Sir,

Acute severe ocular myasthenia in a 92-year-old woman

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The purpose of this report is to document that in severe cases of abrupt onset, the characteristic hallmarks of myasthenia may be absent. Another unique feature is the advanced age of the patient. The average age of onset of ocular myasthenia is 38 years. As ocular myasthenics over the age of 50 years are at much higher risk of developing generalised disease with respiratory crisis or death,[1] prompt diagnosis of this disorder is particularly important.

Case report

The authors describe the case of a 92-year-old woman who was referred with sudden onset of bilateral total external ophthalmoplegia associated with bilateral but asymmetrical ptosis, having awoken that morning unable to open her eyelids. The ptosis was complete on the right and approximately 90% on the left. The patient did not complain of diplopia on lifting her eyelids as the ophthalmoplegia was complete and symmetrical. There was no paresis involving the bulbar muscles. Neurological examination of the upper and lower limbs was normal. There was no prior history of fluctuating ptosis, diplopia, dysphagia or dysarthria, particularly in relation to exertion, or during the course of the day. No fatiguability or variability of the muscle weakness was elicited on clinical testing. The patient had an unremarkable medical history for her 92 years, suffering only from mild hypertension which was controlled with bendrofluazide 2.5 mg once daily.

The differential diagnosis was botulism, Miller–Fisher syndrome, a brainstem infarct or lesion, or advanced chronic progressive external ophthalmoplegia, the initial stages of which, being insidious and symmetrical, may go unnoticed by the patient. Ocular myasthenia was thought clinically unlikely, but in the presence of a normal MRI scan of the brain and orbits and in view of the clinical absence of pain, sensory loss, pupillary involvement and decreased vision, its diagnosis was reconsidered. An ice test was performed. There was a negative result on the right but a positive result on the left lid (Figure 1) suggesting the myopathy to be of a myasthenic nature. This was subsequently confirmed with positive acetylcholine receptor antibodies, a positive Tensilon test and single-fibre electromyographic studies.

Despite maximum dosage with the anticholinesterase drug pyridostigmine (Mestinon, Roche) there was no clinical improvement. As our patient was significantly visually handicapped, immunosuppressive therapy with corticosteroids was considered to be justified. Following initial incremental doses, a period of 3 months on alternate-day dosage of prednisolone 50 mg resulted in gradual improvement followed by complete resolution of both the ptosis and the ophthalmoplegia.

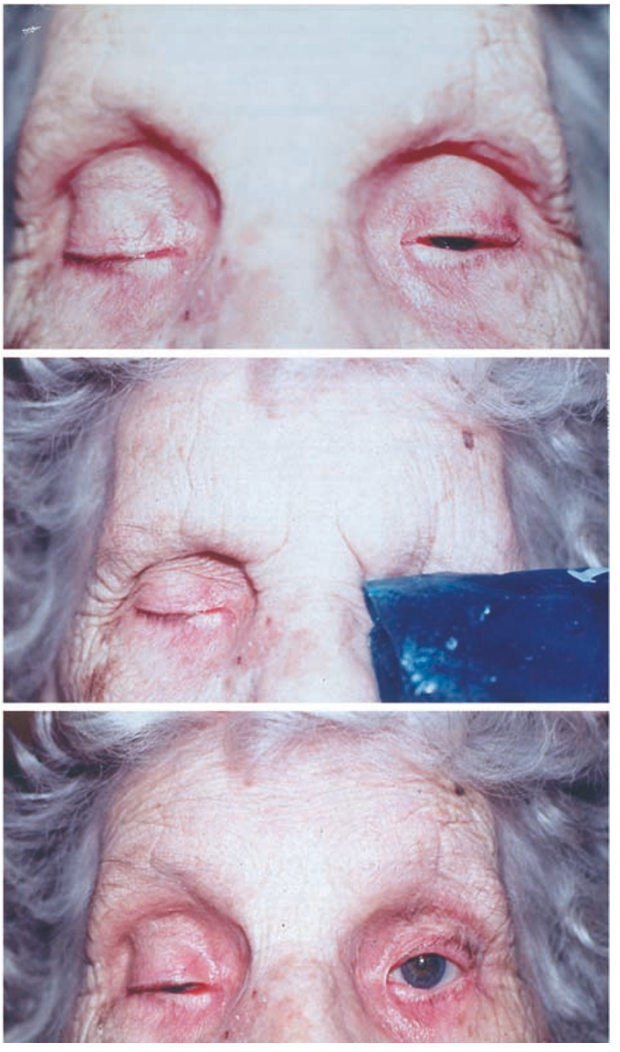

Figure 1 Top: Bilateral marked ptosis. Centre: An ice pack is applied to the left incomplete ptosis. Bottom: A positive ice test with complete reversal of the left ptosis.

Comment

While the generalised form of myasthenia (MG) can present in an acute fulminant form[2] it is most unusual to see a case of ocular myasthenia of such rapid and severe onset. Our case of acute bilateral ophthalmoplegia caused by ocular myasthenia differs from those reported by Keane[3] in that in only one of

his cases was the onset within 24 h and in none did the ophthalmoplegia result in bilaterally fixed globes as was the case here. The presence of severe clinical signs of abrupt onset appearing for the first time on waking, combined with a lack of history of fluctuating weakness and the absence of fatiguability or variability of the myopathy on clinical testing, led to a delay in correctly diagnosing this patient. In retrospect the absence of both the history and the clinical hallmarks of the disease was a function of the extreme nature of the myopathy. It has been reported that severe signs in general[4] but ocular signs[5] in particular may lack diurnal variation.

Extrapolating from reports on the generalised form of myasthenia, acute severe cases are predicted to do badly. In their review of MG, Beghi *et al*[6] found that the outcome of myasthenia was directly related to the severity of the disease at presentation. In general there was a poor response to anticholinesterases but a good response to immunosuppressive therapy, as was the case here. This patient's good response to steroids was also a reflection of her age as well as the purely ocular nature of her signs.[5]

It is possible that the atypical presentation described here may be a function of this patient's advanced years; to our knowledge she is, at 92, the oldest patient diagnosed with this condition. We know that in older patients the pattern of muscle involvement changes, becoming predominantly restricted to the ocular and bulbar groups, as does their response to treatment.[5] Overall older patients have a less favourable outcome in both ocular myasthenia[1] and MG,[6] but we are not aware of any reports that age, *per se*, increases the severity or rapidity of onset of the disease.

In the presence of a marked myopathy the sensitivity index of the ice test is reduced. When levator function is severely affected, cooling the muscle may not produce enough improvement in function to be clinically apparent,[7] leading to a false negative result as occurred here on the completely ptotic right lid.

Acute severe ocular myasthenia should be considered early in the differential diagnosis of sudden-onset complete ophthalmoplegia with ptosis, even in the absence of the characteristic clinical hallmarks of the disease.

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