

Case Report

Thalamic Deep Brain Stimulation for Writer's Cramp

Chul Bum Cho, M.D., Hae Kwan Park, M.D., Kyung Jin Lee, M.D., Hyoung Kyun Rha, M.D.

Department of Neurosurgery, Catholic Neuroscience Center, St. Mary's Hospital, The Catholic University of Korea College of Medicine, Seoul, Korea

Writer's cramp is a type of idiopathic focal hand dystonia characterized by muscle cramps that accompany execution of the writing task specifically. There has been renewed interest in neurosurgical procedures for the treatment of dystonia over the past several years. In particular, deep brain stimulation (DBS) has received increasing attention as a therapeutic option for patients with dystonia. However, to date, limited reporters made investigations into DBS in relation to the Writer's cramp. In this case, unilateral Vento-oralis complex (Vo) DBS resulted in a major improvement in patient's focal dystonic movement disorders. Her post-operative Burke-Fahn-Marsden Dystonia Rating (BFMDR) scale demonstrated 1 compared with pre-operative BFMDR scale 4. We conclude that thalamic Vo complex DBS may be an important neurosurgical therapeutic option for Writer's cramp.

KEY WORDS : Writer's cramp · Deep brain stimulation.

INTRODUCTION

Dystonia is the term for a set of disorders characterized by abnormal postures and unwanted muscle spasms that interfere with motor performance⁷⁾. By definition, writer's cramp is a task-specific focal hand dystonia. It is characterized by an abnormally tight grip while writing with progressive difficulty in performing the task as writing continues. Symptoms appear at a mean age of 38 years and may be painless or accompanied by painful hand and forearm cramping¹⁶⁾. Focal hand dystonias are relatively common, but less frequent than focal dystonia of the neck or eyelids. Remissions are uncommon, and symptoms can progress to the other hand¹⁵⁾. Non-surgical treatment included oral medications, sensorimotor training, immobilization, physical rehabilitations and botulinous toxin injections, but the symptom recurrence is the rule.

Siegfried introduced firstly stereotactic thalamotomy as a treatment for writer's cramp¹⁸⁾. After that, successful treatment by thalamotomy was reported in patients with writer's cramp^{1,6,19)}. Recently, deep brain stimulation (DBS) appears

to be a safe and effective therapeutic option for dystonia. We report a case of writer's cramp treated successfully by thalamic stimulation.

CASE REPORT

A 36-year-old female suffered from right hand tremor and dystonia that were worsened especially during the writing task. Her sensory abnormalities including deficient graph-esthesia and spatial discrimination ability preceded onset of focal hand dystonia. Her family history and genetic abnormalities, including DYT6, DYT7, DYT13, and abnormalities linked to chromosome 18, were unremarkable. Her physical and mental condition was normal. Computed tomography (CT), magnetic resonance images (MRI) and functional MRI of brain revealed no abnormal finding. Other laboratory investigations demonstrated normal results. Electromyography did not showed finding of peripheral neuropathy such as ulnar neuropathy. The causes of secondary dystonia were excluded as far as possible. She has been treated through drug medication for one year, but drug adverse effects made having medication difficult.

Burke-Fahn-Marsden Dystonia Rating (BFMDR) scale was used to evaluate patients' pre-operative and post-operative neurological conditions³⁾. This handwriting scale is set such that normal writing receives a score of 0; slight difficulty, 1; almost illegible writing, 2; illegible writing, 3; and unable to grasp to maintain a hold on a pen, 4. Evaluations

• Received : October 9, 2008 • Revised : December 23, 2008
• Accepted : June 29, 2009
• Address for reprints : Kyung Jin Lee, M.D.
Department of Neurosurgery, Catholic Neuroscience Center, St. Mary's Hospital, The Catholic University of Korea College of Medicine, 62 Yeouido-dong, Yeongdeungpo-gu, Seoul 150-713, Korea
Tel : +82-10-8758-1826, Fax : +82-2-786-5809
E-mail : rhalee@catholic.ac.kr

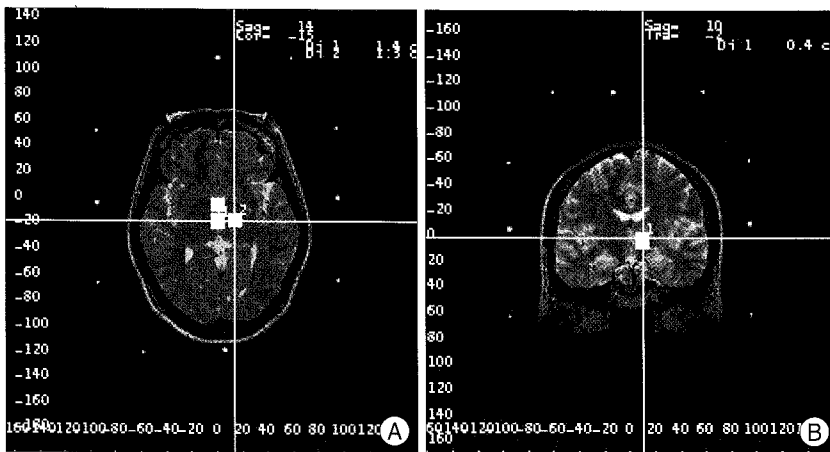


Fig. 1. Preoperative magnetic resonance image guided visual targeting of Vo (nucleus ventrooralis) showing (A) axial and (B) coronal T2-weighted images.



Fig. 2. Letters written by the patient (A) preoperatively, (B) during Vo stimulation, and (C) at 3 month after surgery. She showed remarkable improvement of writing after thalamic stimulation.

were performed using this scale preoperatively, intraoperatively and at 1 week and 3, 12 months after the surgery. Her pre-operative BFMDR scale demonstrated 4 (unable to grasp to maintain a hold on a pen).

We performed left thalamic DBS for right focal hand dystonia. Under local anesthesia, a Leksell G head frame (Elekta Instruments) was fixed to the patient's skull. MRI (1-mm-thick slices) were obtained and the stereotactic target was chosen at 2 mm posterior to the mid-intercommisural point, 0.5 mm dorsal to the intercommisural line and 13.5 mm lateral to the midline (Fig. 1).

According to Schaltenbrand-Wahren atlas¹³⁾, MRI guided visual targeting and intra-operative microelectrode recording, we confirmed the accurate targeting point for ventral thalamic nucleus (Voa : nucleus ventrooralis anterior, Vop : nucleus ventrooralis posterior). All the operative procedures were performed under local anesthesia. A DBS electrode (Model 3387, Medtronic, Minneapolis, MN, USA) with four contact points was placed through the frontal burr hole into the thalamic Vo complex (Voa and Vop). During operation, we confirmed remarkable improvement of writing by thalamic stimulation (2.3 V, 60 usec, 130 Hz). Therefore, we decided to connect the Vo DBS lead with an implantable pulse generator (Medtronic, Inc.) without test simulation period. After the implantation of DBS lead (Medtronic, Inc.), we implanted a pulse generator (Medtronic, Inc.) at left subclavicular area.

She exhibited no surgery-related complications. Her immediate post-operative BFMDR scale demonstrated as 1 (slight difficulty in writing) compared with pre-operative BFMDR scale 4. Her BFMDR scale was 1 at 3 and 12 months after the surgery (Fig. 2). Post-operative brain CT revealed the accurate electrode implantation for pre-operative targeting point (Fig. 3).

DISCUSSION

The dystonias can be classified according to the body part affected. The focal dystonias affect an isolated body region such as the neck (cervical dystonia), eyes (blepharospasm), hand (writer's cramp), or larynx (spasmodic dysphonia). Writer's cramp and laryngeal dystonias are the most common forms of task-specific dystonias.

The pathophysiology of writer's cramp remains unclear. Several speculations have been made. Byl et al.⁴⁾ suggested

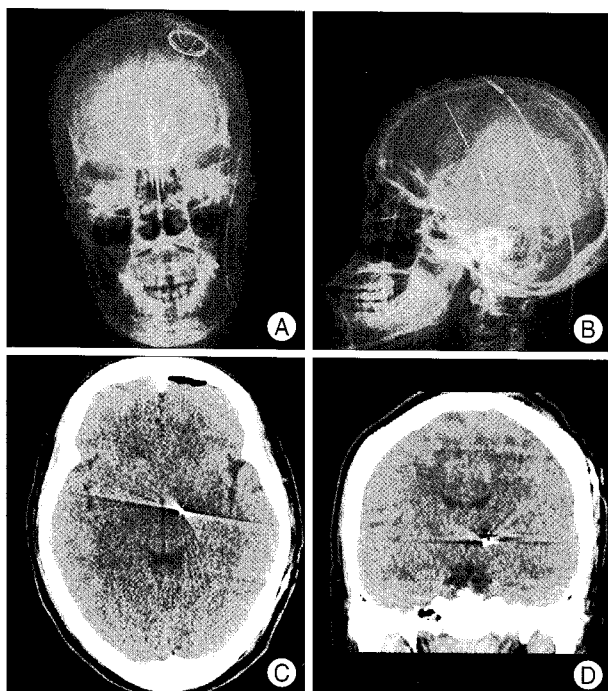


Fig. 3. Post-operative (A and B) simple radiographs and (C and D) computed tomography scans demonstrate proper location of lead.

that focal hand dystonia may reflect a maladaptive response of the brain to repetitive performance of stereotyped movements. Kaji et al.⁹⁾ suggested that a disorder of a motor subroutine might exist in the motor cortex-basal ganglia-thalamus-cortex loop in patients with dystonia. Improvement in dystonia after ablation or stimulation in the motor thalamus and GPi may occur as a result of a disruption or normalization of altered pallidal or thalamic output^{11,14,20)}.

Andrew et al.¹⁾ reported that among 55 cases of dystonia treated by thalamotomy, 1 was a patient with writer's cramp. Mempel et al.¹²⁾ reported successful treatment by thalamotomy in three cases of writer's cramp. Goto et al.⁶⁾ reported that thermocoagulation selectively performed in the Vo complex successfully relieved writer's cramp in a young girl. Taira and Hori¹⁹⁾ reported successful results in 12 patients with writer's cramp using this procedure, and pointed out the usefulness of stereotactic Vo thalamotomy. Shibata et al.¹⁷⁾ also reported a case of writer's cramp in which the patient was successfully treated by stereotactic Vo thalamotomy. The target of Andrew et al. and Mempel et al. seemed to be in the nucleus ventrointermedius, while that of Goto et al. and Taira and Hori was the Vo complex., Krause et al.¹⁰⁾ reported satisfactory results of DBS of the GPi in 17 patients with severe dystonia of different causes. Fukaya et al.⁵⁾ reported a case with which they compared the effects of pallidal and thalamic stimulation and insisted that thalamic stimulation appears to be a valuable therapeutic option for writer's cramp more than pallidal stimulation.

To our knowledge, there are only a few reports in the literature of chronic DBS for the treatment of writer's cramp⁵⁾. In comparison with thalamotomy or pallidotomy, DBS offer the advantages of reversibility, adaptability to clinical situations, well toleration and lower incidence rate of postsurgical neurological deficits²⁾. In addition to these advantages, DBS can cover more wide area in neural elements association with focal dystonia than thalamotomy or pallidotomy. Recently, it has been suggested that the pathophysiology of dystonia is implicated with dysfunction not only of the basal ganglia but also of the cerebellum⁸⁾. Thalamic Vo is associated with the pallidal-thalamic pathway, and thalamic Vop with the cerebello-thalamic pathway. Therefore, lesioning such as thalamotomy can not cover thalamic Vo complex because of minimal lesion but DBS can cover thalamic Vo complex because of stimulating of wide area where DBS lead is located. For these reasons, we suggest that thalamic DBS is more suitable therapy for writer's cramp than thalamotomy.

However, Shibata et al.¹⁷⁾ insisted that patients with writer's cramp are usually young, and consider good cosmetic outcome as very important one and so lesion in Vo complex made by coagulation instead of DBS may be sufficient. In fact, only a few case reports of patients with writer's cramp treated by DBS have been published so far. Therefore, we have to consider the various factors of patients before we decide to perform what kind of operation.

CONCLUSION

Thalamic stimulation appears to be an effective and safe treatment for writer's cramp. In regard of targeting area, although more cases need to be studied, we assure that thalamic Vo complex is a valuable therapeutic target for writer's cramp.

References

1. Andrew J, Fowler CJ, Harrison MJ : Stereotaxic thalamotomy in 55 cases of dystonia. *Brain* 106 : 981-1000, 1983
2. Benabid AL, Pollak P, Gao D, Hoffmann D, Limousin P, Gay E, et al. : Chronic electrical stimulation of the ventralis intermedialis nucleus of the thalamus as a treatment of movement disorders. *J Neurosurg* 84 : 203-214, 1996
3. Burke RE, Fahn S, Marsden CD, Bressman SB, Moskowitz C, Friedman J : Validity and reliability of a rating scale for the primary torsion dystonias. *Neurology* 35 : 73-77, 1985
4. Byl NN, Merzenich MM, Jenkins WM : A primate genesis model of focal dystonia and repetitive strain injury : I. Learning-induced dedifferentiation of the representation of the hand in the primary somatosensory cortex in adult monkeys. *Neurology* 47 : 508-520, 1996
5. Fukaya C, Katayama Y, Kano T, Nagaoka T, Kobayashi K, Oshima H, et al. : Thalamic deep brain stimulation for writer's cramp. *J*

- Neurosurg** 107 : 977-982, 2007
6. Goto S, Tsuiki H, Soyama N, Okamura A, Yamada K, Yoshikawa M, et al. : Stereotactic selective Vo-complex thalamotomy in a patient with dystonic writer's cramp. **Neurology** 49 : 1173-1174, 1997
 7. Hallett M : Dystonia : abnormal movements result from loss of inhibition. **Adv Neurol** 94 : 1-9, 2004
 8. Jinnah HA, Hess EJ : A new twist on the anatomy of dystonia : the basal ganglia and the cerebellum? **Neurology** 67 : 1740-1741, 2006
 9. Kaji R, Shibasaki H, Kimura J : Writer's cramp : a disorder of motor subroutine? **Ann Neurol** 38 : 837-838, 1995
 10. Krause M, Fogel W, Kloss M, Rasche D, Volkmann J, Tronnier V : Pallidal stimulation for dystonia. **Neurosurgery** 55 : 1361-1368; discussion 1368-1370, 2004
 11. Lozano AM, Kumar R, Gross RE, Giladi N, Hutchison WD, Dostrovsky JO, et al. : Globus pallidus internus pallidotomy for generalized dystonia. **Mov Disord** 12 : 865-870, 1997
 12. Mempel E, Kuciński L, Witkiewicz B : [Writer's cramp syndrome treated successfully by thalamotomy.] **Neurol Neurochir Pol** 20 : 475-480, 1986
 13. Schaltenbrand G, Wahren W : **Atlas for Stereotaxy of the Human Brain**, ed 2. Stuttgart : Thieme, 1977
 14. Sellal F, Hirsch E, Barth P, Blond S, Marescaux C : A case of symptomatic hemidystonia improved by ventroposterolateral thalamic electrostimulation. **Mov Disord** 8 : 515-518, 1993
 15. Sheehy MP, Marsden CD : Writer's cramp-a focal dystonia. **Brain** 105 : 461-480, 1982
 16. Sheehy MP, Rothwell JC, Marsden CD : Writer's cramp. **Adv Neurol** 50 : 457-472, 1988
 17. Shibata T, Hirashima Y, Ikeda H, Asahi T, Hayashi N, Endo S : Stereotactic Voa-Vop complex thalamotomy for writer's cramp. **Eur Neurol** 53 : 38-39, 2005
 18. Siegfried J, Crowell R, Perret E : Cure of tremulous writer's cramp by stereotaxic thalamotomy. Case report. **J Neurosurg** 30 : 182-185, 1969
 19. Taira T, Hori T : Stereotactic ventrooralis thalamotomy for task-specific focal hand dystonia (writer's cramp). **Stereotact Funct Neurosurg** 80 : 88-91, 2003
 20. Vitek JL, Chockkan V, Zhang JY, Kaneoke Y, Evatt M, DeLong MR, et al. : Neuronal activity in the basal ganglia in patients with generalized dystonia and hemiballismus. **Ann Neurol** 46 : 22-35, 1999