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Clinical profile and management outcome of ocular myasthenia gravis in South Indian population

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ABSTRACT Myasthenia Gravis, is one of the most well defined forms of autoimmune disease, which involves the neuromuscular junction of skeletal muscles, resulting in weakness of skeletal muscles. The disease can be divided into adult onset and childhood onset. It is the most common disorder that affects the neuromuscular junction.Ptosis is the most common symptom in patients with ocular myasthenia. Females are more frequently affected in age group of less than 15 years and males above 15 years. Cogan's lid twitch sign will be significantly present in myasthenic ptotic eye, irrespective of the grade of ptosis. Thus Cogan's sign can be taken for the initial diagnosis of myasthenic eyes, which can be confirmed by ice pack test, neostigmine test etc. Myasthenia can be affected by environmental temperature. Treatment of myasthenia with pyriodstigmine alone shows significant improvement in patients.

KEYWORDS :

Introduction:
Myasthenia Gravis, is one of the most well defined forms of autoimmune disease, which involves the neuromuscular junction of skeletal muscles, resulting in weakness of skeletal muscles. The disease can be divided into adult onset and childhood onset. It is the most common disorder that affects the neuromuscular junction. Myasthenia is attributable to too few available acetylcholine receptors resulting in impaired transmission across the neuromuscular junction. It is caused by an acquired autoimmunity of the motor endplate, resulting in a decrease in the number of available acetylcholine receptors[1]. When weakness is limited to the extraocular muscles, orbicularis oculi, and levator palpabrae superioris, it is considered to be ocular myasthenia gravis[10].

Aims and objectives:
To determine the presenting complaints, sex preponderance and the age distribution pattern of ocular myasthenia gravis.

Materials and methods:
All clinically proven cases of ocular myasthenia irrespective of age, sex and race were included in the study. The patients were evaluated in detail by torch light and slit lamp biomicroscopic examination for anterior segment. Direct ophthalmoscopy was used for fundus evaluation in all possible cases with reference to supportive indirect opthalmosocpy. Suspected cases of ocular myasthenia gravis were subjected to relevant clinical test viz. eliciting cogan's lid twitch sign. Rest test, Fatigue test. Patient who were clinically suspected for ocular myasthenia gravis, were confirmed by ice pack test (Fig.1 & 2). In cases with doubtful ice pack test, Neostigmine test was used for confirmation. Ptosis of eyelid was evaluated with 15cm plastic ruler. Hess charting and diplopia charting were done wherever applicable to elicit range of limited extra ocular motility and direction of gaze with maximum separation of images. Visual fields were done for patients by confrontation method. Bjerrum's screen was used in cases where field defect was noted by performing confrontation method.

Detailed examination of the motor status of the squint was done wherever applicable.

Results:
Out of 54 patients in the study 37 patients (68.52%) were male and 17 patients (31.48%) were female. The mean age is 39.65 ranging from 8 to 78 years. 53 patients (98.15%) complained of ptosis, 26 patients (48.15%) complained of diplopia, 6 patients (11.11%) complained of strabismus, 33 patients (61.11%) complained of diurnal variation and 5 patients (9.25%) had associated thyroid dysfunction. 41 patients (77.35%) had positive cogan lid twitch sign. Fatigue test was positive in 42 patients (79.24%). 10 (90.9%), 7(80.95%), and 15 (71.42%) patients with mild, moderate and severe ptosis respectively showed positive fatigue test. In patients with bilateral ptosis, eye with worst ptosis was selected and patients who have similar bilateral ptosis, right eye was selected for the test. One patient who didn't have ptosis was negative with cogans sign, fatigue test and rest test showed positive ice pack test with improvement of the ocular movement after 5 minutes of applying the ice pack on his eyes. In our study, 53 patients had ptosis. 37 patients (69.81%) patients were positive for rest test. Maximum number of patients presented to us was in the month of May (10 patients 18.52%) and minimum number of patients presented was in the month of July. The highest cases reported to us correlates with the hottest months in Madurai. In our study, 7 patients were not reviewed at all. 36 patients (76.59%) showed improvement in their symptoms and 11 patients (23.40%) showed no response to treatment. Not even a single patient had worsening of his symptoms.

Discussion:
The ice test has been proposed as a relative simple means of diagnosing ocular myasthenia gravis. The exact mechanism of improvement of myasthenia muscle function has not been completely explained. Cold is believed to affect the neuro – muscular junction both by decreasing cholinesterase activity[1] and promoting efficiency of acetylcholine at eliciting depolarizations at the end plate[2]. Golnik et al[3] investigated subjects who had myasthenic and non-myasthemic ptosis but sued only ice test as the diagnostic tool. They concluded that ice test is a simple, short specific and sensitive test for the diagnosis of myasthemic ptosis. The sensitivity of the ice test in patients with complete ptosis decreases considerably. In our study out of 53 patients with myasthenic ptosis and positive ice test showed no association between the severity of ptosis and logan's did twitch sign, fatigue test or rest test. Simpson (23) has first described the effect of temperature in myasthenia gravis. Ludwig et al[4] has described the deleterious effect of heat on the clinical symptoms and elector – physiological abnormalities of myasthenia gravis. In our study, maximum number of patients attended our clinic were in the month of April (15%) and May (19%) which are considered to be the hottest seasons of the year in this region. The occurrence of myasthenia gravis and hyperthyroidism has been reported[5]. In our study 5 (9%) patients showed associated thyroid disorders.

The treatment for the patients with ocular myasthenia gravis in our study was pyriodostigmate bromide orally. At the end of 4 weeks 36 patients out of 47 (77%) patient who reviewed showed symptomatic improvement, 11 patients (23%) showed no improvement.

Conclusion:
Ptosis is the most common symptom in patients with ocular myasthenia. Females are more frequently affected in age group of less than 15 years and males above 15 years. Cogan's lid twitch sign

will be significantly present in myasthenic ptotic eye, irrespective of the grade of ptosis. Thus Cogan's sign can be taken for the initial diagnosis of myasthenic eyes, which can be confirmed by ice pack test, neostigmine test etc. Myasthenia can be affected by environmental temperature. Treatment of myasthenia with pyriodstigmine alone shows significant improvement in patients.

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