ORIGINAL ARTICLE

Spectrum of Clinical Features of Myasthenia Gravis at The Time of presentation

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ABSTRACT
Objective: To determine the spectrum of clinical features of myasthenia gravis at the time of presentation
Methods: Forty cases of myasthenia gravis presenting to the Neurology Outpatient Department, Mayo Hospital Lahore were registered. After recording demographic data, their clinical features and functional status were noted and recorded on a specified proforma.
Results: Myasthenia gravis, seen roughly equally in both sexes, has a wide range of presentation with various clinical features. The most common initial involvement was ofocular muscles among all followed by bulbar and upper limbs. However facial, lower limb and respiratory muscle involvement was also seen.
Conclusion: Myasthenia gravis has a wide spectrum of clinical features. Clinicians ought to be aware of all these features as well as their relative frequencies in order to be able to aptly recognize and diagnose myasthenia gravis early.

INTRODUCTION
Myasthenia gravis (MG) is an autoimmune disease in which the attack is at the level of postsynaptic acetylcholine receptors of striated muscles and it leads to weakness and fatigability of muscles, which is its characteristic feature[1,2]. In this disease, the antibodies target the postsynaptic nicotinic acetylcholine receptors and it is relatively rare in incidence[3,4]. The exact pathogenesis behind the production of these antibodies is not entirely clear; however, it has been stated that certain genotypes are at greater risk[5]. The specific antibody responsible for the genesis of the disease has been identified and it has been found to be IgG antibody against acetylcholine receptors in about 90% of generalized myasthenia gravis cases[6].

In United States, the annual incidence of disease is estimated to be 2 per 1,000,000; whereas the prevalence varies from0.5 to 14.2 cases per 100,000 people[7]. Earlier diagnosis and increased life expectancy of myasthenia gravis patients has caused this figure to rise over the past 20 years[7].

As regards the sex distribution, the female to male ratio has been described to be 3:2. In the younger age group (i.e. 20-30yrs), the disease shows a female preponderance, whereas in the age group older than 50 years, males predominate[7,8]. As compared to other races, Asians predominate in young onset disease[7].

In a Canadian study conducted on 57 pediatric patients of myasthenia gravis, it was found that ptosis was the most commonly occurring symptom, and it was present in all patients of ocular juvenile myasthenia gravis and in 82% of patients having generalized juvenile myasthenia gravis[9]. Acetylcholine receptor antibodies result came out to be positive in 67% of generalized juvenile myasthenia gravis patients and 44% of ocular juvenile myasthenia gravis patiets[9,10]. It takes over weeks to months for the disease to progress from mild to more severe. The pattern of spread of weakness is from ocular to facial to bulbar muscles, and then to trunk and limb muscles[11].Fluctuating weakness and fatigability of affected muscles marks the clinical features of the disease[7].

Ptosis and/or diplopia are the presenting features in more than half of patients[12,13].About half of patients presenting with ocular manifestations progress to generalized disease within 2 years[14]. Fatigability of chewing develops due to involvement of muscles of jaw closure[15]. Eighty two per cent of patients reached their maximum level of weakness within 2 years in a case series in US that involved 1976 patients having myasthenia gravis[8]. Whereas a

retrospective study in Italy that involved 1152 patients, maximum level of disease was reached in 3 years by 77% of patients[16].

The purpose of our study was to assess the different modes of presentation of myasthenia gravis, the parts of body most commonly involved and the functional status at presentation of patients of myasthenia gravis in our region of the world, as studies in this regard are lacking in our region.

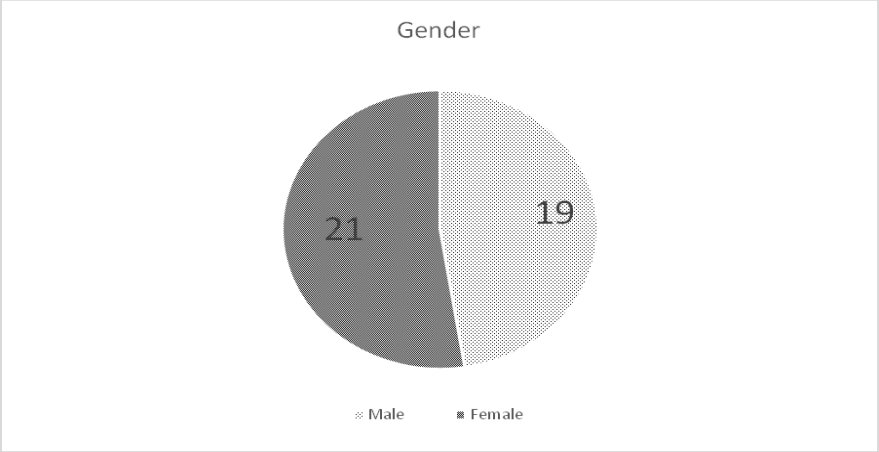
MATERIALS AND METHODS

This study was conducted in the Department of Neurology, Mayo hospital Lahore after permission from ethical review committee. 40 patients of myasthenia gravis were registered for this purpose who presented to the Neurology Outpatients Department, and their biodata was recorded. Confirmation of the diagnosis was done by prostigmine test and repetitive nerve stimulation test performed at the electrophysiology section of Neurology Department, Mayo Hospital Lahore. Their clinical evaluation was done determined regarding involvement of different parts of the body and the functional status of the patients at presentation. All the record was noted on a specified proforma.

RESULTS

Sex Distribution

Male 19
Female 21



First Part Ever Involved

Site	Male (19)	Female (21)	Total (40)	Percentage
Ocular	11	10	21	52.5%
Facial	1	3	4	10%
Bulbar	5	6	11	27.5%
Upper Limb	6	3	9	22.5%
Lower Limb	4	2	6	15%
Respiration	0	0	0	0%

Parts Affected At Presentation

Site	No. of Cases	Percentage
Ocular	40	100%
Facial	20	50%
Bulbar	29	72.5%
Upper Limb	32	80%
Lower Limb	9	22.5%
Respiration	3	7.5%

Functional Status At Presentation

Site	No. of Cases	% age
Ptosis	38	95%
Diplopia	24	60%
Dysphonia/Dysarthria	28	70%
Disability to eat	16	40%
Disability to button	28	70%
Decreased walking	22	55%
Dyspnea	5	12.5%

DISCUSSION

The sex distribution of myasthenia gravis in our study was roughly equal, with a slight female preponderance. The most common presenting symptoms in our patients were ocular which is also the case with other international studies[9,10]. The number of patients presenting with the first symptom as involvement of ocular muscles were more than half (i.e. 52.5%), which again correlates well with international studies[12,13] . Next common presenting features were bulbar (27.5%), followed by upper limb (22.5%). Respiratory symptoms as a mode of presentation are reported to occur in 14-18% cases[17], however it was not seen in a single case in our study. At the time of presentation to us, ocular muscle involvement was seen in all cases, followed again byupper limb and bulbar symptoms. So these three regions of body remain the most commonly involved in patients of myasthenia gravis in our population. As regards the functional status, most common disability was ptosis, followed by dysphonia/dysarthria and disability to button. Here again disability of ocular and bulbar regions remains the most common. However, other regions of body are also commonly involved, including facial, limbs and respiratory muscles.

CONCLUSION

Myasthenia gravis has a wide spectrum of clinical features. Clinicians ought to be aware of all these features as well as their relative frequencies in order to be able to aptly recognize and diagnose myasthenia gravis.

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