

A Rare Case of Ocular Myasthenia Presenting with Uniocular Ptosis in a Patient Attending OPD of a Tertiary Care Centre in North Eastern India

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Abstract: A 62y/F presented with drooping of left upper eye lid since 20 days which improved with rest and associated with generalised weakness. BCVA 6/6, N6 (OU); Left Eyebrows raised, Left Upper Eye Lid drooping reducing the palpebral apertures to approximately 6mm×30mm; L/E MRD 1: 2mm, MRD 2: 4mm, MCD: 8mm; L/E LPS Function: Berke's: 14mm and Putterman's MLD: 8mm. Clinical tests done: Fatiguability test: +, Sleep Test: +, Cogan Twitch Sign: +, Ice Pack Test: +, Neostigmine Test: +. Serum Serum Acetylcholine Receptor Antibody Titre: 6.67nmol/L (HIGH), while other investigations were within normal limits. The patient diagnosed as a case of Ocular Myasthenia involving left eye. The patient was managed conservatively with Oral Pyridostigmine 60mg (TDS, PC) leading to improvement of the condition. Myasthenia Gravis is an autoimmune disease in which there is antibody mediated damage of postsynaptic acetylcholine receptors in neuromuscular junction. Uniocular Ptosis as the only presenting feature in Ocular Myasthenia is a very rare finding: seen in only 11% patients as mentioned in Karri B et al study in 2015.

Keywords: Uniocular Ptosis, Neostigmine Test, Acetylcholine, Neuromuscular Junction, Ocular Myasthenia, Autoimmune

1.Introduction

Myasthenia Gravis is an autoimmune disease in which auto-antibodies damages the post synaptic acetylcholine receptors in neuromuscular junction of skeletal muscles. Patients usually presents with features of generalised weakness associated with weakness of various striated muscles of the body resulting in clinical presentations like ptosis, diplopia, weakness of limbs and also respiratory difficulty in severe cases. Ocular involvement is seen in

almost all the cases of Myasthenia Gravis and it can be the only presenting feature as seen in majority of the cases.

2.Case Report

A 62 year old Female presented to our OPD with complaints of drooping of left upper eye lid since last 15 days. It was insidious in onset and was gradually progressive which was worsened during evening and comparatively less during morning hours. It was associated with generalised weakness.



General Examination revealed no abnormality. **Cranial Nerves** were examined and they were found to be normal.

Best Corrected Visual Acuity was 6/6, N6 (OU). On ocular examination, no obvious abnormality noted in Right Eye. On examining the Left Eye, Eye Brows were raised and Drooping of Left upper eye lid seen. **Palpebral Aperture** was **Narrow** and on measurement, 6mm vertically and 30mm horizontally was noted. **Bell's Phenomenon** was found to be normal.



Since the patient presented with Ptosis, specific test to evaluate the extent of ptosis were performed as mentioned below:

MRD 1: 2mm

MRD 2: 4mm

MCD: 8mm

LPS Function was evaluated:

Berke's Method: 14mm

Putterman's MLD: 8mm



Specific Clinical and Pharmacological tests were done to come at a diagnosis:



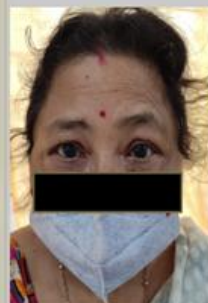



Sleep Test: Ptosis relieved after 30 minutes of sleep and it lasted for 5 mins after which again Ptosis started.

Cogan Twitch Sign was found to be positive, i.e., when the patient looked downwards for some time and then looked straight, there was up shooting of upper lid in left eye followed by twitching.

Fatigability Test: When the patient was asked to look upwards to 5 minutes, there was deterioration of Ptosis, i.e., the drooping of upper eye lid in left eye increased suggesting positive fatigability test.

Ice Pack Test: When ice pack was applied over left upper eye lid, there was improvement in ptosis and it lasted for some time after which the muscle became fatigue and ptosis reoccurred.

CLINICAL TESTS

FATIGUABILITY TEST	SLEEP TEST	COGAN TWITCH SIGN	ICE PACK TEST
<p style="font-size: small;">Patient asked to look up for sometime and thereby, muscle fatigue sets in</p>  <p style="font-size: small;">↓</p> <p style="font-size: small;">Ptosis increased</p> 	 <p style="text-align: center;">IMPROVEMENT OF PTOSIS</p>	 <p style="text-align: center;">POSITIVE</p>	 <p style="font-size: small;">↓</p> 

Neostigmine Test: 10 minutes after intra muscular injection of 1mg of Neostigmine, there was significant

improvement of Ptos. This suggested presence of some neuromuscular disorder.



Investigations: Various Imaging Studies and Blood Investigations were done as shown below:

INVESTIGATIONS		
MRI Brain	CT Thorax	Blood Tests
<ul style="list-style-type: none"> • Diffuse age related cerebral atrophy • A couple of tiny hyper intensities on T2 FLAIR in subcortical white matter of right posterior parietal lobe • Few tiny glottic foci seen in bilateral cerebral peduncles 	<ul style="list-style-type: none"> • No THYMOMA • No Mediastinal lymphadenopathy • No pleural effusion • Clear lung fields 	<ul style="list-style-type: none"> • Acetyl Choline Receptor Antibody titre: 6.67 nmol/L (>0.50 : Significant) • RBS: 119 mg/dL • Serum Creatinine: 0.94 mg/dL • T3: 1.68 nmol/mL • T4: 107.5 nmol/mL • TSH: 5.00 uIU/mL

Based upon the history, findings elicited from examination and investigations, the patient was diagnosed to be suffering from **OCULAR MYASTHENIA INVOLVING LEFT EYE.**

She was managed conservatively with **Oral Pyridostigmine 60mg (TDS, PC)** along with Oral

Pantoprazole 40mg (ODAC, CM). There was **marked improvement** of the symptoms.

3. Discussion

Myasthenia Gravis is an autoimmune disease in which Antibody mediated damage and destruction of

postsynaptic acetylcholine receptors occurs in striated muscles.¹ Weakness is experienced once the number of receptors is 30% or less.² Ocular involvement occurs in 90% of the cases and is the presenting feature in 60%.² 2/3rd of the patients have both ptosis and diplopia.² < 10% of the cases have ptosis alone and <30% presents with diplopia alone.² Ocular Myasthenia has female preponderance in 2nd-3rd decade of life and male preponderance in 6th-7th decade of life. However Juvenile Ocular Myasthenia has no sex predilections.³ Meticulous history taking, thorough clinical observation & examination and a series of hematological, radiological and clinical tests leads to confirm the diagnosis. In patients with Ocular Myasthenia if they continue to have only ocular symptoms for three years, it is very likely that their symptoms will not increase.⁴ Karri B *et al* in a study published in 2015 concluded that UNIOcular PTOSIS was the only presenting feature in Myasthenia Gravis in 11% patients.⁵ Chouhan J K *et al* in 1990 stated that Ophthalmic involvement in myasthenia gravis is not an uncommon feature, but it is very rare to find a case of UNILATERAL OPTHALMIC MYASTHENIA GRAVIS without the slightest involvement of the other eye.⁶

4. Conclusion

Ocular Myasthenia can be very debilitating disease if patient comes in severe stage as it might result in severe visual impairment due to ptosis and diplopia. However, it is easier to diagnose if evaluated thoroughly and can be managed with various pharmacological agents like Oral Pyridostigmine. In severe cases, Immunosuppressant like Prednisolone, Azathioprine, Mycophenolate Mofetil may be necessary. Thymectomy although tried sometimes in cases of Generalised Myasthenia Gravis, it is seldom done in cases of Ocular Myasthenia.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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