Severe aggravation of blepharospasm in Fisher's syndrome

Sir: Essential blepharospasm has been considered a cranial dystonia caused by a biochemical imbalance of the extrapyramidal system.1,2 With no relevant peripheral nervous system contribution,2 Fisher's syndrome is characterised by external ophthalmoplegia, ataxia and areflexia and represents a limited form of acute idiopathic polynuropathy (Guillain-Barré syndrome). Both processes coincided in a 67 year old male. At 20 years of age he began to suffer occasional involuntary lid closure, more prominent in the right eye, but without disability. These spasms had increased slightly in recent years, often triggered by bright light. After an episode of acute febrile rhino pharyngitis 15 days earlier, over a week he developed severe progressive ataxia, complete external and internal ophthalmoplegia, the eyes remaining in neutral position, ptotic but without diplopia, and general areflexia. Consciousness was normal. CSF showed 0 cells, glucose 0.68 g/l and protein 0.63 g/l. In the next few days, transitory breathing and swallowing difficulties developed, as well as mild weakness of the facial musculature. In this situation of complete ophthal-moplegia, the patient made constant gesticulation due to frequent, occasionally sustained, bilateral blepharospasm attacks. This picture regressed to the previous situation after the ophthal-moplegia resolved some months later.

The severe aggravation of facial spasms in our case was striking, well in excess of what could have been expected from the emotional stress of hospitalisation or appearance of new symptoms. A coincidental relation to an improbable midbrain lesion is purely speculative. A lesion located in the midbrain tegmentum was discovered in only one case of Fisher's syndrome2 and had not been confirmed in other necropsy cases. On the other hand, in only one case of blephar-os- spasm was a well-localised upper brain stem lesion found.4 In our patient, the futility of efforts to counter ocular paralysis and palpebral weakness (m. elevator palpebrae) may have accentuated the actions of antagonist muscles (m. orbicularis oculi). The excess of frustrated central excitation and lack of reciprocal inhibition is considered the EMG pattern of dystonia3 and could explain our case.

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References
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