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A RARE CASE OF GENERALIZED MYASTHENIA GRAVIS

General Medicine

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ABSTRACT

Generalized myasthenia gravis is a rare case of autoimmune disorder where the antibodies destroy the postsynaptic acetylcholine receptors at skeletal muscle neuromuscular junctions. The clinical presentation is specific distribution of motor deficit without sensory involvement. It diminishes with rest and worsens with excessive use of the muscles. We report a case of a 24 year Female with symptoms of drooping of eyelids and proximal limb weakness for 3 months Repetitive nerve stimulation showed >10% decrement and prostigmin test was positive. The patient was treated and showed clinical improvement.

KEYWORDS

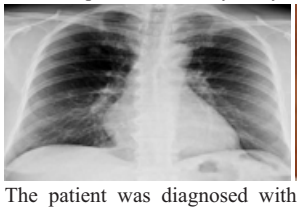
INTRODUCTION

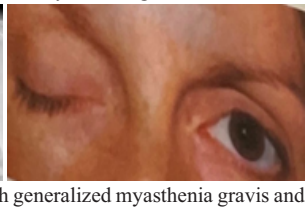
Myasthenia gravis is a rare case of autoimmune in which the antibodies destroy the postsynaptic acetylcholine receptors at skeletal muscle's neuromuscular junctions. It has a prevalence of 1.7-2 per 1 lakh people. The incidence is higher in women than men in ≤ 50 years (7:3), yet at >50 years old, men have more risk (3:2). Clinically, myasthenia gravis is divided into two subgroups : ocular and generalized. Patients with ocular myasthenia gravis only present with diplopia and ptosis. On the other hand, patients with generalized myasthenia gravis manifest as extraocular symptoms such as dysphonia, dysphagia, and even dyspnea.

CASE REPORT

In this case, a 24 year old woman came to Medicine OP with difficulty opening her eyes for 3 months which gradually worsened within 3 days. She also had difficulty gazing right and up, causing her to turn her neck. She could not see clearly due to double vision. She also had slurring of speech, difficulty swallowing liquid and using straws to drink fluids hence often getting choked, difficulty in chewing food and closing her jaws since 2 months ago. Also had proximal limb weakness for 3 months. All of her symptoms were getting worse at midday after activity, yet diminished with rest. She usually felt fine when she woke up in the morning and symptoms are less in the morning. There was no generalized weakness, dysarthria, pain, tingling, nor difficulty of breath. No similar complaints in the family. Vitals are stable. Cranial nerve examination showed she had heavy right eye ptosis 4mm/1mm. The patient had no sensory dysfunction. Motor and deep tendon reflexes were normal. Meningeal signs and pathological reflexes were not found.

Ptosis improved significantly with Ice pack test and edrophonium challenge test and diagnosis of myasthenia gravis was confirmed by Ach Receptor and antibody analysis to myasthenia gravis.





The patient was diagnosed with generalized myasthenia gravis and further diagnostic tests were done. Chest x ray was done - suggestive of thymus mass and it was confirmed by Computed Tomography thorax. Blood tests were done and showed a normal result. Patient was treated with Steroids, Azathioprine and was operated for Thymoma.

Patient gradually improved day by day, she could see without double vision and speak normally. She could also drink from a water bottle without choking. However, she still had difficulty opening her right eye. Physical exam showed improvement of her ptosis 3mm/1mm. Repetitive nerve stimulation test result showed decrement >10% and prostigmin test was positive.

DISCUSSION

Motor weakness without sensory involvement which worsens with repetitive use and diminishes with rest is a specific symptom of myasthenia gravis. In this patient, the acetylcholine receptors (AChR) in the postsynaptic membrane at neuromuscular junction were decreased. This phenomenon was due to the autoimmune response mediated by the anti- AChR by 3 mechanisms : AChR turnover acceleration, AChR active location blockade and damage at postsynaptic muscle membrane. The neuromuscular junction of myasthenic patients had swallow postsynaptic folds and wide synaptic cleft. These mechanisms reduced the efficiency of her neuromuscular transmission, hence causing muscular weakness. The number of released acetylcholine (ACh) will be reduced during repetitive activity (presynaptic rundown), thus causing the patient felt healthy in the morning and making weak after repetitive activity (myasthenic fatigue).

The patient was diagnosed as generalized myasthenic gravis because aside from ocular symptoms, she also had bulbar symptoms. According to her story, she had her first symptoms 3 months ago. Her disease exacerbated 3 days ago due to stress and excessive physical activity. Ice pack eye test is a useful bedside test that can aid in the differentiation of Myasthenia gravis, It is a cheap, safe, and quick test. The test consists of the application of covered ice to the eyes for 2-5 minutes. If positive, the patient no longer has diplopia or a raise of 2 mm of the palpebral fissure. The mechanism behind this test is that by cooling the tissues, more specifically the skeletal muscle fibers , the activity of the acetylcholinesterases are inhibited. Myasthenic Gravis Composite (MGC) scale should have also been done in order to assess the clinical severity of this patient. Increase of ≥ 3 points of MGC scale showed significant clinical improvement. Laboratory examination played an important role in diagnosing myasthenia gravis. The current gold standard are anti-AChR and anti-MuSK antibody tests. Decremental > 10-15% in repetitive nerve stimulation test can be found in myasthenia gravis patients. Prostigmin test was done by intramuscular injection of 3ml prostigmin methyl sulfate and diminished clinical symptoms showed positive result.

The differential diagnoses of this patient were Lambert-Eaton Myasthenic Syndrome (LEMS), botulism and intracranial mass lesion. However, repetitive nerve stimulation test showed incremental results in LEMS, botulism patients present with autonomic symptoms, and intracranial nerve lesions might present with vomiting and other neurological deficits. According to myasthenia gravis algorithm, patients should receive pyridostigmine as a first line drug. Pyridostigmine inhibits acetylcholinesterase in the synaptic cleft thus slowing down the hydrolysis of acetylcholine. If symptoms persisted, clinicians should evaluate

thymectomy. Neck CT scan should be done to detect thymoma. If the patient refused or symptoms still persisted, immunosuppressive drugs should have been given. The patient's prognosis was good. However, most myasthenia gravis patients will not have full remission. Patients should be educated to avoid exacerbating factors. Patients should also be told regarding myasthenic crisis symptoms and management.

CONCLUSION

Generalized myasthenia gravis is a rare clinical entity diagnosed on the basis of clinical and serological findings. Cholinesterase inhibitors are used for symptomatic relief, among which pyridostigmine is most effective. Immunosuppression by thymectomy is effective even in patients with absence of thymoma and shows remission. Immunosuppression can be achieved by corticosteroids or steroid sparing drugs like Azathioprine, Mycophenolate Mofetil, Cyclosporine etc.

Myasthenic crisis is managed by short term treatment with IVIG or plasmapheresis.

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