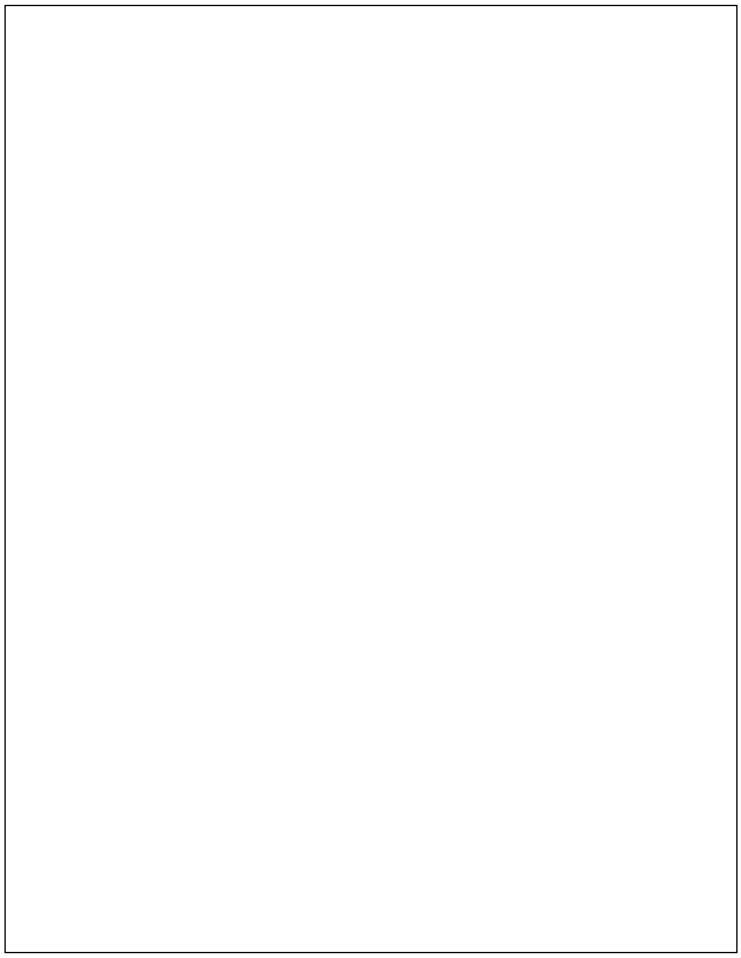

Successful Relief of Abdominal Dystonia After Sequential GPi Pallidotomy with 2-Year Follow-Up

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Key words
- Abdominal dystonia
- GPi pallidotomy

Abbreviations and Acronyms
GPi: Globus pallidus internus

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BACKGROUND: **Abdominal dystonia is very rare. To our knowledge, no clinical study has reported its specific treatment. Stereotactic therapy has been used to treat several movement disorders, including focal and general dystonia. We investigated the use of internal globus pallidum (GPi) pallidotomy for abdominal dystonia after failed oral medication.**

CASE DESCRIPTION: **A 48-year-old man presented with abdominal dystonia and complaints of involuntary undulating and contraction movements of his left abdominal wall for 5 years. Treatment with oral medication for 4 years was ineffective. Lesioning of the right GPi successfully relieved his symptoms. The symptoms recurred at 3 months and right GPi pallidotomy was repeated with complete resolution of symptoms after the second procedure. There was no recurrence or focal deficit at the 2-year follow-up.**

CONCLUSIONS: **GPi pallidotomy is feasible and effective for the treatment of abdominal dystonia that is resistant to standard medical therapy.**

INTRODUCTION

Abdominal dystonia, a form of focal dyskinesia affecting the abdominal wall,[1] was first described by Antonie Van Leeuwenhoek in 1723 when he himself experienced symptoms.[2] Since then, various terms have been proposed for this disorder, including Leeuwenhoek's disease, diaphragmatic myoclonus, diaphragmatic tremor, diaphragmatic flutter, moving umbilicus dyskinesia, abdominal dystonia, and respiratory myoclonus.[3-5]

Abdominal dystonia is extremely rare, consisting of involuntary and repetitive rhythmic movements of the abdominal wall. These movements cannot be suppressed voluntarily, but may be influenced by respiratory maneuvers. Investigations, such as spinal cord and abdominal imaging, usually fail to reveal any local abnormalities to explain this movement disorder.[6,7] Various cases have been reported in the literature with a long list of underlying causes. However, to our knowledge, the exact underlying pathophysiology has not been elucidated. In such cases, symptomatic management is the only available option.[7] The prognosis is unfavorable because no effective treatment exists.[8] We reported our experience with a case of abdominal dystonia treated with internal globus pallidus (GPi) pallidotomy and stereotactic lesioning. A 2-year follow-up is reported.

Video available at www.sciencedirect.com

CASE PRESENTATION

A 48-year-old man presented to our hospital with a 5-year history of involuntary jerking movements of the left anterior abdominal wall. The symptoms gradually worsened with time. The involuntary movements occurred suddenly without any associated aura or obvious trigger. The patient felt the abdominal muscle contractions especially on the left side (see Video 1). The contraction and jerking movements did not improve with resting, sitting, or lying positions. The tightness and pain felt on the abdominal wall caused work and sleep disturbances. The patient had no history of medication use or previous trauma to the abdomen and head. There was no family history of this condition. Neurologic examination and magnetic resonance imaging showed no abnormalities that could explain the symptoms.

The patient received oral medication (including clonazepam, trihexyphenidyl, diazepam, and baclofen) for 4 years without relief. Diagnosis was abdominal dystonia resistant to oral medication. After a brief discussion between the team and patient about any other modality including botulinum toxin injection and surgery, after resistant from oral medication, we performed stereotactic right GPi pallidotomy (Figure 1). The symptoms subsequently resolved completely, and the patient was discharged without any medication. The symptoms recurred after 3 months, and GPi pallidotomy was repeated 3 months later (6 months after the initial pallidotomy) according to the same technique, but in different target locations.

Lesioning was performed using an Inomed ZD (Inomed, Emmendingen, Germany) stereotactic frame. The right GPi target was set at 20.5 mm lateral, 4.0

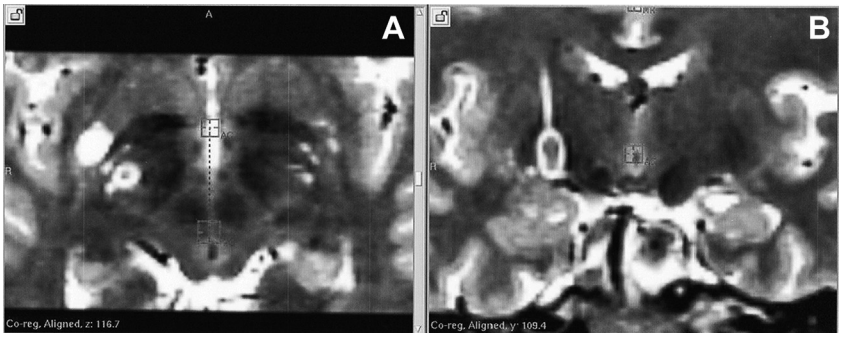

Figure 1. First right GPi pallidotomy. (**A**) Axial section and (**B**) coronal section.

mm inferior, and 2.5 mm anterior from the midpoint of the anterior-posterior commissures, 3 mm 45° anteromedial from the center of the first pallidotomy lesion. We used a 1.1-mm-diameter electrode with a 4-mm active tip. Cosman radiofrequency was used to create a lesion via thermocoagulation at 70°C for 30 seconds (Figure 2). After the second pallidotomy, the symptom resolved completely. The Burke-Fahn-Marsden dystonia rating scale scores were significantly improved from 12 to 0 compared with that before the second GPi pallidotomy, the patient remained in good condition with no symptoms or neurologic deficit at 2 years of follow-up (see Video 2).

DISCUSSION

Our patient, who had a 5-year history of abdominal dystonia, had intractable symptoms even after multimodal medical therapy. Taira et al[9] noted that most cases of dystonia are medically refractory and that surgical treatment results in marked improvement. From physical examination, laboratory findings, and head magnetic resonance imaging, we believed that the etiology in our patient was idiopathic. Krack and Vercueil[10] reported great benefit from surgical treatment, especially in cases of primary (idiopathic or genetic) dystonia and less so with secondary dystonia.

Recently, various neurosurgical interventions have been effective for various types of dystonia.[9] Based on some recent series of patients, lesioning in the GPi has been currently safe and may be the best treatment for dystonia.[10,11] This statement was further supported by Zhuang et al,[12] who reported that neurons in 87% of 367 patients with dystonia exhibited altered neural activity, including grouped discharge in the GPi and subthalamic nucleus, long-lasting neuronal activity, and rapid neuronal discharge in the ventralis oralis posterior or ventralis intermediate nucleus of the thalamus. Neurons in the ventralis oralis posterior, GPi, and subthalamic nucleus were firing at the same frequency as on electromyography during dystonia.[12] Yoshor et al[13] recommended GPi pallidotomy as optimal therapy for most patients with primary dystonia.

Presently, although lesioning has been replaced primary by deep brain stimulation due to its greater safety and the ability to perform adjustable lesions, as Taira et al[14] described and has been shown to improve dystonia,[15] we believed that lesioning remains a good treatment modality for patients who are unwilling to undergo device implantation and/or who cannot undergo deep brain stimulation for economic or geographic reasons. After a brief discussion, our patient underwent lesioning in the right GPi with excellent improvement immediately postoperatively.

The symptoms recurred 3 months postoperatively. After a brief discussion, supported by the study of Krystkowiak et al[16] in 5 patients with focal dystonia, we suggested that the recurrent symptoms may be caused by altered neuronal activity in the right GPi interrupting the cortico-striato-pallido-thalamo-cortical loop induced by the lesion we created at the initial

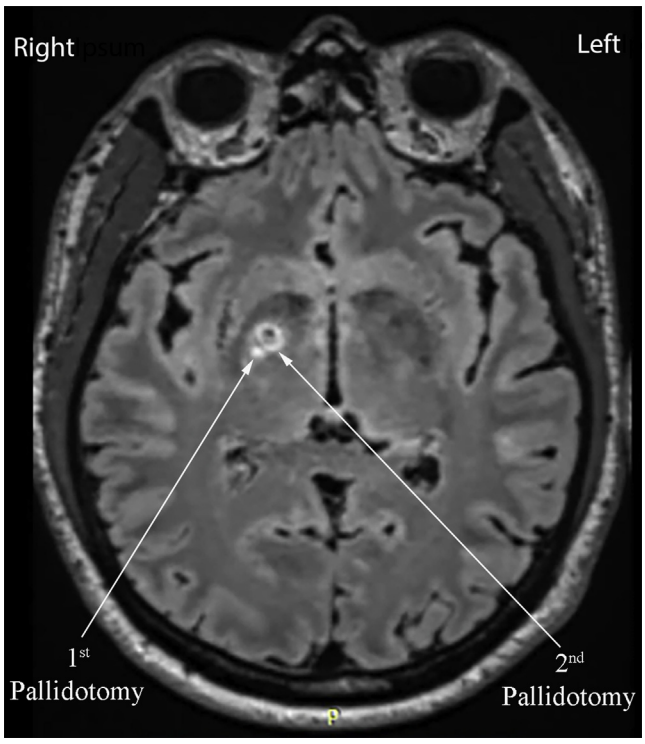

Figure 2. Second right GPi pallidotomy.

pallidotomy. From another study, we concluded that this type of dystonia responds poorly to most medical therapies and surgical therapy yielded the best result.[17] We planned a sequential pallidotomy in the right GPi and symptoms resolved completely after the second procedure. At 2-year follow-up, there was no recurrence or neurologic deficit.

In conclusion, we reported a rare case of abdominal dystonia treated successfully with GPi pallidotomy. This case indicates that GPi pallidotomy can be a safe and effective treatment of choice for patients with abdominal dystonia resistant to medication or those who could not tolerate the medication side effects.

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