

A Case of Myasthenia Gravis Presenting Solely with Bulbar Palsy not associated with Easy Fatigability

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Abstract: Myasthenia Gravis (MG) is a neuromuscular disorder characterized by weakness and fatigability of skeletal muscles as a result of decrease in numbers of available acetylcholine receptors (AChRs) at neuromuscular junction due to an antibody-mediated autoimmune attack. We present here male case of Myasthenia Gravis who presented with complaints of unable to drink water and coming out from nose while swallowing. Gradually he developed inability to eat. 2 months later patient developed slurring of speech followed by total inability to speak. Also developed diplopia on binocular vision. On cranial nerve examination 3, 7, 9, 10th nerve palsies seen in the form of absence of medial & upward movement of left eye (3rd C.N.), inability to close eyes completely (3rd C.N.), reduced power and tone of buccinator muscle (7th C.N.), gag reflex absent (9th C.N.), presence of dysphagia (10th C.N.). Rest of the CNS findings were normal. ANA was (+) with nuclear speckled pattern. VEP study was normal. EMG NCV showed reduced amplitude of bilateral facial nerve, involvement of right oculomotor nerve suggesting bulbar myasthenia > generalized myasthenia. Then we did AChR antibody test which came highly positive. He was started T. Pyridostigmine 60mg TDS and T. Prednisolone 5mg/kg and discharged. On follow up we found marked improvement of symptoms.

Keywords: Myasthenia Gravis, Acetylcholine Receptors (AChRs), Cranial Nerve (C.N.), Muscle-specific tyrosine kinase (MuSK), Dysphagia, Gag Reflex

1. Introduction

Myasthenia Gravis (MG) is a neuromuscular disorder characterized by weakness and fatigability of skeletal muscles⁽¹⁾. The underlying defect is a decrease in number of available acetylcholine receptors (AChRs) at neuromuscular junction due to an antibody-mediated autoimmune attack. So leading to decreased availability of free receptor to bind for Acetylcholine, so its function gets impaired⁽²⁾

These antibodies are against the nicotinic acetylcholine receptor (AChR) at the neuromuscular junction or muscle-specific tyrosine kinase (MuSK).⁽³⁾ This leads to muscular weakness with easy 'fatigability', which is worse on exercise and improves with rest.

2. Case Report

A 30 year old male patient presented with complaints of unable to drink water since 5 months, water coming out from nose. Gradually he developed inability to eat, with no evidence of vomiting. 2 months later he developed slurring of speech which was followed by total inability to speak. Along with that he developed diplopia on binocular vision.

On CNS examination he was conscious oriented with normal plantar and deep tendon reflexes. On cranial nerve examination 3, 7, 9, 10 nerve palsies seen in the form of absence of medial & upward movement of left eye (3rd), inability to close eyes completely (3rd), reduced power and tone of buccinator muscle (7th), gag reflex absent (9th),

presence of dysphagia (10th). Rest of the CNS findings were normal.

CSF examination showed mild increase of protein.

MRI brain was normal.

Vitamin B12, TSH, Calcium, HbA1c, ESR, RA factor were normal. ANA was (+) with nuclear speckled pattern.

Fundus examination was normal with abnormal ocular movement as described. VEP study was normal.

EMG NCV showed reduced amplitude of bilateral facial nerve, involvement of right oculomotor nerve suggesting bulbar myasthenia > generalized myasthenia.

Then we did AChR antibody test which came highly positive. He was started T. Pyridostigmine 60mg TDS and T. Prednisolone 1mg/kg and discharged. On follow up we found marked improvement of symptoms.

3. Discussion

The prevalence of myasthenia gravis in the UK is estimated at about 15 per 100000 population.⁽⁴⁾

For diagnosis, the anti-acetylcholine receptor (AChR) antibody test is highly specific (nearly 100%), though false-positive anti-AChR antibody tests also have been reported. Assays for anti-MuSK antibody, anti-lipoprotein-related protein 4 (LRP4) antibody, anti-agrin antibody,

antistriational antibody may be useful. Other studies include chest CT to rule out thymoma and thymic enlargement and electrodiagnostic studies (repetitive nerve stimulation and single-fibre EMG).⁽⁴⁾

The usual initial complaint is a specific muscle weakness rather than generalized weakness, extraocular muscle weakness or ptosis is present initially in 50% of patients and occurs during the course of illness in 90%. The disease remains exclusively ocular in only 16% of patients. Bulbar muscle weakness is also common. Limb weakness may be more severe proximally than distally. Weakness is typically least severe in the morning and worsens as the day progresses. And is increased by exertion and alleviated by rest. Weakness progresses from mild to more severe over weeks or months, with exacerbations and remissions. Weakness tends to spread from the ocular to facial to bulbar muscles and then to truncal and limb muscles.

MG is a treatable and, at times, curable neurologic disorder. Pharmacologic therapy includes anticholinesterase medication and immunosuppressive agents, such as corticosteroids, azathioprine, cyclosporine, plasmapheresis and intravenous immune globulin (IVIg). Plasmapheresis and thymectomy are also employed to treat MG. Thymectomy is an especially important option if a thymoma is present. Patients with MG require close follow-up care.⁽¹⁾

4. Conclusion

The present case shows that it is important to consider MG even in cases presenting solely with progressive bulbar palsy without easy fatigability.

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