Late-Onset Myasthenia Gravis

A Changing Scene

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The prevalence of myasthenia gravis (MG) among middle-aged and older patients has increased. Patients with early-onset MG live longer than before, but there is also an increase in late-onset MG (onset of the disease after the age of 50 years in patients with no clinical or paraclinical evidence of a thymoma). Epidemiological data support using the age of 50 years to separate early- and late-onset MG. The main immunological difference between early- and late-onset MG is the presence of antibodies to muscle titin, which are detected in approximately 50% of patients with late-onset MG. Treatment of late-onset MG has to be tailored both to the age of the patient and to the immunological findings of that particular form of MG. Arch Neurol. 1999;56:25-27

The disease occurs, for the most part, in the third decade, and is rare before the age of 15, or after 70. In approximately 60 percent of the patients the disease develops between the ages of 20 and 40.1

The concept that myasthenia gravis (MG) mainly affects young adults and is uncommon after the age of 50 years was based on clinical experience and supported by epidemiological data. In 1900, when Campbell and Bramwell2 surveyed the literature and added 1 case of their own, they identified 60 cases, 3 of which involved patients who were older than 50 years at the onset of their disease. By 1953, Schwab and Leland3 reported that 62% of women and 27% of men with MG were younger than 30 years at the onset of the disease. The corresponding figures reported by Simpson et al4 in 1966 were 49% women and 23% men. In both studies, the disease was uncommon among younger men, with the majority of male patients being older than 60 years. Men with MG formed 2 groups: one with the peak age at onset between 25 and 35 years and the other between 60 and 70 years.5 These observations were reflected in standard textbooks of neurology.

Today, some 40 years later, the onset of MG occurring after the age of 50 years is not uncommon. Recent studies have shown an increased prevalence of the disease among middle-aged and older patients.5-8 This may be the result of both an improved prognosis and a more advanced medical diagnosis among elderly patients. Certainly, most patients with MG receive better treatment today and have a longer life span than ever before. However, the data suggesting a higher prevalence of MG cannot be explained by an accumulation of patients with early onset of the disease who live longer, although in the 1990s the life expectancy of patients with early-onset MG who have undergone a thymectomy does not differ from that of the normal population.9

There are also a substantial number of patients who develop the disease much later in life. Three recent studies, from western Denmark,5 central and western Virginia in the United States,6,7 and Croydon, Great Britain,8 have demonstrated an increased incidence of MG in the elderly population. In a Dutch study of 100 consecutive patients with MG who were referred between 1985 and 1989, 33% were older than 50 years at the onset of the disease.10 In an epidemiological study from central and western Virginia, the prevalence was significantly higher in individuals older than 50 years.8 Moreover, when prevalence rates were analyzed, the figures for MG showed an increase over time, compared with rheumatoid arthritis and systemic lupus erythematosus, for which trends were static.11

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Myasthenia gravis has traditionally been regarded as a disorder of young women and older men. The incidence of MG is still higher among younger women than it is among young men, but recent studies have demonstrated that both sexes now show a bimodal curve for age at onset of MG. The age peak for late-onset MG (onset after the age of 50 years) is now the same for both sexes, between 70 and 80 years, mainly because of a relative increase in the disease among older women. Using linear regression techniques, Phillips has calculated that 61.3% of all patients with MG in the United States are now older than 50 years.

The age structure of the population of western societies is changing. Thirty-two percent of the population in the countries of the European Union are older than 50 years. A similar aging process is taking place in the population of the United States. An expansion of the age group from which late-onset MG is recruited will in itself lead to an increase in the number of patients. Neurologists will therefore see more patients with late-onset MG than ever before.

When does late-onset MG start? Compston et al. in their 1980 study, suggested 40 years of age as an arbitrary division. Somnier et al. have pointed out that incidence data are more in favor of separating early- and late-onset MG at the age of 50 years. Late-onset MG is defined herein as the onset of the disease after the age of 50 years. Late-onset MG is more likely to be more severe in patients in whom the disease has developed late in life. He also pointed out, and others have confirmed, that patients with late-onset MG do not improve after thymectomy and that a poorer outcome can be expected if myopathic deterioration already exists. Older patients are especially vulnerable to the many complications of long-term steroid therapy. The choice of immunosuppressive drugs may not be the same as in younger patients. The therapeutic situation in a case of late-onset MG is therefore different from that in a case of early-onset MG, and the treatment offered will have to be tailored both to the age of the patient and to the immunological findings of that particular form of MG.

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REFERENCES


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