

Answer to Ophthalmproblem continued from page 1149

### 3. Benign essential blepharospasm

Benign essential blepharospasm (EB), also called idiopathic blepharospasm, is a form of facial dystonia that involves bilateral contractions of the orbicularis oculi muscle, and other protractors, causing involuntary eyelid closure. The usual presenting symptoms of EB include abnormal, involuntary spasms of the periorcular and facial musculature, resulting in a debilitating disorder known as facial dyskinesia. It is important to note that for certain patients the recurrent, involuntary, and incapacitating nature of the intense blinking renders them functionally blind. The pathophysiology of this disorder remains elusive; however, recent evidence has implicated hyperexcitability of brainstem interneurons secondary to degenerative changes in the basal ganglia as the etiologic factor.<sup>1,2</sup> Essential blepharospasm is usually preceded by eye irritation, photophobia, and increased blinking. Other symptoms that might occur in some patients include facial swelling and stiffness, burning in the mouth, dysarthria or dysphagia, strained or hoarse voice, involuntary jaw opening or closure, nocturnal bruxism, torticollis, writer's cramp, foot inversion, and head or hand tremor.<sup>1</sup> This condition presents predominately in geriatric populations, and might also present with additional neurologic disorders such as essential tremor, parkinsonism, and spasmodic dysphonia.<sup>1</sup> The severity of EB is typically graded on a Jankovic rating scale from 0 (no signs) to 4 (severe incapacitating spasms) in order to characterize the effect of the spasms on the patient's daily functioning.<sup>3</sup>



### Differential diagnosis

Several conditions might present symptoms similar to those of EB. Hemifacial spasm is a myoclonic disorder affecting the muscles innervated by the facial nerve; unlike EB, it almost always presents unilaterally. The pathophysiology might involve an aberrant vessel causing vascular compression of the seventh cranial nerve root, leading to axo-axonal ephaptic transmission and hyperexcitability of the facial motor nucleus.<sup>4</sup> Myokymia of the orbicularis oculi muscle is a benign process presenting in either the lower or upper eyelids of otherwise healthy patients. Facial myokymia differs from benign myokymia in that it is usually unilateral and is characterized by continuous undulating and involuntary movements of the facial muscles as a result of brainstem tumours or multiple sclerosis.<sup>5</sup> In patients with progressive supranuclear palsy, a distinct clinicopathologic entity characterized as progressive parkinsonism that also leads to ocular motility disorders, levodopa has been known to cause jaw-closing mandibular dystonia,

a side effect of EB.<sup>1,6</sup> Essential blepharospasm is often misdiagnosed as keratoconjunctivitis sicca (ie, dry eye syndrome), as both conditions involve symptoms of ocular irritation, photophobia, and blinking. However, keratoconjunctivitis sicca has specific signs, such as blepharitis, decreased tear film, or superficial punctate erosions on fluorescein examination. The symptoms of dry eye often improve with standard treatments such as lubricating eye drops and eyelid hygiene, which have no effect on EB.

### Management

In the past, EB was treated with oral medication, such as benzodiazepines, or with surgery. Currently, however, the preferred first-line management of EB involves an injection of botulinum toxin type A (BtxA) into the periorcular and facial musculature, which is known to be safe and efficacious.<sup>1</sup> Botulinum toxin type A is an effective treatment owing to the high-binding affinity and specificity of the neurotoxin to presynaptic cholinergic nerve terminals, inhibiting acetylcholine release and thereby preventing muscular contractions. Compensatory axonal sprouting begins as early as 4 days postinjection.<sup>7</sup> Partial recovery of function usually occurs 4 to 8 weeks after treatment, at which point the original end plates begin releasing acetylcholine. By 12 weeks, normal function is restored.<sup>8</sup> Although BtxA has been shown to provide substantial relief for patients with EB, the duration of its effects is unknown.<sup>3,9,10</sup> Based on the pharmacology of BtxA, most patients require serial treatments every few months on an ongoing basis.

### Recommendations

Patients with EB should be referred nonurgently to a neurologist or ophthalmologist for assessment and management. These patients might benefit from repeated injections of BtxA to minimize the effects of functional blindness.

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### Competing interests

None declared

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