Treatment of Benign Essential Blepharospasm and Idiopathic Hemifacial Spasm with Vimpat (Lacosamide)
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Abstract

Background: Benign essential blepharospasm and hemifacial spasm are currently treated with botulinum toxin therapy, eyelid protractor myoclonies, microvascular decompression of the facial nerve or pharmacologic therapies including anticonvulsants. Because of limited treatment success in some patients, another treatment option is needed.

Methods: Lacosamide (Vimpat), a recently released, novel anticonvulsant was used to treat one patient with hemifacial spasm and a second patient with both benign essential blepharospasm and hemifacial spasm. Lacosamide selectively enhances the slow inactivation of voltage-gated sodium channels resulting in stabilization of hyperexcitable neuronal membranes and inhibition of repetitive neuronal firing.

Results: Lacosamide provided greater than 90% hemifacial spasm and benign essential blepharospasm in these patients at doses of 200 mg twice per day with sustained symptom relief. Both patients have discontinued botulinum toxin therapy.

Conclusions: Lacosamide provided effective relief in these two patients and may have a role in the treatment of hemifacial spasm and benign essential blepharospasm. The long-term efficacy of lacosamide in hemifacial spasm has yet to be determined.

Key words: hemifacial spasm, benign essential blepharospasm, Vimpat, lacosamide, anticonvulsant

Introduction

Benign essential blepharospasm (BEB) is a syndrome characterized by excessive or continuous bilateral eyelid closure due to spasm of the orbicularis oculi and adjacent muscles. BEB is considered to be a form of focal dystonia caused by basal ganglia dysfunction. Additionally, brain imaging and electrophysiologic studies suggest pathologic changes in excitability in the anterior cingulate, primary and secondary motor areas [1]. BEB is typically a chronic disorder, but up to about 10% of patients may have a spontaneous remission. Most remissions occur within the first 5 years [2]. BEB is often associated with other oromandibular dystonias.

Hemifacial spasm is characterized by a combination of unilateral clonic and tonic spasms of the muscles innervated by the facial nerve. The most common cause of hemifacial spasm (HFS) is now widely recognized as neurovascular contact or compression at the root exit zone of the facial nerve at the lateral pons [3,4]. The movement disorder typically begins in the orbicularis oculi and over the course of years involves the brow, mid and lower face, and neck platysma. The prevalence rate of hemifacial spasm is estimated to be 14.5 per 100,000 in women and 7.4 per 100,000 in men [5,6]. The age of onset is typically between 40 to 50 years of age. Hemifacial spasm, if untreated, is a lifelong condition, and less than 10% of patients experience spontaneous remissions [6]. The medical therapy of choice for HFS is botulinum toxin (BTX).

Patients and Methods

Patient 1

A 52-year-old female occupational therapist presented with a complaint of continuous involuntary twitching of the entire left side of her face. The facial twitching began following a cervical laminectomy procedure in 1999. Initially, only the orbicularis oculi muscle was involved, but the mid and lower face was involved within a month. Treatments with BTX were started in 2000; and these provided temporary partial relief.

Physical examination showed frequent, recurrent spasms of the left side of the face. Contrast MRI of the brain showed a normal brainstem and cranio-cervical junction. Computerized Tomography Angiography (CTA) imaging of the cervical vessels revealed redundancy of the vertebral arteries. Brainstem evoked potential testing showed findings consistent with an ipsilateral lower brainstem lesion between the acoustic nerve and lower pons.

The patient was started on lacosamide on a trial basis for her hemifacial spasm during hospitalization. At a dose of 150 mg twice per day she reported 60 to 70% improvement in her spasms. The lacosamide dose was increased to 200 mg by mouth twice a day, she reported 90%
improvement with deterioration to 70% when under stress. This dose was tolerated well and after one week was increased to 250 mg twice a day. At this dose, the patient reported 99% improvement and was able to sing in the choir again, suck from a straw and whistle without worsening of the spasms. However, at this dose she reported the onset of a hand tremor. The hand tremor interfered with signing her name and texting on her telephone. She also complained that her balance was affected at that dose. When the dose of lacosamide was reduced to 200 mg during the day and 250 mg at bedtime, she experienced significant improvement of the hand tremor and had no hemifacial spasms present the next day. The patient continues to have greater than 90% hemifacial spasm relief.

Patient 2

A 53-year-old female employed as a certified nursing assistant was initially evaluated after being hospitalized for a seizure. Left hemifacial and bilateral blepharospasm were noted during that evaluation. The patient reported that her conditions began in 1999 following a motor vehicle accident and subsequent cervical spine surgery. The symptoms initially involved both eyes. However, the left HFS developed within months. She began BTX treatment in 2002. In addition to BTX therapy, she underwent bilateral protractor myectomies. On months when BTX therapy was omitted she described episodic head shaking that occurred throughout the day and “fender-benders” due to hypersensitivity of her eyes to sunlight. Monthly BTX therapy provided up to 50% symptom relief; but over the past three years that therapy had become less effective.

Initial physical examination showed near continuous twitching of the left face and both eyes. A contrast and non-contrast MRI of the brain showed no abnormalities of the brain or brainstem. The electroencephalogram showed epileptiform discharges in the left posterior temporal and parietal region. Auditory brainstem evoked potentials were normal.

Lacosamide was started as a treatment of her seizure disorder and as an intentional therapeutic trial for the HFS. When taking 100 mg by mouth twice per day, she had no significant change of her BEB and hemifacial spasm, but when the lacosamide dose was increased to 100 mg three times per day, she reported a 60% reduction in blepharospasm and 95% relief of her hemifacial spasm. When the dose was increased to 200 mg twice per day, she had complete blocking of the hemifacial spasm with an estimated 95% blepharospasm inhibition. Due to the extent of her symptom relief since beginning lacosamide she has not returned for BTX therapy. Furthermore, the patient is able to work at a computer and read without holding a book inches from her face. The patient has remained symptom free since beginning lacosamide therapy.

Discussion

Botulinum neurotoxin (BTX) is the medical therapy of choice in BEB and HFS [7,8]. However, the therapeutic effect is sometimes insufficient and repeated BTX injections are often required. Unfortunately, repeated BTX injections have associated limitations. These include high costs and the potential for denervation of the injected muscles. Other treatment options include various oral pharmacologic therapies that have shown limited efficacy [9], or microvascular decompression of the facial nerve [10]. Thus, there is a need for further or alternative treatment options.

Vimpat was initiated because of the success of other antiepileptic drugs (AEDs) in the treatment of HFS [11-16]. Anticonvulsants have been useful for hemifacial spasm most likely due to their ability to inhibit repetitive neuronal firing. However, anticonvulsant therapy is often limited by side effects or a limited response to therapy. Lacosamide is a novel AED licensed as adjunctive therapy for partial-onset seizures with or without secondary generalization [17]. Lacosamide is an agent with a low toxicity profile and has a novel mode of action. It appears to be different from existing AEDs in that it selectively enhances the slow inactivation of voltage-gated sodium channels resulting in stabilization of hyperexcitable neuronal membranes and inhibition of repetitive neuronal firing [18-20]. Lacosamide is generally well tolerated and the most common adverse events are nonspecific central nervous system effects such as dizziness, vertigo, headache and nausea [21,22].

Conclusion

Since lacosamide is not approved by the Food and Drug Administration (FDA) for this condition, treatment of HFS is an off-label use of the medication. Finally, the long-term efficacy of lacosamide in hemifacial spasm has yet to be determined.

Author Contributions

Dr. Gary Mellick provided the case patient, collected reference articles and composed the initial draft of the article. He serves as the corresponding author for this manuscript and its revision.

Dr. Larry Mellick collected reference articles and contributed equally to the writing of the article.

Financial Disclosure Form

Dr. Gary Mellick serves as legal consultant for work related injuries and is retained by Actavis, Lupin, Teva and Sandoz in a joint consulting agreement.

Dr. Larry Mellick is the Editor-in-Chief of Emergency Department Legal Letter and receives a small monthly stipend. He also acts as a consultant in medical malpractice cases.

References

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