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Rare Case of Myasthenia Gravis with Thymoma in Middle Aged Male

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Abstract: ***Introduction: Myasthenia Gravis is a rare Auto Immune disorder with Auto Antibodies against the Acetylcholine receptors in the Skeletal Neuro - Muscular Junction. Case Details: We report a case of 44 years, Male Presented with Complaints of Diurnal variation of Limb weakness, Neck muscle weakness, Ptosis, Dysphagia with Positive Ice - Pack test. Serum Anti - AchR Antibodies were elevated and Repetitive Nerve Conduction Studies showed >10% Decremental Response and Evaluation by CECT showed presence of Thymoma. The Patient was Started on Pyridostigmine which showed Clinical benefit and the Patient was referred to Dept. of CTVS for Surgical Removal of Thymoma. Discussion: Myasthenia Gravis is a rare Auto Immune disorder characterized by motor Weakness with Diurnal Variation with Preserved Reflexes and absent Sensory symptoms. Diagnosis is done by measuring Sr. Anti AchR antibodies, Anti MuSK receptor antibodies and Repetitive Nerve Conduction Studies. Treatment includes Anti - Cholinesterases, Immuno-suppressive agents, Plasmapheresis. Conclusion: Generalized Myasthenia Gravis is a rare disorder that can be seen rarely in a middle aged male associated with Thymoma and is managed primarily with Anti Cholinesterases and Thymectomy has the significant role in Seropositive cases***

Keywords: Myasthenia Gravis, auto immune disorder, limb weakness, thymoma, treatment

1. Introduction

Myasthenia Gravis belongs to a spectrum of Auto Immune disorders in which Anti - AchR antibodies destroys the Post - Synaptic AchR at Skeletal Neuro - Muscular Junction. [1]It has an annual incidence of 7 - 23 cases per million individuals. [2]It has Bimodal distribution with peak in second to third decades among females and the other peak in 60's and 70's among males. Myasthenia Gravis Presents with Weakness and Fatiguability of Skeletal muscles with Diurnal Variation Predominantly involving the Proximal Muscle groups. It can present in 2 forms:

a) Ocular MG with only Ocular manifestations. [3]
b) Generalized MG with Proximal muscle weakness along with ocular findings.

It can also rarely as Distal limb weakness, Isolated Neck weakness, Bulbar weakness like (Dysphagia, Difficulty in Mastication, Dysphonia) and Isolated Respiratory Paralysis

2. Case Presentation

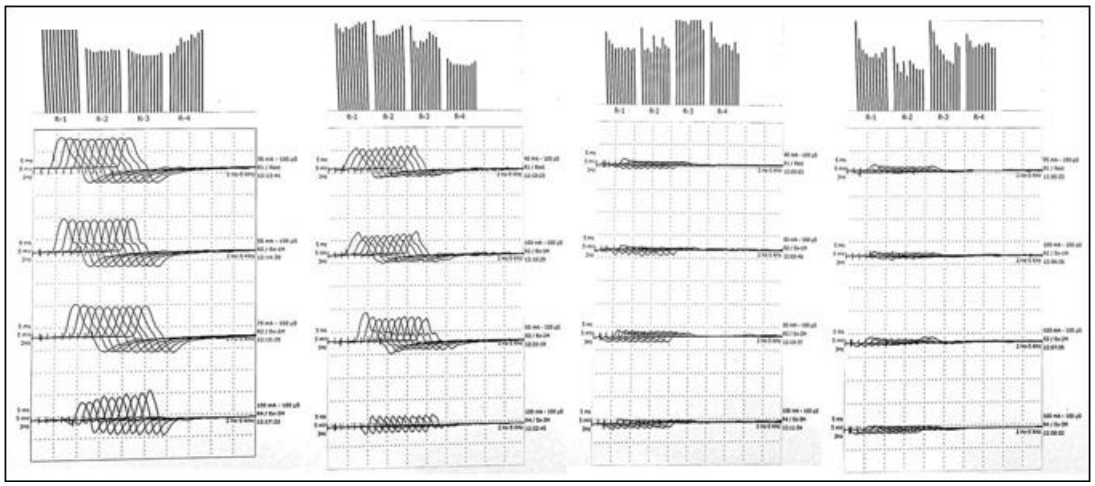
The Clinical Case we Report here is of an 44 year old Male with No Co - morbidities presented with Complaints of Weakness of Bilateral Upper and Lower limbs and unable to hold the neck since one month. Weakness is Sudden in Onset, Patient had difficulty in getting up from Squatting Position, Gripping of Chappals and Climbing Upstairs, Patient also had difficulty in raising the hand above Shoulder, Difficulty in Mixing food, Taking Bath, Buttoning and Unbuttoning the Shirt. Patient also had Difficulty in Opening the eyes, Difficulty in Chewing and Swallowing food

On through History taking, Patient described that the weakness is Progressive and gets worsened as the day Progresses. There is No associated Diplopia, Dysarthria, Dysphonia and Difficulty in Breathing

There is NO previous H/o Trauma, Infection, Consumption of tinned Food. On Examination he had Ptosis of Bilateral eyes 3 mm/3 mm and DTR's were +2 Symmetrical in all joints with Babinski Negative. There is No Sensory Dysfunction. Power in all limbs were examined in morning and evening which showed significant decrease in Power during evening when compared to morning. Later on performing ICE PACK Test, Ptosis got relieved Comparatively

Initial Labs showed No signs of Inflammation and Infection. On CBC - HB: 12.8mg/dl, WBC: 8, 700/mm^3, Platelets: 1.7 lakh/mm^3, Sr. Electrolytes [Na^+, K^+, Cl^- - 140/3.6/101], LFT - Normal, RFT - Normal, HbA1c - 5.6%, CRP - Negative, CPK and CKMB levels - Normal, Sr. Ca^{+2} 9.3 mg/dl, Sr. Mg^{+2} 1.9 mg/dl, TFT - Normal, ANA - IF - Negative with MRI Spine - Normal

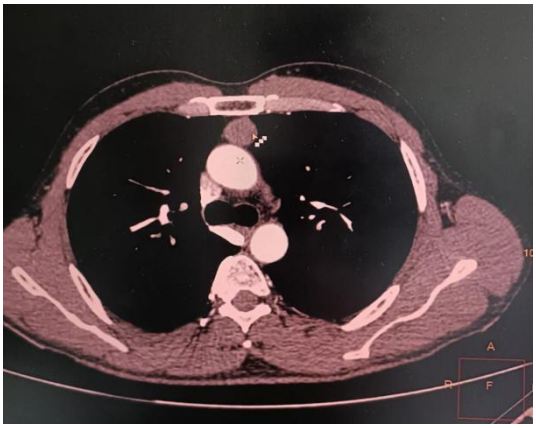
Sr. AchR antibodies was 38.1 nmol/L [Normal 0.4 nmol/L] and Anti MuSK antibodies were Negative. Later the diagnosis of MG is confirmed by Repetitive Nerve Conduction Studies which showed > 10% Decremental response



Repetitive Nerve conduction studies (from left to right)

Left Median nerve, Right Median nerve, Left Orbicularis Oculi and Right Orbicularis Oculi

Then after patient was started on Pyridostigmine 30mg thrice daily His symptoms improved drastically following commencement of therapy and then patient was screened with CECT Chest to look for any Thymus Involvement which showed the presence of Thymoma and then patient was referred to Dept. Of CTVS for further management [Thymectomy]



CECT Chest Image showing Thymoma in the Anterior Mediastinum

3. Discussion

The case described above is the rare presentation of rare disease even though the MG is the commonest NMJ Disorder. Usually in Middle aged population it is commonly seen in Females but where as here It is presented in Male which is unusual

Motor Weakness with Diurnal Variation with Preserved Reflexes and absent Sensory symptoms led to the Suspicion of MG

Mechanism by which it causes Weakness/Fatiguability is by

a) Increases the turnover of Ach Receptor by Receptor Endocytosis
b) Blocking the Active site of Ach receptor
c) Damage to Post Synaptic Muscle Membrane

Diagnosis of MG is done by measuring the levels of Sr. Anti AchR antibodies, Anti MuSK receptor antibodies and Repetitive Nerve Conduction Studies. [4]As the nerve gets Stimulated Continuously the amount of Ach released with each stimulus gets decreased leading to >10% Decremental response in RNS

Differential Diagnosis includes Lambert Eaton Myasthenia Syndrome [LEMS] which shows Incremental response in RNS which is contrary to MG and Botulism which shows Autonomic Involvement with Decreased Reflexes

Whenever MG is confirmed, we should look for other associated disorders like Thyroid Disorders, Autoimmune disorders like [SLE, RA, MYOSITIS] and Thymus involvement so we have done Investigations required and found that the CECT Chest showed the Presence of Thymoma

Thymus Involvement is usually Hyperplastic Thymus but can present rarely as Thymoma as seen in the above case. Other Diagnostic Tests that can be done includes Single Nerve fibre Electromyography which show Increased Jittering. [5]

Treatment for MG includes Anti Cholinesterases like Pyridostigmine started with 30 mg thrice daily and can be increased until maximum dose of 300 mg/day. [6] Other Treatment modalities includes Plasmapheresis and IVIG in case of Severe symptoms with Respiratory Paralysis (Myasthenic Crisis). Immunosuppressive agents like Steroids, Cyclosporine, Tacrolimus, Azathioprine, Mycophenolate mofetil can be used. Rituximab can be used in Refractory MG. [7]Thymectomy has a role in Sero positive cases and decreases the severity of symptoms and Steroid requirement. [8]

4. Conclusion

Generalized Myasthenia Gravis is a rare disorder that can be seen rarely in a middle aged male associated with Thymoma and is managed primarily with Anti Cholinesterases and Thymectomy has the significant role in Seropositive cases

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