Treating benign essential blepharospasm: Could it prevent damages on the anterior segment of the eye?

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Abstract

The aim of this review was to determine the importance of the protective role of benign essential blepharospasm for anterior segment of the eye by analyzing changes of anterior segment structures. In this review, we have emphasized the importance of interventional treatment on benign essential blepharospasm with botulinum toxin. We have also determined with the use of fluorescein on the ocular surface if there were any ocular surface defects. Corneal epithelial defects were statistically less pronounced in the interventional group. To conclude, advanced blepharospasm could have a protective effect on the ocular surface.

Keywords: Benign Essential Blepharospasm (BEB), Botulinum toxin, Dry eye syndrome, Fluorescein.

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Introduction

Blepharospasm has been defined as focal dystonia which is most commonly manifested by involuntary; chronic intermittent or persistent, repetitive eye closure, or bilateral tonic spasms. Orofacial dystonia appears due to spasmodic contractions of the orbicularis oculi muscles [1-5]. Less commonly, blepharospasm could even occur as recurrent clonic spasms and in an even lesser percentage it could appear as eyelid apraxia [1,2].

Blepharospasm could be categorized as primary or secondary blepharospasm. Due to genetic research, almost 25% of all blepharospasm cases have had an autosomal dominant inheritance with reduced penetrance in large families. On the other hand, secondary blepharospasm is even less common and is usually caused by multiple brain lesions located f.e. in thalamus, basal ganglia, lower brain stem cerebellum and cortex, nevertheless it could also be an adverse effect of the lid weakness due to facial palsy and myasthenia [6].

Spontaneous remissions could occur in only 10% of patient's cases and lead to an almost sure subsequent exacerbation [2]. Most cases are idiopathic and blepharospasm is generally a life-long disorder [3].

Leading symptom of blepharospasm is functional blindness due to a reduction in visual acuity while the incidence of blinking is increased. Usual patient's complaints which are emphasized are grittiness in the eye, eye dryness, or photophobia. Clinical examination of the anterior segment is usually normal or the fundamental source of patient's problem could be based upon chronic conjunctivitis, keratitis which could go along with iritis [7].

In order to detect a disruption in the ocular surface, fluorescein is being used. Fluorescein staining is enhanced with cell degeneration or cell death, which leads to increased permeability of cell membrane to fluorescein. Therefore, whenever a cell-cell junction is being disrupted, a fluorescein staining will be increased due to a rapid stromal diffusion [7].

Literature Review

Patients' suffering from Benign Essential Blepharospasm (BEB) has been submitted to various types of procedures in order to provide them with a correct form of medical treatment. Even though a variety of medical and surgical therapies have been tested, since 1989, a Botulinum Toxin (BoT), has been recognized as the best treatment procedure for patients suffering from blepharospasm and has therefore became a gold standard in ophthalmological treatment of this facial dystonia [8].

BoT is considered to be a gold standard for the treatment of blepharospasm and the subsequent effect of BoT is almost the same as in synkinesis after facial nerve palsy. The periodic distance between an injection of the BoT should be wisely adjusted according to the patient's needs and could even be shortened for up to six weeks in case the patient's clinical state is worsening due to an eyelid cramping [9].

Contraindications for usage of BoT are a hypersensitivity to the toxin or an infection (at the exact location where BoT injection should have been placed). There are no known drug interactions with botulinum toxin [10].

Although, a treatment with BoT has become a standard procedure when treating many neurological as well as other medical disorders, adverse effects are still possible. Most common adverse effects which have appeared are muscle

weakness, fatigue, symptoms similar to a flu infection, a dry mouth, vertigo and dermatologicas issues (f.e. skin rash) [11].

Even though BoT is being used in treatment of essential blepharospasm, a quality, randomised, controlled efficacy data to support the use of BoT for blepharospasm was being missed for quite a long time. Earlier studies have provided necessary information that BoT is highly effective and safe for treating blepharospasm and have strongly supported its use, since almost 90% of patients who were submitted to this treatment have ended with successful results [3].

In order to support a usage of BoT in blepharospasm treatment, five studies were conducted and have included a total of 854 patients. Four of the mentioned studies have used onabotulinumtoxin A and one study has used abobotulinumtoxin A. During conduction of studies a BoT has been injected in the pretarsal orbicularis occuli muscle. To be even more precise, the preseptal orbicularis occuli muscle was injected in four mentioned studies, and the preorbital muscle in three studies. Jankovic Rating Scale was used in order to evaluate the effect of medical treatment [12]. Jankovic Rating Scale, also known as spasm grading system is a broady used grading system for detecting the severity of movement disorders prior to a medical treatment. It is used prior to an injection of BoT in order to calculate the correct dosage of BoT needed for the patient and to avoid any possible complications that BoT injection could cause.

Pre-treatment evaluation tool used to calculate propre BoT A dose (two observer's observe the spasm of patient's eyes and grade it as following):

- Grade 0-Normal
- Grade 1-Slight disability, no functional impairment
- Grade 2-Moderate disability, no functional impairment
- Grade 3-Moderate disability, functional impairment
- Grade 4-Incapacitated

In order to grade m. orbicularis oculi weakening, assessment of the closure of the eyes is used:

- Grade 0-Incomplete eyelid closure
- Grade 1-Lids just closing, minimal resistance to overcome
- Grade 2-Closing well, some resistance, easily overcome
- Grade 3-Strong closure, can be overcome with difficulty
- Grade 4-Very strong closure, cannot be overcome or overcome with extreme difficulty

Grade 0, 1, 2 are expected to be good outcomes after the BoT treatment [13].

Adverse events after the injection of BoT were transient, and were corresponding to an injection of the BoT dose. Preseptal orbicularis injections led to ptosis more frequently than pretarsal orbicularis injections of BoT [12].

Another study was conducted on seventy-seven patients with blepharospasm unresponsive to other forms of therapy. They have received an injection of BoT A. BoT was injected into the orbicularis oculi muscle, precisely in both upper and lower eyelids and, some patients have even received an injection into the brows and upper part of the face. Orbicularis muscle spasms decreased markedly over the first five days following injection, both in patients with essential blepharospasm and in those with hemifacial spasm. Benefits of this treatment were rapidly seen, although the positive effect was only temporary and required subsequent injections to achieve similar effects. Complications were, as previously mentioned in another study-local and mild but also transient without any side effects [14].

Discussion

Our study was subsequent to all the prior mentioned studies, but with an even more advanced goal. We were interested to determine whether or not treatment of BEB could also have a positive effect on the eyes, more specifically on preservation of the anterior segment of the eye and its normal clinical state. Since BEB is being treated with BoT, our patients were also the ones being treated the same way and the control group were the ones who did not need interventional therapy with BoT. The procedure of our study consisted out of examination of the periocular area for signs of infection or injuries, as well as to determine the stage of blepharospasm, assessment of visual acuity by Snellen optotypes, Goldmann applanation tonometry to exclude subjects with glaucoma or ocular hypertension and slit lamp biomicroscopy. Staining of ocular surface with 1% fluorescein solution was followed by slit lamp biomicroscopy under cobalt blue light. Fluorescein was used in order to detect if there were any defects of the ocular surface represented by punctate dots. Staining with 1% fluorescein solution, which is usually used in staining corneal epithelial defects and mucous fibrils, could also be used in the diagnosis of dry eye, while fluorescein penetrates the disrupted cell-cell junctions [7].

Male patients were excluded and only female patients were included in this study, while a higher female prevalence of blepharospasm has been observed (female to male ratio, 3:1). By inclusion of only female patients in the study and, since the incidence of dry eye syndrome is quite higher in female patients, possible discrepancies in results were avoided due to possible hormonal influence that could have a negative impact on interpretation of results. It was also detected that fluorescein test score was significantly lower in the interventional therapy group and, since fluorescein stains epithelial defects, it is assumed that the level of clinical expression of blepharospasm could even have a protective role on the ocular surface [7].

Not only that BoT is used in treating BEB, it could also be used in eyelid retraction in Graves' disease, induction of protective ptosis and treatment of crocodile tears syndrome after facial nerve palsy [9].

For those patients who are suffering from BEB, that is refractory to BoT, full myectomy is also a recommendable option, although with a higher possibility of long-term swelling, scar contracture, hollow appearance, and unnatural contour of the eyelids as complications and side-effects. These complications could be somewhat lesser if the simultaneous correction of the ptosis would be performed at the same time [15-17].

Conclusion

Treating a BEB with BoT is a non-invasive and safe procedure. Beneficial effect is rapidly noticeable, although temporary and repeated injections of BoT are required for satisfactory outcome. In patients with advanced BEB that are treated with BoT, blepharospasm has a protective role on the ocular surface, clearly noticeable on fluorescein tests, since epithelial defects were statistically significantly less expressed in the interventional therapy group.

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