MYASTHENIA GRAVIS WITH INTERNAL OPHTHALMOPLEGIA: A RARE CASE REPORT

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ABSTRACT

Involvement of the eye muscles is frequently encountered in myasthenia gravis. In the great majority of cases there is unilateral or bilateral external opthalmoplegia which may be partial or complete. The involvement of the smooth muscles of the pupil seems to be very unusual phenomenon and its presence may even raise doubts that the ocular involvement in such cases is a result of myasthenia gravis. We describe a patient with myasthenia gravis who showed external and internal opthalmoplegia, which responded well to anticholinesterase drugs.

KEYWORDS: Myasthenia gravis; Internal opthalmoplegia.

CASE REPORT

A 40 years (FIG 4) old male presented with history of 2 years duration of difficulty in opening both the eyes, pain in the neck, difficulty in chewing and swallowing the food. He was unable to open both the eyes which worsened as day progressed and was completely relieved in the morning hours. He was comfortable in chewing first few bytes of food, and on chewing further bytes of food, he felt pain and heaviness in the jaw. There was no history of bladder, bowel and sensory involvement.
Neurological examination was normal, except internal ophthalmoplegia. Both the pupils were mid dilated and non reacting to light.

In view of the fluctuating weakness, he got relieved on taking rest Thus a diagnosis of neuromuscular junction disorder probably myasthenia gravis was kept in mind.

On examination, he was conscious. Vitals stable, Pulse rate 84/min, BP 130/80 mm of Hg. Examination of the cardiovascular, respiratory system and abdomen was noncontributory. On neurological examination, higher functions were normal. He had bilateral external ophthalmoplegia, ptosis, and complete paralysis of lateral rectus, superior rectus and inferior rectus on the right side.

All the eye movements were markedly limited on the left side with evidence of ptosis.

Both pupils were semidilated and non-reacting to light on both sides while fundus examination was normal.

Examination of the motor system, deep tendon reflexes, plantar response, superficial reflexes, cerebellar system revealed no abnormality.

Laboratory investigations revealed Hb 14 gm%, Total leucocyte count 7800/mm$^3$, ESR 34, blood urea 27 mg%, serum creatinine 1.0 mg/dl, blood sugar 111 mg/dl. Serum electrolytes, calcium & phosphorus and liver function tests were within normal limits. Thyroid function tests, antinuclear antibodies profile revealed no abnormality.

Serum acetylcholine receptor antibody was 16.06 nmol/L (positive)(<0.25 :negative, 0.25-0.40 : equivalent, >0.40: positive).

X-ray chest showed mediastinal widening (FIG 1).

ECG Normal.

CECT thorax (FIG 2) revealed suspicion of thymoma but the patient refused histopathological examination.

MRI brain (FIG 3) was normal.

Serological test for syphilis was negative.
Therapeutic test with 10mg of edrophonium chloride (tensilon) administration IV. resulted in a temporary improvement of all eye movements which lasted for 4-5 minutes.

A conventional electromyographic examination of maximal voluntary contraction of the masseter and abductor digiti minimi muscles showed a rapid decrease in muscle action potentials.

Following supramaximal tetanic stimuli and the ulnar nerve, there was a progressively rapid decrease in evolved muscle potentials.

The patient’s ocular symptoms including the pupillary abnormalities were partially controlled by 60mg tablets of pyridostigmine bromide every 6 hourly and tablet prednisolone 1mg/kg/day. He was planned for thymectomy and referred to higher centre.

FIG 1: (X-Ray chest PA view: showing mediastinal widening).

FIG 2 CECT Thorax: showing ? thymoma
DISCUSSION

In this patient, external ophthalmoplegia was associated with semidilated pupil, not reacting to light stimulation.

No evidence of any other intracranial or orbital pathology which could be responsible for the pupillary changes was found since MRI brain and fundus examination were normal.

The non-reaction to light of constrictor pupillae and its subsequent fatigue suggested a myasthenia phenomena and it was confirmed by the decrease in size of the myadriatic pupil and the disappearance of pupillary fatigue and extraocular muscle after the administration of anticholinesterase drugs.
Pupillary changes in patients with myasthenia gravis seem to be very rare. However, it has been pointed out that in rare cases the pupillary reflex may be sluggish and may show unusual fatigue.[1,2]

Several years ago, Baptista and Souza[3] described a patient suffering from severe generalized myasthenia gravis who demonstrated pupillary changes similar to those shown by our patient. It seems therefore that the internal ophthalmoplegia in a patient with external ophthalmoplegia excluded myasthenia gravis, should be re-evaluated.

CONCLUSION
Pupillary changes are described in a patient suffering from the ocular form of myasthenia gravis. The pupils were semidilated and non-reacting to light. After a therapeutic test with tensilon and under maintenance therapy with pyridostigmine, the pupils started reacting to light. The pupillary changes in this patient were the result of myasthenic involvement of the constrictor pupillae.

REFERENCES