Review Article

MYASTHENIA GRAVIS

Naila Hamid

ABSTRACT

Myasthenia gravis is a neuromuscular junction disease characterized by severe muscle weakness. It commonly involves muscles of the eye, face and swallowing. In severe cases, respiratory muscles may be affected. It is an autoimmune disease in which antibodies against acetylcholine nicotinic receptors are formed, which destroy these receptors. Thymus is enlarged in many of these patients. These patients are managed by acetylcholine esterase inhibitors (neostigmine), immunosuppression by cortisol and plasmapheresis. Thymectomy is effective in some cases.

Key Words: Neuromuscular junction, Autoimmune disease, Thymus

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INTRODUCTION

Myasthenia gravis is a neuromuscular junction disease manifested by severe weakness of skeletal muscles. Muscles of the eyes, face and swallowing are commonly involved.[1] The name is derived from the Greek term mys meaning muscle, asthenia meaning weakness and the Latin term gravis meaning serious.[2] This disease affects 50-200 per million people.[3,4] Myasthenia gravis is more common in females upto the age of 40 and in males above the age of 60 years.[1] It is an autoimmune disease in which antibodies are formed against acetylcholine receptors at the neuromuscular junction. Decreased number of these receptors results into low voltage of end plate potential (EPP) leading to failure of neuromuscular transmission and muscle contraction.[5,6] Most of the patients have IgG1 and IgG3 antibodies in their plasma.[7]

Babies of myasthenic mothers manifest symptoms of the disease for a few months after birth. The disease has sudden onset. Many patients have an enlarged thymus.[1]

Professor of Physiology, Al-Aleem Medical College, Lahore.

Incidence of other autoimmune diseases is higher in near relatives of myasthenic patients.[8] Due to weakness of extraocular muscles, patients present with double vision (diplopia).[9] Drooping of eye lids (ptosis)occurs due to weakness of levator palpebrae superioris.[10] Patients have difficulty in swallowing, talking and walking.[1] Ocular symptoms get aggravated when the patient watches television, reads a book or drives car, especially in bright light.[11]

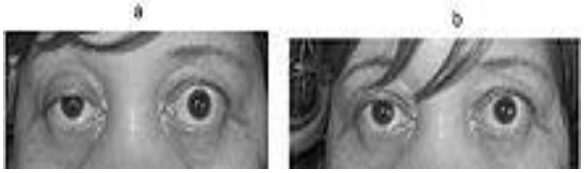

Figure-1. Left photograph shows right partial ptosis in a patient and his left eye compensatory lid retraction. Right photograph is showing improvement in ptosis after edrophonium injection (Figure-1)[12]

Due to dysphagia, food is not completely swallowed; some of it may remain in the mouth after swallowing or regurgitate into the nose. Due to weakness of facial muscles, patients cannot hold the mouth closed. Because of drooping eyelids and facial weakness, the patient may appear sleepy or

sad.[12] In severe cases, respiratory muscles are paralyzed and assisted ventilation is required to sustain life.[13]

Diagnosis of myasthenia gravis is based upon:

Detection of antibodies against the acetylcholine receptors.[12]

A chest CT-scan showing a thymoma.[14] (Figure-2)

Edrophonium test: Intravenous injection of edrophonium or neostigmine (acetylcholine esterase inhibitors) results in improvement.[15]

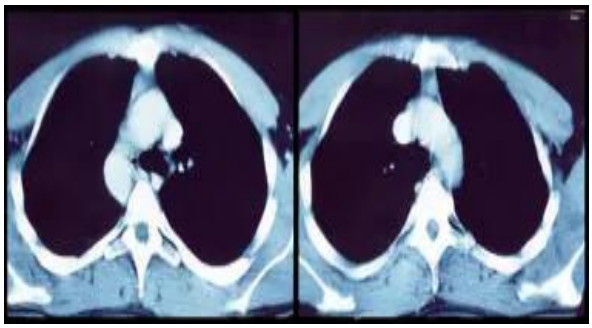

Figure-2: Chest CT scan showing a thymoma

Management

Treatment of myasthenia gravis patients includes medication and surgery.[16] Acetylcholine estrase inhibitors (neostigmine) cause improvement.[17]

Immunosuppressant drugs like prednisolone may be given to obtain a better result.[18] A new Afgartigimod has been approved in Dec 2021, that does not damage the immune system unlike previously used immunosuppressive drugs (Rituxan), hence proved safe when co-administered with COVID-19 vaccine.[19]

Surgical removal of the thymus gland may be beneficial in some patients.[20,21]

To remove antibodies from circulation, plasmapheresis can be used.[22] Patients with myasthenia gravis should be educated about the muscular weakness and exercise. They should be encouraged to perform exercise.[23] With good management patients of myasthenia gravis can live a normal life.

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AUTHOR'S CONTRIBUTION

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