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Varied Presentation of Myasthenia Gravis: A Case Report

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Abstract: Myasthenia gravis is an autoimmune disease in which autoantibodies mediate damage and destruction of acetylcholine receptors in striated muscle causing impairment of neuromuscular coordination. It causes weakness and fatiguability of skeletal musculature sparing cardiac involuntary muscles. It occurs in three forms ocular, bulbar and generalized. Two thirds of ocular myasthenia presents as diplopia and ptosis, but isolated ptosis and isolated diplopia in 10% and 30% respectively. This case presented with isolated uniocular ptosis of right eye with retraction of contralateral upper eye lid. Investigations like ice pack test, fatigue test, neostigmine test and serological tests like acetylcholine autoreceptor antibodies showed positive results. Patient was treated with oral corticosteroid but on maintenance dose symptoms recurred. Tab Pyridostigmine 60mg qid was combined with maintenance dose of corticosteroids showed steroid related complications in a period of 1 month. Patient recovered with immunosuppressant Tab Azathioprine 50mg OD showing neither of the steroid related complications.
Keywords: Ocular myasthenia, Ptosis, Retraction of contralateral upper lid, Corticostriods, Pyridostigmine, Azathioprine.

INTRODUCTION

Myasthenia gravis is an autoimmune disease of postsynaptic acetylecholine receptor of unknown etiology [1]. The prevalance of disease is 1 to 14 / 1, 00,000. Females are more commonly affected than males with male to female ratio of 2:3. It may present as only ocular myasthenia gravis 50 % of time. Incidence of only ptosis which is bilateral is in 20% of cases and 10% of cases present with unilateral ptosis [2]. Spontaneous remissions can occur in any patient. Most patients show a fluctuating course. Age, sex and pattern of onset are not used to predict the onset of the disease [3] . Myasthenia gravis is commonly associated with Grave's disease and Thymomas [4].

CASE REPORT

A 40 year old patient came to Mamata General Hospital, Khammam with the history of drooping of right upper lid since 6 months (Fig. 1). Ptosis was variable in nature with a significant diurnal variation which improved on taking rest. Patient gives history of similar complaints in the past with relief of symptoms on treatment. Patient had no other associated systemic features.

The examination was carried with the prior consent from the patient, on examination best corrected visual acuity both eyes was 20/20. Ptosis in OD upper lid was variable in nature and worsening of ptosis was seen as day progressed (Fig. 2) . Lid crease of OD present at the same level as OS with good LPS function with contralateral lid retraction.

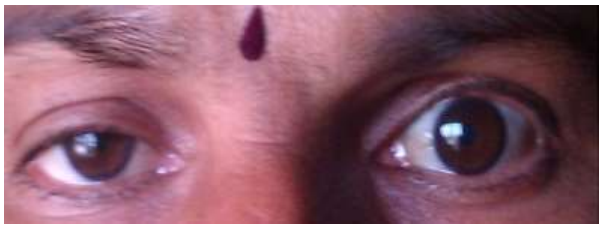

Fig. 1: Drooping of right upper lid with contra lateral lid retraction.

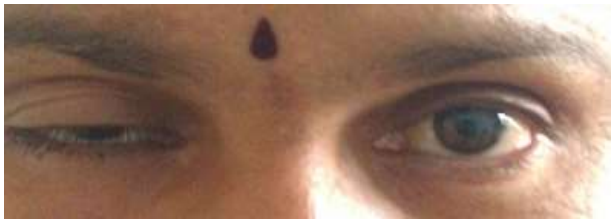

Fig. 2: Diurnal variation in ptosis

There is no restriction of extraocular movements. Ptosis worsened on fatigue test (Fig. 3) and improved with ice pack test which aroused a clinical diagnosis of myasthenia gravis (Fig. 4, 5).

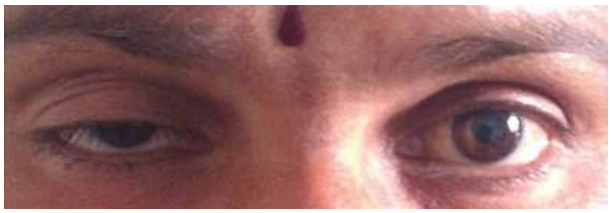

Fig. 3: Fatigue test

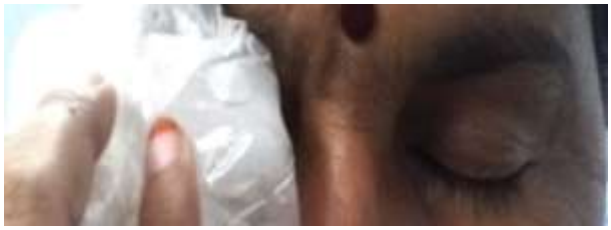

Fig. 4: Ice pack test

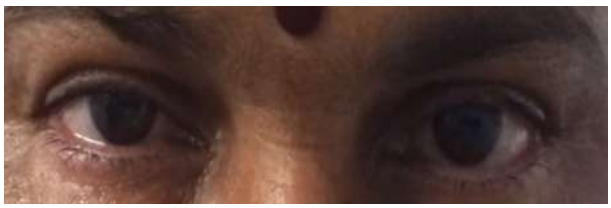

Fig. 5: After ice pack test

Following this patient was subjected to investigations for myasthenia. Neostigmine test showed improvement of ptosis after 30 min of administration of 1.5mg of neostigmine methylsulfate combined with 0.6 mg of atropine sulfate intramuscularly. Acetylecholine receptor antibodies positivity is a highly specific test and is 0.63 nmol/L units(Negative<0.25, Equivocal 0.25-0.40, Positve>0.40) by radioimmunoassay method . Thyroid profile, CT brain and thorax (Fig.6) suggested no abnormality. Electromyography and nerve conduction test showed normal study.

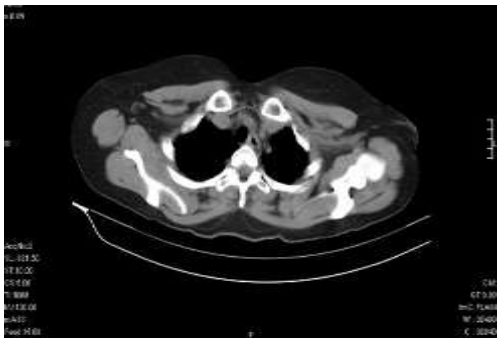

Fig. 6: CT thorax showing normal study

Patient was initially started on oral prednisolone 1mg /kg/day and tapered weekly for 1month and maintained at a dose 5mg/day for 1 month. Following 1 month of maintainence dose patient came with recurrence of ptosis. Patient was then started on oral Pyridosostigmine 60 mg QID dose along with the maintainence dose of oral at 5 mg/day.

Fig. 7: Ptosis relieved on treatment

As the patient showed improvement with this treatment (Fig. 7), patient was followed up monthly for 6 months to evaluate for steroid related complications and any recurrence of ptosis. After 6 months of treatment patient did show Cushingoid features (Fig.8) and raised IOP with no optic nerve changes, hence steroids were stopped and immunosuppressant Tab Azathioprine 50 mg OD was started for the same. Patient is now maintaining well with above treatment.

Fig. 8: Cushingoid features due to long term steroid treatment

DISCUSSION

Myasthenia gravis (MG) is one of the best studied autoimmune diseases. It is due to the formation of auto-antibodies against Acetlcholine receptors located in the neuromuscular junctions. Treatment options for Ocular MG include blepharoplasty or use of lid crutches for providing releif from ptosis, pharmacotherapy includes cholinesterase inhibitors, immunosuppresive therapy using either steroids or other immunosuppresive agents. Thymectomy has also been tried for treating MG. On reviewing the available literature, we could not identify any randomized control trials relating to the treatment options and progression of ocular MG to generalized MG. [5]

In an observational study by Sommer et al, among 178 patients with myasthenia gravis, ocular MG was diagnosed in 78. Thirty one percent(24) of the patients with ocular MG progressed to generalized disease. In the 178 patients, 39 were diagnosed with

concomitant autoimmune diseases, thyroid disease was the most common being present in 33 patients. A correlation was found between progression of ocular MG to generalized MG based on the severity of symptoms. The progression rates in mild, moderate and severe ocular MG were 14%, 36% and 43% respectively. They have also identified that early immuosuppresive treatment helped in reducing the progression from ocular MG to generalized MG. Thymecomy also had a similar beneficial effect. [6]

34 patients with purely ocular signs at the time of presentation were followed-up for a period of 2 years by Monsul et al. The patients who did not receive immunotherapy had a higher rate of progression to generalized disease in comparison to those that received immunotherapy (90.5% and 23.1% respectively). [7]

Paine et al reviewed 56 patients with ocular MG and concluded that use of oral prednisone delayed the onset of generalized MG at the end of 2 years follow-up. Treatment with 60 mg of prednisone daily tapered slowly over 3–6 months significantly reduced the risk of progression. [8]. In a retrospective evaluation of 83 ocular MG patients who were on oral prednisolone (dosage ranging from 10 - 60 mg) tapering doses, Bruce and Kupersmith have identified no significant steroid induced complications at lower dose concentrations. [9]

'EPITOME' study (Efficiency of Prednisolone in treatment of Ocular Myasthenia) is a phase 3 clinical trial in progress. The results of this study would help provide a better understanding of the role of corticosteroids in treatment of ocular MG. [10]

CONCLUSION

It is important that patents with ocular MG (or generalized MG with ocular symptoms) have their eyes checked regularly by an Ophthalmologist. The treating physician may be able to recommend an eye care specialist who is experienced in treating patients with MG. The clinical observations which distinguish ocular myasthenia from generalized MG include: highly restrictive ocular symptoms such as diplopia, ptosis, and weak eye closure. The good news for patients with ocular MG is that if they continue to have only ocular symptoms for two years, there is a very good chance their symptoms will not increase. This case is different in the sense though visual disturbances and diplopia being most common, the patient presented with isolated ptosis and contralateral upper lid retraction which showed remission and exacerbations with steroids but showed relief of symptoms with immunosuppressants Azathioprine.

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