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JUVENILE MYASTHENIA GRAVIS: A CASE REPORT

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KEYWORDS :

INTRODUCTION

Myasthenia gravis (MG) is an autoimmune disease in which antibodies are directed against the postsynaptic membrane of the neuromuscular junction that leads to muscle weakness and fatigability [1]. Juvenile myasthenia gravis (JMG) is a condition of childhood and has many clinical features that are distinct from adult MG. Prepubertal children in particular have a higher prevalence of isolated ocular symptoms, lower frequency of acetylcholine receptor antibodies, and a higher probability of achieving remission.

Diagnosis in young children can be complicated by the need to differentiate from congenital myasthenic syndromes, which do not have an autoimmune basis. Treatment commonly includes anticholinesterases, corticosteroids with or without steroid-sparing agents, and newer immune modulating agents. Plasma exchange and intravenous immunoglobulin (IVIG) are effective in preparation for surgery and in treatment of myasthenic crisis. Thymectomy increases remission rates. Diagnosis and management of children with JMG should take account of their developmental needs, natural history of the condition, and side effect profiles of treatment options.

BACKGROUND

This is a case of 2 year old boy who presented with symptoms of lower respiratory tract infections and ptosis of both upper eyelids and subsequently was diagnosed with juvenile myasthenia gravis. This case demonstrates the importance of early diagnosis, and treating such patients promptly and cautiously to prevent impending grievous complications.

Case Presentation

In this article a child, 2 years old, male, presented in pediatrics OPD with complaints of cough and fever for 2 days, history of fast breathing for 1 day and difficulty in swallowing for 1 day.

His medical history was significant for recurrent respiratory tract infections. His physical examination was significant for the presence drooping of both upper eye lids. It started at one and half year of age which was gradual in onset and worsened over time. The patient was tachypneic with respiratory rate of 50/minute and other vital parameters were within normal limits as per age.

The patient had significant family history as his mother also had ocular symptoms of myasthenia gravis.

The patient was hospitalized and transferred to the intensive care unit. The patient was put on non-invasive mechanical ventilation for respiratory muscle weakness. Antibiotics were added. Ophthalmology consultation was taken to confirm ocular manifestations. Routine blood work up and radiological tests were done to rule out infective etiology.

Serological investigations for AChR antibody, Anti-MuSK antibody and ASKA antibody is negative.

Thyroid profile was done to rule out thyroid diseases associated with Myasthenia Gravis.

Neurology consultation was taken and IVIG at a dose of 0.4mg/kg/day was started and given for five days along with oral pyridostigmine at 2mg/kg/day. Patient's condition improved gradually and he was weaned from ventilator support. After five units of IVIG, the patient was discharged home one week later on oral corticosteroids at 1.5mg/kg/day on alternate days and was gradually tapered over a period of 6 months.

HAEMATOLOGY REPORT	
Hb	12.5gm/dl
Total count	11,900/cumm
Differential count	N72, L24, M03, E01, B00
Platelet count	4.8 lakhs
RBC count	5.3 million
PCV	39.0 %
MCV	73.2 fl
MCH	23.5 pg
MCHC	32.1 g/dl

BIOCHEMISTRY REPORT	
RENAL FUNCTION TEST	
Urea	25.07 mg/dl
Creatinine	0.26 mg/dl
SERUM ELECTROLYTES	
Sodium	135.76 mmol/l
Potassium	4.40 mmol/l
Chloride	104.67 mmol/l

ANTIBODY DETECTION	
AChR	Negative
MuSK	Negative
ASKA	Negative

DISCUSSION

Juvenile myasthenia gravis is a clinical diagnosis with symptoms of weakness and fatigability. It is similar to adult autoimmune disorder but symptoms are often more severe in children. JMG has 2 variants Ocular or general. Ocular MG is due to neurologic involvement is limited to ocular muscles. Approximately 80% of ocular MG progress within first 2 years to systemic involvement presenting as limb girdle and distal muscle weakness. In very young children it is very important to distinguish between auto-immune myasthenia and congenital myasthenic syndromes (CMS) as the treatment options, prognosis, and genetic implications are very different [1].

CMS usually present in the first years of childhood with variable disability. There is often a positive family history, and diagnosis is aided primarily by electrophysiology and DNA analysis and occasionally by muscle biopsy. The CMS are inherited by autosomal recessive mutations that results in loss of function at the neuromuscular junction.

Detection of antibodies to the AChR supports the diagnosis of JMG. In young children where AChR antibodies are negative this can lead to difficulty in differentiating from CMS.

Patients without antibodies to AChR or MuSK are described as having sero-negative myasthenia gravis (SNMG). SNMG patients are phenotypically more similar to AChR seropositive patients than MuSK positive patients, both in clinical presentation and in response to treatment[2].

CONCLUSION

In summary, we described a case of a patient presented with lower respiratory tract infection along with dysphagia and ptosis of both upper eye lids and MG was considered as a possible diagnosis.

There are no internationally accepted standards of care for JMG. The mainstay of treatment usually starts with Cholinesterase inhibitors that are used first line as symptomatic treatment in JMG [3]. Pyridostigmine is a non selective ChE-I and act at the neuromuscular junction. Prednisolone is accepted as the first line immunosuppressive therapy in JMG [1].

Immunosuppressive agents and immune modulators have been used in combination to aid in therapy as well as plasmapheresis and short term IVIg [4-6]. IVIg is an effective treatment for many patients with autoimmune myasthenia gravis. It has been shown to inhibit complement binding, neutralize pathogenic cytokines, downregulate antibody production, enhance remyelination and modulate Fc-receptor-mediated phagocytosis, and T cell function. The most common usage is to treat an exacerbation where a patient has worsening symptoms despite other treatments. IVIg is used to treat patients who are in crisis, or showing signs of impending crisis.

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