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A Case Of Myasthenia Gravis

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Abstract

The prevalence of MG in the world is 12.4 people per 100,000 population1, which indicates the urgent need for attention to this disease for prevention and treatment. Myasthenia gravis (MG) is a neuromuscular disease characterized by weakness in the voluntary muscles. This disease has different symptoms that vary in different patients depending on the degree of involvement of the striated muscles2. The most common type of symptom in patients with myasthenia gravis is ocular symptoms. Here we report a young female who presented with diplopia, ptosis, facial muscle weakness which was insidious in onset, asymmetrical, progressive, variable fatiguability with diurnal variation. Ach Receptor antibody was positive. Patient was started on Pyridostigmine after which her symptoms improved. Thymic hyperplasia was present and the patient was planned for thymectomy.

Keywords: Myasthenia gravis

Introduction

Case Report:

A 19 year old female presented with complaints of difficulty in smiling as well as slurring of speech for past 8 months, double vision and drooping of eyelids for past 15 days. Slurring of speech worsened on speaking continuously and improved on taking rest or not speaking for few minutes. Improvement of complaint noticed on waking up from sleep. For the last 2 weeks, patient noticed seeing single objects as two side by side which was insidious in onset and painless, aggravated on looking to right side , while reading books and relived on closing of left eye and taking rest. Patient had history of drooping of eyelids , difficulty in chewing of food, difficulty in holding air in the mouth. Facial weakness produced a snarling expression when patient attempted to smile. There was no history of sensory disturbance over the facial area. On examination, Cogan lid twitch sign[3] was positive, enhancement of ptosis was present and peek sign was positive.

Fig 1(Left) : Left eye Medial rectus Palsy present when patient was asked to look toward right

Fig 2(Right) : Partial Ptosis present in both eyes

The facial weakness was asymmetrical, progressive aggravated on usage and improved with rest . Diurnal Variation was present .There was no involvement of Sensory system, autonomic system or cerebellar system. Bowel and bladder habit was intact. Other Systemic examination was unremarkable. Routine blood investigations – complete blood count, renal function test , Liver function test , electrolytes, thyroid function test , urine routine, diabetic profile , ANA profile were all normal . MRI brain showed normal study. Ice pack challenge was given to the patient and the ptosis improved. Edrophonium stimulation test was also positive. Ach receptor antibody was Positive (>80.0 nmol/L), Anti Musk Antibody was Negative (0.15u/mL). Repetitive nerve stimulation test showed decremental response.

CT Thorax was done for the patient and it revealed thymic hyperplasia[4]. Patient was planned for Thymectomy. Patient was started on Pyridostigmine 60 mg thrice daily and symptoms improved

Fig 3(Left): CT thorax coronal section showing Thymic hyperplasia , with Thymus of size 6.6 x 4.3 x 2.2 cm.

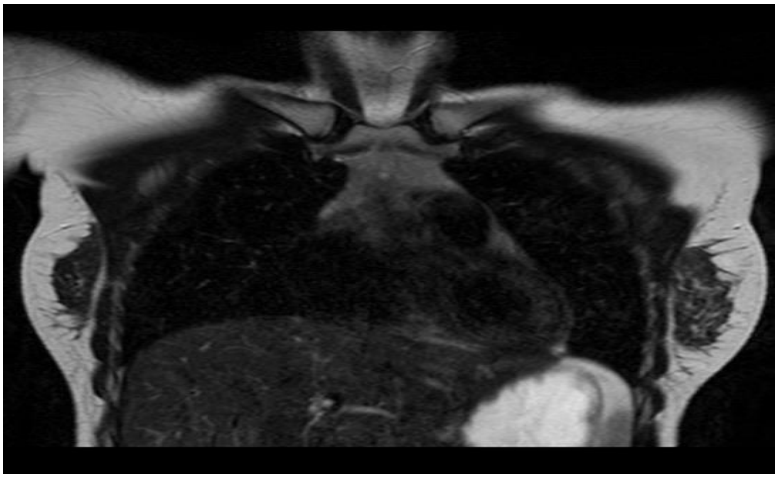
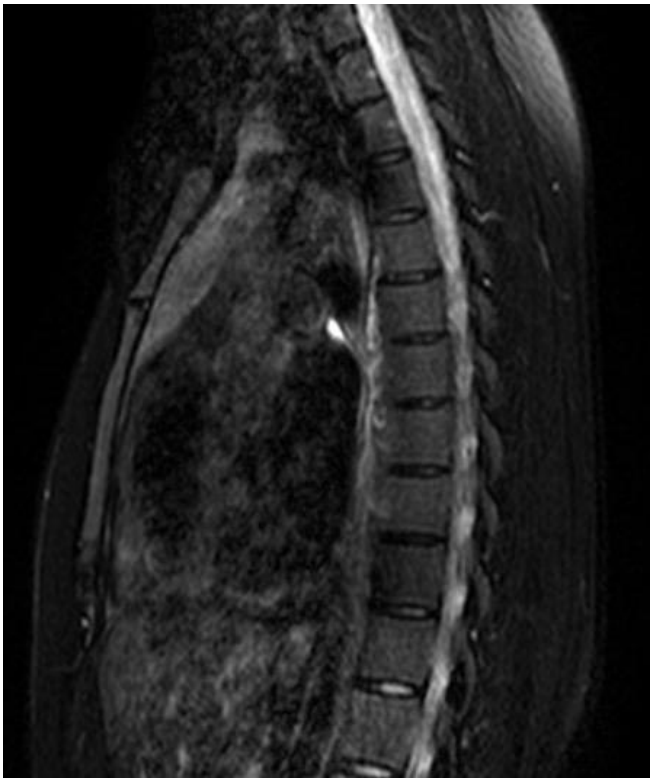


Fig 4(right): CT thorax sagittal section showing thymic hyperplasia



Discussion:

Myasthenia gravis (MG) is a neuromuscular junction (NMJ) disorder characterized by weakness and fatigability of skeletal muscles. The underlying defect is a decrease in the number of available acetylcholine receptors (AChRs) at NMJs due to an antibody-mediated autoimmune attack .More than 80 % receptors are lost before clinical manifestations appear. MG has a prevalence as high as 200 in 100,000.It affects individuals in all age groups, but peak incidences occur in women in their twenties and thirties and men in their fifties and sixties.Overall, women are affected more frequently than men, ratio of ~3:2.

The cardinal features are *weakness* and *fatigability* of muscles. The weakness increases during repeated use (fatigue) or late in the day and may improve following rest or sleep(Diurnal Variation Present).The cranial muscles, particularly the lids and extraocular muscles (EOMs), are typically involved early in the course of MG. Diplopia and Ptosis are common initial complaints. Facial weakness produces a "snarling" expression when the patient attempts to smile. Weakness in chewing, as in chewing meat. Speech may have a nasal timbre caused by weakness of the palate or a dysarthric "mushy" quality due to tongue weakness. Difficulty in swallowing may occur as a result of weakness of the palate, tongue, or pharynx, giving rise to nasal regurgitation. The limb weakness in MG is often proximal and may be asymmetric. Deep tendon reflexes are preserved. Bulbar weakness is especially prominent in MuSK antibody– positive MG. If weakness remains restricted to the EOMs for 3 years, it is likely that it will not become generalized, and these patients are said to have *ocular MG*.

Some specific signs of Myasthenia gravis: (1) Enhanced Ptosis/ Curtain Sign : Partially lifting the ptotic lid may cause the opposite lid to fall. (2) Cogan lid twitch sign: A twitch in the upper eyelid when looking up from a sustained downward gaze. (3) Peek Sign: Sustained tight closure of the eyelids can induce fatigue of the orbicularis oculi muscles resulting in the sclera to be seen.

Antibodies Associated with Myasthenia Gravis: Anti-AChR antibodies are detectable in ~85% of all myasthenic patients but in only about 50% of patients with weakness confined to the ocular muscles. But a negative test does not exclude the disease. The anti-AChR antibodies acts by three distinct mechanisms: (1) accelerated turnover of AChRs (2) damage to the postsynaptic muscle

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membrane (3) blockade of the active site of the AChR. Antibodies to MuSK : They are present in ~40% of AChR antibody–negative patients with generalized MG. MuSK antibodies are rarely present in AChR antibody–positive patients or in patients with ocular MG. Antibodies directed against Netrin-1 receptors and Caspr2 (contactin-associated protein-like 2) often coexist and are associated in patients with thymoma. Anti-striated muscle antibodies directed against titin and other skeletal muscle components seen in ~30% of myasthenic without thymoma ,24% of thymoma patients without myasthenia ,70–80% of patients with both myasthenia and thymoma

Single-fibre electromyography: It is the Most sensitive test (99%).EMG needle is inserted between 2 muscle fibers and action potential is elicitated .Difference between these 2 action potential is called Jitter. Increased Jitter is the diagnostic of MG[5].

Repetitive nerve stimulation Test : Anti Cholinesterase medication should be stopped 12 hours before testing. Electric currents are delivered at a rate of 2-3 Hz to the appropriate nerve and action potential measured.There will be decremental response in the amplitude of evoked potential by more than 10 %.

Ice Pack Test: Cooling facilitates reduced activity of acetylcholinesterase enzyme.

Edrophonium Test (tensilon test): Edrophonium has a rapid onset of 30 seconds and short Duration of 3-5 minutes. Initial Dose of 2mg , if no response addition 8 mg is given .

Treatment:

Anticholinesterase Medication

Pyridostigmine is the most widely used anticholinesterase drug ,initiated at a dosage of 30–60 mg three to four times daily. Action of oral pyridostigmine: begins within 15–30 min and lasts for 3–4 h. The frequency and amount of the dose should be tailored to the patient's individual requirements through- out the day[6] .

Glucocorticoid Therapy

Prednisone should be given single dose rather than in divided doses throughout the day. The initial dose should be relatively low (15–25 mg/d) .Dose is increased stepwise usually by 5 mg/d at 2–3 day intervals. Maintain the dose that control their symptoms for about a month, and then the dosage is slowly tapered to the minimum effective dose [7].

Thymectomy

The thymus is abnormal in ~75% of patients with AChR antibody–positive MG .In ~65% the thymus is hyperplastic .In 10% of patients have thymic tumors (thymomas). Muscle-like cells within the thymus (myoid cells), which express AChRs , may serve as a source of autoantigen and trigger the autoimmune reaction. Available evidence suggest that upto 85 % of the patients experience improvement after thymectomy and of these about 35 % achieve drug free remission. Thymectomy should be carried out in all patients with generalized MG between age of 15 – 55 years. Whether thymectomy should be done in children, age >55 years and in patients with ocular MG is currently resolved [8].

Plasmapheresis and Intravenous immunoglobulin

Plasma, which contains the pathogenic antibodies, is mechanically separated from the blood cells, which are returned to the patient. A course of five exchanges (3–4 L per exchange) is generally administered over a 10- to 14-day period [9].

Intravenous Immunoglobulin(IVIg) : The usual dose is 2 g/kg, which is typically administered >2–5 days. Improvement occurs in ~70% of patients. It is useful as a temporary expedient in seriously affected patients or to improve the patient's condition prior to surgery (e.g., thymectomy).

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