Guest Editorial

Blepharospasm: Is it really a "Benign" or an "Essential" Condition?

Sameera Irfan, FRCS (UK)
Lahore, Pakistan

Blepharospasm means "an involuntary closure of eyelids". It is a type of dystonia (a movement disorder) which starts as an increased frequency of blinking or eyelid twitching; gradually, it worsens into prolonged spasm of the orbicularis oculi muscle and spreads to the adjacent muscles of the upper and mid-face (Jankovic J, 1984; Fahn S, 1985).

It is a common condition, seen mostly in middle-aged/elderly women with a prevalence of 5 in 100,000 in the general population. It mostly occurs sporadically but may also occur in genetically predisposed individuals. Previously, it was thought to be a psychological condition but now it is considered to be a neuropathological disease.

In the scientific literature, it is often labelled as "benign essential blepharospasm" (Graham R, 2019). The term benign means a "harmless" condition (Mifflin H, 2007) which "does not spread to the adjacent body tissues". So, a benign clinical condition should not produce any harmful effect to the patient, physically or otherwise, or alter their quality of life. But in these patients, long-standing blepharospasm results in brow droop, dermatochalasis, entropion. They suffer from extreme visual disturbances as they are unable to keep their eyes open to perform simple tasks like reading, driving, walking outdoors or perform detail-oriented work; they need more time (and concentrated effort) to finish these tasks. Ultimately, they lose their jobs and are restricted to their homes. This extreme degree of disability affects the patient's family, his/her social life, and psychological well-being to the extent that a few patients even develop suicidal tendencies (Bradley EA, Bradley D, Bartley GB, 2006). So such a debilitating condition cannot be considered or labelled as *benign*!

The term "essential" means "of an unknown cause or idiopathic" (Dorland's Medical Dictionary for Health Care Consumers, 2007). However, various studies show that in 96.8% of such patients, an acute triggering event initiates the frequent blinking (loss of spouse, sibling, divorce), while a secondary trigger (meibomian gland dysfunction, tear film instability and chronic dry eyes) worsens it, resulting in blepharospasm, in an attempt to protect the dried ocular surface and to minimise the irritation. With time, the initial depressive episode (the primary trigger) is worsened by the untreated secondary triggers, and a vicious cycle sets in (Reimer J, Gilg K, Karow A, Esser J, Franke GH, 2005). Because of this misconception that it is an essential condition, an attempt to find an underlying cause is often overlooked; patients suffering from blepharospasm are offered a symptomatic treatment, the focus being mainly on the target tissues i.e. the upper facial muscles. They are rendered paralysed temporarily by Botox injections (Cote TR, Mohan AK, Polder JA, Walton MK, Braun MM, 2005). As the effect of Botox disappears in 4-5 months, the blepharospasm recurs and the patients get even more

depressed. Repeated injections are required which can result in complications like ptosis, lagophthalmos, corneal exposure, apraxia of eyelid opening and ultimately, resistance to the Botox injection.

A multidirectional approach needs to be adopted to manage such patients:

1. Identifying and treating the triggering factors (dry eyes, blepharitis), tinted sunglasses to reduce light sensitivity (photo-oculodynia), anxiolytics / anti-depressants. This needs a joint effort by the ophthalmologist and a psychologist.
2. Educating patients and their caretakers regarding the chronic nature of the disease, the need for long-term therapy, how to avoid the triggering factors, and lifestyle modification i.e. to engage in physical activities that interest them to help them get out of their depressive state.
3. Pharmacotherapy to relieve spasms: anticholinergic or dopamine blocking drugs. These are mainly reserved for patients who do not respond to Botox injections and are less effective than Botox.
4. Surgical intervention in the form of a limited orbicular myectomy can be performed simultaneously for both the upper and lower lids (Irfan S, 2015). This should be performed once the ocular surface inflammation has been controlled by medications and the tear-film stability restored.
5. An alcohol injection to paralyse the facial nerve has been tried but results in complications such as loss of facial expressions, movement and eyelid mal-position. This is not a recommended line of action.

Conclusion

Blepharospasm is a debilitating illness, with severe physical and psychological disability. Therefore, it should not be labeled a "*benign*" condition. Since a definite cause is mostly responsible for triggering it initially and then perpetuating it, it should not be considered an "essential" condition. The terminology *benign essential blepharospasm* should be changed to "*blepharospasm" (*Irfan S, 2018). The treating ophthalmologist should make extra time to evaluate such patients, find the underlying cause, the aggravating factors and treat them. The education of patients regarding their disease and the need for long-term therapy cannot be over-stressed.

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