IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 20, Issue 6 Ser.14 (June. 2021), PP 53-58
www.iosrjournals.org

Clinical Profile of Patients with Myasthenia Gravis Admitted In a Tertiary Care Centre in India

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Abstract
*Myasthenia Gravis (MG) is an autoimmune disorder which is characterised by a wide spectrum of clinical symptoms and signs. Aim of the study is to assess the age of onset, gender distribution and clinical presentation of cases admitted in a tertiary care centre in Kerala, India. This was a record based study using medical records of Myasthenia patient admitted in the Neurology department, of Govt. Medical College Hospital Thrissur from August 2012 – July 2017.**Results:** Male: female ratio in our study was in the ratio 1:1.5. There was a single peak of incidence, in the females between 20-30yrs and in males between 60-70 yrs. Ocular symptoms were the most common with ptosis being present in all patients, followed by double vision Diurnal Variation and fatigability most common of the manifestations. **Conclusion:** Myasthenia gravis is a disease of young females with a single peak in the both sexes. Diurnal fluctuation and fatigability are hall marks of the disease. Ocular symptoms especially ptosis predominate followed by diplopia. MGFA classification is of great help in dividing the patients into different sub classes and subgroups.*

***Key Words**: MG (Myasthenia gravis) , MGFA classification(Myasthenia Gravis Foundation of America classification) AchR Ab(Acetyl choline receptor antibodies) , RNS (Repetitive nerve stimulation)*

Date of Submission: 17-06-2021 Date of Acceptance: 02-07-2021

I. Introduction

Myasthenia Gravis (MG) is a relatively rare autoimmune disorder of neuromuscular transmission. The defective neuromuscular transmission is due to the formation of antibodies against acetyl choline (ACh) nicotinic post synaptic receptors at the neuromuscular junction. It is a potentially serious, but treatable disease, characterized by weakness and fatigability of voluntary muscles[1].

The prevalence of MG has increased over the last few decades, because of a) increased life span of patients with the disease b) early diagnosis c) increased availability of investigations to patient[2]. Clinical and epidemiological studies will help us to determine the changing patterns of the disease and provide information for future health care planning. There are few reported studies from India and especially Kerala regarding the epidemiological and clinical profile of myasthenia.

II. Brief review

Acquired Myasthenia can present in two forms. 1) Ocular 2) generalised. Ocular symptoms of MG are caused by weakness of levator palpabrae, orbicularis oculi and other external ocular muscles[3]. Unilateral or bilateral ptosis, horizontal or vertical diplopia, blurring of vision and weakness of eye closure are common symptoms.

When Myasthenia becomes generalised, other muscles like facial palatel, pharyngeal, proximal limb muscles and respiratory muscles become involved. Patients with Myasthenia can be classified into different types using the modified MGFA scale [4]. Fatigability and diurnal variation are the hall marks of the disease and improvement with choline esterase inhibiter neostigmine is an important clue in the diagnosis. Simple clinical test that can be done in the office include. Arm abduction test, lid fatigability test, heel rising test, Icepack test would be great value in the diagnosis of ocular Myasthenia[5].

Repetitive nerve stimulation is usually done at slow rates of 2.3 Hz at room temperature as a diagnostic procedure. Abductor digital minimi, trapezius, nasalis and orbicularis oculi are the muscles tested. RNS is done to look for decremental response of greater than 10% in the baseline to peak amplitude between the first and the fourth response. Serological test like acetyl choline receptor antibody assay and investigations for thymoma like X ray chest and HRCT chest are other investigations employed in the diagnosis of myasthenia. Single fibre

EMG is a very sensitive test but is available only in few sophisticated centers and is very difficult to carry out in ordinary settings[6].

Objectives

To study the clinical presentation, age of onset and gender distribution of patient with myasthenia gravis admitted in neurology department of a tertiary care hospital in Kerala.

III. Materials and Methods

Study design - Retrospective Record based Cross sectional study
Study setting - Department of Neurology, Govt. medical college, Thrissur
Study participants

This is a retrospective study using the Medical case sheets of Myasthenia patients admitted in neurology ward and out patient's tickets of Myasthenia patients attending in the neurology OPD from August 2012 (01/08/2012 to July 2017 (31/07/2017).

Inclusion criteria

All the patients in the study are cases of myasthenia gravis diagnosed on clinical basis and response to cholinesterase inhibitors.

Exclusion criteria

Cases of congenital myasthenia, lower motor neuron syndromes which may mimic myasthenia like Lambert Eaton Syndrome, muscle diseases like congenital myopathies are excluded.

Study period	-	3 months
Simple size	-	All cases during the period 01/08/2012 to 31/07/ 2017
Data collection tool	-	structured proforma
Methodology	-	Data collection Record based Analysis.

Secondary data of patients with MG was collected using a structured proforma. Data on clinical presentation, age of onset, gender and clinical features and diagnostic test employed were collected. Variables included were gender, age of onset and clinical presentation. Patients were grouped according to the MGFA Classification.

Data Analysis

Data was entered in MS excel & analysed using appropriate software like Epi info. Qualitative data was analysed using proportions. It was tabulated & discussed.

Ethical consideration

This is a record based study. It was started after getting the approval from the research and ethical committee. Confidentiality of data will be maintained.

IV. Results

This is a retrospective study using the Medical case sheets of Myasthenia patients admitted in neurology ward and out patient's tickets of Myasthenia patients attending in the neurology OPD from August 2012 (01/08/2012 to July 2017 (31/07/2017).

There were 78 patients, 31 (40%) male and 47 (60%) female patients.

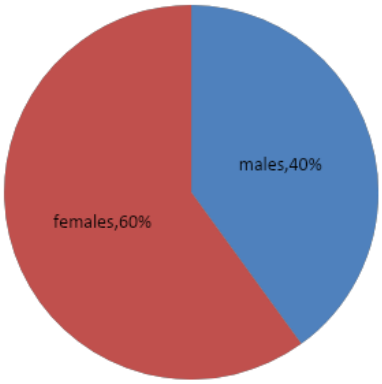

Figure 1 GENDER RATIO

There were no patients below 10 years and above 70 years. The distribution of patients in the various age groups 11–20, 21–30, 31–40, 41–50, 51-60 and 61-70 were 9, 19,12, 12, 11 and 15 respectively; and, their corresponding percentages were 12,24,15 16, 14 and 19 respectively (shown in figure 2).

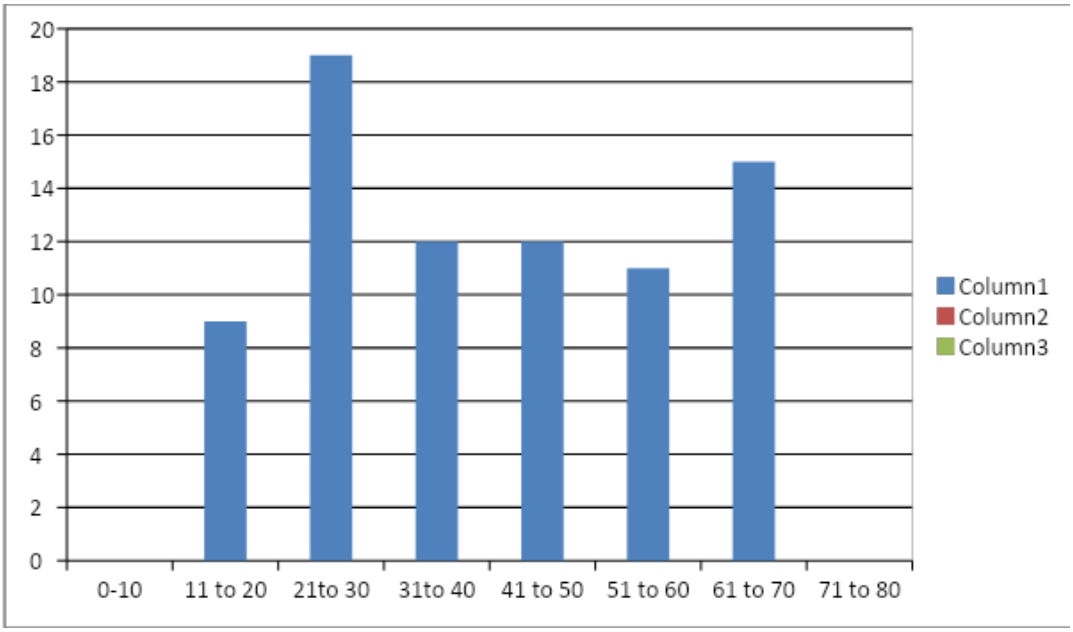

Figure 2 AGEWISE DISTRIBUTION OF PATIENTS

There was unilateral ptosis in 19 patients in the myasthenia group and bilateral involvement in 59 patients. Among the cases with bilateral ptosis, 53 patients had unequal involvement of both eyes. Diurnal variation and fatigability were present in 70 patients (90%). Diplopia was present in 31 (40%) patients in the myasthenia group in addition to ptosis. The incidence of other clinical features, signs, and bedside tests are enumerated in the figures3 and 4.

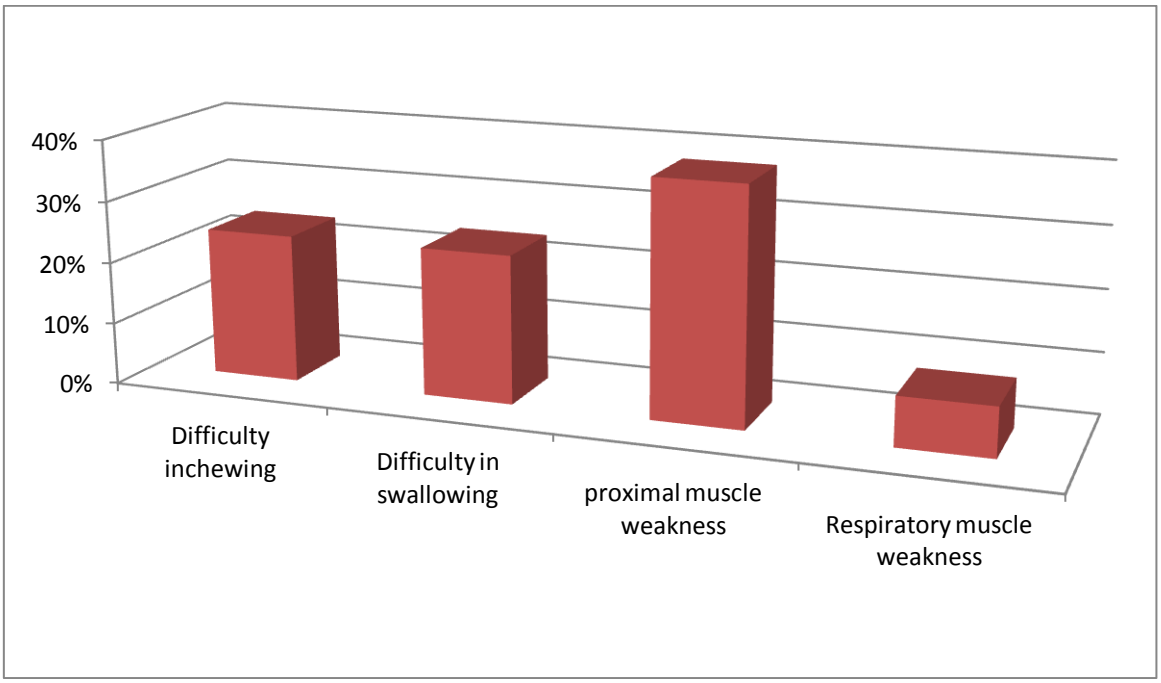

Figure 3 FREQUENCY OF SYMPTOMS

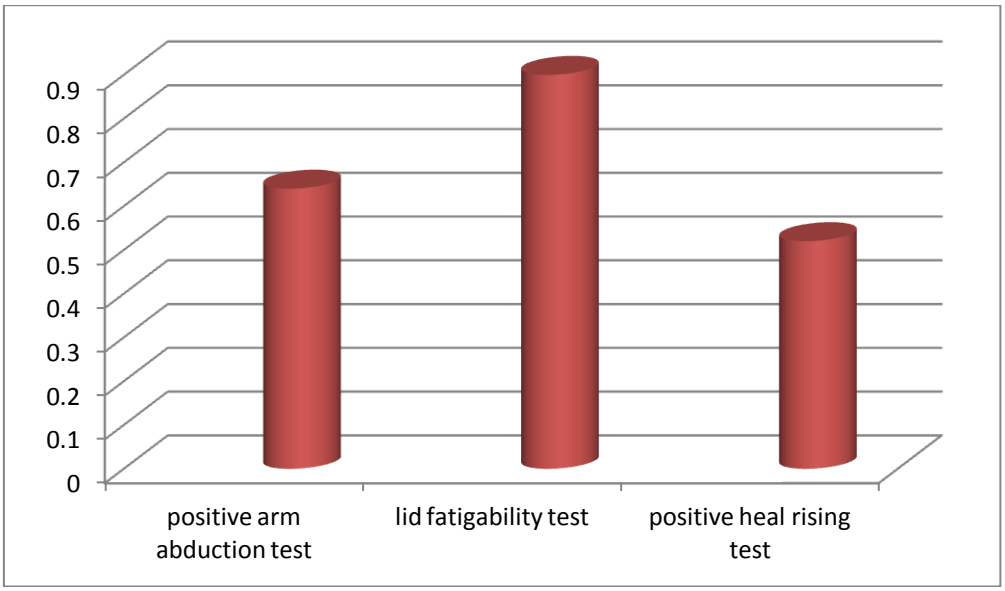

Figure 4 POSITIVITY OF BEDSIDE TESTS

The ice pack test was positive in 70 (90%) patients and negative in 8 patients. RNS was positive in 62 (80%) of the myasthenia patients. Of these, 31 (52%) patients showed a decremental response in the orbicularis oculi and nasalis muscles. Neostigmine test was done in 72 patients, of whom, 60 showed a positive response and 12 showed a negative response. AChR-Abs were done in 27 patients, of whom 10 patients had a positive value. HRCT chest was done in 57 patients and 16(21%) showed a thymic enlargement.

Based on the Myasthenia Gravis Foundation of America (MGFA) classification, 37 (48%) patients belonged to Class I,12 (16%) patients to Class IIa, 5 (4%) patients to Class IIb, 4 (6%) patients to Class IIIa, 5 (6%) patients to Class IIIb, 4 (6%) patients to Class IVa, 5 (6%) patients to Class IVb, and 6 (8%) patients to Class V. Illustration in Fig. 5

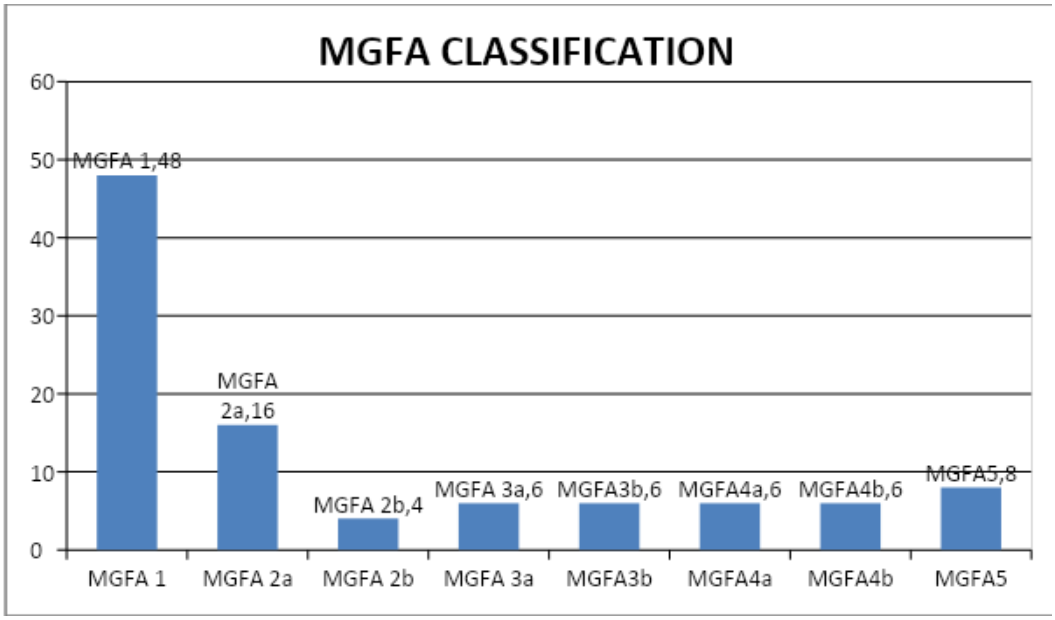

Figure 5 MGFA CLASSIFICATION

V. Discussion

In our study group, incidence of male and female patients were 1:1.5. Myasthenia is considered as being a disease of young females. Two studies from Europe by Poulas et al and Robertson et al reported an incidence of 1:1.4 and 2:1sex ratio[7,8]. In our study there was a single peak in the females between 20to 30 years and in the males between 60 and 70 years. This single peak in females between 20 and 30 is in concordance

with studies of Poulas et al and Saha et al [9]. A similar single peak in the males between 60 and 70 years has been described by Poulas et al and Singhal et al [10]. The mean age of the patients was 34 years in the females and 62 in the males.

There was unilateral involvement in 19 patients (24%) with myasthenia; and, out of the 59 bilateral cases, 53 had asymmetrical involvement of one eye (89%). Asymmetrical ptosis was found in our study. Diplopia in addition to ptosis was present in 31 (40%) patients. The frequency of diplopia was reported as 38% in the population studied by Leeamornsiri *et al*., [11]Extraocular muscles (EOMs) are needed to sustain the gaze in all directions. Twitch fibers in them have an increased frequency of synaptic firing than limb muscles. They develop tension faster .This makes them susceptible to fatigue. ACh receptors are fewer in EOM which makes them more susceptible to receptor damage. Another contributing factor for the susceptibility of EOMs may be the differential expression of type of ACh receptor when compared to the skeletal muscle this may be the reason for the increased incidence of ocular involvements in our study .

Fatigability and diurnal variations were present in 67(86%) patients and also form a hallmark of the disease. Along with the presence of auto antibodies, reduction in the number of ACh receptors also results in the characteristic pattern of progressively reduced muscle strength with repeated use of muscle and recovery of muscle strength following periods of rest.

Difficulty in chewing, speaking, and swallowing was present in 24% of the myasthenic patients. In 36% of patients there was affection of the proximal muscle.Respiratory difficulty was present in 4 (8%), bulbar weakness in 12 (24%) and proximal muscle weakness in 18 (36%) patients. In another study, the incidences of bulbar and proximal muscle weakness were 56.25% and 32.25%, respectively[12]. Six (8%) patients were in myasthenic crisis in our study. Similar incidence (9%) of myasthenic crisis has been reported in an Indian study[13]. Orbicularis oculi weakness was present in 44 (88%) patients of the myasthenia group.

Eye fatigability was positive in90% of the patients . Arm abduction was positive in 64% of the patients. Heel rising test was positive in 52% of the patients.Ice pack test was positive in 90% of the patients. Ertas *et al* reported an ice pack test positivity of 80% in his study[14]. RNS was positive in 80% of the patients. RNS positivity was reported to be 76.5 % and 79.5% respectively in the studies of Leeamornsiri et al and Singhal et al. Neostigmine test had a positivity of 83%. Saeed et al has reported a positive Neostigmine response in his patients . AChR-Abs were found to be positive in 10 (28%) patients out of 57 patients. Meriggioli et al has reported positivity 40-50 % for the AChR-Abs [15]. HRCT was done in 57 patients and 16 patients showed thymic enlargement.

As myasthenia gravis is a disease with variety of clinical symptoms there are many systems of classification of MG patients. Of these the most widely excepted one is the Myasthenia Gravis Foundation of America (MGFA) clinical classification. This classification divides the patients in to 5 classes and several subtypes depending upon the clinical feature and the severity.

Patients with only ocular symptoms were more frequent 48% in our series. In the studies of Mantegazza et al the incidence of ocular symptoms comes to 39.3%[16]. Eight percent of patients in our series had respiratory difficulty and required intubation . Saha et al has reported nine percentage incidence of myasthenic crisis requiring ventilatory support in his studies.

VI. Conclusion

Myasthenia gravis is more common in the young females. There is a single peak of incidence in both the sexes (female population between 30-40 years), and in the (males between 60-70 years).

Diurnal variation and fatigability are hallmarks of the disease Ocular symptoms are more common in the study population Ptosis followed by diplopia is the commonest symptoms

MGFA classification helps in a better way to arrange patients depending upon the symptoms and severity.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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