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Commentary

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A Short Commentary on Globus Pallidus Internus Deep Brain Stimulation in Primary Meige Syndrome

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Abstract

Meige syndrome is a type of segmental dystonia that manifests a combination of blepharospasm and oromandibular dystonia and is often associated with other types of craniocervical dystonia. Although the precise pathogenesis of primary Meige syndrome remains to be elucidated, it has been suggested that this movement disorder might be a basal ganglia disorder. Multiple single case reports and open-labeled small case series have shown that globus pallidus internus deep brain stimulation (GPi-DBS) could result in therapeutic benefits in patients with severe Meige syndrome. However, randomized and controlled trials are necessary to accurately assess the safety and therapeutic efficacy of GPi-DBS in treating patients with Meige syndrome. This short commentary introduces the current use of GPi-DBS in the treatment of medically refractory Meige syndrome.

Keywords: Meige syndrome; Deep brain stimulation; Globus pallidus internus; Blepharospasm; Craniocervical dystonia; Segmental dystonia

Introduction

Meige syndrome was first described in detail by Henri Meige [1], the French neurologist. This movement disorder is a type of dystonia, and is also known as Bruegel's syndrome [2] and oral facial dystonia. Meige syndrome is characterized by the presence of blepharospasm in combination with oromandibular dystonia and is often associated with other types of craniofacial dystonias [3]. Meige syndrome affects women more often than men (2:1 ratio) and its symptom onset is typically between 30 and 70 years of age. In general, Meige syndrome responds poorly to oral pharmacotherapy [4-6]. A double-blind study [7] has proven that trihexyphenidyl, an antimuscarinic agent, is effective in the treatment of segmental craniocervical dystonias that include Meige syndrome. However, only 1 of 9 patients with Meige syndrome responded to trihexyphenidyl in this study. The United States Food and Drug Administration (FDA) has approved botulinum toxin type A (BTA) as a first-line treatment for blepharospasm and cervical dystonias [3,8]. Since blepharospasm associated with Meige syndrome is usually more severe than isolated blepharospasm, it is difficult to obtain satisfactory results with BTA injections in the treatment of Meige syndrome [8-10]. In addition, BTA treatments have the potential to cause adverse events such as diminished responses, facial weakness, dysphagia, dry mouth, flu-like syndrome, and the development of antibodies against botulinum toxin [8,11-13]. Despite these best medical attempts, intractable disabling dystonias often persist in some patients with primary Meige syndrome.

The precise cause of primary Meige syndrome is poorly understood, however it is thought to be a variant of idiopathic torsion dystonia [14,15]. As in other dystonia syndromes, multiple single case reports and open-labeled small case series have shown that bilateral deep brain stimulation (DBS) of the globus pallidus internus (GPi), which is the major basal ganglia output nucleus, can produce powerful therapeutic benefits in medically intractable primary Meige syndrome [16]. The therapeutic efficacy of GPi-DBS as determined by Burke-Fahn-Marsden dystonia rating scale total movement score (BFMDRS-M) showed an improvement of 64.3% (SD 29.5) [17]. In the selected Meige syndrome patients with longer follow-up periods, that is, for more than 5 years (mean ± SD, 80.7 ± 21.9 months), the therapeutic efficacy of GPi-DBS resulted in an improvement of 70.9 ± 17.1% (mean ± SD), as determined by BFMDRS-M [18-21]. Overall clinical outcomes suggest that GPi-DBS might be more efficient in patients with Meige syndrome than in those with other types of focal and segmental dystonias [17]. The beneficial effects of GPi-DBS in patients with Meige syndrome could be maintained for more than 5 years after surgery. However, the true benefits of GPi-DBS are currently unclear due to the lack of randomized controlled trials in patients with Meige syndrome. The stereotactic coordinates for active contacts of the DBS lead usually are usually located in the posteroventrolateral portion of the GPi [22-26]. Stimulation parameters used in patients with primary Meige syndrome were highly variable with amplitude of 1.0–5.2 volts, frequency of 60–235 Hz, and pulse width of 90–500 μs [16,18-29]. In our experience, a long pulse width (e.g. 450 μs) usually yielded good clinical results [21,27,28]. Using the meta-regression analysis on patients with dystonias that include Meige syndrome, Andrews et al. reported that the only variable showing a trend towards association with improved percentage outcome by GPi-DBS was a shorter time duration between the disease onset and DBS initiation (p=0.06) [17].

GPi-DBS should be considered only if the patients experienced markedly disabling motor symptoms associated with Meige syndrome even though they had received other treatments options, administered by skilled and experienced neurologists [3]. The exclusion criteria for the use of GPi-DBS in patients with Meige syndrome are essentially the

same as those applied to patients with other primary dystonias. They include the presence of brain atrophy and/or other organic lesions, marked cognitive impairment, acute psychiatric changes, severe depression, and other coexisting medical disorders that would increase surgical risk [30]. Perioral tightness [18] and stimulation-induced severe parkinsonism [29] have been reported as un-adjustable complications caused by GPi-DBS in patients with Meige syndrome. Ostrem et al. reported difficulty with coordination and slowness in motor function including worsening of handwriting, typing, balance, and walking in 4 of 6 patients with Meige syndrome [22]. We also should pay a careful attention to depression, which can be a potential risk factor for suicide after GPi-DBS [31]. The parallel organization of functionally segregated cortico-basal ganglia-thalamo-cortical feedback loops invokes the possibility that GPi-DBS could influence motor as well as the other functional loops including associative or limbic loops [32,33]. Ventral two-thirds of the posterior GPi is the primary motor cortex-related territory and dorsal one-third of the posterior GPi is the prefrontal cortex-related territory [34,35]. More dorsal and anterior to the motor cortex-related territory is the supplementary motor area-related territory, while the most medial part of the GPi corresponds to the limbic cortex-related territory [34,35]. For those reason, correct positioning of the active electrodes and careful follow-up assessments of non-motor symptoms including psychiatric conditions after GPi-DBS should be required. In summary, bilateral GPi-DBS could produce a significant improvement of medically refractory dystonia symptoms in patients with Meige syndrome. In the near future, well-designed studies with randomized and controlled trials will be required to establish GPi-DBS as an effective and tolerable treatment option for Meige syndrome.

Conflict of Interest Statement

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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