



Case Report

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Anti ACh Receptor Ab Positive late-onset Bulbar Myasthenia Gravis – Predominantly presented as Laryngeal Myasthenia

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Abstract

Though the report of late onset myasthenia gravis is seen frequently in literature, isolated bulbar myasthenia in elderly age group is rarely found. Moreover, dysphonia as an initial manifestation of bulbar myasthenia has not been emphasized in late-onset MG. Here we report an elderly man who presented with bulbar symptoms in whom diagnosis was made with clinical clues as well as serological test like anti acetylcholine receptor (AChR) antibody. A high index of suspicion is required to exclude other common causes of acute bulbar weakness like stroke, demyelinating neurological diseases, toxicity, infection and other causes. This case illustrates one of the uncommon presentations of late-onset MG.

Keywords: Myasthenia gravis, Late onset bulbar myasthenia, Anti Ach receptor Ab, Dysphonia.

INTRODUCTION

Myasthenia Gravis (MG) is an autoimmune disorder. This is the commonest neuromuscular junction disorder, results from antibody mediated impairment in neuromuscular transmission. In 70 to 80% of MG patients, anti acetyl choline receptor (AChR) antibodies are found which act against nicotinic acetylcholine receptors present in the post synaptic membrane.^[1] This leads to defective function of neuromuscular junction and results in classic features of myasthenia gravis like fluctuating muscle weakness and fatigability.

Estimated worldwide prevalence of myasthenia gravis is between 15 and 179 per million people. ^[2] MG occurs both in men and women. Onset of the disease in women is bimodal, that is one in younger age group which is between 20-30 years and another after the age of 50 years. But in men, commonly occurs after 50 years with peak incidence around the age of 70 years.^[3] Myasthenia gravis occurring in older age group, that is after the age of 50 years, is referred to as "late onset myasthenia gravis" (LOMG). Recently patients presenting with LOMG is being noticed more.^[4,5] This LOMG comprises about one third of all cases of myasthenia gravis. According to a study, LOMG comprises 24.3% of total MG patients.^[6] In early-onset MG, female to male ratio is 4:1 whereas in late-onset MG it is 1:3.^[8]

Among the late-onset myasthenia gravis patients, isolated bulbar onset myasthenia is rare one. As per a study report, the isolated bulbar presentation in late-onset MG is around 21%.^[6] Even in isolated bulbar myasthenia gravis patients, manifestation as dysphagia, dysarthria and chewing difficulty (indicative of pharyngeal, oesophageal, palatal and lower facial weakness) are more common than dysphonia (laryngeal myasthenia). Here, we report a late-onset myasthenia gravis patient whose main symptom was dysphonia. As LOMG presenting predominantly as laryngeal myasthenia is rare, this case is being presented here.

CASE REPORT

A 74 years old gentleman, admitted with C/O difficulty in speaking of one week duration. Patient was apparently normal one week ago. He first noticed hoarseness and low tone in voice while talking over phone. While starts speaking, voice was good. Within one to two minutes voice became

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low and husky, as well as getting slurred. After that, patient noticed similar speaking difficulty whenever he talked to people. He did not find any diurnal variation in his symptoms. While probing the history, patient told there was no difficulty in swallowing both solid food as well as liquids. But he felt as if food gets stuck in his throat. Also while eating developed cough often. But anyway he managed to drink and eat slowly. No nasal regurgitation. No difficulty or fatigability while chewing hard food. No double vision or ptosis. No difficulty in raising arms or getting up. Onset of symptoms: nearly sudden onset, till previous day he didn't have any symptoms. No fever, injury, headache, recurrent vomiting or altered sensorium. No similar illness in the past. Patient was not a smoker or alcoholic or any substance abuser. No history of diabetes, hypertension or heart disease. Not on any medications. With these complaints he consulted an ENT surgeon who examined him with video laryngoscopy. But as he could not find any abnormality referred the patient to our neurology OPD.

On examination, vital signs: BP:116/80 mmhg, pulse rate: 82/min, temperature and respiration were normal. No neurocutaneous markers. No thyromegaly. Higher mental functions were normal. Cognition normal, no emotional lability. No diplopia or ptosis. Extraocular movements were full. Sensation over the face was normal. No facial weakness. Hearing normal. Speech: Patient had dysphonia and voice fatigability. Starts speaking well with normal volume and tone, but within one to two minutes voice becomes low, hoarse, slurry and becomes aphonic. Bilateral palatal movements normal. Gag reflex was slightly diminished. No neck muscle weakness. No wasting, weakness or fibrillation in tongue. Examination of spinomotor system: power and reflexes were normal in limbs. No sensory or cerebellar abnormalities.

As the symptoms were nearly acute in onset we wanted to rule out stroke first. Basic blood and urine investigations were normal. ECG was within normal limits. MRI scan brain taken which was normal and did not show stroke or demyelination features (Fig.1&2).

Our next differential was myasthenia; hence we did repetitive nerve stimulation (RNS) of orbicularis oculi and nasalis bilaterally. But there was no significant decremental response. As the patient's attenders were not willing for edrophonium (acetyl cholinesterase inhibitor) test we went for antiacetylcholine receptor (AChR) antibody assay which was about 17.4 nmol/litre (positive: > 0.4 nmol/litre). As Ach receptor antibody test was positive, we started the patient on tablet pyridostigmine 60 mg three times a day. Patient showed good response. Dysphonia and voice fatigability disappeared, voice clarity improved and which made us to make a final diagnosis of late onset bulbar myasthenia, predominant presentation as laryngeal myasthenia. CT scan of mediastinum was advised which did not show any thymic abnormality (Fig.3&4). Considering his age, distribution of symptoms and response to medical management, thymectomy was not advised. Patient is under our follow up now with good control of symptoms.

DISCUSSION

There are many causes for acute bulbar palsy, like stroke, myasthenia gravis, central demyelination (eg. multiple sclerosis), peripheral demyelination (eg. Guillain-Barré Syndrome (GBS), toxicity (like botulism, snake bite), infective causes like Lyme disease. Causes of dysphonia are laryngitis (due to allergy, infections, smoking, voice abuse), functional dysphonia, growth (neoplasm) of vocal cord, neurogenic (vocal cord paralysis), myasthenia gravis, multiple sclerosis, hypothyroidism, amyloidosis, acromegaly, psychogenic. One of the important differential diagnosis of husky voice with dysphonia, especially in elderly age group is physiological, that is age related atrophic changes that occur in vocal fold muscles which results in incompetence of glottis. In our patient the video laryngoscopy study was normal and he had typical fatigability of voice. When he starts speaking, able to speak well, subsequently dysphonia occurs.

Moreover, apart from dysphonia, he had given the history of cough while eating with reduced gag reflex suggestive of minimal palatal weakness. As the onset was nearly acute, we initially evaluated with MRI brain to rule out stroke. MRI brain was normal. The voice fatigability gave us clue to diagnosis. Hence we considered the possibility of myasthenia gravis.

Ocular symptoms are the commonest manifestation in myasthenia gravis, including LOMG. About 50 to 75% of myasthenia gravis patients present with ocular muscle weakness in the form of double vision and drooping of eyelids.^[7] However, about 20.73% of MG patients may present with isolated bulbar features. Likewise about 23.17% patients may present with both bulbar and ocular symptoms as their initial manifestation.^[7]

In patients with bulbar myasthenia, palatal weakness causes nasal twang in voice and laryngeal weakness produces dysphonia. Pharyngeal, tongue and lower facial muscle weakness result in dysphagia and dysarthria. Our patient's predominant feature was dysphonia which indicates laryngeal weakness.

Montero-Odasso M, in his report on LOMG, recorded dysphonia as the first and exclusive manifestation of isolated bulbar weakness.^[8] Mao et al. analysed 40 patients with laryngeal myasthenia gravis and noticed dysphonia was the initial and primary complaint of those patients. They also found most of those patients were seronegative and only one was seropositive for anti AChR antibody.^[9]

Diagnosis of myasthenia gravis is made by history, clinical examination, edrophonium test (Tensilon test), repetitive nerve stimulation (RNS) test, single fibre electromyography (SFEMG), serological tests like anti acetyl choline receptor antibody, anti muscle specific kinase antibody. Hence we did repetitive nerve stimulation of orbicularis oculi and nasalis. As RNS was negative, we planned for edrophonium test but as we couldn't get patient's relatives consent, we proceeded with anti AChR Ab test, which was positive, by that diagnosis of late onset MG was arrived. And patient was started on treatment with acetylcholinesterase inhibitor and showed good recovery.

Earlier and prompt recognition of this treatable disorder is essential as bulbar weakness in LOMG may progress to become generalised and worsen with respiratory insufficiency and may need aggressive and immunomodulating treatment along with respiratory care to reduce the morbidity and mortality.^[10]

CONCLUSION

When an elderly man comes with acute bulbar symptoms especially with dysphonia that will lead to diagnostic dilemma as there are good number of causes, apart from laryngeal myasthenia which is an uncommon aetiology. This case report stresses the significance of keeping LOMG in the differential diagnosis of acute onset of dysphonia in elderly people.

Conflict of Interest

Authors declare no conflict of interest.

REFERENCES

1. Aydin Y, Ulas AB, Mutlu V, Colak A, Eroglu A. Thymectomy in Myasthenia Gravis. *The Eurasian Journal of Medicine*. 2017;49(1):48-52.
2. Statland JM, Ciafaloni E. Myasthenia gravis: Five new things. *Neurology: Clinical Practice*. 2013;3(2):126-133.
3. Gattellari M, Goumas C, Worthington JM. A national epidemiological study of Myasthenia Gravis in Australia. *Eur J Neurol*. 2012; 19(11):1413-20.

4. Aragonés JM, Bolibar I, Bonfill X, Bufill E, Mummy A, Alonso F, Illa I. Myasthenia gravis a higher than expected incidence in the elderly. *Neurology*. 2003;60(6):1024-6.
5. Alkhawajah NM, Oger J. Late-onset myasthenia gravis: a review when incidence in older adults keeps increasing. *Muscle Nerve* 2013;48:705-10.
6. Porras-Yaurivilca AL, Bertorini TE, Duenas-Vicuna VJ, Mays WH. Late onset myasthenia gravis: A review and report of 24 cases. *Brain Nerves* 2018;2(1):1-3.
7. Sawal N (2017) The Iced Drink Test - A Bedside Test for Diagnosis of Bulbar Myasthenia. *J Neurol Disord* 5:333.
8. Montero-Odasso M. Dysphonia as first symptom of late-onset myasthenia gravis. *J Gen Intern Med*. 2006;21(6):C4-6.
9. Mao VH, Abaza M, Spiegel JR, Mandel S, Hawkshaw M, Heuer RJ, Sataloff RT. Laryngeal myasthenia gravis: report of 40 cases. *J Voice*. 2001; 15(1):122-30.
10. Sakai W, Matsui N, Ishida M, et al. Late-onset myasthenia gravis is predisposed to become generalized in the elderly. *eNeurological Sci*. 2016;2:17-20.

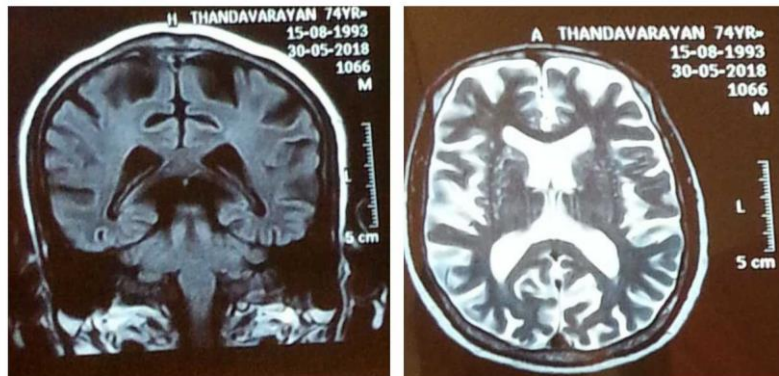


Figure 1: MRI Brain showing normal study

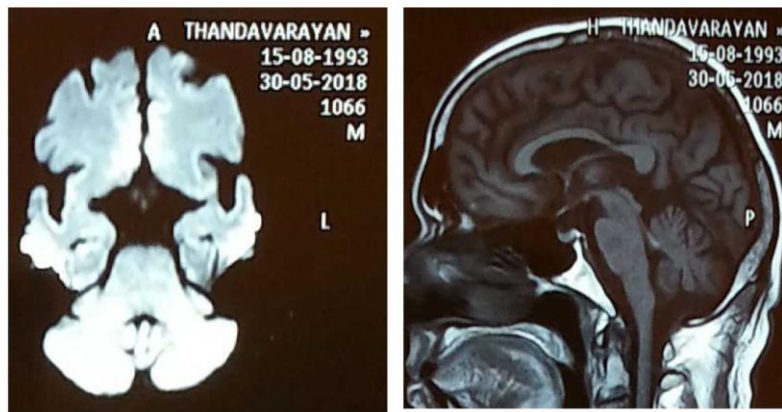


Figure 2: MRI Brain showing normal study

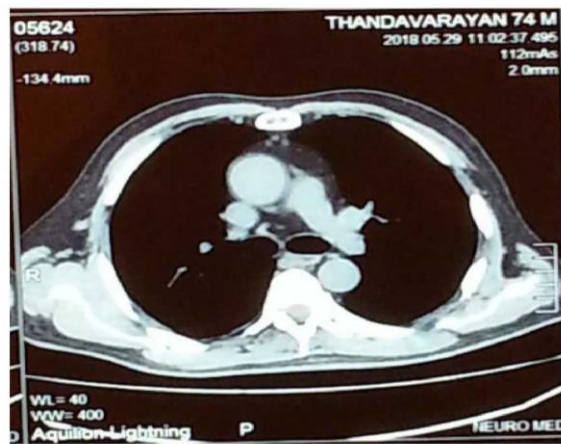


Figure 3: CT scan mediastinum: normal study; no thymus abnormality

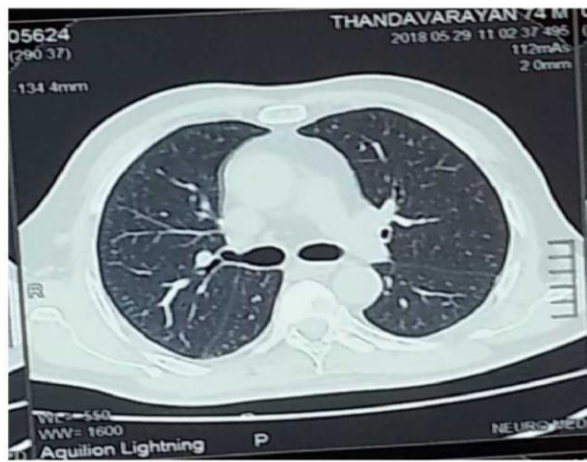


Figure 4: CT scan mediastinum: normal study; no thymus abnormality