Introduction

Myasthenia Gravis has a special implication in the neuromuscular field as it is a potentially treatable disease among many others. The onset of the disease may be from early infancy to late elderly. Diverse clinical and laboratory characteristics of the disease make the accurate diagnosis and treatment simple but impossible in many instances. Furthermore, while different MG subtypes may impart special prognostic and therapeutic characteristics, the course of the disease specifically its maximal severity and response to treatment cannot be fully predicted in all cases. Altogether, the puzzle of MG is presumably defined by different demographic, clinical, serologic, electrophysiological and genetic properties. Each of these factors may affect disease severity and treatment outcomes alone or in combination. In this study, we studied Iranian MG patients from various diagnostic and therapeutic perspectives. We specifically investigated the relationships between these factors and disease severity and response to treatment over a follow-up period of more than 5 years.

Method

Patients with the diagnosis of MG, referred to the neuromuscular clinic of Shariati Hospital were enrolled. MG diagnosis was considered in clinically indicated cases who had one or more of the following findings: positive anti-AChR antibody, positive Anti-muskel antibody, positive 3H RNS (>10% decremental response), positive single fiber EMG, significant improvement after a trial of oral AChEI or Edrophonium test. All patients had a chest CT with IV contrast to evaluate the state of thymus. MGFA clinical classification was used to determine maximum disease severity, Class I: Ocular; Class II: Mild generalized; Class III: Moderate generalized; Class IV: Severe generalized. MGFA post-intervention status was used as the overall outcome of the therapy as follows: Clinical Stable Remission (CSR), Pharmacologic Remission (PR) and Minimal Manifestation (MM) as ‘good’ response, Improved (I) as ‘intermediate’ response and Unchanged (U) or Worse (W) status as ‘poor’ response. The study was approved by the research committee of Tehran University of Medical Sciences.

Results

In our series of 146 patients, MG was more severe in older, anti-Muskel positive, and thymomatous cases (Table 1). Despite differences in the course of the disease and treatment options (Figure 1), the overall outcome was favorable in the majority of patients. Seropositivity to Musk antibody and the presence of thymoma determined the need to immunosuppressive drugs. Nevertheless, these patients did not show further resistance to the conventional immunosuppressive therapies.

Conclusions

We tried to depict a holistic picture of MG in 146 patients over a long course of the disease (6 -37 years). Our data not only include diagnostic evaluations but also disease severity, treatment choices, clinical course and response to treatment in Iranian MG patients. In our series, maximum disease severity was significantly related to age, antibody status and thymus pathology. Despite the differences in the disease course and severity, the overall outcome was favorable in the majority of patients with MG. In contrary to the previous reports anti-Muskel positive patients in our series did not need a more vigorous treatment comparing anti-acetylcholin receptor positive or seronegative patients.

References