ISSN: 2754-4737

Journal of Neurology Research Reviews & Reports

SCIENTIFIC Research and Community

Case Reports Open Access

Reporting an Unusual Case of Childhood Myasthenia Gravis

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Received: January 09, 2020, **Accepted**: February 07,2020, **Published:** February 28, 2020

Keywords: Myasthenia Gravis, Neostigmine, Mitochondrial Cytopathy

Introduction

A Three year old boy, Master R, presented at Apollo Children's hospital, Chennai, in June 2019 with ptosis, ophthalmoplegia, diplopia, difficulty in walking, dysphagia and nasal quality in his voice. He is a known case of Myasthenia Gravis, diagnosed in May 2019, already on Neostigmine.

In May 2019, his parents noticed his voice becoming hoarse and he was choking on food. He had asymmetrical ptosis. He was initially diagnosed elsewhere as Mitochondrial Cytopathy as his serum lactate levels were high. A thorough work up was done. His CT Brain and blood metabolic parameters were normal. Thymoma was ruled out with CT chest. Clinical Exome done shows no pathogenic or likely variants causative of the reported phenotype. He was seropositive for anti-ACH receptor antibody. Based on this, the diagnosis was revised to Juvenile Autoimmune Myasthenia Gravis at their native place. With therapeutic doses of Neostigmine the child went into Cholinergic crisis. The same was managed, dose of Neostigmine reduced. The child developed all symptoms again; hence came to Apollo children's hospital for further treatment.

The child was given Neostigmine at dose of 0.4 mg / kg 6th hourly, (previously getting 8th hourly dose) and Injection Methyl prednisolone co-prescribed. He improved slightly. On the next day, he developed increased salivary secretions and bronchorrhoea, excessive drooling and poor respiratory effort warranting immediate intubation and mechanical ventilation.

On ventilator, he was noticed to have persistent low heart rate, constricted pupils, and copious endo-tracheal and oral secretions. The bradycardia and miosis could very well be due to brain stem dysfunction. This posed a management dilemma as to whether the recent worsening is due to Myasthenic crisis or cholinergic crisis. Since Neostigmine is not expected to offer any benefit in a ventilated child, it was withheld to avoid confusion with cholinergic crisis. The child

was then started on Atropine to manage cholinergic crisis, IV Immunoglobulin and steroids as per the treatment protocol in view of seropositivity for Anti ACH-R antibodies. After stabilisation, a couple of days later, Neostigmine was added in a sub-therapeutic dose and Prednisolone added. The myasthenic symptoms became better. However there was residual ptosis .In the meantime the Clinical Exome was reported as " no pathological mutation ".As Steroid Sparer, Mycophenolate mofetil was added. The residual symptoms also resolved. The child is being monitored for steroid toxicity and side effects of MMF. Since child is developing a cholinergic crisis every time at therapeutic doses, he is presently on maintenance with sub-optimal doses of Neostigmine. The symptoms abated only with addition of steroids and Mycophenolate Mofetil.

Discussion

Juvenile Myasthenia as such is a rare condition with incidence of 1 in one million per year, rarely diagnosed at the age of three years. In Autoimmune Myasthenia Gravis, patients generally tolerate Pyridostigmine at therapeutic doses; It has to be combined with steroids in Anti ACH-R seropositive subjects. The benefits have to be weighed against the risk of steroid toxicity. In this child there was steroid toxicity hence the prednisolone is maintained at a very low dose. A second drug Mycophenolate Mofetil was added. At present the child is symptom free with Mycophenolate Mofetil and one third maintenance dose of steroids. Dosage of Pyridostigmine is titrated against the chief ocular symptom which is mainly ptosis but the child has cushingoid features with a normal ocular and bone health and normal blood pressure.

The lesson learnt from this child's case is:

1. A very narrow gap between therapeutic dose and toxic dose of Pyridostigmine existed, which is very special in this child .While trying to address the myasthenic symptoms the child gets pushed into to a cholinergic crisis which is a life threatening event .Hence the dosage adjustment had to be very slow asking the parents to accept a mild ptosis.
2. One has to keep in mind the three factors:
3. Myasthenic symptoms, steroid toxicity And Mycophenolate Mofetil toxicity in the long term follow-up of such patients. This child has a low threshold for a cholinergic crisis. Whole Genome Study is done, reports are awaited. This study may throw more light.

Myasthenia Gravis- Cholinergic crisis.

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