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Neurology

Clinical profile of patients with Myasthenia Gravis followed up in Madurai Medical College in 2015 to 2016

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ABSTRACT Twenty five patients were diagnosed as myasthenia in our institution over the last 1 year. There were eleven males and fourteen females. Five were in pediatric age group. Overall there were 6 patients with ocular and 19 with generalized myasthenia. In the pediatric age group 2 patients had ocular and 3 had generalized myasthenia. All patients were evaluated with CT chest and 2 adult patients were found to harbor a thymoma. Both successfully underwent thymectomy and had decreased need for cholinesterase inhibitors for symptom control after surgery. Three patients suffered from acute myasthenic crisis and required mechanical ventilation. One patient with myasthenic crisis along with comorbid uncontrolled diabetes and ketoacidosis, succumbed despite intensive care. Our series highlights the importance of screening for thymoma in all patients with myasthenia. Management of comorbid diabetes can be challenging in the setting of steroid therapy and needs to be focused on.

KEYWORDS : Myaesthenia Gravis, Repetitive Nerve Stimulation (RNS), Neostigmine challenge test (NCT).

Introduction:

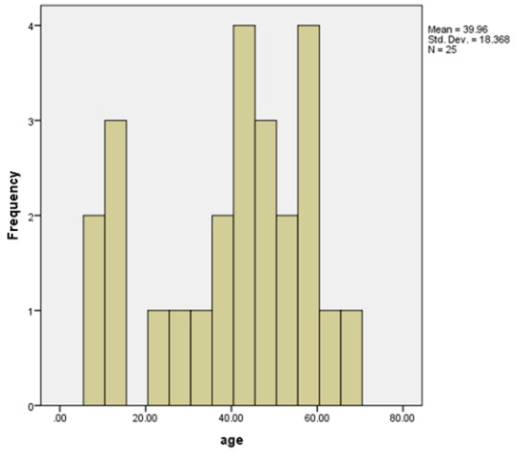
Myaesthenia Gravis is a disorder of neuromuscular transmission. In normal neuromuscular transmission, the motor nerve terminal releases acetyl choline in discrete packages (quanta). The acetyl choline binds to acetyl choline receptors in the muscle membrane (AChR). As a result, the muscle depolarizes and muscle contraction occurs. The acetyl choline is rapidly metabilised by cholinesterase, which terminates the action of acetyl choline. In Myaesthenia Gravis, there are antibodies to acetyl choline receptor which bind to the receptors and degrades them. This leads to neuromuscular transmission failure and muscle weakness. Myaesthenia Gravis can be clinically diagnosed based on the typical distribution of weakness and the characteristic diurnal variation and fatiguability of weakness. The diagnosis can be confirmed by simple electrophysiological studies which include repetitive nerve stimulation to demonstrate a significant decremental response. A demonstrable clinical response to an Intramuscular injection of cholinesterase inhibitor, Neostigmine (Neostigmine Challenge Test – NCT) can also help to confirm the diagnosis. AChR antibody assay can be confirmatory if available.

The thymus gland is abnormal in three fourths of patients with myasthenia. According to existing literature, ten percent of patients harbor a thymoma.

Patients diagnosed with Myaesthenia Gravis from 2015-2016:

Twenty five patients were diagnosed as myasthenia in our institution over the last 1 year. There were eleven males and fourteen females. Five were in pediatric age group. Overall there were 6 patients with ocular and 19 with generalized myasthenia. In the pediatric age group 2 patients had ocular and 3 had generalized myasthenia.

Figure 1: Age distribution of patients with Myaesthenia Gravis



Diagnosis:

Patients were clinically diagnosed based on typical pattern of weakness with fatiguabilty and positive ice pack test. Repetitive nerve stimulation (RNS) used to confirm the diagnosis. A decrement of greater than 10% was taken as significant. Intramuscular Neostigmine challenge test was used to comfirm the diagnosis and look for therapeutic response.

RNS protocol:

3 Hz RNS was done in the involved muscles. RNS was done at rest and 1 and 3 minutes after exercise. Decremental response of greater than 10% was taken as significant. Reproducibility was ensured by repeating the test when necessary

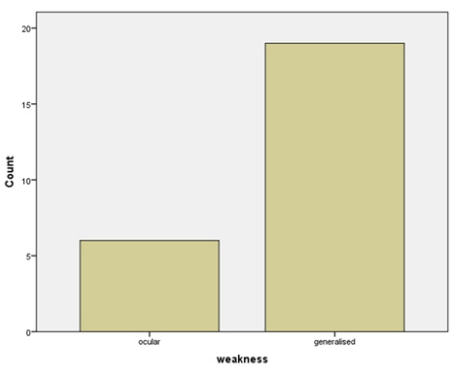
Neostigmine challenge test (NCT) protocol:

At first the baseline ptosis, eye movements and limb weakness were documented prior to the test. Intramuscular Neostigmine was given at a dose of 0.05mg/kg along with atropine. The patient was monitored for 30 minutes and any improvement in ptosis, eye movements and muscle power was documented.

Distribution of weakness:

Overall there were 6 patients with ocular and 19 with generalized myasthenia. In the pediatric age group 2 patients had ocular and 3 had generalized myasthenia.

Figure 2: Distribution of weakness



Screening for Thymoma:

All patients were evaluated with CT chest and 2 adult patients were found to harbor a thymoma. Both successfully underwent thymectomy and had decreased need for cholinesterase inhibitors for symptom control after surgery. Thymoma has been reported in 10% of patients with myaesthenia Gravis in previous literature and it appears to be more common in males and patients older than 40 years. In our series, thymoma was found in 16% (4 patients). In our series, npone of

the pediatric patients had thymoma.

Acute crises:

Three patients suffered from acute myasthenic crisis and required mechanical ventilation. They were treated with IVIG and supportive care. Two of them improved and were weaned form ventilation within 1 week. Patients who improved were switched back to oral medication. One patient with myasthenic crisis along with comorbid uncontrolled diabetes and ketoacidosis, succumbed despite intensive care.

Conclusion:

Our series highlights the importance of screening for thymoma in all patients with myasthenia. Our data show that Myaesthenia Gravis can be effectively managed with clinical diagnosis and electrophysiology. This is important in resource poor settings where antibody testing and genetic testing may not be possible. Management of comorbid diabetes can be challenging in the setting of steroid therapy and needs to be focused on.

References:

1. Bradley's Neurology in Clinical Practice. 7th edition
2. Thymoma in Myaesthenia Gravis; from diagnosis to treatment. Fredrick Romi. Autoimmune Dis. 2011; 2011: 474512