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 Ventro-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 cramp1,6,19). 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 graphesthesia 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 show 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 conditions10). 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
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-intercommissural point, 0.5 mm dorsal to the intercommissural line and 13.5 mm lateral to the midline (Fig. 1). According to Schaltenbrand-Wahren atlas, 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.
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 Hori\(^19\) 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 cramp\(^9\). 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 deficits\(^3\). 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 cerebellum\(^8\). Thalamic Voa 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.

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