

## Juvenile myasthenia gravis

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### I. Introduction:

Juvenile myasthenia gravis is a rare disorder acquired in childhood, representing 10% to 15% of all cases of myasthenia gravis<sup>1</sup>. It is generally characterized by an autoimmune attack on acetylcholine receptors at the neuromuscular junction. Here, we report a patient with juvenile myasthenia gravis, presented to us with ptosis and drooling of saliva.

### II. Case description

A 5 year old female child presented with cough, cold, fast breathing and drooling of saliva. Child was born out of non consanguineous marriage, without any perinatal insult during delivery or developmental delay. No history of sibling death or autoimmune disorders in the family. On examination, there was ptosis (Figure 1) which was more significant during the late in the evening and other neurological and systemic examination was unremarkable. Ophthalmological examination was done revealed ptosis without any gaze evoked nystagmus. Routine blood investigations were within normal limits. With progressive ptosis towards the end of the day, Juvenile Myasthenia gravis was suspected. Acetyl cholinesterase (AChE) test was done with subcutaneous injection of neostigmine which showed a dramatic response (Figure 2). Repetitive nerve stimulation RNS study showed significant decremental response (>10%) in orbicularis oculi, Right and Left trapezius muscle, suggestive of post synaptic neuromuscular junction (NMJ) receptor disorder. Computerized tomography CT scan of thorax was done to rule out thymic hyperplasia. We diagnosed this child as a case of juvenile myasthenia gravis and treated with pyridostigmine. The child improved symptomatically and under the follow up (Figure 3).



Figure 1: A 5 yr old girl with clinically evident ptosis and compensatory head elevation.



Figure 2: Same girl showed dramatic response to neostigmine test



Figure 3: During follow up the child had reduced degree of ptosis.

### III. Discussion

Myasthenia gravis (MG) in childhood and adolescence is rare comprising 10 to 20 % of all myasthenic patients<sup>2,3</sup>. Myasthenia gravis is a chronic disease characterized by rapid fatigability of striated muscle. There are 3 clinical forms of MG in children: juvenile myasthenia gravis, congenital myasthenia gravis, and transient myasthenia gravis. The most common cause is an autoimmune mediated blockade of acetyl choline receptors (AChR) with elevated anti AChR antibodies. Girls are more commonly affected than boys (1.3 -1.8)<sup>4,5</sup>. In Morita et al, Most of the cases presented between 9 to 12 years of age<sup>6</sup>. Our case was also a female child presented in the early age.

In Morita et al study most of the patients 94.4% had ptosis as one of the initial symptoms. Ptosis was bilateral in 88% patients and unilateral in 12%. Other symptoms like diplopia, bulbar signs, bulbar and limb weakness were present in 61%, 50%, 55%, 66.6% cases. Similarly we also had the child with ptosis and drooling of saliva. The diagnosis is based essentially on the patient's history and clinical examination with a positive anticholinesterase (edrophonium or neostigmine) test<sup>7</sup>. EMG with repetitive stimulation and single-fiber EMG are useful for the diagnosis<sup>3</sup>. Serum acetylcholine receptor antibodies (AChR Ab) are important to confirm the diagnosis but are more frequently negative when compared to adults. Antibody-negative myasthenia gravis were not rare in children<sup>3-5</sup>. We diagnosed this child based on clinical and neostigmine test, but we could not do AChR auto antibodies level which was our limitation.

In morita et al study, patients were treated with oral doses of pyridostigmine bromide, 30 to 300 mg per day. 12.5% had a complete remission in 1 to 3 months, 18.7% cases improved, 12.5% cases remained the same, and 56.2% worsened<sup>6</sup>. Other treatment of myasthenic children includes immunosuppressants (mainly prednisone and azathioprine), plasmapheresis, intravenous high-dose immunoglobulins (IVIg), and thymectomy<sup>7</sup>. We treated the child with pyridostigmine, his ptosis and other symptoms resolved. The patient is under the follow up for 6 months without any of the symptoms.

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