



Case Report

Seven-year resolution of cervical dystonia after unilateral pallidotomy: A case report

Shiro Horisawa, Takakazu Kawamata, Takaomi Taira

Department of Neurosurgery, Tokyo Women's Medical University, Tokyo, Japan.

E-mail: *Shiro Horisawa - neurosurgery21@yahoo.co.jp; Takakazu Kawamata - tkawamata@twmu.ac.jp; Takaomi Taira - ttaira@twmu.ac.jp



*Corresponding author:

Shiro Horisawa,
Department of Neurosurgery,
Tokyo Women's Medical
University, Tokyo, Japan.

neurosurgery21@yahoo.co.jp

Received : 12 September 2022

Accepted : 26 November 2022

Published : 23 December 2022

DOI

10.25259/SNI_840_2022

Videos available on:

https://doi.org/10.25259/SNI_840_2022

Quick Response Code:



ABSTRACT

Background: Reports on the long-term effects of pallidotomy for cervical dystonia remain scarce.

Case Description: We report a case of cervical dystonia successfully treated by unilateral pallidotomy. The patient was a 29-year-old man without past medical and family history of cervical dystonia. At the age of 28 years, neck rotation to the right with right shoulder elevation developed and gradually became worse. After symptoms failed to respond to repetitive botulinum toxin injections and oral medications, he underwent left pallidotomy, which resulted in significant improvement of cervical dystonia and shoulder elevation without surgical complications. At the 3-month evaluation, the symptoms completely improved. The Toronto Western Spasmodic Torticollis Rating Scale score dramatically improved from 39 points before surgery to 0 points at 7-year postoperative evaluation.

Conclusion: This case suggests that unilateral pallidotomy can be an alternative treatment option for cervical dystonia.

Keywords: Cervical dystonia, Globus pallidus internus, Pallidotomy

INTRODUCTION

Cervical dystonia is an involuntary movement disorder, in which involuntary muscle contractions in the cervical musculature cause postural abnormalities.^[5] Surgical treatments that can be performed to treat cases of refractory cervical dystonia after conservative treatments, such as medications and botulinum toxin injections, may illicit minimal response. Surgical treatments for cervical dystonia include selective peripheral denervation, deep-brain stimulation (DBS), and radiofrequency (RF) thermocoagulation.^[19] Among surgical treatments for cervical dystonia, DBS of the globus pallidus internus (GPi), which comprises the output portion of the basal ganglia, is currently the most commonly used and established method.^[20,39] Cervical movements, such as anterior-posterior flexion and lateral rotation, are achieved by combined movements of the bilateral cervical muscle groups. Accordingly, the muscle groups involved in cervical dystonia are thought to involve motor output from the bilateral cerebral hemispheres. Therefore, GPi-DBS for dystonia is, in principle, performed on the bilateral hemispheres. The Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS), which assesses cervical dystonia symptoms on a 0–85 point scale, has a 50–60% improvement with bilateral GPi-DBS.^[14,17,18] Similar efficacy has been reported with bilateral pallidotomy, although limited.^[1,9,10,21,27,34,35] In contrast, reports on

the treatment of cervical dystonia with DBS or pallidotomy to the unilateral GPi are very limited. We report the case of a patient with cervical dystonia that was treated with unilateral pallidotomy and showed complete resolution of cervical dystonia over a 7-year period.

CASE REPORT

A 29-year-old man was diagnosed with cervical dystonia at the age of 27 years after developing symptoms of cervical rotation to the right and consulted a neurologist. He received medications (baclofen 15 mg, trihexyphenidyl 6 mg), but his symptoms were not improved. In addition, he was treated several times with botulinum toxin injections with no response. He was referred to our hospital for surgical treatment. The patient had right cervical rotation of 45–60°, associated neck pain, and elevation of the right shoulder: dystonic symptoms were alleviated by holding the posterior neck with the hand (sensory tricks). On awakening, dystonia symptoms were relatively mild, but gradually worsened from noon to evening (early morning effect). No other movement disorders were observed. Based on the characteristic findings of dystonia, such as sensory tricks and early morning effect, cervical dystonia was diagnosed [Figure 1a and Video 1]. The preoperative TWSTRS score was 39 points.

As the patient's cervical dystonia symptoms were accompanied by symptoms of the right shoulder elevation, selective peripheral denervation was not a treatment option. The patient strongly rejected implantation of the instrument and preferred to undergo RF surgery. As the affected area of dystonia was the right shoulder and right cervical rotation, we decided to perform left pallidotomy based on the symptomatic predominance of involuntary motor output from the left hemisphere. Under local anesthesia, a head magnetic resonance imaging (MRI; T1-axial/T2-coronal image) examination was performed with a Leksell frame, and a surgical plan was made using a Leksell surgiplan (Elekta AB; Stockholm, Sweden). The left GPi was set 19.5 mm

to the left lateral, 3.0 mm inferior, and 2.0 mm anterior to the midpoint of the anterior commissure and posterior commissure. A coagulation probe with a heat-conducting part of 1 mm in diameter and 4 mm in length was used, and a Leksell Neurogenerator (Elekta AB) was used for electrical stimulation and thermal coagulation. Before thermal coagulation, electrical stimulation (130 μ s/100 Hz/2–5 mA) was performed to confirm that there was no proximity to the optic tract or internal capsule. Thermal coagulation was performed at 70°C for 40 s. Then, the electrode was withdrawn in 3-mm increments to increase the lesion size, producing two contiguous lesions. Electrical stimulation and thermal coagulation were also performed 3 mm posterolaterally from the target. Therefore, in total, four lesions were made using two trajectories. The volume of the lesions was measured using a 3D slicer on a T1-weighted MRI performed immediately postoperatively. The measured volume was 99.3 mm³. The first trajectory lesion was confirmed at 19.7 mm lateral, 2.3 mm anterior, and 3.0 mm inferior to the midpoint of AC-PC. The second trajectory lesion was confirmed at 20.5 mm lateral, 0.3 mm posterior, and 2.0 mm inferior to the midpoint of AC-PC.

From the day after surgery, the neck was maintained in the midline position, and right shoulder elevation was improved to the point where there was no difference between the left and right sides [Video 2]. The only remaining symptom was neck pain with a TWSTRS score of 4 points at 1 week postoperatively. There was no postoperative complication. At 3 months postoperatively, neck pain had also improved, and the TWSTRS score was 0 points [Figure 1b]. ¹²³I-IMP-SPECT showed reduced cerebral blood flow of extensive bilateral hemispheres from preoperative [Figure 2a] to 3 months postoperatively [Figure 2b]. Post-operative T1-weighted MRI images showed coagulated lesion in the left GPi on the day of the surgery [Figure 3a] and old scar in the left GPi 3 months

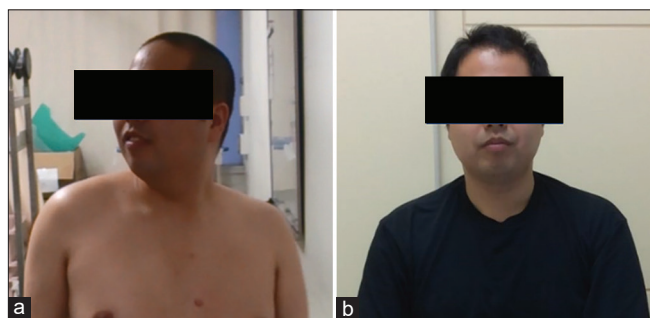


Figure 1: Pre- and post-operative neurological conditions, (a) Neck rotation to the right with right shoulder elevation before left pallidotomy is presented, (b) Complete resolution of cervical dystonia and right shoulder elevation at 3 months after left pallidotomy.



Video 1: Preoperative condition.

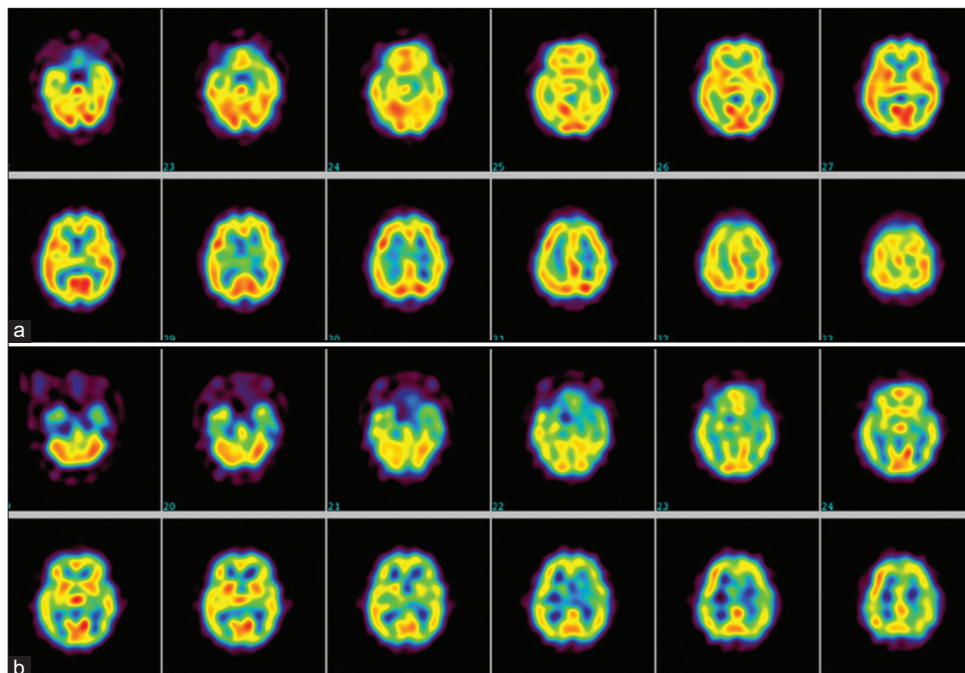


Figure 2: Pre- and post-operative cerebral blood flow (^{123}I -IMP-SPECT), ^{123}I -IMP-SPECT showed reduced cerebral blood flow of extensive bilateral hemispheres from preoperative (a) to 3 months postoperatively (b).

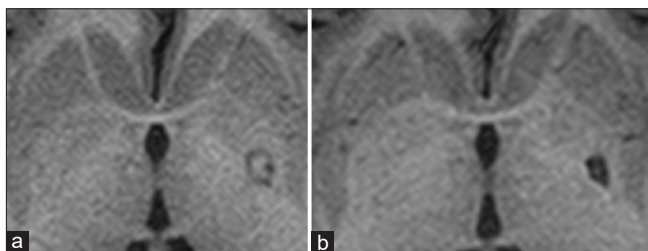


Figure 3: Post-operative T1-weighted MRI (a) showing coagulated lesion in the left globus pallidus internus on the day of the surgery. (b) Showing old scar on the left globus pallidus internus 3 months after left pallidotomy.

after the surgery [Figure 3b]. During the 7-year postoperative follow-up, there was no recurrence of cervical dystonia without medications (TWSTRS = 0), and the patient, who had to quit his job due to the impact of his condition, was able to return to work and has a normal social life [Video 3].

DISCUSSION

In this case, unilateral pallidotomy for cervical dystonia resulted in complete symptomatic improvement over 7 years. This is the report with the longest follow-up period of pallidotomy for cervical dystonia. The current standard surgical treatment is bilateral GPi-DBS, but before the advent of DBS, thermal coagulation surgery using various targets was used in the 1960s–1980s.^[26] In the mid-1980s, after it was reported that dyskinesia and dystonia in Parkinson's disease



Video 2: 1-day postoperative condition.

were dramatically improved by pallidotomy, the GPi became the standard therapeutic target for dystonia.^[6,32] However, the advent of DBS led to the gradual disuse of pallidotomy as a surgical treatment for dystonia.^[2,6] GPi-DBS has been established as a surgical treatment for dystonia with high-quality evidence, including randomized trials.^[39] In contrast, pallidotomy lacks high-quality evidence, and its long-term efficacy and safety remain unclear.

GPi-DBS for cervical dystonia is generally performed bilaterally. Cervical dystonia is often accompanied by



Video 3: 1-year postoperative condition.

symptoms of the left/right rotation/tilting, and involuntary muscle contractions in the cervical muscle groups are often not symmetrical. The condition may also be associated with involuntary movements, such as shoulder elevation, upper extremity dystonia, or tremor, and one of the left or right cerebral hemispheres is often presumed to be the more symptomatically dominant hemisphere. Indeed, interhemispheric differences in neural activity in GPI have been reported to be present in rotational cervical dystonia but absent in symmetrical retroflex cervical dystonia. Abnormalities in cortical excitability and inhibition with transcranial magnetic stimulation have been identified in dystonia cases.^[24,31] In rotational cervical dystonia, these abnormalities are predominantly found in the hemisphere contralateral to the direction of rotation.^[16] Cervical dystonia secondary to stroke, head trauma, or intracranial tumors often results in neck rotation in the opposite direction from the hemisphere of the lesion.^[22,28,33] These facts indicate that in rotational cervical dystonia (asymmetrical cervical dystonia), the neurophysiological abnormalities in the left and right cerebral hemispheres are not equal, suggesting the existence of a “symptomatic hemisphere” predominantly involved in the development of symptoms, similar to our case. Recently, a possible mechanism has been proposed, in which a focal brain lesion causes abnormalities in a network that includes the cerebellum and somatosensory cortex — which are involved in the generation of cervical dystonia.^[3] It is possible that amelioration of the dysfunction of the network involved in the development of cervical dystonia does not necessarily require treatment of the bilateral GPI. In the future, examination of such network abnormalities may be a critical factor in determining which side of the brain to operate on, the left or right.

In the case of such asymmetrical rotator cervical dystonia, there has been no debate concerning whether treatment

should be directed to the bilateral cerebral hemispheres. The effects of unilateral GPI-DBS for rotatory cervical dystonia are scattered in case reports.^[4,15,37] Escamilla-Sevilla *et al.* performed bilateral GPI-DBS for the left rotator cervical dystonia and reported a greater treatment effect with the right GPI-DBS alone (BFMDRS = 94.1%) than with bilateral GPI stimulation (BFMDRS = 41.2% improvement).^[4] Valalik *et al.* performed unilateral pallidotomy on the more symptomatic side for cervical dystonia with blepharospasm and reported a nearly 90% improvement in symptoms after 2 years.^[38] Lai *et al.* performed unilateral pallidotomy in 12 patients with cervical dystonia who were refractory to selective peripheral denervation.^[21] Interestingly, four of the 12 patients showed 100% improvement after evaluation using the TWSTRS at 5 years postoperatively. The treated side was the GPI contralateral to the contracting splenius-complex group.^[21] The surgical results and surgical side determination were similar to those of our patient. Hassler and Dieckmann demonstrated that pallidothalamic stimulation induced the electromyography activity in the ipsilateral sternocleidomastoid muscle.^[7] Based on these results, Hassler and Dieckmann performed unilateral ablation on the Forel’s field where pallidothalamic fibers pass through to treat cervical dystonia.^[7] The present case also had right rotatory cervical dystonia with elevation of the right shoulder; thus, pallidotomy was performed on the left GPI, and the patient has been completely cured for 7 years. The preoperative ¹²³I-IMP-SPECT showed no significant difference of elevated or decreased blood flow between the left and right hemispheres, and the relationship to clinical symptoms was unclear. Postoperative SPECT showed decreased blood flow throughout the bilateral cerebral hemispheres and bilateral cerebellum, with a slight left-sided predominance in the cerebral cortex, suggesting an effect of coagulation surgery on the left GPI. Increased blood flow in the basal ganglia and motor cortex has been noted in dystonia, which may reflect the pathophysiology of this condition of excessive motor output.^[23,36] GPI-DBS for dystonia has been reported to correlate symptom improvement with decreased blood flow, and this was also the case for this patient.

Complications of pallidotomy are known to increase in frequency and severity, especially when performed bilaterally.^[13] Pallidotomy for Parkinson’s disease has been reported to cause dysarthria, dysphagia, and decreased voice volume.^[8,13] Recent reports of pallidotomy for dystonia have reported delayed cerebral infarction posttreatment, postural reflex disturbance, frozen gait, dysarthria, and dysphagia that appear after bilateral pallidotomy.^[10,25] We recently reported that, due to concerns regarding late cerebral infarction, surgery was performed on the pallidothalamic tract (which are output nerve fibers from the GPI to the thalamus) and obtained symptomatic improvement for cervical dystonia similar to unilateral pallidotomy without late cerebral

infarction.^[11,12] If good long-term improvement is achieved after unilateral pallidotomy, as in this case, there is no need to treat the other side, and the risk of bilateral pallidotomy can be avoided. In cases of dystonia, in which unilateral pallidotomy alone does not provide sufficient improvement, a decision should be made as to whether to operate on the contralateral side using RF thermocoagulation or DBS, considering the risk of complications. In principle, RF thermocoagulation surgery is performed after an interval of at least 1 year from the initial coagulation surgery, and the target of treatment on the contralateral side should be different from that of the initial coagulation surgery (pallidothalamic tract). However, patients with decreased voice volume and dysarthria may still present even after this manner. Matsumoto *et al.* recommended that bilateral thalamotomy be performed safely in a staged fashion, with no complications from the initial surgery, a long surgical interval, and an age <70 years.^[29,30] We believe that bilateral coagulation, including bilateral pallidotomy, bilateral thalamotomy, and unilateral pallidotomy with contralateral pallidothalamic tractotomy, may carry a certain risk of dysarthria and reduced voice volume. DBS is always a treatment to be considered in terms of avoiding these complications. In the future, it is necessary to verify which cervical dystonia can be effectively treated with unilateral thermal coagulation alone.

CONCLUSION

This case suggests that unilateral pallidotomy can be an alternative treatment option for cervical dystonia.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

This work was supported by JSPS KAKENHI Grant Number JP21K09113.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

- Boutet A, Madhavan R, Elias GJ, Joel SE, Gramer R, Ranjan M, *et al.* Predicting optimal deep brain stimulation parameters for Parkinson's disease using functional MRI and machine learning. *Nat Commun* 2021;12:3043.
- Cif L, Hariz M. Seventy years of pallidotomy for movement disorders. *Mov Disord* 2017;32:972-82.
- Corp DT, Joutsa J, Darby RR, Delnooz CC, van de Warrenburg BP, Cooke D, *et al.* Network localization of cervical dystonia based on causal brain lesions. *Brain* 2019;142:1660-74.
- Escamilla-Sevilla F, Mínguez-Castellanos A, Arjona-Morón V, Martín-Linares JM, Sánchez-Álvarez JC, Ortega-Moreno A, *et al.* Unilateral pallidal stimulation for segmental cervical and truncal dystonia: Which side? *Mov Disord* 2002;17:1383-5.
- Fahn S, Bressman SB, Marsden CD. Classification of dystonia. *Adv Neurol* 1998;78:1-10.
- Gross RE. What happened to posteroventral pallidotomy for Parkinson's disease and dystonia? *Neurotherapeutics* 2008;5:281-93.
- Hassler R, Dieckmann G. Stereotactic treatment of different kinds of spasmodic torticollis. *Confin Neurol* 1970;32:135-43.
- Higuchi Y, Iacono RP. Surgical complications in patients with Parkinson's disease after posteroventral pallidotomy. *Neurosurgery* 2003;52:558-71.
- Horisawa S, Fukui A, Kohara K, Kawamata T, Taira T. Unilateral pallidotomy in the treatment of cervical dystonia: A retrospective observational study. *J Neurosurg* 2019;134:1-7.
- Horisawa S, Fukui A, Takeda N, Kawamata T, Taira T. Safety and efficacy of unilateral and bilateral pallidotomy for primary dystonia. *Ann Clin Transl Neurol* 2021;8:857-65.
- Horisawa S, Fukui A, Tanaka Y, Yamahata H, Kawamata T, *et al.* Pallidothalamic tractotomy (Forel's Field H1-tomy) for dystonia: Preliminary results. *World Neurosurg* 2019;129:e851-6.
- Horisawa S, Kohara K, Nonaka T, Fukui A, Mochizuki T, Iijima M, *et al.* Unilateral pallidothalamic tractotomy at Forel's field H1 for cervical dystonia. *Ann Clin Transl Neurol* 2022;9:478-87.
- Hua Z, Guodong G, Qinchuan L, Yaqun Z, Qinfen W, Xuelian W. Analysis of complications of radiofrequency pallidotomy. *Neurosurgery* 2003;52:89-99.
- Hung SW, Hamani C, Lozano AM, Poon YY, Piboolnurak P, Miyasaki JM, *et al.* Long-term outcome of bilateral pallidal deep brain stimulation for primary cervical dystonia. *Neurology* 2007;68:457-9.
- İşlekel S, Zileli M, Zileli B. Unilateral pallidal stimulation in cervical dystonia. *Stereotact Funct Neurosurg* 1999;72:248-52.
- Kaňovský P, Bareš M, Streitová H, Klajblová H, Daniel P, Rektor I. Abnormalities of cortical excitability and cortical inhibition in cervical dystonia. *J Neurol* 2003;250:42-50.
- Kiss ZH, Doig-Beyaert K, Eliasziw M, Tsui J, Haffenden A, Suchowersky O. The Canadian multicentre study of deep brain stimulation for cervical dystonia. *Brain* 2007;130:2879-86.
- Krause M, Fogel W, Kloss M, Rasche D, Volkmann J, Tronnier V. Pallidal stimulation for dystonia. *Neurosurgery* 2004;55:1361-8.
- Krauss J. Surgical treatment of dystonia. *Eur J Neurol* 2010;17:97-101.
- Kupsch A, Benecke R, Müller J, Trottenberg T, Schneider GH, Poewe W, *et al.* Pallidal deep-brain stimulation in primary generalized or segmental dystonia. *N Engl J Med* 2006;355:1978-90.
- Lai Y, Huang P, Zhang C, Hu L, Deng Z, Li D, *et al.* Unilateral pallidotomy as a potential rescue therapy for cervical dystonia after unsatisfactory selective peripheral denervation. *J Neurosurg Spine* 2020;33:658-66.

22. LeDoux MS, Brady KA. Secondary cervical dystonia associated with structural lesions of the central nervous system. *Mov Disord* 2003;18:60-9.
23. LeDoux MS, Rutledge SL, Mountz JM, Darji JT. SPECT abnormalities in generalized dystonia. *Pediatr Neurol* 1995;13:5-10.
24. Lee JR, Kiss ZH. Interhemispheric difference of pallidal local field potential activity in cervical dystonia. *J Neurol Neurosurg Psychiatry* 2014;85:306-10.
25. Lim JY, De Salles AA, Bronstein J, Masterman DL, Saver JL. Delayed internal capsule infarctions following radiofrequency pallidotomy. Report of three cases. *J Neurosurg* 1997;87:955-60.
26. Lohrer TJ, Pohle T, Krauss JK. Functional stereotactic surgery for treatment of cervical dystonia: Review of the experience from the lesional era. *Stereotact Funct Neurosurg* 2004;82:1-13.
27. Lozano AM, Kumar R, Gross RE, Giladi N, Hutchison WD, Dostrovsky JO, *et al.* Globus pallidus internus pallidotomy for generalized dystonia. *Mov Disord* 1997;12:865-70.
28. Münchau A, Mathen D, Cox T, Quinn N, Marsden C, Bhatia K. Unilateral lesions of the globus pallidus: Report of four patients presenting with focal or segmental dystonia. *J Neurol Neurosurg Psychiatry* 2000;69:494-8.
29. Matsumoto K, Asano T, Baba T, Miyamoto T, Ohmoto T. Long-term follow-up results of bilateral thalamotomy for parkinsonism. *Stereotact Funct Neurosurg* 1976;39:257-60.
30. Matsumoto K, Shichijo F, Fukami T. Long-term follow-up review of cases of Parkinson's disease after unilateral or bilateral thalamotomy. *J Neurosurg* 1984;60:1033-44.
31. Moll CK, Galindo-Leon E, Sharott A, Gulberti A, Buhmann C, Koeppen JA, *et al.* Asymmetric pallidal neuronal activity in patients with cervical dystonia. *Front Syst Neurosci* 2014;8:15.
32. Moro E, Gross RE, Krauss JK. What's new in surgical treatment for dystonia? *Mov Disord* 2013;28:1013-20.
33. Naumann M, Becker G, Toyka K, Supprian T, Reiners K. Lenticular nucleus lesion in idiopathic dystonia detected by transcranial sonography. *Neurology* 1996;47:1284-90.
34. Ondo WG, Desaloms JM, Jankovic J, Grossman RG. Pallidotomy for generalized dystonia. *Mov Disord* 1998;13:693-8.
35. Teive HA, Sa DS, Grande CV, Antoniuk A, Werneck LC. Bilateral pallidotomy for generalized dystonia. *Arq Neuropsiquiatr* 2001;59:353-7.
36. Thobois S, Ballanger B, Xie-Brustolin J, Damier P, Durif F, Azulay JP, *et al.* Globus pallidus stimulation reduces frontal hyperactivity in tardive dystonia. *J Cereb Blood Flow Metab* 2008;28:1127-38.
37. Torres CV, Moro E, Dostrovsky JO, Hutchison WD, Poon YY, Hodaie M. Unilateral pallidal deep brain stimulation in a patient with cervical dystonia and tremor: Case report. *J Neurosurg* 2010;113:1230-3.
38. Valalik I, Jobbagy A, Bognar L, Csokay A. Effectiveness of unilateral pallidotomy for meige syndrome confirmed by motion analysis. *Stereotact Funct Neurosurg* 2011;89:157-61.
39. Volkmann J, Mueller J, Deuschl G, Kühn AA, Krauss JK, Poewe W, *et al.* Pallidal neurostimulation in patients with medication-refractory cervical dystonia: A randomised, sham-controlled trial. *Lancet Neurol* 2014;13:875-84.

How to cite this article: Horisawa S, Kawamata T, Taira T. Seven-year resolution of cervical dystonia after unilateral pallidotomy: A case report. *Surg Neurol Int* 2022;13:586.

Commentary:

Despite a long history of destructive interventions in the surgical treatment of movement disorders, it remains unclear if the disease progression, natural healing and brain plasticity translate into gradual worsening of surgical outcomes with longer duration of follow up. Here the authors of the report present a complete resolution of cervical dystonia in a patient treated with radiofrequency thermoablation of conventional pallidal target with torticollis rating (TWSTRS) decreasing from 39 before surgery to 4 immediately after surgery and 0 at the time of 7-year follow up. The correct matching of clinical indications with defined surgical target and appropriate patient and procedure selection are likely the reasons for such positive outcome. This result supports validity of considering targeted lesioning along with an option of deep brain stimulation in carefully selected patients and in presence of an experienced surgical team.

Konstantin Slavin, MD
Chicago, USA

Disclaimer

The views and opinions expressed in this article are those of the authors and do not necessarily reflect the official policy or position of the Journal or its management. The information contained in this article should not be considered to be medical advice; patients should consult their own physicians for advice as to their specific medical needs.