

Case Report

Unilateral Ocular Myasthenia Gravis

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Abstract

Myasthenia gravis, an acquired auto immune disease of the neuromuscular junction, has ophthalmic manifestations as integral part. Herewith we report a rare uniocular involvement of the eye in a 38yrs old male. He presented with the drooping of right upper eye lid since two years, worsening by evening and after exertion. Though the blood investigations and Ach antibody titres were normal, he responded well to 1.5mg neostigmine intramuscularly.

Key Words: Ocular myasthenia gravis, Neostigmine

Introduction

Myasthenia gravis (MG) is an acquired auto-immune disease of the neuromuscular junction which causes rapid muscle fatigue and weakness¹. Ocular manifestations are an integral part of Myasthenia gravis. Ptosis and diplopia are the initial complaint in around 75% to 90% cases. Ocular MG, in which weakness remains confined to the ocular muscles only, is encountered in up to 17% to 58% of all myasthenics⁴. Some less common or rare ocular signs like paradoxical reversal of ptosis, enhanced ptosis, total external ophthalmoplegia, internuclear ophthalmoplegia, chronic progressive external ophthalmoplegia and lid retraction contralateral to the ptotic eye⁵. Although bilateral ophthalmological involvement of disease is quite common, it is rare to find a case of unilateral Ocular Myasthenia Gravis.

Case Report

A 38 year old male presented to Ophthalmology OPD, Chettinad Health and Research Institute with a chief complaint of drooping of right upper eyelid since two years which was worsened by evening and on excessive work. He has no other systemic illness and no ocular complaints of diplopia.

On examination, visual acuity was 6/6 and extra ocular movements were full. Right eye revealed ptosis of 4mm, Bell's phenomenon was normal, Levator palpebral action 7 mm by Berke's method with rest of the anterior segment and posterior segment being normal. Anterior and posterior segments were found to be in normal limits in left eye except mild lid retraction. Ice pack test was positive. The routine blood investigations were found to be in normal limits except the mild elevation of ESR levels. USG abdomen and HRCT chest were found to be in normal limits. The acetyl choline receptor antibody titres were found to be in normal titres. The patient was subjected to

neostigmine test after obtaining anaesthetic fitness and with cardiac monitoring. After injection neostigmine 1.5 mg intramuscularly, the ptosis showed significant improvement after 30 minutes, confirming the diagnosis of Unilateral Ocular Myasthenia gravis. Neurologist opinion was obtained and advised tablet Pyridostigmine 60 mg thrice daily 8.

Discussion

Myasthenia gravis is a disease characterized by fatigue and weakness of the striated muscle within the body. Although the exact cause is unknown it is thought to be an acquired autoimmune disorder the defect lies at the postsynaptic membrane. 70 percent of patients with myasthenia gravis have visual symptoms in the initial presentation. Generally the female: male ratio in generalized myasthenia is 3:2 or higher. But on the contrary ocular myasthenia is more in especially after the age of 40 years. In addition, the average age of onset for generalized myasthenia is 33 years, while that of ocular myasthenia is 38 years. 2 Common clinical features seen in patients with ocular MG are ptosis (droopy eyelids), diplopia (double vision) and incomplete eye closure. Environmental, emotional, and physical factors also found to influence the symptoms for the patients. The major factors found to affect them are bright sunlight, extreme temperature, emotional stress, illness, surgery, menstruation, and pregnancy. Symptoms were found to be worse at the end of the day during the evenings or after exertion of the muscle.

Diagnostic Criteria For Ocular Myasthenia ³	
1	Ptosis present in one or both upper lids not due to local eyelid disease, preferably that is fatigue and recovers with rest.
2	Extraocular muscle weakness in one or both eyes but does not follow strict third nerve muscle innervation pattern.

3	Weakness in one or both orbicularis oculi but no weakness of other head and neck muscles
4	No pupillary abnormality
A Plus B or C or D or E	
A	Fatigue of the affected muscle with clear cut worsening of the ptosis after upward gaze for 30 to 60 seconds or worsening of the monocular duction after 120 seconds of gaze in the direction of action.
B	Recovery of the upper lid ptosis to almost normal after 30 seconds to 10 minutes of eyelid closure. Recovery of the monocular duction after 120 – 180 seconds of gaze in the direction of the antagonist muscle.
C	A positive edrophonium/ neostigmine test (fig 1 to fig 3).
D	Abnormal repetitive stimulation electromyography with a minimum decrement of 10%
E	Abnormal serum acetylcholine receptor binding level.



Fig 1: Patient before the test



Fig 2: Patient showing improvement in ptosis -30 min after the test

Elevated Ach receptor antibody confirms the diagnosis of Myasthenia gravis in 80 % to 90% cases of Myasthenia gravis and 30-77% cases of Ocular myasthenia gravis. Of these patients, 30% will have autoantibodies against muscle specific kinase (anti MuSKAb) expressed on skeletal muscle. Patients who are

negative for both Ach and MuSK antibodies are classified as "seronegative" Myasthenia gravis. CT scan should be done to rule out thymic hyperplasia. Thyroid function tests should be done to rule out autoimmune thyroid disease⁴.

Treatment is chiefly medical and aims at improving muscle weakness (thereby alleviating symptoms of diplopia and ptosis), achieving disease remission, minimizing drug induced side effects, and slowing or preventing progression to generalized Myasthenia². Medications include cholinesterase inhibitors such as Mestinon, pyridostigmine, steroids such as Prednisone, or other immunosuppressants used alone or in combination⁷. Other options include plasmapheresis, or IV IG therapy. These treatments offer only a temporary improvement and repeated treatments are necessary to sustain the effect. Supportive measures include the use of prisms or occlusion therapy for those with persistent diplopia and crutch glasses for severe ptosis².

Conclusion

Eventhough Myasthenia gravis is a common neuromuscular junction disorder. Ocular myasthenia in the early stages is often misdiagnosed. So clinicians should examine each case with a high degree of suspicion. The diagnosis can be made with simple clinical and pharmacological tests.

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