maintaining balance.

1) The patient's complete blood count was normal with no eosinophilia.

- HB 11.5mg/dl, wbc 7500 plt 1.89 lakh.
- electrolytes were normal range s.na 137,s.k 4.3,s. creat 0.8 s.urea 34 as was his thyroid function test (tsh 5.5 , s.t3 1.1 s.t4 4.6), liver function test, coagulation profile, pulmonary function test.

3) 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 persist with presence of other myasthenic signs and symptoms (of the generalized form).

DISCUSSION

Myasthenia Gravis is a rare neurological disorder involving neuromuscular junctions and most commonly affects young females or older men.

The most common initial presentation of MG is ocular weakness. Ocular weakness most commonly manifests itself as ptosis in 90% of cases. However an accommodative/vergence insufficiency or a concomitant or Non-commitant oculomotor paresis may also be seen . 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 .

The second most common presentation of MG is bulbar weakness. However, bulbar weakness usually progresses to generalized myasthenia gravis. Fatigue is the hallmark of any kind of myasthenia . 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 MG that develops after the age of 50 is said to be late onset MG .

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 30 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 , Isolated 9th cranial nerve palsy was provisionally diagnosed. 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 on the Myasthenia Gravis Foundation of America's Clinical Classification (5,6,7)

- 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.

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 continued absence of ocular weakness, fatigue, and generalized symptoms in his 3 month follow-up visit is astonishing, as far as early onset myasthenia is concerned. Further follow-up of 6 months or more is indicated for this patient

Consent

Written informed consent was obtained from the patient.

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