A 63-year-old woman with persistent periocular twitching

Commentary by **JAMES COLEBATCH** MB BS, DSc, FRACP

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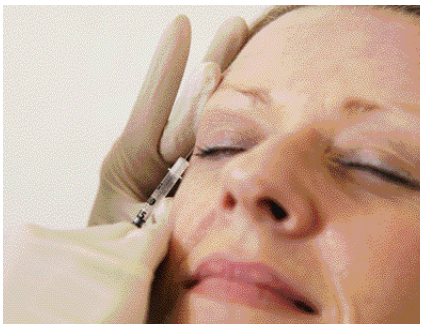

PHOTOLIBRARY

Figure. Injection of botulinum toxin for treatment of hemifacial spasm.

Case scenario

A 63-year-old woman presented with a four-month history of continuing rhythmic twitching underneath her left eye. She could not feel it and was not aware of it unless she observed it in a mirror. The area concerned appeared to be near the point of contact of her spectacles on the inferior margin of her orbicularis oculi muscle. The twitching was regular at about 120 per minute. She was not tired, stressed or taking medication and she did not drink coffee. She had no other health problems, felt well and energetic and did not suffer from any other muscle spasms and cramps.

Why is the twitching happening and can she be treated?

Commentary

Unilateral, persistent periocular twitching in this age group is commonly due to hemifacial spasm (HFS), a condition characterised by involuntary, episodic twitches and spasms of muscles innervated by the 7th cranial nerve. However, involuntary facial movements can have a wide range of other causes.

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History and examination

When taking a history, the patient should be specifically questioned about previous facial weakness (Bell's palsy), as this condition may be followed by synkinesis (overflow contractions) or, rarely, hemifacial contracture. Simple benign fasciculations of orbicularis oculi can occur but are self-limited and unlikely in this patient given the chronicity and stereotyped nature of her symptoms. Facial twitches can occur in patients with focal epilepsy, but this is unlikely to cause twitches of just part of the orbicularis oculi muscle. Blepharospasm affects both eyes and consists of prolonged contractions. HFS rarely causes constant unremitting twitching, at least initially.

During an examination, any associated facial nerve weakness and the presence of any other neurological abnormalities, particularly affecting the 5th or 8th (hearing) cranial nerves, should be noted. If other cranial nerve abnormalities are found, additional investigations will be required. The possibility of ocular myokymia needs to be considered, particularly in a younger patient. Ocular myokymia has a characteristic clinical appearance and consists of undulating or rippling, whereas HFS, in its initial stages at least, consists of sudden, discrete twitches. Ocular myokymia may be symptomatic of serious neurological disorders and needs full investigation.

Episodes of HFS can be brought on by asking the patient to forcibly close the eyes and then relax. In more severe cases, the eyebrow characteristically elevates during involuntary episodes of eye closure (the 'other' Babinski's sign). There is an association with hypertension and the patient's blood pressure should, therefore, be measured. For HFS, few investigations other than cerebral imaging are required. Cerebral imaging is advisable to exclude rare central causes of this condition. Formal electromyography studies are generally not necessary but can confirm HFS by the presence of typical bursts of high-frequency discharges. Mild facial weakness on the affected side may be present. Any pre-existing facial weakness needs to be noted as it will make treatment with botulinum toxin more difficult.

HFS usually progresses unilaterally to involve other facial muscles and the short myoclonic twitches can transform into prolonged spasms. The condition is usually not indicative of other serious pathology, but can be irritating and socially embarrassing. The twitching can make it difficult to read or perform other activities requiring close concentration.

Treatment

Treatment of hemifacial spasm can be by medication, botulinum toxin injections or surgery.

Medication and botulinum toxin injections

A trial of carbamazepine (Carbamazepine Sandoz, Tegretol, Teril) or gabapentin is worth considering, particularly if there is no ready access to botulinum toxin.

Unless there is a very good response to

carbamazepine or gabapentin, botulinum toxin treatment should be considered. The are two preparations of botulinum toxin type A available in Australia – Botox and Dysport – but only Botox is approved under the S100 scheme for the treatment of HFS. Botulinum toxin, the same as that which causes botulism, is taken up by presynaptic nerve fibres. Here it blocks neuromuscular transmission by breaking down SNARE proteins, which are essential for the release of acetylcholine.

The periocular twitching is usually the feature most easy to treat with botulinum toxin and one can be fairly confident of a good result in the patient described. Lower facial involvement is more difficult to treat because satisfactory control of the twitching nearly always leads to some facial asymmetry. This is quite noticeable when smiling and tends to be commented upon. One option, if the patient is concerned by this, is to treat the unaffected side of the face as well. The injections need to be repeated at intervals of three to six months. Patients need to be prepared to receive between two and 20 injections into facial muscles, depending upon severity.

Complications of botulinum toxin include the effects of excessive muscular weakness – specifically ptosis and diplopia. In older patients, lower lid ectropion can limit the dosage given. The development of neutralising antibodies causing secondary failure of treatment is very rare. Anticoagulation does not exclude treatment, as long as the international normalised ratio is within acceptable limits, but does increase the risk of bruising.

Surgery

Surgery, which nowadays means microvascular decompression, is also a possibility. The pathology underlying HFS in most cases is thought to be the result of vascular irritation of the nerve near where it exits from the brainstem. Microvascular decompression requires a craniotomy and exploration of the 7th nerve exit zone and should only be performed by surgeons with specific expertise. It is difficult to be certain prior to the operation that a responsible artery will be identified. High-resolution MRI scanning should be performed in all patients prior to surgery.

Although surgery is the only permanently curative treatment, even in expert hands it carries a risk of significant morbidity (notably damage to the cochlear or vestibular components of the 8th nerve) and is not always successful. It is not clear who should proceed to surgical treatment. My own practice is to make younger patients aware of this treatment option, but to suggest that they undergo at least a one-year period of treatment with botulinum toxin, during which this treatment can be optimised, before making a final decision. Most patients appear to be satisfied with nonsurgical treatment. MT

DECLARATION OF INTEREST: None.