Cukurova Medical Journal

Olgu Sunumu / Case Report

Investigation of the Effect of Deep Brain Stimulation Surgery on Mobility, Emotional Status and Quality of Life in a Patient with Dystonia: Case Report

Distonili Bir Hastada Derin Beyin Stimülasyon Cerrahisinin Mobilite, Emosyonel Durum ve Yaşam Kalitesi Üzerine Olan Etkisinin İncelenmesi: Olgu Sunumu

Filiz Altuğ[1], Ayşe Ünal[1], Gönül Kılavuz[1], Erdoğan Kavlak[1], Feridun Acar[2], Uğur Cavlak[1]

[1]Pamukkale University, School of Physical Therapy and Rehabilitation, DENİZLİ
[2]Pamukkale University, Faculty of Medicine, Department of Neurosurgery, DENİZLİ

Cukurova Medical Journal 2015;40(2):358-363.

ABSTRACT

The aim of our study is to examine the effects of deep brain stimulation (DBS) surgery on severity of dystonia, mobility, emotional status, and health-related quality of life (QOL) in patient with dystonia. This study was carried out on a case of 41-year-old female with complaints of neck and arm pain, involuntary contraction for 30 years. GPİ-DBS was applied to the patient bilaterally in 2010. Global Dystonia Scale (GDS) was used to assess the severity of the patient's dystonia, effects on body parts and functions. Beck Depression Inventory (BDI) was applied to detect emotional status. 12- Meter Walking Test (mobility) and Short Form-36 (SF-36) were used to evaluate quality of life. All assessments were repeated prior to surgery, after surgery 1st year and 3rd years. Prior to surgery, GDS was 112. Postoperative 1st and 3rd years, GDS was respectively 69/84. Prior to surgery, the total BDI score was 31. Postoperative 1st and 3rd years, GDS was respectively 28/40. 12-Meter Walking Test results were respectively 18/10/15 sec. for the prior to surgery, postoperative 1st. and 3rd years. General health status and physical function subscales of SF-36, respectively, prior to surgery were 30/75, postoperative 1st year was 60/85 and 3rd years were 20/35. GPI-DBS is a surgical procedure, which has positive effect on mobility, emotional status and QOL, in patient with dystonia.

Key words: Dystonia, Deep Brain Stimulation, Mobility, Emotional Status, Quality of Life.

ÖZET

Çalışmamızın amacı distonili bir hastada derin beyin stimülasyon cerrahisinin mobilite, emosyonel durum ve yaşam kalitesi üzerine olan etkilerini incelemektir. Çalışma, 30 yıldır boyun ve kolda istemsiz kasılma ve ağrı şikâyetleri olan 41 yaşında bayan bir olgu üzerinde gerçekleştirilmiştir. Hastaya 2010 yılında Bilateral Globus Pallidus İnterna (GPİ) yerleşimli derin beyin stimülasyon cerrahisi yapılmıştır. Hastanın distoni şiddetini, etkilediği vücut fonksiyonları ve kısımlarını değerlendirmek için Global Distoni Skoru kullanılmıştır. Emosyonel durumu belirlemek için Beck Depresyon Ölçeği, yaşam kalitesi için Kısa Form-36(SF-36) ve mobilite durumunu değerlendirmek için 12 metre yürüme testi uygulanmıştır. Hastamızda bütün değerlendirmeler cerrahi öncesinde, cerrahiden sonra 1. yıl ve 3. yılda tekrar edilmiştir. Hastanın cerrahi öncesinde Global Distoni Skoru 112 iken cerrahi sonrası 1. yılda 69 ve 3. yılda 84 olarak kaydedilmiştir. Emosyonel durum toplam puanı cerrahi öncesinde 31 iken cerrahi sonrası 1. ve 3. yılda sırasıyla 28/40 kaydedilmiştir. 12 metre yürüme test sonuçları cerrahi öncesinde 18 sn iken, 1. yılda 10 sn

ve 3. yılda 15 sn kaydedilmiştir. Yaşam kalitesi SF-36 alt ölçeklerinden genel sağlık durumu ve fiziksel fonksiyon cerrahi öncesinde sırasıyla 30/75 iken cerrahi sonrası 1.yıl 60/85 ve 3. yılda 20/35 olarak kaydedilmiştir. Distonili bir hastada GPİ derin beyin stimülasyon cerrahisi mobilite, emosyonel durum ve yaşam kalitesi üzerine olumlu yönde etkisi olan cerrahi bir yöntemdir.
Anahtar kelimeler: Distoni, Derin Beyin Stimulasyonu, Mobilite, Emosyonel Durum, Yaşam Kalitesi.

INTRODUCTION

Dystonia is a clinical syndrome characterized by sustained muscle contractions causing twisting and repetitive movements and/or abnormal postures[1,2,3]. Primary generalized dystonia describes patients in whom dystonia is the sole phenotypic manifestation and involves a combination of segmental curial dystonia which spreads to any other segment Primary generalized dystonia causes physical and social incapacity in patients with normal cognitive function[4,5].

Pharmacologic treatments have limited efficacy, and injections of botulinum toxin are useful only in restricted areas (e.g., the face and neck)[6]. Severe forms of dystonia respond poorly to medical treatment. Deep Brain Stimulation (DBS) is a remarkable therapy for dystonia, succeeding when all manner of medications and botulinum toxin injections have failed[7]. Deep-brain stimulation of the globus pallidus (GPİ-DBS) has been used successfully in primary generalized dystonia[8,9,10]. Bilateral pallidal neurostimulation has been shown, in both controlled and open-label studies to improve motor symptoms, motor disability, and quality of life without cognitive or affective adverse effects[11].

The aim of this study was to examine the effects of deep brain stimulation (DBS) surgery on severity of dystonia, mobility, emotional status, and health - related quality of life (QOL) in patient with dystonia.

CASE REPORT

This study was carried out on a case of 41-year-old female with complaints of neck and arm pain, involuntary contraction for 30 years. She started to move her head and neck to the left and backwards involuntarily at the age of nine. After a few years, bending towards the left had occurred on her trunk involuntarily. Therefore, she used various medications, but she could not get enough benefits. She had a right thalamotomy operation in 1997. After it, bending to the left on her trunk was getting lower, but dystonia on head and neck which was on the background cause of intensity of trunk dystonia began to be disturbing. It had been taken poor response to medical treatment. Over the last 13 years, Botox was applied her once every three months. However, after the last few injections the response to treatment began to decrease gradually. In her physical examination, there was a dystonic movement composed of a mixture of lateropulsiyon to the left, retropulsiyon and anteropulsiyon. It was seen external rotation and extansion on right hand and arm while she was writing. Dystonia was not observed on other extremities. Walking was normal. Deep Brain Stimulation of the Globus Pallidus Internus (GPİ-DBS) was applied to the patient bilaterally in 2010.

MATERIALS and METHODS

Patients were assessed prior to surgery and 1st and 3rd years after surgery. The assessments included Global Dystonia Scale (GDS), Beck Depression Inventory (BDI), 12-Meter Walking Test, and Short Form-36 (SF-36). Global Dystonia Scale (GDS) was used to assess the severity of the patient's dystonia, effects on body parts and functions. Beck Depression Inventory (BDI) was applied to detect emotional status. 12-Meter Walking Test was used to assess mobility, and Short Form-36 (SF-36) was used to assess quality

of life. All assessments were repeated prior to surgery, 1st and 3rd years after surgery.

RESULTS

Prior to surgery, GDS was 112. Postoperative 1st and 3rd years, GDS was 69/84, respectively. Prior to surgery, the total BDI score was 31. Postoperative 1st and 3rd years, GDS was 28/40, respectively. 12-Meter Walking Test results for prior to surgery, postoperative 1st and 3rd years, were 18/10/15 sec., respectively (table I). General health status and physical function subscales of SF-36, prior to surgery were 30/75, postoperative 1st year was 60/85 and 3rd year were 20/35, respectively (table II).

Table.1. GDS, BDI and 12-Meter Walking Test Results

	Before Surgery	Postoperative 1st year	Postoperative 3rd year
Global Dystonia Scale	112	69	84
Beck Depression Inventory	31	28	40
12-Meter Walking Test (sec.)	18	10	15

Table.2. Assessment of SF-36 Sub-Parameters

SF-36 Sub-parameters	Before Surgery	Postoperative 1st years	Postoperative 3rd years
General Health	30	45	45
Physical Functioning	75	60	35
Role limitations due to physical problems	25	50	0
Role limitations due to emotional problems	33.33	33.33	0
Social Functioning	50	62,5	65
Bodily Pain	22.5	45	0
Vitality, energy or fatigue	35	62.5	25
Mental Health	24	56	48

DISCUSSION

Deep brain stimulation (DBS) is a viable treatment alternative for patients with dystonia. This disorder has detrimental effects on the severity of dystonia, mobility, emotional status; health related quality of life (HRQoL).

We discussed the effects of deep brain stimulation (DBS) surgery of Globus Pallidus İnternus (GPi) on mobility, emotional status, and

health-related quality of life (QOL) in a patient with dystonia. All assessments were repeated prior to surgery, 1st and 3rd years after surgery. Prior to surgery, GDS was 112. Postoperative 1st and 3rd years it decreased to 69/84, respectively. Prior to surgery, the total BDI score was 31. Postoperative 1st year it decreased to 28 but the 3rd year it increased to 40. 12-Meter Walking Test results were 18 sec at first and it decreased to 10 sec postoperative 1st year and to 15 sec. postoperative 3rd year. General health status and physical function subscales of SF-36 were 30/75 prior to surgery, 60/85 postoperative 1st year, and 20/35 postoperative 3rd year, respectively.

Kiss et al. performed a prospective, single-blind, multicenter study assessing the efficacy and safety of bilateral GPİ-DBS in 10 patients with severe, chronic, medication-resistant cervical dystonia. Two blinded neurologists assessed patients before surgery and 12 months post-operatively using the Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS), Short Form-36 and Beck depression inventory. The TWSTRS severity score, Quality of life as measured by SF-36, and Beck depression scores improved at 12 months post-operatively (p= 0.003)[12,13]. Our results at 12 months are similar to those reported in the literature. Vidailhet et al. investigated the effects of bilateral pallidal stimulation on motor impairment, disability, quality of life, cognitive performance, and mood in a prospective multicentre 3 year follow-up study. The improvement in quality of life (SF-36 questionnaire) observed at 1 year was maintained at 3 years (p=0·05). Relative to preoperative status, the SF-36 questionnaire showed improvements in general health (p=0·02), physical functioning (p=0·008), and pain (p=0·01) after 3 years of follow-up. Mood (Beck depression inventory) was unmodified at 3 years relative to baseline and 1 year. 11 We found similarly results with literature. In a study performed by FitzGerald et al. patients were assessed using the Burke-Fahn-Marsden (BFM) Dystonia Rating Scale prior to surgery, 6 months after implantation and thereafter at 1 year, 2 years and 5 years follow-up. The group showed mean improvements in the BFM severity and disability scores of 43% and 27%, respectively, by 6 months, and this was sustained[14].

Vidailhet et al. assessed 22 consecutive patients with primary generalised dystonia who underwent bilateral GPi-DBS at 3, 6, and 12 months with the Burke-Fahn-Marsden dystonia scale (BFMD) and the SF- 36. There were significant improvements in mean BFMD and disability score at 12 months. The SF-36 showed significant improvements at 12 months in measures of general health (16%), physical function (21%), and vitality (10%). There were no changes in mood or cognition[6]. In a study performed by Kupsch et al. four patients with generalized dystonia and one with segmental dystonia were treated with bilateral and unilateral GPi-DBS, respectively. The BFMD, EuroQol 1, EuroQol 2, and PDQ- 39 were assessed at 3–12 months. There was a 43% (p,0.02). improvement in BFMDS at follow up. The EuroQol 1 and EuroQol 2 improved by 56% (p,0.05) and 400% (p,0.02), respectively. Using a modified PDQ-39 (there was a 65% improvement (p,0.05) in PDQ-39SI. Dimension subscores were not available[15]. In the double blind trial of DBS for 40 patients with segmental or generalized dystonia, there was a significant improvement in the SF-36 physical composite score at 3 months[16].

These results indicate that there is an improvement in the severity of dystonia, mobility, emotional status, and health-related quality of life measures after deep brain stimulation (DBS) surgery of globus pallidus internus (GPİ), especially in the 1st year after surgery in patient with dystonia. Postoperative 3rd year the scores was better than preoperative measures but a little higher than postop 1st year measures.

We believe that; further research in larger samples is needed to make about the effects of DBS surgery on the severity of dystonia, mobility, emotional status, and health-related quality of life

(QOL) in patient with dystonia. And researches which have high level of evidence should be planned.

CONCLUSION

GPI-DBS is a surgical procedure, which has positive effect on mobility, emotional status and QOL in patient with dystonia.

This article is accepted as poster presentation at 18th International Congress of Parkinson's Disease and Movement Disorders. 8-12 June 2014. Stockholm- SWEDEN.

REFERENCES

1. Vercueil L, Pollak P, Fraix V, Caputo E, Moro E, Benazzouz A, et al. Deep Brain Stimulation in the Treatment of Severe Dystonia. J Neurol. 2001;2:695–700.
2. Manji H, Howard RS, Miller DH, Hirsch NP, Carr L, Bahtia K, et al. Status Dystonicus: the Syndrome and its Management. Brain. 1998;121:243–52.
3. Balash Y, Giladi N. Efficacy of Pharmacological Treatment of Dystonia: Evidence-Based Review Including Meta-analysis of the Effect of BTX and Other Cure Options. Eur J Neurol. 2004;11:361–70.
4. Fahn S, Bressman SB, Marsden CD. Classification of dystonia. Adv Neurol. 1998;78:1-10.
5. Fahn S. Concept and classification of dystonia. Adv Neurol. 1998;50:1–8.
6. Vidailhet M, Vercueil L, Houeto JL, Houeto JL, Krystkowiak P, Benabid AL, et al. Bilateral deep-brain stimulation of the globus pallidus in primary generalized dystonia. N Engl J Med. 2005;352:459–67.
7. Thobois S, Danaila T, Polo G, Simon E, Mertens P, Broussolle E. Deep-Brain Stimulation for Dystonia: Current Indications and Future Orientations. Future Neurology. 2014;9:77-87.
8. Benabid AL, Benazzouz A, Hoffmann D, Limousin P, Krack P, Pollak P. Long-term electrical inhibition of deep brain targets in movement disorders. Mov Disord. 1998;13:119-25.
9. Lozano AM, Abosch A. Pallidal stimulation for dystonia. Adv Neurol. 2004;94:301-8.
10. Cif L, El Fertit H, Vayssiere N, Hemm S, Hardouin E, Gannau A, Tuffery S, Coubes P. Treatment of dystonic syndromes by chronic electrical stimulation of the internal globus pallidus. Neurosurg Sci. 2003;47:52-5.
11. Vidailhet M, Vercueil L, Houeto JL, Krystkowiak P, Lagrange C, et al. Bilateral, pallidal, deep-brain stimulation in primary generalised dystonia: a prospective 3 year follow-up study Lancet Neurol. 2007;6:223–29.
12. Kiss ZHT, Doig-Beyaert K, Eliasziw M, Tsui J, Affenden A, Suchowersky O. et al. The Canadian Multicentre Study of Deep Brain Stimulation for Cervical Dystonia. Brain. 2007;130:2879-86.
13. Alterman RL, and Tagliati M. Deep Brain Stimulation for Dystonia: Patient Selection, Surgical Technique, and Programming the Open Neurosurgery Journal, 2011;4:29-35.
14. FitzGerald JJ, Rosendal F, De Pennington N, Joint C, Forrow B et al. Long-term outcome of deep brain stimulation in generalised dystonia: a series of 60 cases. J Neurol Neurosurg Psychiatry.2014; doi:10.1136/jnnp-2013-306833.
15. Kupsch A, Klaffke S, Kuhn AA, Meissner W, Arnold G, et al. The effects of frequency in pallidal deep brain stimulation for primary dystonia. J Neurol. 2003;250:1201–5.
16. Mueller J, Skogseid IM, Benecke R, Kupsch A, Trottenberg T, Poewe W et al. Pallidal deep brain stimulation improves quality of life in segmental and generalized dystonia: results from a prospective, randomized sham-controlled trial. Mov Disord. 2008;23:131-4

Yazışma Adresi / Address for Correspondence:
Dr. Filiz Altuğ
Pamukkale University,
School of Physical Therapy and Rehabilitation
Kinikli Campus, 20070
DENİZLİ
E-mail: fkural@pau.edu.tr

Geliş tarihi/Received on : 19.03.2014
Kabul tarihi/Accepted on: 21.04.2014