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The Epidemiologic, Clinical and Laboratory Findings of Patients with Myasthenia Gravis in Southern Iran

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Abstract

Introduction: Myasthenia gravis (MG) is an autoimmune disorder of neuromuscular junction associated with presence of antibodies against nicotinic acetylcholine receptors (nAChRs). The pattern of the MG varies in different ethnical and geographical regions. Data regarding the pattern of the disease in Iran is scarce. Thus we performed this study in order to describe the epidemiologic, clinical and laboratory characteristics of MG in Iranian population. **Method and Material**: This was a retrospective study being performed in Nemazee Hospital, a tertiary health care center affiliated with Shiraz University of Medical Sciences, from 2001 to 2010. The medical records were reviewed and the data were entered into a computer database. Data are presented as mean ± SD and proportions as appropriate.**Result**: Overall we included 208 patients with MG among whom there were 62 (29.8%) men and 146 (70.2%) women. The mean age of the patients was found to be 33.19 ± 15.75 (range 1-85) years. The median age at onset was 28±2.3 years. Eight (3.9%) patients had family history of MG, 1.9% and 2.4% of patients had a background of Diabetes Mellitus and Rheumatoid disease in family, respectively. Thymoma was observed in 9.1% of patients. The presenting symptom was found to be ocular ones detected in 67 (34.9%) patients out of which 59 (30.7%) had bulbar and 12 (6.3%) had appendicular involvement. Edrophonium test was done for 28 patients out of whom 78.6% tested positive. The most common histopathology finding was thymic hyperplasia and the second most common was thymoma.
Conclusion: This is the first study describing MG in an Iranian population. The pattern of disease was found to be much more similar to North America and Europe. MG was found to be more common in females consistent with western studies. **[GMJ. 2012;1(1):20 -23]**

Keywords: Myasthenia Gravis – Epidemiology – Clinical findings – Thymectomy – Iran

Introduction

Myasthenia gravis (MG) is a relatively rare autoimmune disorder of neuromuscular transmission in which antibodies form against acetylcholine (ACh) nicotinic postsynaptic receptors at the neuromuscular junction. A reduction in the number of ACh receptors (AChR) results in a characteristic pattern of progressively reduced muscle strength with repeated use of the muscle and recovery of muscle strength following a period of rest.
The prevalence of MG has increased over the past two decades, primarily because of the increased life span of patients with the disease but also because of earlier diagnosis (1). Epidemiological studies which improve

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advances in laboratory investigations allow to determine the changing patterns of disease, testing of etiological hypotheses and provide information for health care planning (2). In an African study, the data of 190 under 20 years MG individuals were analyzed and found that Ocular MG (26%) happened among younger children (mean 5.1 years), compared to those developing generalized MG (74%) (mean 10.2 years) (p=0.0004) (3). Dr. Kalb and his coworkers in a resemble study estimated the prevalence and selected characteristics of MG in the county of Stockholm, Sweden and concluded that the mean age at onset for men and women was 48.5 and 34.9 years, respectively also generalized MG was found to be in 79% of patients, and 10% had severe symptoms (4). There is a lack of epidemiological data from Iran. We present findings from our data (collected over 10 years 2001-2010) from a single tertiary referral center in Iran.

Methods and Materials

This was a retrospective study was performed in Nemazee hospital a tertiary health care center affiliated with Shiraz University of Medical Sciences from February 2001 to March 2010.

All the patients were cases of myasthenia gravis according to the Medical Scientific Advisory Board (MSAB) of the Myasthenia Gravis Foundation of America (MGFA) (5) diagnosed on clinical manifestations that were reviewed in a 10 years period. The review and ethical board of Shiraz University of Medical Sciences approved the study.

We did this survey retrospectively by reviewing extracted data from patient's history in hospital. We collected data of gender distribution, age at onset, duration of admission, age, family history of MG and other autoimmune diseases, manifestations of disease, Thymectomy and response to it for each patient. Also we reported the medications beside thymectomy.

Statistical Analyses

The data was recorded by a brief checklist entered to a computer data base. Data was analyzed by Statistical Package for the Social Sciences version 15.0 (SPSS Inc., Chicago, IL). Descriptive results are presented as mean value ± standard deviation (SD) for 95% confidence interval (CI) with or proportions.

Results

All the 208 patients had acquired Myasthenia and we couldn`t find any congenital one. Of this number 62(29/8%) were males and 146(70/2%) were females. The F:M ratio was 2/35:1. The median age at onset was 28±2.3 years (interquartile range 19-38 years); in females it was 28 years (interquartile range 19-38 years) and in males it was 25/75 years (interquartile range 20/5-31/5). The mean age of the patients was 33/19 years, (females-33/29 years, males-32/98 years), (range 1-85).

Just 8(3/9%) patients had family history of MG. 1/9% and 2/4% of the patients had a background of DM and Rheumatoid disease in family respectively.

Thymoma was observed in 9/1 % of the patients. Eleven (5/3%), eight (3/8%), four(1/9%) and tow(1%) of the patients had thyroid disease DM, Lupus, and Rheumatoid disease respectively. The first presenting symptoms (onset symptoms) was found to be Ocular ones detected in 67(34/9%) patients out of which 30/7% had Bulbar and 12(6/3%) had Appendicular involvements.

Between the patients that had Ocular symptoms interval the disease 151 (72/9%) patients had Ptosis (dropping of eyelid) and 98 (47/3%) patients had Diplopia (double vision) involvements; while of the patients that had bulbar manifestations 112(54/4%) ones had Dysphagia (difficulty in swallowing), 101(49%) ones had Dyspnea (respiratory disturbance) and 105 (51%) had Dysphonia (disability and disturbance in speaking and pronunciation). Nasal speech was the most common claim of the patients with dysphonia involvement. Also some manifestations like eye movement, anorexia, odynophagia, horseness, frozen eye and lip dropping were observed. We could find 12 (5/7%) cases of regurgitations that 8 of them were nasal; aspiration was found in just 3 (1/44%) cases. Most of the patients (93/8%) were admitted just once in the ER ward while there was a case that was admitted for 4 times. All the autoimmune disorders that we sur-

veyed were approximately alike in both gander and the manifestations were not highly different except dyspnea that was more common in females; also dysphagia had same result but it was not significant (P= 0/082).
Edrophonium test was done for 28 patients out of whom 78/6% tasted positive. Myasthenia crisis was found in 23/6% of the patients and 90(43/3%) of them were admitted to Intensive Care Unit (ICU). The most common histopathology findings was thymic hyperplasia and the second one was thymoma. Plasmapheresis was done for 55/3% of the patients, whereas IVIG (another therapy) had given just to 16/1% of them; with a mean dose of 26/65 gr (more common dose was 20 gr). Thymectomy is the most effective way to treat MG; we can say it according statistics. This operation was done for 152 patients from whom 93/1% of them improved dramatically. Despite thymectomy for whom that has thymoma is dangerous it was done for 16 ones of 19 cases. Mestinon is an acetylcholinesterase inhibitor drug that had given to 91/7% of the patients; while prednizolone, azathioprine and cellcept that are immunosuppressive drugs had given to 42/7%, 27/2% and 6/8% of the the patients, respectively.
Overall 197 (94/7%) patients were ambulate and able to continue to ordinary life after treatment but by some limitations.
Eleven cases in this survey expired that three of them were due to heart block, respiratory-heart block and infarction. From eight ones that were dead in result of MG, seven cases had respiratory disturbances. In one case first diagnosis was Rheumatism instead of MG so neurologists may be has wrong diagnosis with alike manifestations.

Discussion

In our study the most frequent manifestation were ptosis,72.6%, and other features such as dysphonia, dysphagia and dyspnea were relatively common. Similarly the commonest presentation inojini(6) study was ptosis followed by diplopia and limb weakness, but other features such as dysphonia, dysphagia and dysarthria were relatively uncommon .in other study(10) It has been established that the ocular form of myasthenia gravis is more prevalent in Uzbekistan than the pharyngeo-facial one.
29.8% of the patients were male and 70.2% were female and also 71.6% were between 20 and 60 years old relatively the same as one study (7) that demonstrated more than half of all patients with myasthenia gravis were female and almost four fifth fell it before the age of 50 years. This might well reflect the general belief in the second half of the 20th century that MG occurred in young women.
Acetylcholiensterase blocker are still used as the first line treatment of myasthenia gravis. In our study acetylcholinsteras blocker (Mestinon) was used in most of the patients, in about 90.9%, however immune suppressive drugs and IVIG were used in 42% and 14.4% respectively.
Pathology of thymus showed hyperplasia in 23.1%, thymoma in 6.7% and in 1.4% thymoma and hyperplasia were seen together, in another study (8) CT scans of the thymus showed thymus enlargement, thymoma and thymus hyperplasia in 51.5% (22/43), 11.6% (5/43) and 2.32% (1/43) of the patients, respectively.
Thymectomy was carried out on 63.7% of the patients in MantegazzaR (8) study, and in our study 73.1% had operation and13.0% of patients had postoperative complications and 1% didn`t have and in other study(9)the postoperative complications were found in 13.1% of patients.

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References

1. Evoli A, Batocchi AP, Tonali P. A practical guide to the recognition and management of myasthenia gravis. Drugs 1996;52:662-70.
2. Robertson N P,Deans J, Compston D A S. Myasthenia gravis: a population based epidemiological study in Cambridgeshire, England. Neurol Neurosurg Psychiatry 1998;65:492–6.
3. Heckmann JM, Hansen P, Van Toorn R, Lubbe E, Janse van Rensburg E, Wilmshurst JM. The characteristics of juvenile myasthenia gravis among South Africans. South African Medical Journal. 2012 May 23;102(6):532-6.
4. Kalb B, Matell G, Pirskanen R, Lambe M. Epidemiology of Myasthenia gravis: A Population-Based Study in Stockholm, Sweden. Neuroepidemiology. 2002; 21: 221–5.
5. Jaretzki A, Barohn R J, Ernstoff R M, Kaminski H J, Keesey J C, Penn A S. Myasthenia Gravis: Recommendations for Clinical Research Standards. The Annals of Thoracic Surgery. 2000;70:327-34.
6. Ojini FI, Danesi MA, Ogun SA. Clinical manifestations ofmyasthenia gravis: review of cases seen at the LagosUniversity Teaching Hospital. Niger Postgrad Med J 2004;11:193-7.
7. Rastenyte D, Vaitkus A, Neverauskas R, Pauza V .Demographic and clinical characteristics of patients with myasthenia gravis.Medicina (Kaunas). 2002;38(6):611-6.
8. Notash AY, Salimi J, Ramezanali F, Sheikhvatan M, Habibi G.Clinical features, diagnostic approach, and therapeutic outcome in myasthenia gravis patients with thymectomy.Department of Surgery, Medical Sciences/University of Tehran, Tehran, Iran. ActaNeurol Taiwan. 2009 Mar;18(1):21-5.
9. Mantegazza R, Baggi F, Antozzi C, Confalonieri P, Morandi L, Bernasconi P, Andreetta F, Simoncini O, Campanella A, Beghi E, Cornelio F .Myasthenia gravis (MG): epidemiological data and prognostic factors. Ann N Y Acad Sci. 2003 Sep;998:413-23.
10. SamibaevMKh, Gabrielian MI, Allazogly N.Features of the clinical picture and results of the treatment of myasthenia in Uzbekistan.1985;85(11):1658-61.