ORIGINAL RESEARCH PAPER
Volume - 9 | Issue - 6 | June - 2020 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

ACQUIRED GENERALIZED MYASTHENIA GRAVIS IN INFANT: A RARE CASE REPORT.

Neurology

Dr Arvind Vyas Sr Professor and Unit Head. Department of Neurology, SMS Medical College, Jaipur.

Dr Jaypalsing Ghunawat* Senior Resident Department of Neurology, SMS Medical College, Jaipur. *Corresponding Author

Dr Jivan Kinkar Senior Resident Department of Neurology, SMS Medical College, Jaipur.

ABSTRACT

Myasthenia gravis is an acquired autoimmune disorder leading to abnormal fatiguability of muscles due to deficiency of acetylcholine receptors caused by circulating antibodies directed against them.1 Ocular myasthenia is a form of myasthenia gravis clinically involving levatorpalpebraesuperioris, the orbicularis oculi, and the extraocular muscles.Ptosis and ophthalmoplegia, both unilateral and bilateral, constitute the only signs in about 20% cases while in about 70% cases, ocular symptoms mark the onset of generalised myasthenia gravis

KEYWORDS

INTRODUCTION:

Infantile autoimmune myasthenia gravis is rare but case reports are available.[4]Congenital myasthenia is common in infantile age; however, cases of acquired autoimmune myasthenia are reported at this age. Ocular myasthenia is common presentation of infantile myasthenia. Here we report case of generalised autoimmune myasthenia in infant.

Case report

A 14-month child admitted with history of drooping of bilateral eyelids from 8-month age with diurnal variation, being increased during evening hours. Simultaneously he developed difficulty in standing from sitting position, worsening in evening hours. He could stand of his own during morning hours but requires support in evening time. He also had history of poor cry and decreased volume of speech in evening hours as compared to morning time. He never had respiratory difficulty or regurgitation of liquids and did not required admission. His antenatal as well as birth history was uneventful and developmental milestones were normal. There was no similar history of illness in family member.

Examination revealed a child with normal physical appearance and age appropriate motor and language milestones. Neurological assessment suggested bilateral symmetrical ptosis with normal pupillary reflex in both eyes (Figure 1). Tone, cerebellar examination, and deep tendon reflexes were normal. He was examined during morning and evening time which revealed his ability to stand of his own during morning time which he was unable to in evening hours. Other systemic examinations were unremarkable. With a clinical diagnosis of myasthenia in child of a non-myasthenic mother, he was investigated. Repetitive nerve stimulation (RNST) was done in right median nerve showed decremental response. Serum anti- acetylcholine receptor antibody titre was 3.45nmol/L (normal less than 0.25nmol/L). Chest X- ray was normal. MRI mediastinum showed no evidence of thymoma. Genetic panel for congenital myasthenia was negative.

Figure 1.

DISCUSSION:

MG is characterized byfatigability aftersustained muscle activity and subsequent improvement after rest. Typical age of onset is in young adults but can occur in infancy and childhood. Different clinical manifestation in childhood are transient neonatal myasthenia in an infant of myasthenic mother, juvenile myasthenia and congenital myasthenia.[2]Our patient was normal till 8 month of life and no feeding difficulty in early infancy. His weakness was gradually progressive with diurnal variation. Hence the possibilities of acquired versus congenital myasthenia were likely. His genetic panel for congenital myasthenia were negative and anti Ach-R antibody were positive with significant titre of 3.45 nmol/L.

Treatment includes repetitive doses of cholinesterase inhibitor. Steroids, immunosuppressants, plasmapheresis or thymectomy are not helpful. Our case was started on pyridostigmine(7mg/kg/d) and oral prednisolone with significant clinical improvement. Medications like succinylcholine, quinine, neomycine and other neuromuscular blocking agents should be avoided.[2] Juvenile myasthenia gravis comprises approximately 1% of all cases of myasthenia gravis.[3]Infantile autoimmune myasthenia gravis is rare but case reports are available.[4]In our case, high level of anti Ach receptor antibody were detected which are extremely rare.

Long-term steroid therapy is effective due to autoimmune basis of the disease. Temporary remissions can be achieved by plasmapheresis and or intravenous immunoglobulin (IVIG) in patients with high titre of anti Ach receptor antibodies. Resistant cases may require rituximab, a monoclonal antibody to B cell CD antigen.[3]

CONCLUSION:

Infantile autoimmune myasthenia is rare entity. Early recognition and timely intervention result in good prognosis. Anticholinesterase antibody are helpful in differentiating acquired myasthenia from congenital cases.

REFERENCES

1. Weinberg DA, Lesser RL, Vollmer TL. Ocular myasthenia: a protean disorder. SurvOphthalmol 1994; 39:169–209.
2. Dubowitz, V. Myasthesia gravis. In Schaffer AI, Markowitz M eds. "Muscle disorders in childhood,"Major Problems in Clinical Practice," Philadelphia and London. W.B. Saunders Company Ltd., 1978; 191-201
3. Kini PG. Juvenile myasthenia gravis with predominant facial weakness in a 7-year-old boy. Int J PediatrOtorhinolaryngol 1995; 32: 167-169.
4. Thymoma Associated with Myasthenia Gravis in Infancy. Indian J Pediatr 2009; 76 (10) :1057-1058.