

Pattern of Ocular Myasthenia Gravis in a Tertiary Eye Care Hospital, Bangladesh

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Abstract

Purpose: The purpose of this study was to evaluate pattern of ocular myasthenia gravis patients in a tertiary eye care hospital in Bangladesh. **Methodology:** This observational study was conducted in National Institute of Ophthalmology and Hospital (NIOH), Dhaka from January 2019 to December 2019. A total of 33 cases were purposively selected among ocular myasthenia gravis patients attending in the Department of neuro ophthalmology of NIOH irrespective of age, gender, residence. Study variables were age, gender, clinical features, presence of thymoma, clinical, pharmacological and serological test findings. A proforma was prepared to record the data on particulars of the patients. **Results:** Among the cases, 45.5% were male and 54.5% were female where the mean age was 38.7 years. All patients presented with ptosis (100.0%) and most of them had fatigability (87.9%), cogan twice sign (81.8%) and orbicularis weakness (81.8%) while 63.6% had diplopia. Diabetes mellitus was observed in 18.2% patients and 15.1% had thyroid associations. Most of them had positive ice test (87.9%), 51.5% had positive serological test and 6.1% had positive neostigmine test. Thymoma was present in 6.1% cases. **Conclusion:** Ptosis was the most common clinical presentation for ocular myasthenia. Clinical diagnosis was confirmed by ice pack test and neostigmine test. Ocular myasthenia gravis patients may have other systemic diseases.

Key Words: Myasthenia Gravis, ptosis, tertiary eye care hospital.

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Introduction

Myasthenia gravis (MG) was first mentioned by Sir Thomas Willis in 1672. Later in 1890, it was described in detail by three German physicians Goldflam, Erb, and Jolly. The name myasthenia gravis (MG) is derived from the Greek. meaning "gravely weak muscles"¹.

Myasthenia gravis is an autoimmune neuromuscular disease leading to fluctuating muscle weakness and fatigability. In MG, weakness is caused by circulating antibodies that block acetylcholine receptors at the postsynaptic neuromuscular junction, inhibiting the excitatory effects of the neurotransmitter acetylcholine on nicotinic receptors throughout neuromuscular junctions². MG have two types of onset, adult and childhood. Myasthenia gravis may be ocular, bulbar or generalized. In ocular myasthenia gravis, weakness is limited to extra ocular muscles, orbicularis oculi and levator palpebrae superioris³.

The most common symptoms are ptosis and diplopia which are found in 70-75% of patients initially and eventually in 90% of individuals. Immunologic, endocrine and genetic factors contribute disease severity. Patients with MG have a higher incidence of other autoimmune diseases, such as thyroid diseases, polymyositis, systemic lupus erythematosus, diabetes mellitus. Sjogrens, rheumatoid arthritis, ulcerative colitis, pemphigus vulgaris and ankylosing spondylitis⁴. In Bangladesh, national institute of ophthalmology and hospital poses only neuro ophthalmology department in government sector. Now a days

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ocular myasthenia patients' occupancy is gradually emerging in that department. OM leads to development of visual morbidity along with compromised patients' life style.

The purpose of this study was to evaluate pattern of ocular myasthenia gravis in a tertiary eye care hospital in Bangladesh.

Methods

This observational study was conducted in National Institute of Ophthalmology and Hospital (NIOH), Dhaka from January 2019 to December 2019. Study population was taken among clinically proven cases of ocular myasthenia gravis in Department of neuro ophthalmology of NIOH irrespective of age, gender, residence. Patients with other neuro ophthalmological disease in combination with ocular myasthenia were excluded from this study. Variables were age, gender, clinical features, clinical, pharmacological and serological test findings. Sampling method was purposive sampling. Informed written consent was taken from each patients and attendants. Detailed history was taken. Clinical examination was performed and properly recorded. Both eyes were examined. Ocular examinations were done by torch light and Slit lamp. Visual acuity were measured by Snellen's chart. Intraocular pressure was evaluated by Goldmann Applanation Tonometer. Ophthalmoscopy was performed. Suspected cases of ocular myasthenia gravis were subjected to relevant clinical test viz. eliciting, fatigue test, cogan's lid twitch sign. Measurements were taken in department of neuro ophthalmology when patients came for clinical evaluation and treatment. Patient who were clinically suspected for ocular myasthenia gravis, were confirmed by ice pack test and neostigmine test for confirmation. Ptosis

of eyelid was evaluated with 15cm plastic ruler. Diplopia charting were done wherever applicable to elicit range of limited extra ocular motility. Measurements were taken in out patient department of neuro ophthalmology when they were referred from department of ophthalmology. Complete blood count, RBS, Thyroid profile were done in all cases. Anti-nuclear antibody, RA test, X-ray of chest and neck and CT-Scan or MRI of chest were done in selected cases. A proforma was prepared to record the data on particulars of the patients. Ethical principles were followed accordingly.

Statistical Analysis

All collected data were recorded properly. All statistical analyses were completed using SPSS 26 version and windows excel.

Results

A total number of 33 cases were taken as study population where both eyes were examined. Among the 33 patients, 15 (45.5%) patients were male and 18 (54.5%) patients were female. Mean age was 38.7 years. Diabetes mellitus was observed in 18.2% patients and 15.1% had thyroid associations (table 1). All patients had ptosis, 21 (63.6%) patients had diplopia, 23 (69.7%) patients had variability, 29 (87.9%) patients had fatigability, 27 (81.8%) patients had cogan twice sign, 27 (81.8%) patients had orbicularis weakness, 06 (18.2%) patients had diabetes mellitus and 05 (15.1%) patients had thyroid association (table 2). Clinical test (Ice test) was positive in 29 (87.9%) patients, serological test (antibody test) was positive 17 (51.5%) patients while pharmacological test (neostigmine test) was positive 02 (6.1%) patients (figure 1).

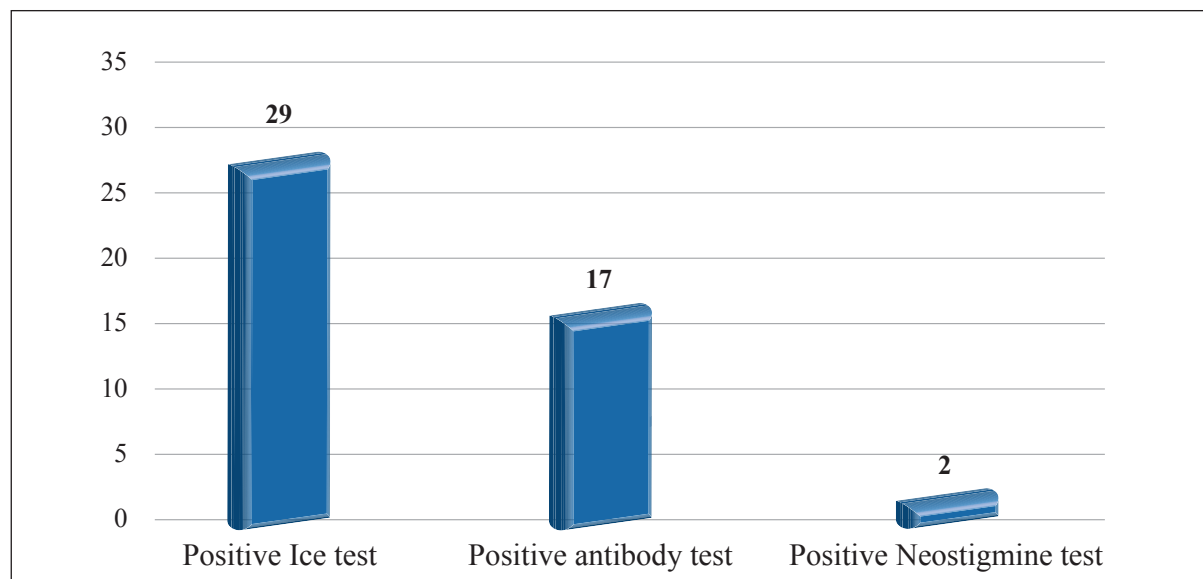
Table 1: Distribution of patients by baseline characteristics

Criteria	Number	Percentage
Gender		
Male	15	45.5
Female	18	54.5
Age (mean ± SD)	38.7±10.3 years	
Comorbidity		
Diabetes mellitus	6	18.2
Thyroid associations	5	15.1

Table 2: Distribution of patients by clinical features (n=33)

Clinical feature	Number	Percentage
Ptosis	33	100.0
Fatigability	29	87.9
Cogan twice sign	27	81.8
Orbicularis weakness	27	81.8
Variability	23	69.7
Diplopia	21	63.6
Diabetes mellitus	06	18.2
Thyroid association	05	15.1

Multiple response

**Figure 1:** Distribution of patients by positive clinical, pharmacological and serological tests

Discussion

This study was conducted in National Institute of Ophthalmology and Hospital (NIOH), Dhaka from January 2019 to December 2019 to evaluate pattern of ocular myasthenia gravis in National Institute of Ophthalmology and Hospital in Dhaka, Bangladesh.

Ocular myasthenia gravis (OMG) in a form of myasthenia gravis which the muscles that move the eyes and control the eye lids are easily fatigued and weakened⁵. The diagnosis of OMG is usually made by a combination of patient history,

clinical findings and other diagnostic procedures. The first suspicion of OMG should come to mind during the case history if the patient manifests symptoms of ptosis, diplopia, and/or blur, which increases with use of the ocular muscles, or as the day progresses. Variability of symptoms should further trigger suspicion. The gold standard for diagnosis of OMG is the tensilon test/ neostigmine test. However, there are a number of non invasive tests that can also be used to make the diagnosis; all are simple, inexpensive, and highly sensitive⁶.

In this study, male patient was 45.5% and female was 54.5% and mean age was 38.7 years. OMG can occur at any age, but has a bimodal distribution that affects women below age 40 years and men more than 60 years of age. Other predominant study showed that this autoimmune disease more prevalent in female and more common in third decade of life^{3,7,8,9}.

In our study, out of 33 patients, all patients were suffering from ptosis and they all were positive for fatigue test. Variability of ptosis and cogan twitch sign positive in 69.7% and 81.8% cases respectively. Various study showed that two thirds of all cases of MG initially manifest ptosis and/or diplopia. A history of a variable ptosis makes MG the most likely differential diagnosis⁶. In fatigue test involves "fatiguing" the extraocular muscles in upgaze. The patient attempts to sustain extreme upgaze for 30 seconds, then quickly returns to primary position. Patients with OMG often demonstrate either a lid-lag or an increase in ptosis (Darple's sign)⁶.

In this study, respectively 81.8% and 63.6% patients were suffering from variable degree of ophthalmoplegia and diplopia. In myasthenia gravis diplopia is frequently vertical although any or all of the extra ocular muscle may be affected³.

In this study, 87.9% patients were positive for Ice test. The ice-pack test has been shown to be both highly sensitive and specific for OMG in more than 90% of the cases¹⁰. The palpebral fissure is measured, and then an ice pack is applied to the ptotic lid for a minimum of 3 minutes. An increase in the palpebral fissure of 2 mm is considered a positive response. Nonmyasthenic patients do not demonstrate such a change. The ice-pack test can also be performed to look for a decrease in diplopia; however, the results may be less dramatic¹¹.

6.1% OMG patients were positive for Neostigmine test. If the diagnosis of MG is still suspected but unconfirmed, a neostigmine/tensilon test may be performed. Neostigmine/tensilon (edrophonium chloride), an anti cholinesterase drug, inactivates the enzyme that

breaks down acetylcholine. This results in an excess of acetylcholine in the neuromuscular junction, thus producing transimproved muscle function. Immediately following the injection, either the patient's ptosis or eye-muscle function will improve in a myasthenic patient. It should be noted that some patients with MG will have a negative neostigmine/tensilon result. However, more than 90% of patients with OMG will have a positive neostigmine/tensilon test (atropine should be readily available in case of a hypersensitivity reaction)¹².

In this study, 51.5% cases had positive serological test. In various studies around 50% cases have positive serological test findings¹³.

In our study, 18.2% patients presented with diabetes mellitus and 15.1% with hypothyroidism. Patients with MG have a higher incidence of other autoimmune diseases, such as thyroid diseases, systemic lupus erythematosus, diabetes mellitus, Sjogrens, rheumatoid arthritis, pemphigus vulgaris and ankylosing spondylitis¹⁴.

Conclusion

This study stated some clear features of ocular myasthenia. Ptosis was mentioned as the most common symptom. Females were more frequently affected. Cogan's sign can recognize disease clinically at initial stage. Diagnosis can be confirmed by ice pack test, neostigmine test, serological test etc. Early diagnosis and proper treatment is necessary to ensure patients' well being.

Conflict of Interest: Nothing to declare.

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