

Ocular myasthenia gravis (OMG) Steroid Hackneyed: A Case study

Abstract

Ocular myasthenia gravis (OMG) is a subtype of the systemic disease which affects only the eye but many patients do progress to generalized form. The mainstay of treatment is acetylcholine esterase inhibitors along with corticosteroids and/or immunosuppressant drugs which is aimed at disease remission and prevention of progression to the generalized form. We report a case of a 9 year old female patient who was diagnosed as OMG. She was on oral steroids, unmonitored from the past 4 years and came to us for persistent variability of symptoms and was found to have disc changes suspicious of glaucoma with IOP on higher side in both eyes along variable ptosis and diplopia. Systemically she had mild stunting of growth for which we involved a team of pediatrician and endocrinologist who advised tapering and regular monitoring. We hope to stress upon the importance of ophthalmic and systemic monitoring of patients on steroids for ophthalmologic conditions, specially the pediatric age group.

Introduction:

Myasthenia Gravis is an auto-immune disorder that affects the post-synaptic neuromuscular junction membrane. Acetylcholine receptor (AChR) antibodies are frequently present, and the number of functioning postsynaptic receptors is reduced. Muscle-specific kinase antibodies may be identified in individuals with and without AChR antibodies.^[1] It is called the great masquerader owing to its varied clinical presentations. Myasthenia may affect any age group and shows no geographic predilection.^[2] Onset of symptoms in the first decade or after the age of 70 years is less common.^[3] The incidence ranges from 0.04 to 5/100 000/year and prevalence estimates of 0.5-12.5/100 000/year.^[3] Myasthenia is a disease of neuromuscular

junction. Anti acetylcholine receptor antibodies (AChR Abs) have been demonstrated in up to 99% of patients with generalized myasthenia and 40-77% of patients with OMG. Approximately half of the patients are first seen with purely ocular myasthenia gravis (OMG) with ptosis and diplopia being; of these, 53% develop generalized Myasthenia gravis (GMG) within 2years (80% in the first year). Furthermore, a spontaneous remission rate of 30% has been reported in patients with OMG during a 15-year period. ^[4]

Case Report:

A 9 year old male patient presented to the opd with chief complaints of intermittent ptosis and diplopia from past few months. He was a diagnosed case of Ocular Myasthenia gravis, with positive Anti-Ach receptor antibodies and was using Oral Acetyl Cholinesterase (Tab. Dystinon 60 mg OD) and steroids (Tab. Omnacortil 20 mg OD) for the same from the past 4 years at the time of presentation. He did not follow up with the treating doctor and was on treatment unmonitored. On examination, His distant visual acuity was 6/6 in both eyes and near visual acuity was N6. Extra-Ocular movement assessment revealed mild restriction on dextro-elevation. [Fig.1] He had diplopia in extreme right and left gazes. Left eye showed mild ptosis in the primary gaze along with slight head tilt ad

head turn towards right side. Icepack test and Cogan twitch sign were negative at the time of presentation. On slit lamp examination, anterior segment was normal in both eyes. Intraocular pressure was 18 mmHg in both eyes with applanation tonometer. His visual fields were within normal limits on Humphrey's perimetry. [Fig.2]

On dilated fundus examination he had an increased C:D ratio of 0.5:1 in both eyes which was suspicious of glaucomatous changes. [Fig.3] Rest of the fundus appeared normal. He was referred to pediatrician and was diagnosed with mild stunting of growth attributable to the prolonged steroid usage. However, His blood investigations and X-ray were within normal limits. A multidisciplinary approach was opted for the patient. Revised treatment regimen was advised by a team of endocrinologist, pediatrician and ophthalmologist, which included tapering of oral steroid (Tab. Omnacortil-5 mg, along with syrup shelcal and Tab. mestinon 60 mg OD and was advised regular follow up with pediatrician and ophthalmologist. This case stresses upon the importance of regular monitoring of patients on steroid usage for ophthalmic conditions.



Figure 1

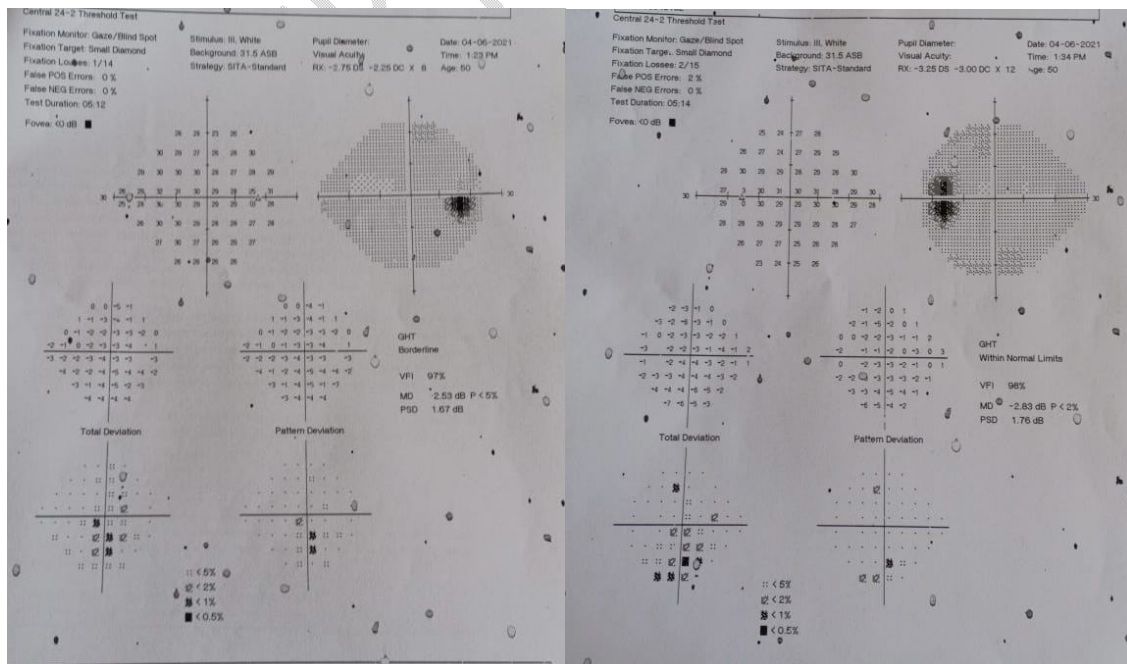


Figure 2



...t
...s
...le
...ph
...d
...e
...c

...affecting the ocular surface, anterior and posterior segments of the eye due to their anti-inflammatory, anti-edematous, and anti-neovascularization properties. The ophthalmic use of steroids makes a big list including diseases of both anterior and posterior segments like allergic conjunctivitis, Scleritis,

Episcleritis, Uveitis, herpes zoster, traumatic optic neuropathy, optic neuritis e.t.c, .^[6] The route of administration can be systemic or local. Conventional topical application to the eye is the route of choice when targeting diseases affecting the ocular surface and anterior segment, while periocular, intravitreal, and suprachoroidal injections can be potentially effective for posterior segment diseases. However, the steroid usage doesn't come without any side effects. The systemic side effects include growth retardation, musculoskeletal osteoporosis, Myopathies, endocrine and metabolic abnormalities, immunosuppression, Tuberculosis.^[7]

The most common ocular side effects are cataracts with topical and glaucoma with systemic steroid usage. Although oral corticosteroids are the treatment of myasthenia gravis (MG), the possibility of steroid-induced exacerbation of symptoms, especially during the initial course of steroid therapy, has limited their use patients with severe MG.^[8] Apart from the long term side effects; the initial high dose therapy is known to cause acute exacerbation especially in bulbar type.

^[8] Many studies showed that steroid therapy causes retardation of linear growth in pediatric age group.^[9] However, the tendency is more with high dose steroid therapy indicated for conditions like nephrotic syndrome. Our patient although was on low dose steroid manifested mild stunting of growth. In a

previously reported study, the authors found steroid-induced glaucoma to account for one-fourth of all acquired glaucoma's in children.^[10] Increasing trends of steroid-induced glaucoma in children over the last few years have been reported, probably signifying increasing use of unmonitored steroid use.^[11] Similar is the case in our current patient. We would like to stress upon the importance of counseling and regular monitoring of patients on steroid therapy to prevent adverse local and systemic adverse effects.

UNDER PEER REVIEW

References:

1. Hoch W, McConville J, 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(3):365-368.
2. López-Cano M, Ponseti-Bosch JM, Espin-Basany E, Sánchez-García JL, Armengol-Carrasco M. Clinical and pathologic predictors of outcome in thymoma-associated myasthenia gravis. *Ann Thorac Surg* 2003;76:1643-9.
3. O'Brien MD. The miracle at St Alfege's: Seventy years on. *J R Soc Med* 2007;100:257.
4. Oosterhuis HJ. Observations of the natural history of myasthenia gravis and the effect of thymectomy. *Ann N Y Acad Sci*. 1981;377:678-690.
5. Ferrara G, Petrillo MG, Giani T, Marrani E, Filippeschi C, Oranges T, Simonini G, Cimaz R. Clinical Use and Molecular Action of Corticosteroids in the Pediatric Age. *Int J Mol Sci*. 2019 Jan 21;20(2):444. doi: 10.3390/ijms20020444. PMID: 30669566; PMCID: PMC6359239.
6. Dinning WJ. Steroids and the eye--indications and complications. *Postgrad Med J*. 1976 Oct;52(612):634-8. doi: 10.1136/pgmj.52.612.634.

7. STANBURY RM, GRAHAM EM Systemic corticosteroid therapy—side effects and their management *British Journal of Ophthalmology* 1998;82:704-708.
8. Bae JS, Go SM, Kim BJ. Clinical predictors of steroid-induced exacerbation in myasthenia gravis. *J Clin Neurosci*. 2006 Dec;13(10):1006-10.
9. Kochar, Dhanpat & Shubhakaran, Khichar & Kumawat, BI & Thanvi, Indu & Joshi, A & Vyas, SP. (1998). Ophthalmoscopic abnormalities in adults with falciparum malaria. *QJM: monthly journal of the Association of Physicians*. 91. 10.1093/qjmed/91.12.845.
10. Kaur S, Dhiman I, Kaushik S, Raj S, Pandav SS. Outcome of ocular steroid hypertensive response in children. *J Glaucoma*. 2016 Apr;25(4):343–347.
11. Gupta S, Shah P, Grewal S, Chaurasia AK, Gupta V. Steroid-induced glaucoma and childhood blindness. *Br J Ophthalmol*. 2015 Nov;99(11):1454–1461