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Ocular Myasthenia Gravis (OMG) Steroid Hackneyed: A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

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. It is called the great masquerader owing to its varied clinical presentations. 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 male patient who was diagnosed as OMG. He 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 normal side in both eyes with variable ptosis and diplopia. Systemically she had mild stunting of growth for which the pediatrician and the endocrinologist 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. With increasing trends of steroid-induced glaucoma in children over the last few years, probably signifying increasing use of unmonitored steroid use, 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.

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1. 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; of these, 53% develop generalized Myasthenia gravis (GMG) within 2 years (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].

2. CASE REPORT

A 9 year old male patient presented to the outpatient department 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 Tablet Mestinon 60 mg OD and Tablet Omnacortil 10mg 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 and head turn towards right side. Cogan twitch sign was positive although Icepack test was 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 in 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 in view of short stature and repeated infections 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 Tablet 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.

The patient was followed up after 5 months, and a drastic improvement in diplopia along with ptosis was observed. The mild restriction in extraocular movement in dextroelevation had improved.

3. DISCUSSION

Systemic steroid therapy is the primary treatment modality in many conditions like JIA (Juvenile Idiopathic arthritis), Scleroderma, ulcerative colitis, crohn disease, Asthma, Nephrotic syndrome, demyelinating disorders and skin conditions like atopic dermatitis etc [5]. "Corticosteroids also remain the mainstay of the treatment for various ocular conditions 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 etc" [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].

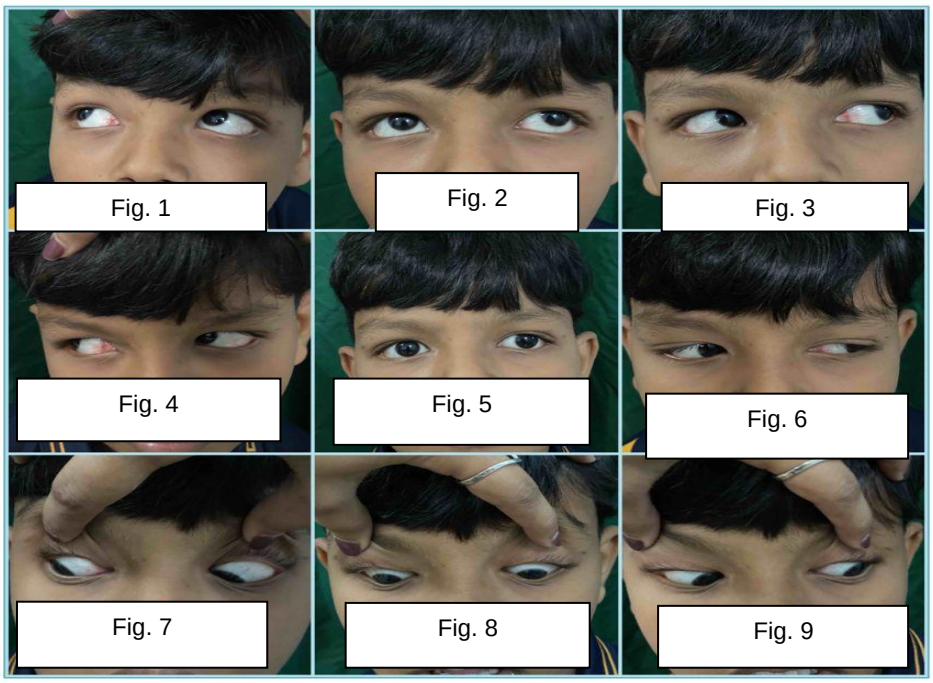

Fig. 1. Dextroelevation (mild restriction)
Fig. 2. Elevation
Fig. 3. Laevoelevation
Fig. 4. Dextroversion
Fig. 5. Primary position
Fig. 6. Laevoversion
Fig. 7. Dextrodepression
Fig. 8. Depression
Fig. 9. Laevodepression

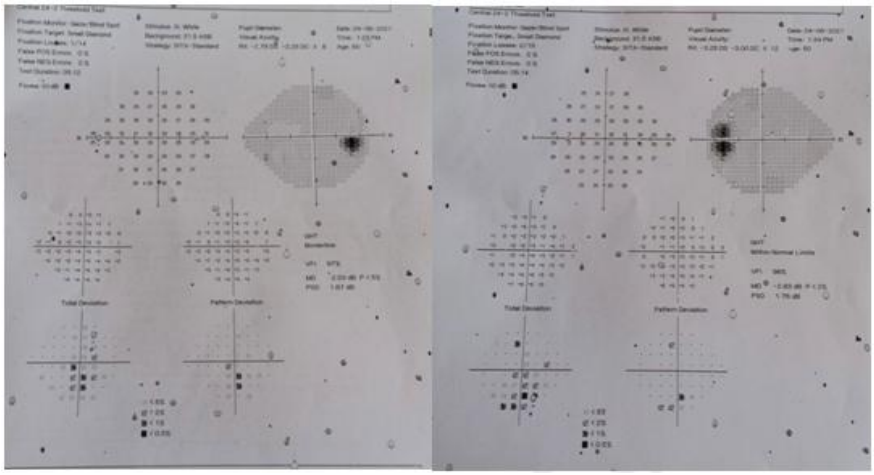

Fig. 10. Showing humphreys perimetry report of the patient that was within normal limits

Fig. 11. Showing the fundus photograph of right eye and left eye of the patient with glaucomatous cupping of 0.5:1

"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 in 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 of Myasthenia Gravis [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 therapyto prevent adverse local and systemic adverse effects.

"The first line of treatment, once the diagnosis of OMG is made, is pyridostigmine in children or neostigmine in neonates" [12]. "The next line of treatment is corticosteroids, such as prednisone. Sparing corticosteroid therapy, such as azathioprine, can be added. If there is no adequate response to this treatment, management with cyclosporine, methotrexate, or cyclophosphamide can be initiated, and there are even reports in literature on the efficacy of tacrolimus (FK 506) in ocular symptoms refractory to treatment and use of plasmapheresis and immunoglobulins in acute phases" [13]. "Thymectomy should be avoided in childhood by immunosuppression, especially in children with negative antibodies; it is a rare measure in the pediatric population due to the low frequency of thymomas" [14].

The prognosis of OMG is benign and may present remission after 2 years of treatment [15,16].

4. CONCLUSION

This case emphasizes the importance of routinely monitoring patients on steroid use for ophthalmic conditions. The most common ocular side effects are cataracts with topical and glaucoma with systemic steroid usage. Numerous studies have demonstrated that steroid therapy in children hamper their linear growth. With high dose steroid therapy prescribed for illnesses like nephrotic syndrome, the propensity is more pronounced.

CONSENT

As per international standard, parental written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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