

Myasthenia Gravis a Case Report and a Review of Literature.

Maryam Rashid¹, Aysha Babar², Muhammad Saeed Khan³, Shujaat Hussain⁴

ABSTRACT

Juvenile myasthenia gravis is a rare autoimmune disease acquired in childhood that represents about 10% to 15% of all cases of myasthenia gravis. Ocular myasthenia gravis presents as ptosis with extraocular movement restriction and leads to misdiagnosis when bilateral as third nerve palsy or congenital ptosis. Early diagnosis helps to prevent worry of anxious parents and prevent further progression of disease. 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.

KEYWORDS: Juvenile Myasthenia Gravis; Autoimmune Disease; Neuromuscular Junction; Congenital Ptosis

HOW TO CITE THIS:

Rashid M, Babar A, Khan MS, Hussain S. Myasthenia Gravis a Case Report and a Review of Literature. *Isra Med J.* 2018; 10(1): 53-55.

This is an Open Access article distributed under the terms of the Creative Commons Attribution-NonCommercial 4.0 International License (<http://creativecommons.org/licenses/by-nc/4.0/>), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

INTRODUCTION

Myasthenia gravis is the most common disorder of neuromuscular junction with prevalence about 20 cases per 100,000 populations.¹ The disease is caused by antibodies directed to various proteins at the neuromuscular junction, most commonly the acetyl choline receptors. Like the adult form, it is caused by antibody-mediated attack on acetylcholine receptors at the neuromuscular junction. Most patients present with clinical fatigue which means weakness that worsens with muscle use and improves with rest, ptosis and diplopia. Ocular weakness is the most common complaint presenting in over half of the patients.^{1,2} This leads to variable weakness of various extraocular muscles including levator palpebrae superioris and orbicularis oculi.³ Childhood ocular myasthenia is a rare entity which is prone to be misdiagnosed.⁴ Patients can present with ptosis, strabismus,

and diplopia. Early onset childhood ocular myasthenia is even rarer and a diagnostic challenge to the clinician. The advanced cases may also have bulbar and limb weakness. Left untreated, the disease may progress to paralysis of the respiratory muscles. Early recognition of this disease helps to prevent the progression of symptoms and anxiety of parents. We herewith report a rare case of bilateral juvenile myasthenia gravis in which 8 years old girl was misdiagnosed with simple congenital ptosis and was also advised surgery for the same.

CASE REPORT

The child's grand mother told that the patient suddenly developed ptosis of right eye and diplopia without any obvious cause. Two weeks later ptosis on the left side also appeared. Ptosis was negligible in the morning after night's sleep but got aggravated as the day passed. There was no weakness of any other part of the body. There was no history of fever, headache, vomiting or insomnia. There was no history of trauma. There was no history of difficulty in chewing and swallowing. There was no history of such episode in the past. 3 sisters and 2 brothers, she was the eldest, all other members were healthy. She was a student of class III. She was sitting comfortably in the bed with marked bilateral ptosis, her temperature was 98 F, pulse 80/min regular, respiratory rate 16/min, BP 110/70mmHg. She was well oriented in time and place. All the cranial nerves were intact. Fundus oculi were normal and speech was not affected. There was weakness of all the extraocular muscles and there was marked ptosis of both eyes (Figure - 1). There was no weakness of any other muscles and there was no wasting or fasciculation of any muscle. All the reflexes were normally elicitable and plantar flexors in response. The patient was already examined by other hospital where he was diagnosed to have congenital ptosis and was advised to undergo tarsofrontalis sling surgery to clear the visual axis and prevent amblyopia. The patient came to our hospital after 2 months for second opinion.

1. Assistant Professor of Pharmacology
Akhtar Saeed Medical & Dental College, Lahore
2. Assistant Professor of Physiology
Yusra Medical College Rawalpindi
3. Associate Professor Medicine
4. Professor of Medicine

Al-Nafees Medical College & Hospital
Isra University Islamabad Campus Pakistan

Correspondence to:

Maj Gen(R) Prof. Dr. Shujaat Hussain
Professor of Medicine
Al-Nafees Medical College & Hospital
Isra University Islamabad Campus Pakistan
Email: shujaat.hussain@yahoo.com

Received for Publication: 28-05-15

Accepted for Publication: 10-01-18



Figure – 1: Showing bilateral ptosis (Juvenile myasthenia gravis patient)

Since this child has history of bilateral ptosis of sudden onset. Provisional diagnosis of Juvenile Myasthenia gravis was made. Laboratory investigations showed a highly raised serum acetyl choline receptor antibody level. Ice pack test was performed which shows very little improvement (around 1 mm) only. Then we planned to give 0.5 mg neostigmine inj i/m and within 15 minutes the child ptosis disappeared but the effect lasted for 5-6 hours only and then given tab neostigmine 15mg tds and the patient responded well. She was thoroughly investigated and thymus gland localized by CT scan chest. Then subjected to surgical treatment i.e. thymectomy done under general anesthesia utilizing transternal approach, midline incision extending from manubrium to xiphisternum. Biopsy report of thymus gland revealed calcification of the Hassalls corpuscles. Thymus gland. No germinal centres were seen as there is no evidence of thymoma. Tab neostigmine discontinued on the third post operative day but the child again developed ptosis so drug continued upto 3 months. Stitches were removed on 10th postoperative day and the patient discharged on 07-06-2014 with definitive improvement.

DISCUSSION

Myasthenia gravis is known ocular myasthenia gravis when weakness is limited to eyes only especially to extraocular muscles.³ Childhood myasthenia has three types which are Neonatal, juvenile, and congenital myasthenic syndrome.⁴ There is transplacental transmission of autoantibodies in Neonatal myasthenia.⁵ Neonatal myasthenia has good recovery. Congenital myasthenic syndrome (CMS) is an inherited neuromuscular disorder caused by defects of several types at the neuromuscular junction.⁶ Myasthenia gravis if presented before the age of 19 years is called juvenile myasthenia gravis.⁷ The prevalence in childhood of Chinese population is about 50% of all myasthenia cases⁸ and in Caucasians about 10% cases in prepubertal age.⁹ Vander Pluym et al. reported 18 cases in which 8 were below 3 years of age and 10 patients have ocular myasthenia.¹⁰ Gradient

et al. reported three cases of Juvenile myasthenia gravis in which one case was not ocular and less than three years.¹¹ Evoli et al. reported five ocular cases in which age between 1.5 and 9.2 years.¹² There is marked resemblance of myasthenia gravis with other neuromuscular disorders. This is the reason that early age ocular juvenile myasthenia is usually misdiagnosed with congenital ptosis. The chance of misdiagnosis especially very high if patient has bilateral ptosis as presented in the current case report. As a rule in medicine, the proper history and examination help early diagnosis of disease and prevent its complication. The sensitivity of Acetyl choline receptor (AChR) antibodies are almost 55% in ocular myasthenia⁷ and 70% of AChR-negative patients have Anti-MuSK (muscle-specific tyrosine kinase) antibodies.⁴ Electrophysiological testing is not helpful in diagnosing juvenile myasthenia gravis. Treatment of juvenile myasthenia gravis is same like adult myasthenia (anticholine esterase inhibitors, steroids and immunosuppressant).⁷ Since our patient showed dramatic response to neostigmine, addition of steroids was not considered. Prepubertal form of juvenile myasthenia gravis has good prognosis.¹³ Our case was well maintained on neostigmine at 3 months of follow-up without developing any symptoms of generalized myasthenia.

CONCLUSION

The present case report gives the importance of myasthenia gravis to be in the differential diagnosis of any child presenting with bilateral ptosis with or without diplopia. A proper history and examination are the key to the diagnosis and better treatment in all such cases.

CONTRIBUTION OF AUTHORS

Rashid M: Manuscript Writing
Babar A: Manuscript Writing
Khan MS: Literature Search
Hussain S: Literature Review, Manuscript final reading and approval

Disclaimer: None.

Conflict of Interest: None.

Source of Funding: None.

REFERENCES

- 1 Grob D, Brunner N, Namba T, Pagala M. Lifetime course of myasthenia gravis. *Muscle Nerve*. 2008; 37(2):141-49.
- 2 Grob D, Arsura EL, Brunner NG, Namba T. The course of myasthenia gravis and therapies affecting outcome. *Ann N Y Acad Sci*. 1987;505:472-99.
- 3 Juel VC, Massey JM. Myasthenia gravis. *Orphanet J Rare Dis*. 2007; 2:44.
- 4 Parr JR, Jayawant S. Childhood myasthenia: clinical subtypes and practical management. *Dev Med Child Neurol*. 2007;49(8):629-35.
- 5 Téllez-Zenteno JF, Hernández-Ronquillo L, Salinas V, Estanol B, da Silva O. Myasthenia gravis and pregnancy: clinical implications and neonatal outcome. *BMC Musculoskelet Disord*. 2004, 16;5:42.

- 6 Engel AG, Ohno K, Sine SM. Congenital myasthenic syndromes: progress over the past decade. *Muscle Nerve*. 2003;27(1):4-25.
- 7 Finnis MF, Jayawant S. Juvenile myasthenia gravis: A pediatric perspective. *Autoimmune diseases*. 2011;404101.
- 8 Zhang X, Yang M, Xu J. Clinical and serological study of myasthenia gravis in HuBei Province, China. *J Neurol Neurosurg Psychiatr*. 2007;78(4):386-90.
- 9 Kalb B, Matell G, Pirskanen R. Epidemiology of myasthenia gravis: a population-based study in Stockholm, Sweden. *Neuroepidemiology*. 2002;21(5):221-25.
- 10 VanderPluym J, Vajsar J, Jacob FD. Clinical characteristics of pediatric myasthenia: a surveillance study. *Pediatrics*. 2013;132(4):939-44.
- 11 Gadiant P, Bolton J, Puri V. Juvenile myasthenia gravis: three case reports and a literature review. *J Child Neurol*. 2009;24(5):584-90.
- 12 Evoli A, Batocchi AP, Bartoccioni E. Juvenile myasthenia gravis with prepubertal onset. *Neuromuscul Disord*. 1998;8(8):561-67.
- 13 Andrews PI, Massey JM, Howard JF Jr. Race, sex, and puberty influence onset, severity, and outcome in juvenile myasthenia gravis. *Neurology*. 1994;44(7):1208-14.