CASE REPORT / CAS CLINIQUE

SPASMODIC DYSPHONIA MAY RESPOND TO BILATERAL THALAMIC DEEP BRAIN STIMULATION

DYSPHONIE SPASMODIQUE : EFFET BENEFIQUE DE LA STIMULATION THALAMIQUE BILATERALE

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RESUME

Introduction
La dysphonie spasmodique est une dystonie focale primitive caractérisée par une perte de contrôle des muscles de la voix secondaire à un spasme des muscles laryngés. La physiopathologie est mal connue. Une stimulation cérébrale profonde pour d'autres dysphonies focales a été rapportée.
Méthodes
Nous présentons le premiers cas d'une stimulation thalamique profonde bilatérale ayant amélioré une dystonie spasmodique chez un patient atteint d'un tremblement essentiel.
Résultat
Ce cas démontre l'effet bénéfique d'une stimulation thalamique profonde bilatérale à la fois pour le tremblement essentiel des mains et les adducteurs des cordes vocales.
Conclusion
Le mécanisme physiopathologique de cette constatation est discuté.

SUMMARY

Background
Spasmodic dysphonia is a primary focal dystonia manifested by loss of control of the vocal muscles during speech secondary to laryngeal muscle spasms. The pathophysiology is not well understood. Deep brain stimulation surgery (DBS) for other focal dystonias has been well reported.
Methods
We report the first case of bilateral thalamic DBS improving spasmodic dystonia (SD) in a patient with essential tremor.
Results
This case demonstrates the beneficial of effects of bilateral thalamic DBS for both ET of the hands and AdSD of the vocal cords.
Conclusions
The potential pathophysiologic mechanisms of this finding are discussed.

INTRODUCTION

There are few case reports of thalamic deep brain stimulation surgery (DBS) improving voice tremor in essential tremor (ET) patients (6). There are no published reports of DBS improving spasmodic dysphonia (SD) or dystonic tremor associated with SD. We report a case of a patient with ET of the hands and adductor SD with vocal tremor who responded to bilateral thalamic DBS.

CASE MATERIAL

A 72 year old female presented to our institution with a 30 year history of hand tremor diagnosed as essential tremor. She was refractory to medications including beta-blockers, primidone, gabapentin, clonazepam and mirtazapine. She also developed adductor SD at age 64 and received regular botulinum toxin type A (BoNT-A) injections with moderate results. Chronic BoNt-A injections, however, made her voice severely hypophonic and high-pitched for a few weeks following each injection. Just prior to DBS surgery, she had a moderate postural tremor and mild resting tremor of both hands, as well as a moderate adductor SD with vocal tremor.
Bilateral thalamic DBS was performed using the COMPASS® stereotactic system and the Medtroinc® DBS electrodes. The surgical procedure was performed under local anesthesia, with intravenous administration of minimal amounts of conscious sedation and analgesic medications. A magnetic resonance (MR)-compatible frame was applied to the patient's head and MR imaging was performed using a 1.5 tesla machine. The thalamic target was chosen in the ventralis intermedius (Vim) nucleus. The initial target coordinates were targeted based on the anterior and posterior commissures and thalamic height and 11.5 mm lateral to the ipsilateral wall of the third ventricle. Microelectrode recordings were done beginning 20 mm above the thalamic target using 3 concentric bipolar tungsten microelctrodes driven simultaneously by a hydraulic Alpha-Omega ® microdrive at incremental depths of 0.3 to 0.5 mm. Intraoperative electrophysiology was performed to locate and confirm the ventral posterior border of the Vim nucleus adjacent to the sensory nucleus of the thalamus. Microelectrode recordings were followed by microstimulation in order to localize the best depth and trajectory. Subsequent implantation of Medtronic® quadripolar 3387S DBS electrodes was performed. Macrostimulation was done to further refine final electrode placement based upon intraoperative effect. Each DBS electrode was then connected to a Medtronic Soletra® implantable pulse generator (IPG) in an infraclavicular pocket. Postoperative brain MR confirmed placement of the electrode tips in the Vim nucleus of the thalamus in both sides.

RESULTS

Six months following DBS surgery, the patient rated her hand tremors as 100% improved, and her SD 75% improved. Blinded ratings of voice and laryngo-videostroboscopic examinations were performed, comparing stimulators ON versus stimulators OFF. With the stimulators OFF, the voice was rated as moderate SD with

moderate to severe vocal tremor. Videostroboscopy showed intermittent aperiodicity with laryngeal tremulousness and intermittent incomplete vocal cord closure with sustained phonation. With the stimulators ON, the voice was rated as mild SD with no vocal tremor; videostroboscopy showed no laryngeal tremulousness and completed closure of the vocal cords with sustained phonation. Pulse generator settings at 6 months post-DBS were as follows: for the right IPG, settings were contact 3(+), contact 0(-), 2.3 volts amplitude, 90 microseconds pulse width, and 130 Hz frequency; for the left IPG, settings were contact 2(+), contact 0(-), 3.2 volts amplitude, 90 milliseconds pulse width, and 130Hz frequency. Since DBS, the patient has not required any oral tremor medications nor vocal cord injections of BoNT-A.

DISCUSSION

Spasmodic dysphonia is considered a primary focal dystonia. Involuntary laryngeal muscle spasms resulting in loss of voluntary control of the vocal cords during speech production is the hallmark of the disease (2). The disorder most commonly presents as the adductor type (AdSD) manifested by spasmodic bursts especially during vowel pronunciation (4,7). Our patient presented with AdSD. The focal dystonias are generally believed to be due to basal ganglia abnormalities, however, the pathophysiology of spasmodic dysphonia is poorly understood (1,5). Recent findings by Simonyan et al. using a combined diffusion tensor imaging with neuropathological study have suggested altered microstructural integrity of the corticobulbar and corticospinal tracts (9). They postulate that the findings may represent the primary neurological changes in spasmodic dysphonia. Direct projections from the laryngeal motor cortex to the phonation nuclei (nucleus ambiguus) occur via the corticobulbar and corticospinal pathways. The putamen receives input from the laryngeal motor cortex which it in turn relays back to the laryngeal motor cortex via the globus pallidus and ventral lateral thalamus (9). This forms part of the striato-pallido-thalamo-cortical loop. The microstructural abnormalities identified by Simonyan and colleagues could play a role in affecting the voluntary laryngeal control in patients with spasmodic dysphonia (9).

Klostermann et al. identified the effect of subthalamic nucleus (STN) deep brain stimulation on the development of dysarthrophonia in Parkinson's Disease patients (6). They postulate the dysarthrogenic effects following bilateral STN DBS arise from effect on the adjacent structures. It is estimated that the current spread from a DBS electrode is approximately 3 mm depending upon the voltage of the stimulator settings (3,8). Perhaps the effect noted in our patient is the result of such current spread or downstream effect along the striato-pallido-thalamo-cortical loop identified by the studies of Simonyan (9). This hypothesis needs to be confirmed with further studies.

CONCLUSIONS

This case demonstrates the beneficial of effects of bilateral thalamic DBS for both ET of the hands and AdSD of the vocal cords. Whether thalamic DBS is a reasonable treatment option for isolated AdSD or AdSD associated with ET requires further study.

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