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An Atypical Presentation with Extraocular Muscle Atrophy in Myasthenia Gravis Delaying Diagnosis

Case Report

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Abstract

A 14 year female presented with 10 year history of slowly progressive asymmetric bilateral ptosis and complete bilateral external opthalmoplegia without involving pupil or vision, 1 month history of dysphagia, nasal intonation of voice and proximal muscle weakness without diurnal variation or Fatigability. MRI showed normal brain with atrophied extraocular muscles. RNS hinted towards decremental response. Neostigmine challenge, Serum anti-AChR antibody was positive. There was no thymic enlargement. Usually generalization of ocular myasthenia occurs within 2-3 years of onset but in this case it took almost 10 years. Lack of significant diurnal variation may go unnoticed by patient sometimes and may mislead the physician to a very close differential diagnosis of CPEO. In this case so many atypical features are there like lack of significant variation of symptoms, generalization after a stable disease course of 10 years and associated extraocular muscle atrophy.

Keywords: Myasthenia gravis; Chronic progressive external opthalmoplegia(CPEO); Anti-AChR antibody (Anti-Acetylcholine receptor antibody)

Key Messages: Learning point from this case is that every case of opthalmpoplegia deserves to be screened for possibilities of myasthenia gravis and extraocular muscle atrophy should not be taken be indicative of CPEO without excluding possibilities of a treatable disease like myasthenia gravis.

Introduction

Ptosis and external opthalmoplegia are common initial presentation of myasthenia gravis. In long standing slowly progressive cases diurnal variation and Fatigability may go unnoticed if mild which can easily mislead to chronic external opthamoplegia especially when associated with extraocular muscle atrophy in neuro-imaging. Extraocular muscle atrophy can be rarely present in myasthenia gravis. Usually generalization of ocular myasthenia occurs within 2-3 years but the duration varies in different studies.

Case Report

A 14 year female presented with 10 year history of slowly progressive asymmetric bilateral ptosis. The symptoms started at the age of 4 years. The family member noticed that she has a fix gaze and unable to move her eyeballs. On ophthalmic examination there was bilateral ptosis (interpalpebral fissure of 4mm on right, 3 mm on left) with complete bilateral external opthalmoplegia without involving pupil or vision. There was no significant history of diurnal variation or Fatigability. On Suspicion of chronic progressive external opthalmoplegia, MRI was done which showed a normal brain with bilaterally atrophied extraocular muscles (Figure 1). Within 1 month of initial presentation she again presented with dysphagia, nasal intonation of voice and proximal muscle weakness. There was grade 4 power of proximal muscles, otherwise rest of motor and sensory examinations were normal. This time we decided to do repetitive

nerve stimulation which hinted towards decremental response. Neostigmine challenge was given and ptosis as well as other symptoms improved remarkably (Figure 2). All blood investigations including CPK were normal. Her serum anti-acetylcholine receptor antibody was positive (>8 nmol/l), anti-MuSK antibody negative ((0.2 U/ml). There was no thymic enlargement in CT scan of chest. She improved with pyridostigmine and prednisolone. Glycopyrrolate 0.5 mg twice a day was started along with pyridostigmine 30 mg three times daily orally with dose increase according to response. Patient was started with 20 mg oral prednisone with 10 mg dose increase in every 2 weeks till 1 mg/kg/day and maintained till improvement.

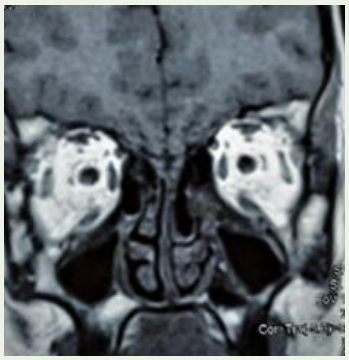

Figure 1: Atrophied extraocular muscle.

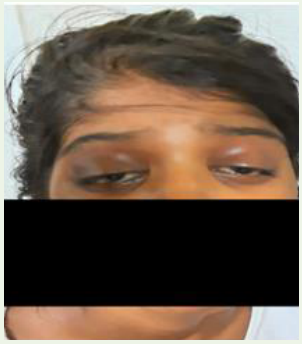

Figure 2: Bilateral ptosis.

Discussion

Usually generalization of ocular myasthenia occurs within 2-3 years of onset but in this case it took almost 10 years [1]. Lack of significant diurnal variation may go unnoticed by patient sometimes and may mislead the physician to a very close differential diagnosis of CPEO. Though extraocular muscle atrophy is uncommon finding in myasthenia gravis, it may be misleading sometimes. In this case so many atypical features are there like lack of significant variation of symptoms, generalization after a stable disease course of 10 years and associated extraocular muscle atrophy [2,3].

Conclusion

Learning point from this case is that every case of Opthalmoplegia deserves to be screened for possibilities of myasthenia gravis and extraocular muscle atrophy should not be taken as indication of CPEO without excluding possibilities of a treatable disease like myasthenia gravis.

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