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Seronegative Myasthenia Gravis Presenting as Dysphagia in a Young Male- A Diagnostic Dilemma

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ABSTRACT

21year old gentleman presented with insidious onset, gradually progressive dysphagia for 2 weeks. There was no other associated signs or symptoms. Initial blood workup, radiological investigations and diagnostic interventions were non-contributory. In the meantime, dysphagia worsened and was found to be progressive with advancement of the day. Oesophageal manometric studies showed gradual weakening of peristaltic contractions after a string of repetitive swallows. Repetitive nerve stimulation test and edrophonium test were suggestive of Myasthenia Gravis; Anti-acetylcholine receptor antibody and anti-MuSK antibody were negative. He was initiated on Pyridostigmine and had improvement of dysphagia after a few days. He has now achieved remission. Hence, we present a curious case of Seronegative Myasthenia Gravis presented with only dysphagia and had complete recovery after proper diagnosis and treatment.

KEYWORDS: seronegative myasthenia gravis in young, dysphagia

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INTRODUCTION

Myasthenia Gravis is an auto-immune disorder of the neuro-muscular junction in which there is destruction or blocking of the Nicotinic Acetylcholine Receptors by auto-antibodies in the post-synaptic membrane of the myo-neuronal junction. This prevents nerve impulses triggering muscle contractions resulting in weakness [1]. It most commonly starts with ocular weakness manifesting as ptosis and diplopia and can progress to generalised disorder which can manifest as dysarthria, dysphonia, nasal regurgitation, dysphagia, limb weakness, respiratory difficulties and so on and is associated with diurnal variation of symptom severity [2]. Diagnosis is by: Clinically- Ice pack test and Edrophonium test; Serologically- Anti-acetylcholine receptor antibody, anti-MuSK antibody, LRP 4 antibody and so on; Radiologically- CECT or MRI of neck and chest; Electrodiagnostic tests- Repetitive nerve stimulation test and EMG [3]. Treatment is by: Acetylcholinesterase inhibitor like Pyridostigmine for symptomatic treatment, Corticosteroids and other immunomodulatory drugs to target underlying immune dysregulation, IVIg and Plasma exchange in Myasthenic crisis and Thymectomy [4].

CASE REPORT

21year old gentleman reported to the outpatient department with a brief history of dysphagia acute on onset and progressive in nature for last 2 weeks. He did not have any history of known comorbidities or significant past illness. Dysphagia was mild initially, equally intense for both solid and liquid, but more with liquids at the time of presentation. There were no constitutional or any pathognomonic or other systemic symptoms.

On Examination, HR- 82/min; BP- 110/80 mm of Hg. General examination was normal. On Gastrointestinal system examination- no abnormality was found. Other systemic examination was within normal limits.

After admission in ward, routine blood count and other pertinent investigations were sent. They showed Haemoglobin 14, TLC 7000, Platelet 2.2lac, ESR 9mm in 1st hour, Creatinine 1, Sodium 139, Potassium 3.7, Magnesium 2.1. He also underwent radiological investigations to detect the etiology of dysphagia- Barium swallow, Xray of chest and CECT of neck, chest and upper abdomen were normal. Then, he underwent diagnostic interventions: Upper GI Endoscopy was normal, and Fibreoptic Laryngoscopy showed unhindered motion of bilateral vocal cords with no space occupying or pathological lesion. EMG was normal.

Meanwhile, his dysphagia was worsening gradually, and it was noted that dysphagia was least during morning and became worse as the day progressed. Oesophageal manometric studies showed reduced peristaltic contractions which went on decreasing with successive swallows with a gross diminution after a string of repetitive swallows. Neurological consultation was taken. He underwent further tests- Repetitive nerve stimulation test showed significant decrement of muscle contraction in both pre and post exercise stimulation in bilateral facial and spinal accessory muscles; Edrophonium test was positive, both suggestive of Myasthenia Gravis. Blood tests for Anti-acetylcholine receptor antibody and anti-MuSK antibody were negative.

Figure 1

Figure 2

Figure 3

Figure 1, 2 & 3: Cect Neck With Chest

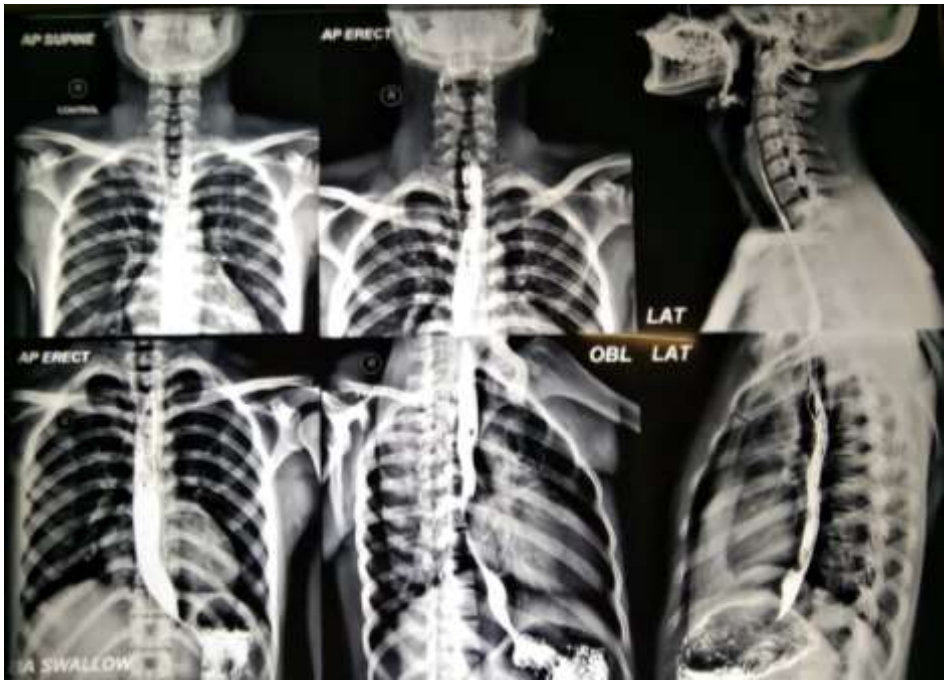

Figure 4

Figure 4: Barium Swallow

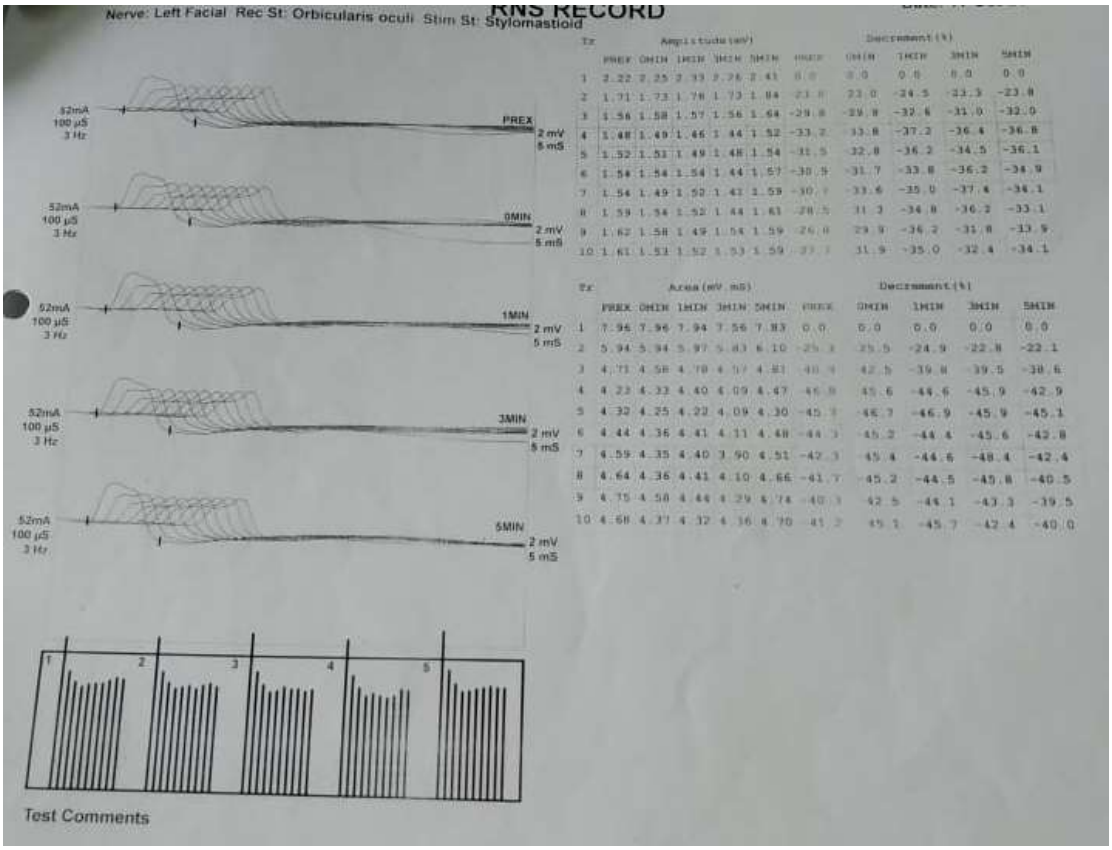

Figure 5

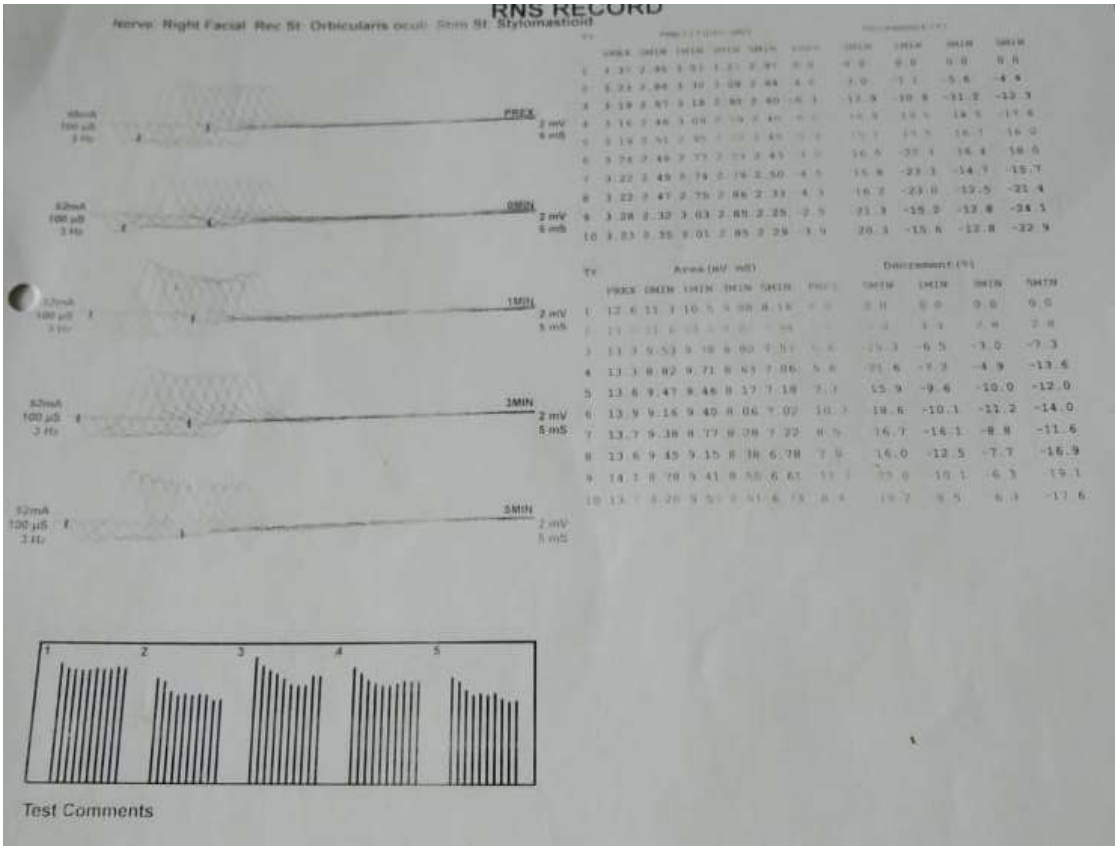

Figure 6

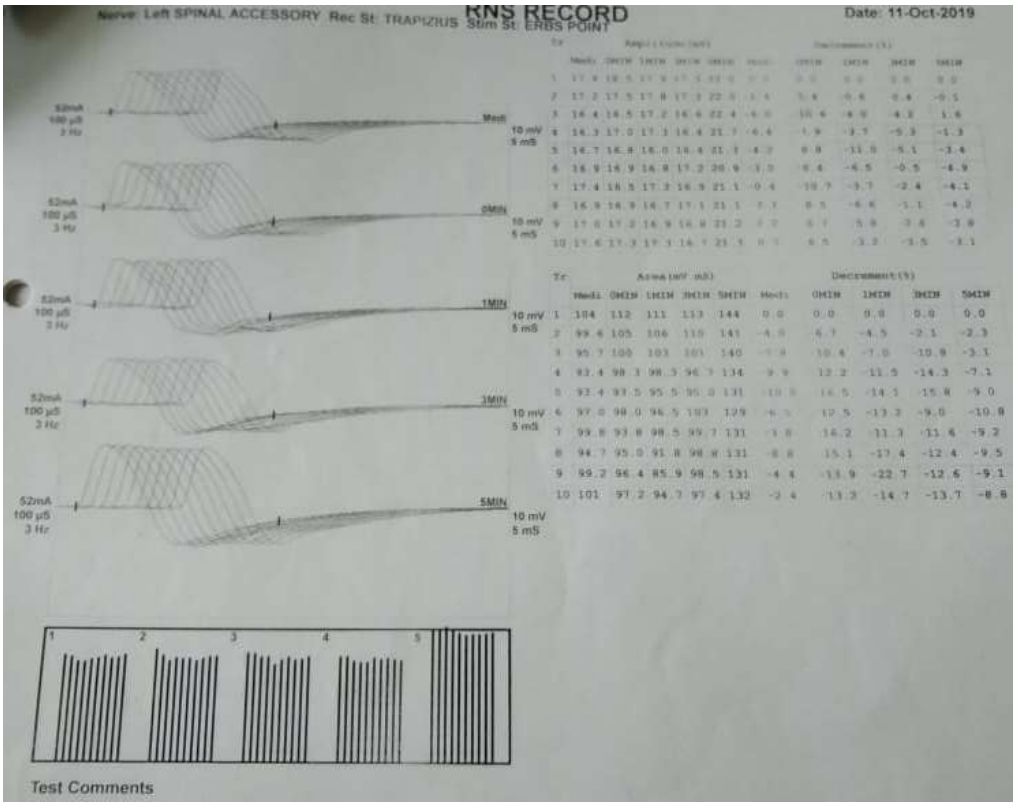

Figure 7

Figure 5, 6 & 7: Repeatative Nerve Stimulation Test Of Bilateral Facial And Spinal Accessory Nerves

He was initiated on Acetylcholine esterase inhibitor, Pyridostigmine. His dysphagia improved after a few days of starting the medication. Pyridostigmine was continued and was aided by swallow therapy and swallow exercises on daily basis and has now achieved remission. He has now returned to his normal life.

DISCUSSION

Dysphagia is defined as difficulty in swallowing- maybe to solids or liquids or both [5]. It is caused mainly by obstructive lesions which maybe present in the lumen, wall or extraluminal of oesophagus and rarely can be caused by neurologic disorders of stroke, dementia or Parkinson's disease [6]. Dysphagia is very rare in young adults and is usually seen at extremes of age [7].

Our patient was admitted for evaluation of dysphagia- which did not detect any biochemical or radiological abnormalities. His dysphagia kept on worsening and was found to have diurnal variation of symptoms. Oesophageal manometry showed gradual decreasing trend of contractions on successive swallows. Repetitive nerve stimulation test showed decremental response on successive contractions and Edrophonium test was positive. Both are suggestive of Myasthenia Gravis [8]. Both Anti-Cholinesterase antibody and Anti-MuSK antibody were negative, hence patient is predicted to have a less severe course of illness [9]. Pyridostigmine is an anticholinesterase agent used for the symptomatic treatment of myasthenia gravis [10].

For seronegative Myasthenia Gravis with only bulbar symptom of dysphagia, Edrophonium test was used successively for diagnosis in a patient by Kim et al [11]. There was another case report by Meena et al, where an Elderly woman presented with only dysphagia which later was diagnosed as Myasthenia Gravis [12].

Dysphagia in Myasthenia Gravis is common in patients with Generalised form with incidence of 15-40% [13]. It is uncommon to be the sole manifestation of the disease and is found in elderly [14]. Interestingly, patients of myasthenia with dysphagia usually have Anti-MuSK antibody positive but was negative in our patient [15].

CONCLUSION

Hence, we report a rare case of Seronegative Myasthenia Gravis in a young gentleman who presented with only Bulbar symptom of dysphagia- was diagnosed through careful history taking aided by clinical examinations and investigations and had complete recovery after administration of Pyridostigmine.

CONSENT OF PATIENT: Taken.

CONFLICT OF INTEREST: None.

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