Journal of Ophthalmology & Visual Sciences

Open Access

Austin Publishing Group

Case Report

Unilateral Isolated Inferior Rectus Palsy: A Rare Presentation of Ocular Myasthenia Gravis

Yadav Y*, Dhar S, Singh P, Mohapatra T, Gupta R and Mishra SK

Department of Ophthalmology, Army Hospital Research & Referral, India

***Corresponding author:** Yadav Y, Department of Ophthalmology, Army Hospital Research & Referral, New Delhi - 110010, India

Received: September 30, 2021; **Accepted:** October 22, 2021; **Published:** October 29, 2021

Abstract

Ocular Myasthenia Gravis with unilateral isolated Inferior Rectus palsy and no ptosis is rarely encountered. We present a case of 25-year-old male with sudden onset binocular diplopia and no features of ptosis. Deviations in primary gaze were 3-8 Prism Dioptre (PD) with left hypertropia. Right and left head tilt deviations were 5PD and 3PD left hypertropia respectively with increase in hypertropia in left gazes. Acetylcholine receptor binding antibodies were found to be raised. Repetitive Nerve Stimulation showed decreased amplitude. Patient was managed with Oral Pyridostigmine with excellent symptomatic relief and drug tolerance. No features of Myasthenia Gravis (Systemic or Ocular) were noted on follow-up.

Keywords: Acetylcholine receptor binding antibody; Diplopia; Inferior Rectus Palsy; Ocular Myasthenia Gravis

Introduction

Ocular Myasthenia Gravis (OMG) presents diagnostic challenge in isolated unilateral Extra Ocular Muscle (EOM) involvement particularly in absence of any features of ptosis or lid involvement.

Materials and Methods

A 25-year-old male, presented with sudden onset, intermittent and non-progressive binocular diplopia of one month duration. Diplopia aggravated in outdoor conditions. Diplopia was relieved on resting and tilting head to left. Respiratory, chest and neurological examination was within normal limits. No weakness of extremities or difficulty in swallowing was noted. Distant visual acuity was 20/20 in both eyes with normal pupillary reactions. Margin to reflex distances 1 and 2 in both eyes were 4cm and 5cm respectively. Anterior and posterior segment examination revealed no abnormality.

On orthoptic workup patient had head tilt towards left. Ocular movements were normal and no limitation of movement was seen in any of the gazes. On Hirschberg test there was approximately 7 degree of left hypertropia. On sensory evaluation, his stereopsis without head posture was 120sec of arc with TNO, there was symmetric crossed response on bagolini striated glass test and diplopia on worth four-dot test. On Prism Bar Cover Test (PBCT) with right eye fixing, he had 3-8 Prism Dioptre (PD) left hypertropia in primary and dextro-depression gazes (Figure 1). Deviation was variable depending on the time of day it was measured. Deviation on right head tilt- 5-6 PD L/R and on left head tilt- 3 PD L/R was noted. Three-step test was performed by tilting the head to 45° to each side and measuring the deviation with prism bar cover test with patient maintaining distance fixation. Increase in hypertropia was seen on left gazes. Therefore left inferior rectus (IR) was found to be the affected muscle. Diplopia charting was as depicted in Figure 2.

Patient was diagnosed as a case of Isolated left IR Palsy.

Patient was evaluated further to find the etiology. Hematological investigations, thyroid profile, carotid doppler and MRI brain came out to be normal. In order to investigate further Myasthenia Gravis (MG) was thought to be ruled out.

Serum level for Acetylcholine Receptor binding Antibodies (AChR-Ab) were found to be raised {(18.22nmol/L) (normal value-<0.25nmol/L)} (Figure 3a). Repetitive Nerve Stimulation (RNS) revealed decreased amplitude (Figure 3b). Computed Tomography of Chest (to rule out thymic hyperplasia) was normal.

On basis of clinical and laboratory findings patient was diagnosed as a case of Ocular Myasthenia Gravis with Isolated left IR palsy.

Management

On diagnosis of OMG, patient was reviewed by neurologist to rule out systemic MG. He was managed with oral Pyridostigmine 30mg twice a day (no side effects observed). After 2 months of Pyridostigmine therapy, patient became asymptomatic (diplopia free) and ocular examination was normal (Figure 4). At last follow up (two years), patient is asymptomatic with well tolerance to drug and no features of ptosis, EOM palsy, contralateral eye involvement or any systemic features.

Discussion

Review of literature showed that MG presenting as isolated IR palsy without ptosis is very rare. Most of the patients of isolated IR palsy with MG have ptosis as associating feature. In a retrospective study, it was observed that out of 35 patients of Isolated IR Palsy studied 15 had MG and out of these 15, 14 patients had variable degree of clinical ptosis and only one patient had isolated palsy without ptosis [1].

Isolated unilateral IR muscle involvement in OMG is not a common finding. In a review study of ten-year duration only two patients out of 49 cases of newly detected OMG had IR involvement [2]. In a prospective study of OMG patients in south India, none of the 40 patients had involvement of IR [3].

Citation: Yadav Y, Dhar S, Singh P, Mohapatra T, Gupta R and Mishra SK. Unilateral Isolated Inferior Rectus Palsy: A Rare Presentation of Ocular Myasthenia Gravis. J Ophthalmol & Vis Sci. 2021; 6(4): 1059.

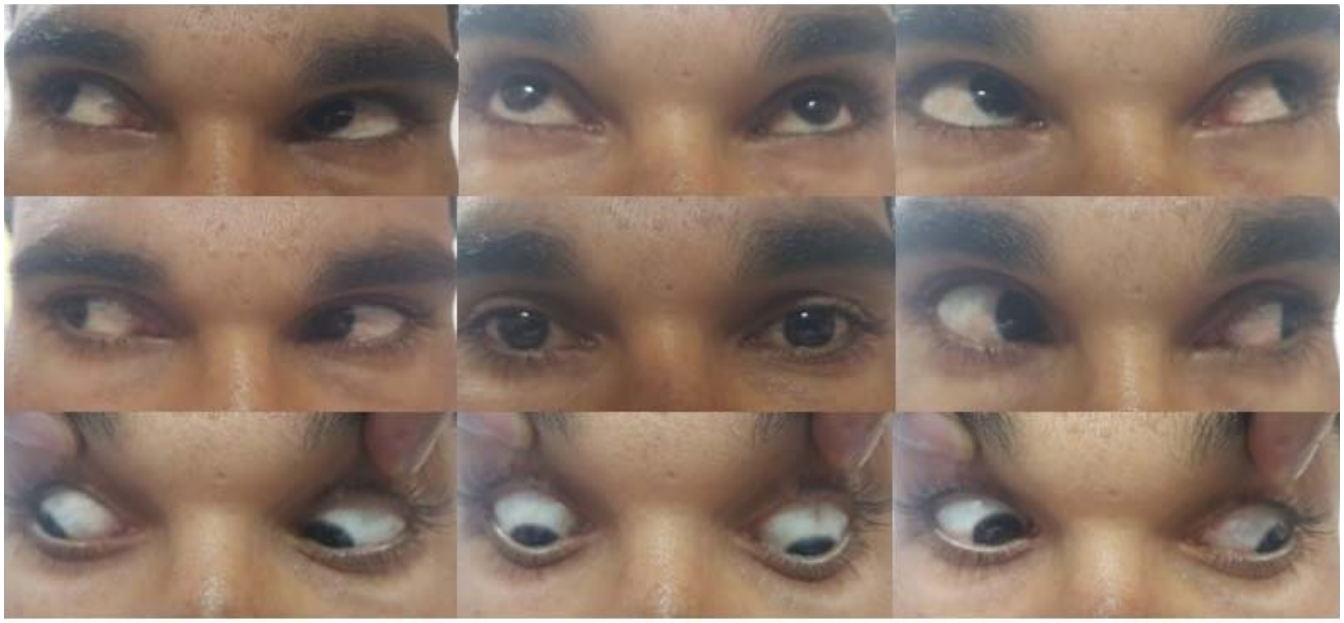

Figure 1: Nine gaze clinical pictures on arrival (before start of treatment).

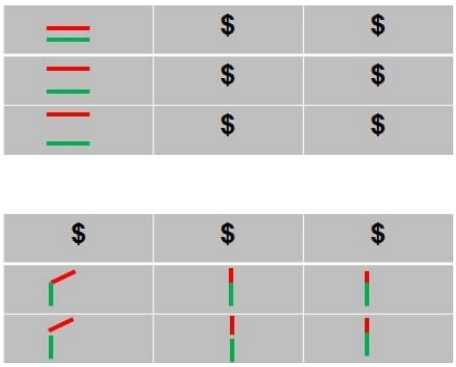

Figure 2: Diplopia Chart (Patient's View).

Neuroimaging (if not contraindicated) should be conducted in all cases of sudden onset diplopia in young individual to find any vascular abnormality leading to oculomotor nerve dysfunction. Causes of IR palsy in decreasing order are micro vascular abnormality, trauma, inflammatory causes, MG and thyroid ophthalmopathy.

Raised AChR-Ab levels generally confirms OMG but does not rule out OMG if found to be negative. It is also helpful in differentiating acquired and congenital OMG. In AChR-Ab negative patients, muscle-specific kinase (MUSK) antibodies may be present in 70% of cases [4].

RNS can help in confirming OMG in cases of AChR-Ab and MUSK antibody seronegative patients. Characteristic decrease in amplitude of action potential of muscle is noticed after 4th or 5th

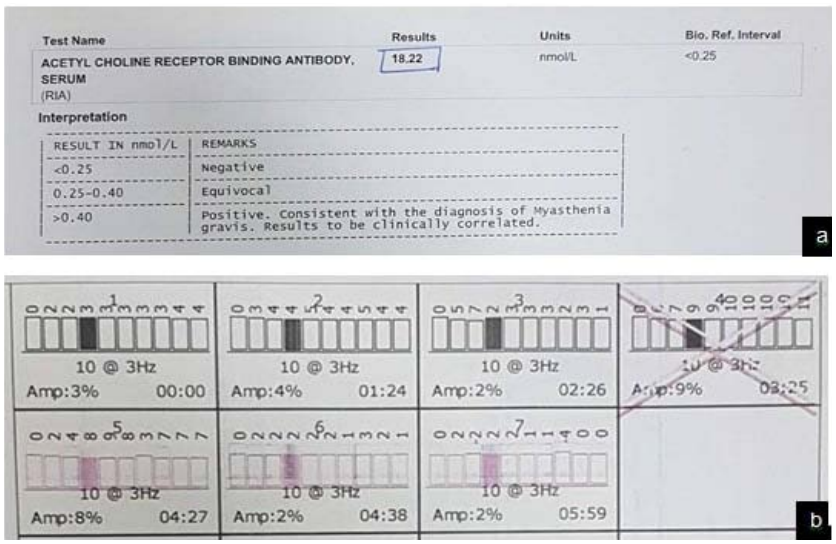

Figure 3: a: Serum Acetyl Choline Receptor Binding Report of patient; b: Repetitive Nerve Stimulation Report of patient.

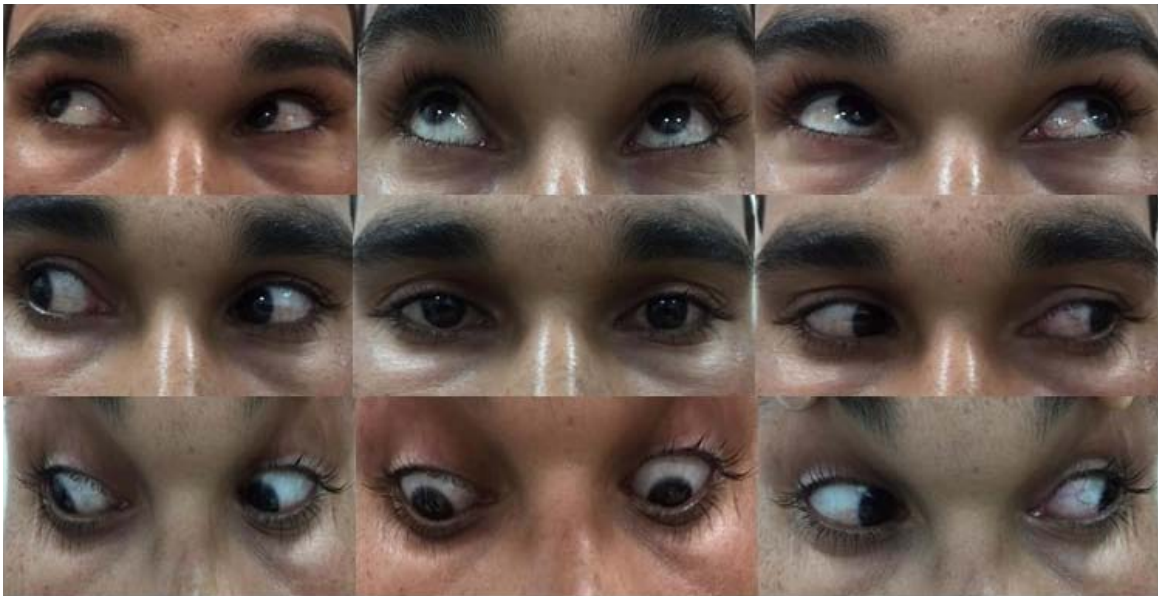

Figure 4: Nine gaze clinical pictures after 2 months of treatment.

stimulus.

Acetylcholinesterase inhibitors are mainstay of treatment in MG. It is a very well tolerated drug, reduces the need of steroid treatment and their associated side effects. These provide great symptomatic relief and their response to ptosis is better than diplopia [5].

Conclusion

OMG can present as diplopia of varying nature without eyelid involvement. MG should always be considered in the differential diagnosis of any acquired and isolated unilateral EOM palsy even if there is no ptosis. Neurologist should be involved to look for any systemic involvement and treatment thereof.

Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

References

1. Louise M. Garnham, John P. Lee. Isolated inferior rectus palsy-Is it myasthenia gravis. Jaapos. 2009; 13.
2. Marie Cleary, Graema John Williams, Richard Andrew Metcalfe. The pattern of extra-ocular muscle involvement in ocular myasthenia. Strabismus. 2008; 16: 11-18.
3. Chithra P, et al. Ocular Myasthenia in South India-A prospective cohort study. JMSCR. 2018.
4. Hoch WMJ, Helms S, Newsom-Davis J, Melms A, Vincent A. Auto-antibodies to the receptor tyrosine kinase MuSK in patients with myasthenia gravis without acetylcholine receptor antibodies. NAT MED. 2001; 7: 365-368.
5. Mark J Kupersmith, Robert Latkany, Peter Homel. Development of generalized disease at 2 years in patients with ocular myasthenia gravis. Arch Neurol. 2003; 60: 243-248.