Treatment of Hemifacial Spasm

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Carlos Singer

Case

The patient is a 36-year-old man, working at high managerial and customer relation levels, who started noticing bothersome contractions of the left lower eyelid. His past medical history included an inguinal hernia, obstructive sleep apnea under control with oral appliance (no obesity), right rotator cuff tear, and dyslipidemia. His only medication was atorvastatin. He had no history of Bell's palsy.

Exam disclosed synchronous brief contractions of the left lower eyelid that would come in flurries in a spontaneous fashion and that would not cause palpebral fissure closure. The rest of his neurological examination was unremarkable.

It was initially thought this abnormal movement represented either eyelid myokymia or early hemifacial spasm. Injections with botulinum toxin were initiated with success. Over the ensuing months, eyelid contractions were associated with partial palpebral fissure closure and a few months later with left mid-cheek contraction.

MRI of the brain including MRA revealed a tortuous and ectatic left distal vertebral artery that impinged and posteriorly displaced the root entry zone of the left cranial nerves 7/8 complex and mild mass effect on the left hypoglossal and left abducens nerves. Neurovascular surgery consultations were obtained, but the patient preferred to continue receiving botulinum toxin injections every 4–6 months, which have provided excellent relief.

Discussion

Hemifacial spasm (HFS) is a syndrome of partial or complete and painless involuntary contractions of one side of the face. The movements come on spontaneously or are triggered by facial movements (smiling, lip pursing) or emotion. The contractions can be sustained (tonic) or brief and repetitive (clonic). HFS can rarely be bilateral (less than 5%), in which case contractions on one side are asynchronous with the ones on the other side.

Prevalence of HFS is 11 per 100,000 persons. HFS affects individuals of all ages with a peak in the middle age. There is female preponderance (M/F: 1/2). The only acknowledged risk factor is hypertension. Rare familial cases have been reported. A less vigorous association has been postulated with migraine and trigeminal neuralgia.

HFS may start as a very restricted phenomenon, such as lower eyelid twitch, as seen in our patient, before it extends to the rest of the face. HFS is a stand-alone movement disorder, and it should not be accompanied by any symptom outside facial symptomatology, and there are no sensory features.

HFS originates in the transition zone of central and peripheral myelin or the more proximal root entry zone of the seventh cranial nerve, where it is being compressed by an aberrant or tortuous loop of a vessel of the vertebrobasilar system. The most common culprit vessels are the anterior-inferior cerebellar artery (AICA), the posterior-inferior cerebellar artery (PICA), and the vertebral artery. HFS, therefore, belongs to the family of neurovascular compressive syndromes that also include trigeminal neuralgia, glossopharyngeal neuralgia, and vestibular nerve compression syndrome (postulated as a potential cause of dizziness and vertigo). The vessel pressure generates ectopic firing in the facial nerve or hyperexcitability of the facial motor nucleus.

Other etiologies of HFS are much less common. They include cerebellopontine angle tumors, brain stem gliomas, multiple sclerosis, and CNS vascular insults. A very small number are familial, but the genetic abnormality is unknown.

HFS requires differentiation from other facial dyskinesias, especially facial synkinesis following Bell's palsy and other facial nerve injuries (trauma, surgery). A key distinguishing feature of HFS is its spontaneous contractions

C. Singer, MD
Department of Neurology, University of Miami School of Medicine, Miami, FL, USA
e-mail: csinger@med.miami.edu

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as opposed to the tight triggering by voluntary facial movements seen with facial synkinesis. HFS mimics include psychogenic hemifacial spasm, craniofacial dystonia (blepharospasm, orofacial dystonia, oromandibular dystonia), facial tic, facial myokymia, hemi-masticatory spasm, and oculomasticatory myorhythmia.

Diagnosis may begin and end with the clinical exam if the clinician feels comfortable with an otherwise typical presentation and where surgical decompression is not being entertained in the near future. Magnetic resonance imaging of the brain can be done if the clinician is concerned about nonvascular etiologies, while brain MRA may pinpoint the culprit vessel and the site of compression. It is not unusual that only at the time of neurovascular surgery can the compression be visualized. Cerebral angiography is only needed if surgery is seriously being considered and is required for surgical planning.

Our patient had clear evidence of vertebrobasilar dolichoectasia specifically of the left vertebral artery. Of interest was his substantially younger age than most reported cases (mean age in the 50s), which raises the question of other predisposing factors (genetic, small posterior cranial fossa) that might be of academic interest to pursue but that would have no bearing on treatment at this time.

Botulinum toxin injections represent the treatment of choice with a high rate of success (85–95%) accompanied by a low rate of reversible complications (ptosis, facial weakness). Repeat injections are usually required every 3–6 months.

I inject subcutaneously in four sites around the involved eye with doses between 1.25 and 5 units of onabotulinumtoxinA (or other neurotoxin equivalents). Two sites are located in the tarsal portion of the upper eyelid, one site in the lateral canthus and one site in the outer half of the lower eyelid. I might also inject additional sites including the corrugator and the midface (zygomaticus complex, nasalis, risorius) as well as the perioral muscles and mentalis (see Fig. 75.1).

Oral pharmacology with carbamazepine, clonazepam, baclofen, and gabapentin is notoriously disappointing. Surgery represents the only chance for a permanent solution. Under general anesthesia, a retrosigmoid craniotomy is performed, and a Teflon sponge is placed between the vessel and the facial nerve. Figures of recurrence are in the low single digits, but there are risks of temporary or permanent deafness and or facial paralysis (also in the single digits).

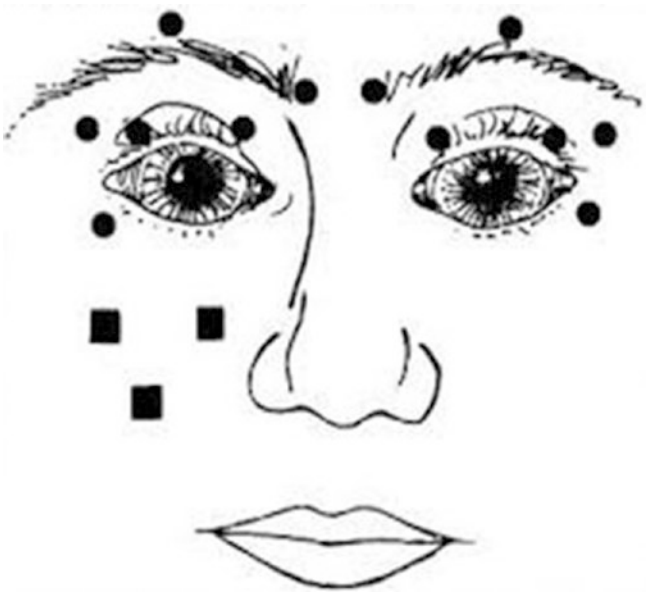

Fig. 75.1 Diagram of possible injection sites for a patient with a right hemifacial spasm. Two sites are located in the right upper lid staying away from the midline. One site is located above the right lateral canthus and another one in the lateral third of the lower eyelid. Other potential sites include the frontalis muscle above the eyebrow, the corrugator, and the midfacial area. The sites on the left side of the face would be considered in rare cases of alternating hemifacial spasm. (Reprinted from Taylor JD, Kraft SP, Kazdan MS, Flanders M, Cadera W, Orton RB. Treatment of blepharospasm and hemifacial spasm with botulinum A toxin: a Canadian multicentre study. Can J Ophthalmol. 1991 Apr;26(3):133–8. With permission from the Canadian Journal of Opthalmology)

Suggested Reading

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