Myasthenia Gravis (MG) in the Elderly

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Abstract: Myasthenia Gravis (MG) is an autoimmune disease of the neuromuscular junction. It is often misdiagnosed in elderly due to atypical presentations. Bulbar symptoms are the most common presentation seen in elderly. We report a case of elderly male presenting with acute onset bulbar and ocular symptoms, decremental response to CMAPs and a good response to a therapeutic trial of pyridostigmine.

Keywords: Myasthenia Gravis, elderly, late onset myasthenia gravis, bulbar symptoms.

INTRODUCTION
Myasthenia gravis is a neuromuscular disorder characterised by fluctuating ocular, bulbar, respiratory and limb weakness. It affects the post synaptic membrane at the neuromuscular junction, as the T cell antibodies are directed against the Acetyl choline receptors or receptor binding proteins there. Over the age of fifty, the incidence in males is higher than in females, and it is the reverse in patients below the age of forty (1). Over a third of the cases are found in people over the age of seventy (2). However, in another study only 6% - 20% of individuals with myasthenia gravis are over 60 years of age (3). This may be the result of racial differences in the populations studied (3). Limb weakness as the initial complaint is seen in 14-27% of the elderly (2). Thymomas are more common in the elderly (2). Antibodies against the Acetyl choline receptor (AChR ab) and the muscle specific tyrosine kinase antibody (MuSK ab) are the polyclonal antibodies found in this disease. The AChR abs are present in approximately 80 to 90 percent of patients with generalized disease. 98 to 100 percent of the patients with thymoma are positive for these antibodies. Repetitive nerve stimulation resulting in a decremental response to CMAPs is characteristic of MG.

CASE REPORT
A sixty three year old gentleman presented to the Geriatric Medicine Outpatient Department of our institution with a sudden onset of dysphagia, nasal regurgitation and dysphonia for two weeks, associated with ptoosis of the right eye for one week prior to admission. He did not complain of fatigability or fluctuation of symptoms. There was no diplopia, hemiparesis, giddiness or vomiting. He did not have any diarrhea preceding the event. There was no history of prolonged use or recent change in any of his medications. There was a history of an episode of nasal regurgitation which lasted for one week a year ago. This resolved spontaneously. There was a history of weight loss, which was not quantified. He had impaired glucose tolerance and chronic obstructive airway disease. He was cognitively well and independent for activities of daily living. On general physical examination, there was no pallor, icterus, clubbing or lymphadenopathy. His vitals were stable, with a pulse rate of 72/minute and blood pressure of 110/80mmHg. There was no postural fall in blood pressure. He had a respiratory rate of 22/minute. Clinically, there were no features of SLE or hypothyroidism. On central nervous system examination, he had ptosis of right eye; the pupils were equal and reacting to light. The extraocular movements were normal, with no apparent diplopia. The gag reflex was absent bilaterally, but uvula was central. The rest of the cranial nerve examination was normal. Motor system revealed normal strength in all four limbs (no proximal weakness noted) and there were no fasciculations or wasting noted. The deep tendon and plantar reflexes were bilaterally elicitable. There were no significant sensory deficits or cerebellar signs. There were no signs of Progressive Supranuclear Palsy or Parkinsonism. His gait was normal. Initially, a posterior circulation stroke was considered, in view of the sudden onset bulbar symptoms. His MRI brain did not show any focal lesions to explain above neurological findings. Other differentials that were kept in mind with above neurological features were Motor Neuron disease and Myasthenia Gravis. Rest test showed improvement in ptosis.

The Edrophonium test was not done in view of the potential complications of this test in view of his age. His Nerve Conduction Study showed a decremental response at 3 Hz with repetitive nerve stimulation, which confirmed the diagnosis of Myasthenia Gravis. His Acetylcholine Receptor Antibodies were negative. His Chest X-Ray showed bilateral upper lobe fibrosis with traction bronchiectatic features in favour of post inflammatory changes. A CT scan of the chest showed similar findings and there was no evidence of a thymoma. ESR was 48 in one hour and three sputum AFBs were negative. His haemoglobin and thyroid function tests were normal. In view of the history of weight loss, a paraneoplastic workup (including an ultrasound abdomen, CSF cytospin and onconeural antibodies) was done. This was negative. He was seen by the ENT specialists, and was diagnosed to have left vocal cord palsy on NPL scopy. His palatal movements were normal bilaterally. He was initiated on naso-gastric feeds, in view of the history of nasal...
regurgitation. On initiating oral pyridostigmine, there was improvement noted in his voice, but his gag reflex was weak and he was discharged on nasogastric feeds. On follow up in the outpatient department one month later, his dose of pyridostigmine was increased and he was advised to come for regular follow up. However, he was lost to follow up after this.

DISCUSSION
Clinical manifestations of MG in the elderly are different compared to the younger age group. Ocular disease with or without bulbar symptoms are more common in males in any age group(1). A majority of the elderly patients with MG present with ocular myasthenia, which may be associated with peripheral weakness. Diagnosis of MG is often difficult in the elderly due to subtle symptoms, associated co morbidities and many of the presenting symptoms have a broad differential diagnosis in the elderly(4). LOMG can mimic a posterior circulation stroke because of bulbar onset and associated vascular risk factors in the older age group. Hence, one should have a high degree of suspicion to diagnose MG in older adults(5). In an earlier study(6) , early onset MG was associated with presence of acetylcholine receptor (AChR) and anti SM antibodies. In contrast, LOMG was associated with the presence of anti SM antibodies and low antibody titre to acetylcholine receptor(7). Treatment of Myasthenia Gravis and response to treatment is similar in elderly and younger adults. However, complications of immuno-suppressive therapy are seen more in elderly patients. Acetylcholinesterase inhibitors are the first line of treatment. In general, bulbar symptoms are more amenable to treatment, rather than ocular symptoms. Chronic immunomodulating agents like steroids or steroid sparing agents (azathioprine, mycophenolate or Cyclosporine) are reserved for patients not responding to pyridostigmine or patients with a relapse while on treatment with acetylcholinesterase inhibitors. Rapid immunotherapy in form of plasmaphresis and IV immunoglobulin is reserved for Myesthenic crisis or as a “ bridge therapy” in severe MG. Surgical therapy in the form of thymectomy can be considered in elderly,, however this is associated with high postoperative complications(8). Thus decision for surgical treatment is based on overall assessment of patient’s comorbidities and functional status. Immunizations with pneumococcal vaccine and annual influenza vaccine is recommended for patients with MG, especially in those with ocular myasthenia.(9).

CONCLUSION:
We have presented this case as there were many atypical features in this patient. He had had an episode of nasal regurgitation one year ago, which subsided on its own. He may have had intermittent miniscule problems between this and his acute presentation, which may have been ignored. Historically there were no fluctuations or fatigability in his symptoms. The two major points which clinched the diagnosis were – a decremental response to repetitive stimulation on nerve conduction studies and a positive response to pyridostigmine. We had also ruled out the various differential diagnoses considered either on a complete neurological evaluation or on laboratory testing. One should have a high degree of suspicion of MG in the elderly, even in the event of acute neurological symptoms, in the absence of any vascular or local pathology.

REFERENCES: