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 with no relevant peripheral nervous system contribution. 

However, this syndrome is characterised by external ophthalmoplegia, ataxia and areflexia and represents a limited form of acute idiopathic polyneuropathy (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 rhinopharyngitis 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 ocular paralysis, the patient made constant gesticulation due to frequent, occasionally sustained, bilateral blepharospasm attacks. This picture regressed to the previous situation after the ophthalmoplegia 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 syndrome and had not been confirmed in other necropsy cases. On the other hand, in only one case of blepharospasm was a well-localised upper brain stem lesion found. 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 dystonia and could explain our case.

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References


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Orthostatic tremor: diagnostic entity or variant of essential tremor?

Sirs: Heilman1 described three patients with orthostatic tremor, that is, tremor of the legs and trunk which commenced shortly after standing but disappeared when walking, leaning against a support, sitting or lying. Heilman considered orthostatic tremor to be a distinct neurological entity related to the maintenance of static posture.

Certain features distinguish orthostatic tremor from classical essential tremor. The oscillation frequency of orthostatic leg tremor has been reported to be between 14 and 18 Hz,2,3 far higher than the frequency range of classical essential tremor. Furthermore, it has been reported that propranolol, the drug of choice in essential tremor, is ineffective in orthostatic tremor.4 Clonazepam (2–4 mg/day) has been found to be beneficial in orthostatic tremor5 6 but to be of little benefit in essential tremor.7

There is, however, some overlap in the phenomenology of orthostatic tremor and essential tremor. One of Heilman's orthostatic tremor patients had a concurrent postural tremor of the hands and a family history of essential tremor.8 Wee et al8 described a family in which some members had a 7–8 Hz tremor of the hands which was responsive to propranolol, and others had a 6–7 Hz tremor of the legs on standing which was responsive to clonazepam but not to beta-blockers.

We describe a patient with a diagnosis of essential tremor of the hands and legs. Leg tremor, but not hand tremor was successfully treated with clonazepam.

A 53 year old women had a 15 year history of trembling of the legs when standing unsupported. The tremor disappeared on walking or when leaning against a support. There was no tremor at rest or when seated with the legs held outstretched against gravity. For the last 3 years a postural tremor of the hands had been also present. On examination, there were no general or neurological abnormalities other than tremor (see below). In particular there were no cerebellar or Parkinsonian signs. Her father had a 6 year history of hand tremor. Objective (accelerometric) measures showed a 7-0 Hz tremor of the distal muscles of the upper limbs when held outstretched in pronated posture. A 6-4 Hz tremor was present in the proximal muscles of the lower limbs when standing but was absent during sitting and walking. Diazepam had been tried without success. Propranolol (30 mg/day) was ineffective but higher doses were not tried. Primidone initially produced a marked

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