



## AN UNUSUAL PRESENTATION OF MYASTHENIA GRAVIS

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### ABSTRACT

Myasthenia gravis (MG) is a rare, autoimmune neuromuscular junction disorder. Prevalence rates approach 1/5,000. MG presents with painless, fluctuating, fatigable weakness involving specific muscle groups. It occurs more common in younger individuals. Late-onset MG is more frequent in elderly men and is often misdiagnosed. We present a case of an 80 year old male presented with ocular weakness and breathlessness who was apparently normal earlier & started on betablockers as adjuvant anti hypertensive a month back. Acetylcholine receptor antibody were high indicating MG.

**KEY WORDS:** Myasthenia gravis, Acetyl choline receptor antibodies, plasmapheresis



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## CASE HISTORY

An 80 year old male came with complaints of inability to open eyes for 10 days which was insidious onset, progressive, worsening during course of day and better in the mornings. Breathlessness for 3 days sudden onset Progressive Worsening during course of day. No complaints of chest pain, fever, orthopnea, PND. known case of systemic hypertension for 3yrs on regular medication .He was a Vegetarian with normal bowel and bladder habits ,a Non smoker and non alcoholic his father had diabetes mellitus and hypertension. On examination Patient conscious oriented afebrile. Nopallor, icterus, clubbing, cyanosis, Lymphadenopathy, Pedal edema. PulseRate of 78/min regular, Blood pressure of 130/80 mm Hg. CNS: - B/L ptosis

.Cranial nerves normal. Myasthenia gravis was suspected. Routine investigation, acetylcholine receptor antibody, anti-musk antibody, CPK were sent. Acetyl choline receptor antibodies levels were very high – 26.95 NMOL/L Cpk was 244. All the other blood investigations were within normal limits. CT chest was normal Patient shifted to ICU for observation & further management. Patient was started on Pyridostigmine. Patient's respiratory distress worsened. He was intubated. Plasmapheresis was planned .Patient received 5 cycles of plasmapheresis. Patient was subsequently intubated and discharged with Pyridostigmine and mycophenolate mofetil.



**PRE PLASMAPHERESIS    POST PLASMAPHERESIS**

## DISCUSSION

Myasthenia gravis is a potentially serious but treatable muscle disease caused by autoantibodies directed at the acetylcholine receptor. On the postsynaptic membrane of the neuromuscular junction. It affects individuals in all age groups, but peaks of incidence occur in women in their twenties and thirties and in men in their fifties and sixties. <sup>1</sup> There is anecdotal evidence that the diagnosis is sometimes missed in older patients. <sup>2</sup> A 10-year (1991 to 2000) prospective study of MG in the county of Osona (Barcelona, Spain) reveals an annual incidence rate of 21.27 cases per million inhabitants which states Myasthenia's incidence is higher than expected in the elderly<sup>3</sup>. This is true for many reasons, such as the difficulty of recognizing the typical

symptoms and signs owing to the physiologic changes that occur with aging, and the tendency to attribute the symptoms to more common diagnoses such as cerebrovascular disease.<sup>4</sup> ageing causes a decrease in the total eyelid area with sagging of the lower eyelids, a ptosis may be more difficult to diagnose in the elderly. Almost one third of all cases present in the elderly population. In men, the highest incidence is among those older than 50 years, with a peak age around 70. Women have 2 incidence peaks: 1 between the ages 20 and 40 years and 1 at approximately 70 years of age.<sup>5</sup> Similar to other forms of myasthenia, the symptoms are less obvious upon awakening or after rest and become progressively worse later in the day, as demonstrated by this case. Acetylcholine

receptor antibody test serum levels are the most specific diagnostic test for MG; however, the titers can be lower or undetectable in older patients.<sup>5</sup> Myasthenia gravis is a masquerade in elderly persons, as the fluctuating weakness, a cardinal feature for diagnosis, can be mistaken for age-associated changes or another acute comorbid condition. While MG most commonly occurs in younger people, almost a third of all patients are seen in the seventh decade of life or beyond, with a preponderance of men.<sup>6, 7.</sup> Onset of MG in the older population is increasingly being recognized and is designated as "late-onset MG"<sup>5, 8</sup>. Limb weakness is a rare initial complaint, occurring in 14% to 27% of cases, and should be differentiated from nonspecific generalized fatigue.<sup>9</sup> Late onset myasthenia gravis differs from the typical form in several ways. The HLA DR3 haplotype is uncommon in late onset disease, while thymoma are more common. It is said that late onset disease is more severe and less likely to remit, and that bulbar involvement is more common.<sup>10</sup> Because of confusion with signs of the aging process or from age-related comorbidities, it has been suggested that MG might be underdiagnosed or misdiagnosed in older people. The problem of underdiagnoses is believed to be of greater importance in patients older than 80 year<sup>11</sup>. A recent cohort study suggested that myasthenia gravis might be underdiagnosed in older people. In that study, 2000 asymptomatic individuals aged 60 and over, who participated in the Oxford healthy aging project, were screened for AchR-abs. surprisingly, 0.71% were seropositive. Four of eight seropositive subjects had a diagnosis of "stroke" or "transient ischaemic attacks", and the authors concluded that these patients might have been misdiagnosed<sup>11</sup>. For most neurologic diseases, PP presumably removes pathogenic antibodies from the immunoglobulin fraction of serum. Only in myasthenia gravis (MG),

however, has this presumption been shown,<sup>3,4</sup> in that patient improvement is associated with a drop in antibody titers as a result of PP.<sup>12,13</sup> The role of thymus in the pathogenesis of myasthenia gravis is well-known. High antibody titers have been reported in patients with thymoma in MG.<sup>14</sup> The mean age at onset of MG for thymoma cases is 50-60 years. Approximately 10-15% of all MG patients have a thymoma, and around 40% of all thymoma cases are associated with MG.<sup>4</sup> during normal aging, the thymus tissue becomes atrophic and replaced with fat. Recent data on MG thymus pathology suggest that lymphocyte accumulation indicating residual thymus may also be found in the elderly, and that there is little qualitative difference between the young and the old thymus from MG patients. The mean concentration of antibodies to acetylcholine receptor (AChR) is lower in MG in the elderly than in early-onset or thymoma-associated MG. Seronegative MG is less common among older patient.

## CONCLUSION

Myasthenia gravis are quite a rare autoimmune disorder which should be considered in elderly people with unexplained bulbar symptoms. Most patients also develop some form of fluctuating weakness. The diagnosis is based on clinical features, antibody tests and further investigations including electromyography. Cholinesterase inhibitors are the basic treatment. Steroids such as prednisolone are usually used for maintenance therapy they can be used alongside cholinesterase inhibitors for rapid control of symptoms. Failure to identify myasthenia gravis early may lead to myasthenia crisis which is treated with Intravenous immunoglobulins and plasmapheresis.

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