RESEARCH ARTICLE / ARAŞTIRMA MAKALESİ

DOI: 10.4274/atfm.galenos.2019.51523
Journal of Ankara University Faculty of Medicine 2019;72(2):209-213

SURGICAL SCIENCES / CERRAHİ BİLİMLER

Stereotactic Radiofrequency Deep Brain Lesioning in Treatment of Dystonia

Distoni Tedavisinde Stereotaktik Radyofrekans Derin Beyin Lezyonları

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Abstract

Objectives: Movement disorders are neurologic diseases that affect speed, quality and feasibility of muscle contractions. Aim of this study is, to assess the surgical outcomes of radiofrequency lesioning through deep brain stimulation (DBS) in a group of patients and comparing these results to the formerly known conventional procedures.
Materials and Methods: This study involved 25 dystonia patients in whom radiofrequency lesioning through DBS procedures were performed between 1997 and 2003. All of the patients were examined by the same neurologist and examined due to UPDRS, Fahn-Burke-Marsden dystonia rating scales pre and post operatively.
Results: Eighteen male (72%) and seven female (28%) patients (range=14-65 years, mean=26.6 years) with diagnosis of general dystonia (n=11,44%), hemidystonia (n=8, 34%) and dystonic tremor (n=6, 24%) enrolled in the study. Twenty patients with secondary dystonia involved 18 cerebral palsies, one multiple sclerosis and one poststroke dystonia cases.
Conclusion: Gpi DBS seems to be a more considerable approach for primary dystonia patients. Radiofrequency deep brain lesioning can be defined as a successful method for secondary dystonia treatment and must be implicated among treatment options.
Key Words: Dystonia, Thalamotomy, Campotomy, Movement Disorders, Stereotaxic

Öz

Amaç: Hareket bozuklukları kas kasılmalarının hızı, niteliği ve fizibilitesini etkileyen nörolojik rahatsızlıklardır. Bu çalışmanın amacı, talamotomi ve kampotomi kombinasyonunun bir grup hasta üzerinde cerrahi sonuçlarının değerlendirilmesini ve daha önce bilinen uygulamalarla karşılaştırılmasıdır.
Gereç ve Yöntem: Bu çalışmaya 1997 ve 2003 yılları arasında talamotomi-kampotomi yapılan 25 distoni hastası dahil edilmiştir. Tüm hastalar aynı nörolog tarafından UPDRS ve Fahn-Burke-Marsden distoni skalalarına göre cerrahi öncesi ve sonrası dönemlerde değerlendirilmiştir.
Bulgular: Çalışmaya 18 erkek (%72) ve yedi kadın (%28) hasta (14-65 yaş, ortalama 26,6 yıl) dahil edilmiş olup genel distoni (n=11, %44), hemidistoni (n=8, %34) ve distonik tremor (n=6, %24) tanıları ile takip edilmişlerdir. Sekonder distoni tanısıyla izlenen 20 hastanın 18'i serebral palsi, bir multiple skleroz ve bir inme sonrası distoni olarak kabul edilmiştir.
Sonuç: Gpi DBS primer distoni hastalarında daha uygun bir uygulama olarak görülmektedir. Talamotomi-Kampotomi kombinasyonu sekonder distoni tanılı hastalarda başarılı bir metod olarak kabul edilebilir ve tedavi alternatifleri arasında değerlendirilmesi gereken bir seçenek olarak bulunmalıdır.
Anahtar Kelimeler: Distoni, Talamotomi, Kampotomi, Hareket Bozuklukları, Stereotaksi

Introduction

Movement disorders are neurologic diseases that affect speed, quality and feasibility of muscle contractions. They were firstly defined by Gowers in 1888 and Oppenheim in 1911 (1,2). Dystonia is characterized by involuntary, repetitive, sustained muscle contractions that result in twisted bending, abnormal torsion movements. Simultaneous contraction of agonist and antagonist muscle groups occur. Two different

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Received/Geliş Tarihi: 05.07.2019 **Accepted/Kabul Tarihi:** 08.07.2019

axes were suggested regarding distonia classification. The first based on clinical signs and onset whereas the second based on histopathologic and genetic issues (3,4).

Dystonia is the third most frequently seen movement disorder following Parkinson and tremor diseases. Due to lack of complete understanding of dystonia pathophysiology, a standard surgical procedure has not been still established.

The aim of this study is to assess the surgical outcomes of radiofrequency deep brain lesioning in a group of patients and comparing these results to the formerly known conventional procedures.

Materials and Methods

This study involved 25 dystonia patients in whom radiofrequency deep brain lesioning procedures were performed between 1997 and 2003 in Ankara University Faculty of Medicine, Department of Neurosurgery. Registered data belonging to these patients were evaluated retrospectively in order to reveal surgical outcomes and group statistics. The most frequent surgical indications were resistant spasticity and tremor due to secondary or primary dystonia. All of the patients were examined by the same neurologist and examined due to Unified Parkinson's Disease Rating Scale (UPDRS), Fahn-Burke-Marsden dystonia rating scales pre and post operatively. Also combined radiofrequency deep brain lesioning operations were performed by the same surgeon in order to provide uniformity.

Patient Distribution and Features

Eighteen male and seven female patients (14-65 years) with diagnosis of general dystonia (n=11), hemidystonia (n=8) and dystonic tremor (n=6) enrolled in the study. Five patients were diagnosed with primary idiopathic dystonia. Twenty patients were classified as secondary dystonia [18 cerebral palsies (CP), 1 multiple sclerosis and one post-stroke dystonia]. Neonatal hypoxic damage, hyperbilirubinemia, encephalitis, trauma, stroke and neurodegenerative diseases were implicated in the etiology of CP cases. Postop follow-up visits were conducted at first week, sixth month and in the end of the first year. Mean follow-up duration was 8.5 years and ranged between 16 years -6 months. Patients were distributed into five groups according to degree of postsurgical improvements yielded in dystonia and tremor scores. Group A involved 40% and more, group B 40-20%, group C 10-20% and group D involved 10% and lesser score improved patients. Patients with lack of any improvement fell into group E. The initial two groups were regarded as highly benefitted from surgery.

Surgical Procedure

All patients were performed computer tomography/magnetic resonance imaging (CT/MRI) examinations prior to frame fixation and target site determination. Computer based brain atlas' images were applied in site targeting. While thalamotomy was performed by 4 mm monopolar basic electrodes, curled tips were used in performance of campotomy. Coordinates of the site to be damaged were defined according to a line between anterior and posterior commissure which has a length of 25-27 mm.

Target coordinates were obtained by indirect Cartesian measuring based on schaltenbrand stereotactic brain atlas. Vim coordinates of thalamotomy were defined as: 4 mm posterior to the midpoint of anterior commissure (AC) and posterior commissure (PC) line and 13 mm lateral and 1 mm superior to AC-PC line. Target sites regarding Forel and zona inserta regions were calculated as 3 mm inferior and 1-8 mm front of thalamic target. Campotomy was performed by curled electrodes. During detecting target coordinates, we administered benzodiapens and some other anxiolitics for sedation. In case of severe tremor we preferred to give propofol and midazolom. Provided determination of target site, injection of local anesthetic, 1% lidocain plus 1/200.000 epinephrine followed by formation of a burr hole, 1 cm front and 3 cm lateral to coronal sutura. Insertion of an isolated stimulator electrode through nucleus ventralis intermedius and Forel, zona insertia regions was carried out under impedans monitorization. Then it was tested gradually from lower (2-4 hz) frequency to the higher (100 hz) limits as a stimulation. Lesion formation has occurred at 42°C and lasted 60 seconds. During this process patients were examined and registered for contralateral tremor, strength, coordination, speech and memory alterations. Patients who provided no occurrence of neurologic deficit have undergone permanent radiofrequency deep brain lesioning. Permanent ablations were performed at 70°C-78°C and have lasted 60 seconds. Neurologic re-examination was performed after lesion formation. In case of residual tremor after thalamotomy, probe location was adjusted to solve tremor. Bipolar stimulation enabled observation of tremor, rigidity and some other irregular movements. Radius of coagulated site was ranging between 3 to 5 mm. Each patient was performed routine MRI examination postoperatively.

Statistical Analysis

UPDRS and FBMD scores were obtained as numeric values. Student's t-test, Mann-Whitney U test and chi-square tests were performed for comparison of pre and post op functional percentages. $P<0.05$ was accepted as significance value.

Results

Eighteen males (72%) and seven females (28%) patients (range, 14-65 years, mean 26.6 years) with diagnosis of general dystonia (n=11, 44%), hemidystonia (n=8, 34%) and dystonic tremor (n=6, 24%) enrolled in the study. Duration of preop

disease suffering period varied between 1.5 years and 29 years (mean 15.6 years). Mean follow-up duration for all patients was 8.5 years and ranged as 16 years -6 months. All patients were performed combination therapy of radiofrequency deep brain lesioning. Two patients (8%) developed non-permanent surgical complications. One had temporary hemiparesis and other had minor asymptomatic cerebral hematoma which resolved during follow-up postoperatively. Five patients who were diagnosed as primary dystonia were rediagnosed as primary idiopathic torsion dystonia. Primary dystonia group consisted of four generalized dystonia and one dystonic tremor cases.

Twenty patients with secondary dystonia involved 18 CP, 1 multiple sclerosis and 1 poststroke dystonia cases. Neonatal hypoxic damage and trauma were the most frequent two causes of CP in our patient group. While 90% of secondary dystonia patients were highly benefitted from surgery, 75% of primary dystonia patients fell in to benefit group A and B. Low level surgical benefit was encountered in a primary dystonia patient. Dorsal Root Entry Zone was performed to a patient due to lack of benefit from combined surgery. Encephalitis, trauma, stroke and neurodegenerative diseases consisted the etiology of CP cases. Surgical success was detected to be 83% in CP cases ($p<0.05$). Overall, highly beneficial surgery outcomes occurred as 80% ($p<0.05$). Surgical benefit percentages regarding dystonia types exhibited 72% for 11 generalized dystonia cases ($p<0.05$), 100% for 6 dystonic tremor patients ($p<0.05$) and 75% for eight hemidystonic patients ($p<0.05$). Contrast to generalized dystonia patients involved in the least beneficial group most beneficial results were seen among dystonic tremor patients. In secondary tremor patients, the pre-op mean tremor score of 2.8 transformed to 1 point, posing a 64.3% improvement post operatively ($p<0.05$). Indicating a 40% improvement ($p<0.05$), mean dystonia score of secondary generalized tremor cases of group A and B transformed from 38.9 to 23.4 postoperatively. When we look to mean dystonia scores of secondary dystonia patients a reduction of 40.8 point to 26.4 point has indicated a 35.5% improvement ($p<0.05$). Regarded as secondary dystonia patients, all hemidystonia cases had an improvement of 54.2% ($p<0.05$) (change from 37.1 to 17 postoperatively). Two of 4 patients with generalized primary dystonia had scores of high success A and B groups. Patients of these A-B groups exhibited a change of mean dystonia score from 56.5 to 44.2, indicating a 22% improvement ($p<0.05$). The only patient with dystonic tremor belonging to primary dystonia group showed a decrease of mean tremor score from 2.5 to 1 which led to 60% improvement ($p<0.05$). Generalized dystonia patients including primary and secondary types developed a mean dystonia score change from 46.5 to 32.9. Improvement in this group was measured as 30% ($p<0.05$). A reduction of mean tremor scores from 2.75 to 1 occurred in six primary and secondary dystonic tremor patients. Score improvement was detected as 63.6% in this group ($p<0.05$). Overall surgical benefit involving all patients was detected to be 43% ($p<0.05$) in the study.

Discussion

Movement disorders are neurologic diseases that affect speed, quality and feasibility of muscle contractions. Many studies have reported an increased cortical activation during dystonia (5). In a study of Ikoma et al. (6), during dystonia, increased M waves of motor-evoked potentials responses belonging to flexor carpi radialis were detected while conduction of transcranial stimulation. This increase has been suggested as enhanced cortical activation during dystonia. Furthermore, a lentiform-thalamic metabolic disassociation has been detected during 18-flourodextrose PET scanning. It is proposed to be an indication of over activation of putamenopallidal inhibitor tract which leads to dystonic diseases (7).

Up to now numerous surgical procedures were defined for primary and secondary dystonia treatments. Cooper became the first performer of thalamotomy in primary dystonia (8). He had performed thalamotomy to 208 generalized dystonia patients between 1955 and 1974 which were reported to yield 69.7% success. In a study involving 29 patients, Tasker et al. (9) obtained 68% surgical success by thalamotomy.

According to literature complications due to thalamotomy in seconder dystonia patients vary between 16% and 47%. Cardoso et al. (10) have reported a complication rate of 35%; most of them were hemiparesis incidents. There is limited data about pallidotomy conducted over secondary dystonia cases. Iacono et al. (11) had reported no benefit from the pallidotomy they have performed on a 24-year secondary generalized dystonia patient. In a similar study, Lin et al. (12) have performed bilateral pallidotomy on 18 seconder dystonia patients. As a result, except minimal improvements about distonic symptoms none of patients could have exhibited improved movement scale assessments.

Thalamic deep brain stimulation (DBS) is implicated as another option for secondary dystonia patients. Vercueil et al. (13) had performed DBS to eight secondary dystonia patients and followed them for 6 months-11 years. Despite occurrence of a minimal functional advancement, these patients could not develop an improvement for Fahn-Marsden scores. Campotomy which is another treatment option for secondary dystonia were reported to be succesfull by Spiegel et al. (14). In 1969 Wycis had performed campotomy to a patient with myoclonus and obtained favorable results (15). Imer et al. (16) studied effects of thalamo sub thalamotomy combinations in treatment of dystonia. There are some debates over DBS treatment in secondary dystonia cases. Due to variable outcomes by DBS expectance of treatment success is not as much as obtained in

primary dystonia cases. Some contraindications of DBS include major depression, drug addiction, demans and mostly psychotic diseases.

In our study 5 primary and 20 secondary patients enrolled and were performed radiofrequency deep brain lesioning. Complying with the literature, most of our secondary dystonia cases were sufferer of CP. In Tasker et al. (9) series mostly encephalitis patients consisted the secondary dystonia group. Our follow-up duration was relatively longer than other studies in the literature. When compared to primary dystonia patients, many studies indicated higher surgical success of radiofrequency deep brain lesioning for secondary dystonia sufferers. Similarly, in our study patients with secondary dystonia exhibited much more clinic improvements than primary dystonia cases. Surgery appeared to be most beneficial for cases with dystonic tremors. As expected, dystonia scores of primary generalized dystonia patients were higher than secondary ones. Long exposure to dystonia disease and formation of contractures were poor prognostic factors regarding surgery. To our knowledge hemidystonia cases are mostly related to trauma incidents. Coherent with the literature half of our eight hemidystonia patients had experienced trauma before. During whole follow-up, we did not encounter a mortality. In our study two patients developed temporary surgical complications; one had temporary hemiparesis and other had minor asymptomatic cerebral hematoma. These complications are more considerable than complications of sole thalamatomy or campotomy. Indirect Cartesian minimum convex polygon calculation of target site by reference of CT/MRI images and calculating the anterior commissure AC and PC coordinates is a conventional method. However, variations related to brain atrophy, ventricular dilatation and hemisphere symmetry may be confounding conditions prior to surgical site calculation. Another limitation for MR-stereotactic imagination is unstability of dystonia patients. Also, magnetic impact may contribute to this instability. Although in awake patient's confirmation of target site by stimulation is frequently applied as a control step, its false positive or negative interpretation may be encountered and requires high attention. While a variety of brain atlases are available for target site estimation, we prefer Schaltenbrand stereotactic atlas in our clinics. Recent computer-based 3D program based on these atlases are capable of morphologic imagination. Since we do not have any evidence about technical superiority of such programs, a long period of time is required for maturation of these applications. Experience of the surgeon is still a major factor in post op success. Radiofrequency deep brain lesioning treatment was a rarely performed method up to now. Surgical outcomes indicate this combination therapy as a safe and considerable method especially in secondary dystonia cases.

Conclusion

Radiofrequency deep brain lesioning treatment was not a routinely performed method before. While experience of the surgeon is still a major factor, advanced neuroradiologic and stereotactic programs enable more precise site targeting and more favorable post op prognosis. At all, Gpi DBS seems to be a more considerable approach for primary dystonia patients.

Dystonia is a severe disease effecting life quality and expectancy which is not completely understood. Stereotactic methods appear to be the most considerable treatment in these cases. Advanced technology and accumulation of literature data should enlight etiology of dystonia and promise new surgical techniques. Radiofrequency deep brain lesioning can be defined as a successful method for secondary dystonia treatment and must be implicated among treatment options.

Ethics

Ethics Committee Approval: This is a retrospective archive study that does not need ethics approval.

Informed Consent: Informed consent was taken from each patient preoperatively.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: A.S., Concept: Ü.E., M.C.A., A.S., Design: Ü.E., A.S., Data Collection or Processing: Ü.E., M.C.A., A.S., Analysis or Interpretation: Ü.E., M.C.A., A.S., Literature Search: Ü.E., Writing: Ü.E., A.S.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

Acknowledgments

This study was abstracted from data of a specialty thesis in neurosurgery at Ankara University School of Medicine, given by the first author (UE).

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