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 metabolised 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.

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 from 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 Myasthenia 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:
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