Blepharospasm

Diagnosis

Involuntary facial movements have been recognized for long time and were depicted by artists who were fascinated how these movements distorted the facial expression. For example, in the 16th century, the Flemish artist Brueghel painted a woman with apparent blepharospasm and involuntary jaw opening. Although the eponym "Meige's syndrome" is still occasionally used to designate the combination of blepharospasm (involuntary eye closure) and oromandibular dystonia (involuntary jaw and mouth spasms), a more appropriate term to describe this combination is cranial dystonia. The term dystonia means involuntary sustained, patterned, twisting, pulling or squeezing movements. One of the most common types of adult-onset focal dystonia is blepharospasm. Blepharospasm is a neurological, not an ophthalmological, disorder as there is usually no abnormality with the eyes on careful eye examination. Many patients are initially misdiagnosed as having "dry eyes" but eye drops or lubricants rarely provide any benefit. About one-third of blepharospasm patients report increased frequency of blinking, commonly associated with a feeling of irritation in the eyes and light sensitivity (photophobia). The blinking usually progresses to more sustained contractions of the orbicularis oculi (eyelid muscles) leading to forceful closure of eyelids. Blepharospasm is usually exacerbated by bright light (photophobia) and, as a result, many patients wear sunglasses both outside and inside. The spasms may be transiently alleviated by pulling on an upper eyelid or an eyebrow, pinching the neck, talking, humming, yawning, singing, sleeping, relaxation, reading, concentration, looking down and other alleviating maneuvers. Blepharospasm may vary in severity from only a slightly annoying or embarrassing
condition to a disabling disorder that may render the patient functionally blind and interfere with daily activities such as reading, watching television, and driving. Blepharospasm is seldom an isolated condition; this form of dystonia is often associated with dystonia in other facial, jaw, or neck muscles leading to "cranial-cervical dystonia". In some patients, the dystonia spreads to involve the vocal cords (spasmodic dysphonia), trunk and limbs.

**Cause**
Blepharospasm may be associated to exaggerated neuroplasticity and although usually sporadic (without family history), genetic mechanisms may be involved, as they often are in other forms of dystonia

**Treatment**
Botulinum toxin (BTX) injections into the eyelids and eyebrows are now considered the treatment of choice for blepharospasm, providing moderate to marked improvement in over 90% of patients. The average latency from the time of the BTX injection to the onset of improvement is 2-5 days and the average duration is 3-4 months, after which time the treatment must be repeated in order to sustain the benefit. In addition to the marked functional improvement, there is usually a meaningful amelioration of discomfort and, because of less embarrassment, the patients' self-esteem also frequently improves. Although about 10-15% of all treatment sessions are followed by some side effects (droopiness of eye lid, blurring of vision or double vision, tearing, and local hematoma), the complications only rarely affect patient's functioning and usually resolve spontaneously in less than two weeks. There is no apparent decline in benefit and the frequency of complications actually decreases after repeat BTX treatments. Aproclonidine, an eye drop that can cause contraction of the upper eyelid muscle, can be used as a temporary measure while the BTX effect has not started or when it is wearing off.
Some patients with blepharospasm have partial relief from medications such as clonazepam, trihexyphenidyl, lorazepam, baclofen and tetrabenazine. However, given their side effects and the significant improvement with BTX, oral medications are not frequently used.

In very severe cases in patients with significant functional impairment who have not responded well to botulinum toxin, surgery may be another option. A myectomy, which involves removal of some or all of the muscles responsible for eyelid closure, may be considered. The suspension of the a muscle in the forehead with synthetic threads evaluated in refractory cases with good results in a small study, but the use of this technique in a larger numbers or patients is warranted. The use of deep brain stimulation was also reported in small studies, with success in refractory cases of blepharospasm associated or not with other craniocervical dystonias.

Selected References


four patients and a literature-based analysis of its treatment effects. 

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