IMMUNOMODULATION IN MYASTHENIA GRAVIS: NEUROPHYSIOLOGICAL AND LABORATORISTIC SERIATE ASSESSMENT

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Objectives

Myasthenia gravis is an autoimmune disease characterized by excessive muscular fatigue due to a neuromuscular junction disorder. Single fiber electromyography (SFEMG) and the presence of specific antibodies are useful in diagnosis, with high sensitivity (SFEMG), and high specificity (antibodies). The aim of this work is to show an utility of these exams in order to evaluate the clinical response to Intravenous immunoglobulin (IVIG).

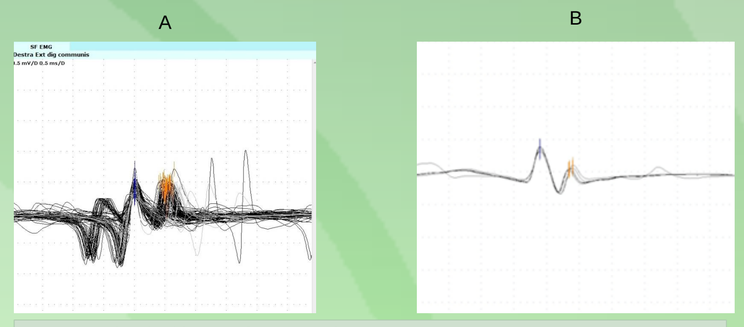

Figure 1. Single fiber EMG potentials
A abnormal jitter *B* normal Jitter

Table 1. Characteristics of 5 patients with MG

Patients	sex	age at onset	Kind of onset	Osserman classification	Thymic alteration	Anti achR	anti musk	anti titine
1	f	41	diplopia, weakness	IIa	yes	yes	no	no
2	f	51	diplopia,dyspnea	IIb	yes	yes	no	yes
3	f	60	dysphagia/ dysarthria,ptosis	IIb	no	yes	no	yes
4	f	28	ptosis	IIb	no	no	yes	no
5	m	69	hypophonia	IIIb	no	yes	no	yes

Materials and methods

Five patients with moderate/severe generalized myasthenia gravis, with a poor response to classical treatment (pyridostigmine, prednisone, azathioprine), underwent to IVIG treatment 0,4 gr/kg die for 5 days every 4 week, for three times. At baseline and 4 weeks after each treatment we performed a clinical evaluation using the Myasthenia Gravis-Specific Activities of Daily Living Profile (MG-ADL), SFEMG with concentric needle on extensor digitorum communis and the dosage of anti AchR, anti Musk e anti Titine antibodies.

Results

The mean age at diagnosis was 49,8±16 years; the mean MG-ADL score was 7,25±5,44 at baseline and 3,25±4,03 after 4 weeks from the third cycle of IVIG; the mean jitter was 64,6 µs at baseline and 41,78 µs after treatment; the mean percentage of pathological jitter was 80% at baseline and 44% after treatment. 4 patients have anti AchR antibodies at baseline and three of them were also anti Titine positive. Only one has anti Musk antibodies. The mean titer of anti AchR antibodies was 12,73 nmol/L at baseline and 8,82 nmol/L at the last follow-up. In patients with ab anti Titine, the mean titer was 32,0 at baseline and 23,6 at last follow up.

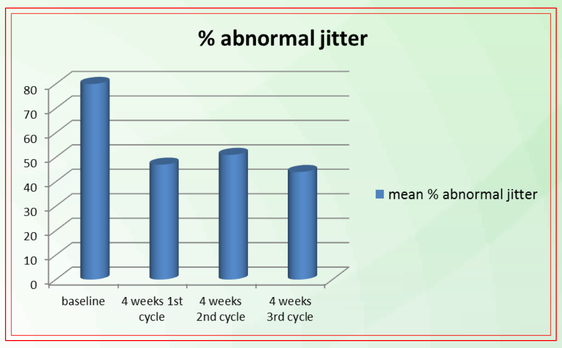

Figure 2. % abnormal jitter at single fiber EMG changes during treatment for 5 patients

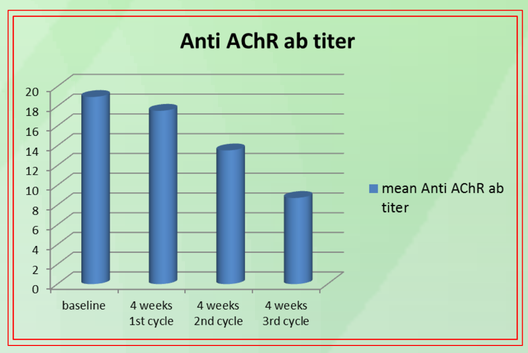

Figure 3. Mean anti AchR antibodies titers changes during treatment for 4 patients

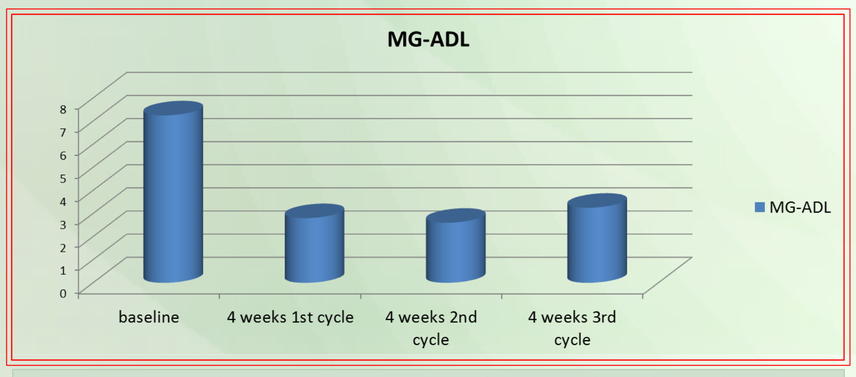

Figure 4. Myasthenia Gravis-Specific Activities of Daily Living Profile (MG-ADL) changes during treatment for 5 patients

Discussion

Despite the small number of patients, both SFEMG parameters and antibodies titers decreased with IVIG in 5 patients with moderate/severe generalized myasthenia gravis, according to the clinical course.

Conclusion

SFEMG and Ab titers are useful in myasthenia gravis diagnosis, but they lack of prognostic and predictive value. EMG could be very useful in right hands, but it shows a great variability due to operator experience. The absolute titers of specific antibodies seems to not correlate with clinical severity of disease in other studies, but the variability of their titers after immunomodulation with IVIG could correlate with clinical course. This little work shows a possible correlation between SFEMG, ab titers and clinical course of myasthenia gravis. A larger patients number could be helpful to assess the utility of these biomarkers.

References:

1- *Correlating extent of neuromuscular instability with acetylcholine receptor antibodies.* Farrugia ME, Jacob S, Sarrigiannis PG, Kennett RP. Muscle Nerve. 2009 Apr;39(4):489-93.

2- *Predictors of response to immunomodulation in patients with myasthenia gravis.* Katzberg HD, Barnett C, Bril V. Muscle Nerve. 2012 May;45(5):648-52.

3- *Biomarker development for myasthenia gravis.* Kaminski HJ, Kusner LL, Wolfe GI, Aban I, Minisman G, Conwit R, Cutter G. Ann N Y Acad Sci. 2012 Dec;1275:101-6

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