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Sir,

Myasthenia Gravis with Pupillary Involvement

Myasthenia gravis is an autoimmune disorder affecting the nicotinic acetylcholine receptors at the neuromuscular junction of skeletal muscle. The resulting loss of functional receptors impairs transmission of signals at the neuromuscular junction leading to weakness and increased fatiguability in those muscles. Myasthenia gravis can occur in generalised or ocular forms, but patients with both types frequently present to the ophthalmologist initially with symptoms of diplopia and/or ptosis. We present a case where atypical signs confounded the diagnosis.

Case Report

A 15-year-old girl presented with a 5 day history of vertical and horizontal diplopia and right ptosis. She was otherwise fit and well and had no past medical history of note.

On examination visual acuity was 6/6 right and left. The right pupil (3.5 mm) was larger than the left (2 mm) and was slow to constrict (and redilate) on direct and consensual light stimulation. There was limitation of right elevation, adduction and depression, but left extraocular movements were full. She had 4 mm of ptosis of the right lid. The optic discs and posterior poles were healthy.

Neurological examination (including deep tendon reflexes) was otherwise normal. Full blood count and film were normal, as was random blood glucose. Autoantibody screen including acetylcholine receptor antibody assay and syphilis serology were all negative, and thyroid function tests were normal. Magnetic resonance imaging (MRI) of the brain and orbit was normal; so too was an MRI angiogram. A lumbar puncture was performed, and results of cerebrospinal fluid analysis were unremarkable.

Her symptoms gradually improved over the Christmas period, but on her return to school the ptosis and diplopia worsened, especially by the end of the day. A test dose of edrophonium was given intravenously with immediate but temporary resolution of the ptosis and diplopia, confirming our suspicion of myasthenia gravis. She has been asymptomatic on oral physostigmine for the last 3 months. Her right pupil is decreased to 2.5 mm in diameter but is still sluggish in its reaction to light. No hypersensitivity to g. Pilocarpine 0.125% has developed. Right extraocular movements are full and there is only 1 mm residual ptosis.

Comment

Myasthenia gravis, in its generalised or ocular form, is understood not to affect the pupil or accommodation since these functions are subserved by non-striated muscle.[1] However, pupillographic analysis[2] and slit lamp biomicroscopy studies[3] have demonstrated that a high proportion of myasthenic patients have abnormal pupillary function. Furthermore Dutton *et al.*[4] have demonstrated fatiguability of pupil constriction in bright light in a series of 11 patients with myasthenia gravis. The absence of nicotinic receptors on the iris musculature means pupillary involvement is difficult to explain. The site of dysfunction may lie elsewhere, perhaps at the nicotinic receptors of the ciliary ganglion. Research by Watanabe *et al.*[5] suggests possible dysfunction at the neuronal nicotinic acetylcholine receptors in the central nervous system of patients with myasthenia gravis. It has been suggested that seronegative myasthenia gravis affecting only oculobulbar musculature may represent a separate disease entity in which an autoimmune response is not implicated.[6] In such a case symptoms and signs may not be limited to those attributed to dysfunction at the neuromuscular junction of skeletal muscle. Clearly 'myasthenia gravis' is incompletely understood and further research is being undertaken.

In the meantime we note that pupillary involvement does not exclude the diagnosis of this treatable disease. Its early consideration as a differential diagnosis may prevent unnecessary and invasive investigations and avoid delay in the administration of appropriate therapy.

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