SL Evans[1], N Kumar[2], MH Rashid[2] and DS Hughes[2]

[1]SpR Singleton Hospital, Swansea, UK

[2]Royal Gwent Hospital
(Cardiff Road) Newport, UK

Correspondence: DS Hughes
Tel: +44-1633-238443
E-mail: anfiander@doctors.net.UK

Sir,

A novel temporary treatment remedy for blepharospasm

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Blepharospasm is an involuntary spasmodic bilateral eyelid closure that may be accompanied by dystonia of the facial, oromandibular, and cervical muscles.[1] It is associated in 7% of cases with eyelid apraxia,[1] a nonparalytic motor abnormality characterised by difficulty in initiating the act of elevation in the absence of sensory loss or ataxia.[2,3]

Blepharospasm can be a debilitating condition that may render some patients functionally blind. We present a case of blepharospasm associated with apraxia of eyelid opening and describe a new treatment approach that proved to be an excellent temporising measure resulting in significant improvement in visual function.

Case report

A 45-year-old man was referred to the eye department with a 3-year history of episodic weakness and muscle pain in both eyelids. This had become much worse 3 months prior to presentation. His eyelids would undergo spasmodic closure, but after the spasm subsided, he would still be unable to spontaneously open them (Figure 1a). He could, however, prise them open with his fingers. He also reported that if he held his eyelids open with his fingers, they would remain open for a few minutes during which spontaneous closure was not possible. These symptoms were typically worse in the morning and got worse during the day.

Magnetic resonance imaging of the brain and muscle biopsy was normal, and investigations for myasthenia gravis were negative. A diagnosis of blepharospasm with an apraxia of eyelid opening was made.

Initial treatment with 70 U of Botulinum Toxin type-A (BTA) (Botox) injected into the eyelids (60 U laterally and 10 U medially) had a positive therapeutic effect but lasted only 3 weeks. Another injection of Botox using 80 U was carried out but was ineffective. This was followed by 200 U of Dysport, which still made little difference to his symptoms. All injections were given by the usual subcutaneous approach and no preseptal injections were administered.

At this time, it was felt that any further increase in dosage would only lead to more side effects, as he was a poor responder. In an attempt to improve his eyelid opening, he was referred to the contact lens clinic within the next 3 weeks where he was fitted with scleral contact lenses with ptosis props (Figure 1b). This resulted in marked improvement of his symptoms and he is now able to maintain binocular visual function consistently. Although, he was given another injection of Dysport within the next 3 months of the contact lens fitting, he did not report any unusual benefit following which further BTA therapy was withheld. His improvement was to such a degree that he has resumed his full-time occupation as an engineer. This treatment has been used as a short-term measure while the patient has been referred to a specialist oculoplastic service to be considered for an eyelid myectomy procedure. His follow-up period after the contact lens fitting was 10 months and is still being followed up at Hull Royal Infirmary.

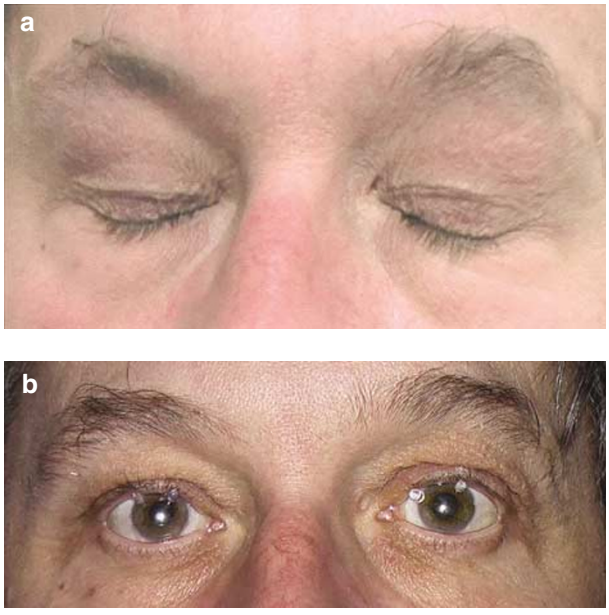

Figure 1 (a) Photograph showing inability to spontaneously open both eyes after an episode of spasmodic closure. (b) Eyelid appearance with scleral lens and ptosis props *in situ*. The pegs on the front surface prevent spontaneous eye closure and facilitate lens handling.

Comment

It is important to recognise the association of blepharospasm and eyelid apraxia because the implications for successful treatment of both conditions may vary when they coexist in the same patient.[1,2] Although they are both preceded by an increased frequency of blinking, patients with blepharospasm have forceful orbicularis spasms following which the lids spontaneously open. However, in patients with apraxia, the lids will not open even in the absence of any orbicularis spasm; instead, the brows are elevated secondary to frontalis overaction.[2] Although patients with apraxia of eye opening often complain of involuntary lid closure, the problem is actually in overcoming the levator inhibition. It is a transient phenomenon, which can cause backward thrusting of the head to interrupt the inhibition of levator muscles, following which lids open suddenly.[4]

Anderson *et al*[1] have proposed a defective circuit theory to explain the pathogenesis of blepharospasm: an afferent arm with impulses travelling via the trigeminal nerve, a control centre in or near the basal ganglia, and an efferent arm via the facial nerve to the eyelid protractors (orbicularis, corrugator superciliaris, and procerus). Treatment can be aimed at all arms of the circuit, but is predominantly directed at the efferent arm.

The two most successful treatment options for blepharospasm are BTA and myectomy of the eyelid protractor muscles.

BTA is effective in up to 86% of patients with blepharospasm, but the effect lasts less than 4 weeks in 13% of responders.[1] It does not treat the eyelid apraxia component when this is present. Approximately 50% of patients with blepharospasm and eyelid apraxia show no therapeutic response to BTA.[1] The reason for this is unknown, but may be related to immune-mediated phenomena.[5,6] Myectomy has been reported to be successful in relieving symptoms in 88% of these cases, but more than one operation is sometimes necessary.[1,2]

Hirayama *et al*[7] have reported the beneficial effects of goggles in patients with apraxia of the lids. Eyelid opening was improved in two patients with apraxia from Parkinson's disease. They suggested that the improvement resulted from the additional proprioceptive input leading to a modulation of dystonic impulses from the basal ganglia.

We have been able to treat the apraxia component of this condition using a scleral contact lens. In theory, this is possible because of the increased proprioceptive input as a result of the eyelids resting on the lens. The lens has also been effective in preventing complete closure of the lids during episodes of blepharospasm. This may be because of the mechanical effect of the lens on lid excursion and also possibly because of reducing stimuli from the afferent arm of the 'defective circuit' as reported by Anderson *et al*.

This treatment has been very successful in providing temporary benefit and restoration of binocular function in this patient with blepharospasm and eyelid apraxia. To the best of our knowledge, this has not been reported before.

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A Salam, AJ Singh, JR Innes and B Melia

Department of Ophthalmology, Hull Royal Infirmary
Anlaby Road, Hull HU3 2JZ, UK

Correspondence: A Salam
Tel: +44 1482 328 541
Fax: +44 1482 605 327
E-mail: aysha_salam@yahoo.com

If there are any queries about the procedures used for designing or fitting the scleral lenses, please write to brian.melia@hey.nhs.uk

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Sir,

Acute angle-closure glaucoma and pupil-involving complete third nerve palsy as presenting signs of thrombosed cavernous sinus aneurysm

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Complete third nerve palsy (CTNP) with a fixed and dilated pupil is in most cases the presenting sign of intracranial aneurysm, usually at or near the junction of