

Poststroke fixed dystonia of the foot relieved by chronic stimulation of the posterior limb of the internal capsule

Case report

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The authors report the case of an adult patient with irreducible fixed dystonia (inward rotation) of the right foot that arose after cardioembolic ischemia of the left putamen and globus pallidus externus. Given the resistance of such symptomatology to all of the attempted conservative treatments (including botulinum toxin), the authors decided to perform deep brain stimulation, positioning the intracerebral electrode in the left internal capsule at the level of the motor fibers controlling the right foot, as confirmed by intraoperative electromyography. After the intervention, the patient was able to perform voluntary movements of outward rotation and abduction in the right foot and begin gait rehabilitation. Deep brain stimulation of the posterior limb of the internal capsule could be an alternative target used to treat poststroke fixed dystonic conditions. (DOI: 10.3171/2009.4.JNS08785)

KEY WORDS • fixed dystonia • internal capsule • deep brain stimulation

FIXED postures and focal fixed dystonia may appear after ischemic lesions of the basal ganglia and thalamus.^{7,8} The mostly recognized of these syndromes is the so-called "thalamic hand," which is due to thalamic and/or corona radiata infarctions;⁵ rarely, fixed postures involve the inferior limb and foot. These conditions are usually highly debilitating and refractory to drugs. Botulinum toxin is currently the first choice of treatment,⁸ but impressive results have been obtained by chronic MCS within the "hand knob" area in the treatment of arm and hand fixed postures, whereas in our experience GPi DBS was ineffective.⁶ In this paper we report the case of a young female patient who developed fixed posture and dystonia of the right foot after an ischemic lesion of the left putamen and of the external portion of the left globus pallidus. Botulinum toxin provided relief from the disease for ~2 years; thereafter, the patient became resistant to such treatment and confined to a wheel chair due to the fixed posture of inward rotation of the right foot. Due to the difficulties in stimulating the foot motor cortex region, which is located on the brain medial surface (anterior portion of the paracentral lobule), we decided to implant a deep brain electrode within the left posterior

limb of the internal capsule close to the fibers controlling the right foot. Neuroimaging, surgical planning, and treatment results are reported and discussed.

Case Report

History and Presentation. This 31-year-old woman was admitted to the hospital when she was 28 years old because of the gradual onset of the fixed posture of inward rotation of the right foot with slight hyperextension of the ipsilateral toe, which developed after the occurrence of an ischemic lesion of the left putamen. The brain infarct was considered secondary to the patency of the foramen ovale with the coexistence of an aneurysm of the interatrial septum. Magnetic resonance imaging revealed a signal alteration (hypointense and hyperintense on T1- and T2-weighted sequences, respectively) at the level of the left putamen and external portion of the globus pallidus (Fig. 1A). The patient then successfully underwent surgical closure of the interatrial defect.

Unfortunately, the inward rotation of the right foot gradually worsened and the patient started to develop difficulty in standing and walking. Autonomous deambulation became impossible, and the patient required the use of a wheelchair. A neurological examination disclosed irreducible inward rotation and flexion of the right foot (Fig. 1B) and

Abbreviations used in this paper: DBS = deep brain stimulation; EMG = electromyography; GPi = globus pallidus internus; MCS = motor cortex stimulation.

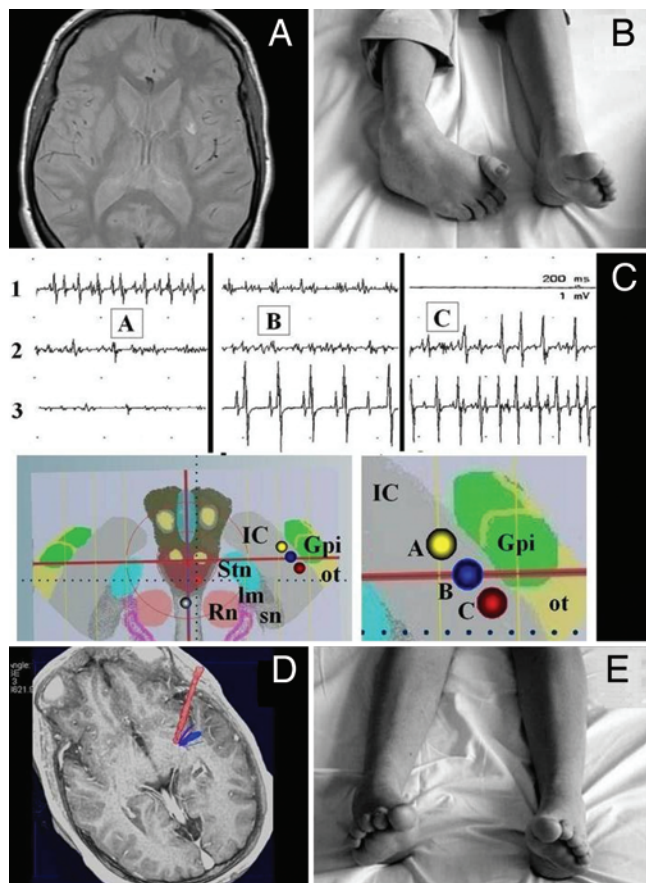


FIG. 1. Images and recordings obtained in the patient before, during, and after the course of treatment with DBS. A: Preoperative brain MR image showing the ischemic lesion at the left putamen and globus pallidus externus. B: Preoperative photograph of the patient's lower limbs. The fixed posture of inward rotation of the right foot is evident. C: Intraoperative EMG recordings (upper) from the right hand muscles (Track 1), the right leg muscles (Track 2), and right foot muscles (Track 3), for each of the 3 targets (lower, A, B, and C) in which macrostimulation was attempted. The stimulation at Target C resulted in a motor response consisting of extension and outward rotation of the right foot, eliminating the pathological fixed inward rotation. The definitive electrode was positioned at Target C because stimulation at Targets A and B also elicited involuntary contractions of the ipsilateral hand and leg at the same stimulation parameters. IC = internal capsule; Im = medial lemniscus; ot = optic tract; Rn = red nucleus; sn = substantia nigra; Stn = subthalamic nucleus. D: Postoperative volumetric MR image merged with preoperative tractography demonstrates the location of the definitive electrode (red) with respect to the fibers of the posterior limb of the left internal capsule (blue). E: Postoperative photograph of the patient's lower limb, demonstrating the reversal of the previously fixed posture.

confirmed the diagnosis of fixed dystonia, ruling out other conditions potentially involved in the differential diagnosis such as poststroke spasticity. The EMG procedure of the right lower limb disclosed tonic and synchronous clonic activity of the flexor and adductor muscles of the right foot, with coactivation of antagonist muscles. Laboratory determination of the *DYT1* gene, which was performed to rule out a genetic predisposition to develop such a condition, was negative. A monitored test using general anesthesia to eliminate the dystonic posture showed complete reversal of the pathological fixed posture of the right foot and its reap-

pearance before return of the conscious state, ruling out a somatoform disorder. Neuropsychological and psychiatric examinations also ruled out somatoform disorders as well as other psychopathological conditions potentially leading to anomalous somatic postures. A brain FDG-PET scan disclosed an area of diminished captation at the level of the left putamen, whereas brain PET examination with analysis of the density of the striatal dopamine transporter with ioflupane was normal. The patient then began prolonged medical therapy with baclofen (~5 months), etizolam, and levodopa, which was completely unsuccessful in controlling symptoms. Repeated botulinum toxin injections after the first 2 years only led to transitory and partial improvement of symptoms, as did motor rehabilitation therapy. Taking into account the severity of the clinical picture and high resistance to conservative therapy, in accordance with the agreement of the patient and her relatives, we decided to perform DBS of motor fibers controlling the right foot at the level of the internal capsule.

Operative Procedure. Under local anesthesia the patient underwent brain CT during stereotactic conditions with the Leksell frame (Elekta AB). The stereotactic coordinates of the targets were obtained through the probabilistic Franzini atlas, which stores previous data of internal capsule stimulation and somatotopy.² The obtained X, Y, and Z coordinates were confirmed, placing the target on the MR imaging tractography of the patient. The examination had been obtained a few days before surgery and merged with the CT scan performed during stereotactic conditions. Typically, the definitive electrode (QUAD 3387, Medtronic) was placed at the estimated target through a coronal bur hole. The definitive coordinates were 18 mm lateral to the intercommissural line (X), 1 mm behind the midcommissural point (Y), and 4 mm below the commissural plane (Z).

Macrostimulation via the definitive implanted electrode (bipolar stimulation between the 0 and 1 contacts) elicited motor responses of the right foot at 200 Hz, 0.8 V, and pulse width 200 μ sec; the motor response consisted of extension and outward rotation of the foot, eliminating the fixed inward rotation. Intraoperative EMG confirmed the selective activation of the right foot and inferior limb. This method was used to assess the proximity of the DBS electrode to the internal capsule at low-amplitude stimulation and to assess the improvements of the motor symptoms. Electromyographic recordings were performed using the Nicolet Viking System (Nicolet Inc.) and monopolar needle electrodes. Subdermal electrodes were placed in a bipolar fashion with active and distal reference needles in close proximity (1–3 cm) and were grounded by a ground electrode situated in the shoulder. Electrodes were placed on the symptomatic limb and hand, contralateral to the stimulating DBS lead, in the abductor pollicis brevis, anterior tibialis, and extensor digitorum longus. An additional 2 more anterior DBS tracks (Y = +1 and +2) led to the same motor response of the inferior limb, but EMG disclosed motor activation of the arm and hand with the same current parameters (Fig. 1C). The neurophysiological evaluation excluded the presence of stimulation artifacts with the current delivered at the target, taking into account the ac-

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tual increase in discharge frequency of the involved muscle segments (when stimulating the most posteriorly explored track) with respect to the 2 more anterior tracks, at the same stimulation parameters.

The electrode was then definitively implanted in the first target and trajectory that were used at the intervention. Merged images between preoperative MR imaging tractography and postoperative CT were used to obtain 3D reconstructed images (Dextroscope), which confirmed the correct placement of the electrode within the posterior limb of the internal capsule (Fig. 1D).

During general anesthesia, a subclavicular internal pulse generator (Solettra, Medtronic) was connected to the deep brain electrode and stimulation parameters were tested a few hours after surgery. The threshold for the motor response of the capsular lower limb was determined to be 1 V for 60 μ sec at 100 Hz. Continuous stimulation was initiated at 0.5 V for 60 μ sec at 100 Hz; such parameters did not reach the threshold for eliciting any motor response. The choice of the electrical parameters was determined by our previous experience with posterior limb stimulation of the internal capsule.⁴ The patient was unaware of the beginning of the electrical stimulation and no subjective sensation was reported when she was fully awake and in the following days after the procedure.

Postoperative Course. Beginning the second day after surgery the patient gradually became able to perform voluntary outward rotation of the right foot (Fig. 1E), which was absolutely impossible before surgery with the foot in the stable fixed condition. In the following days the patient attempted autonomous gait, leaving the wheelchair for short periods. One week later, at discharge from our institute, the dystonic posture of the right foot had almost disappeared and voluntary movements became possible, allowing gait rehabilitation.

After 3 months of follow-up the clinical benefit of the procedure has continued to be maintained and the patient does not show any neurological signs associated with a stimulation-related side effect.

Discussion

This case illustrates the role of the pyramidal motor system in the pathogenesis of focal fixed postures and dystonia due to small ischemic lesions of the basal ganglia and thalamus. We believe that the analogy with hand knob motor cortex stimulation, which relieves the thalamic hand syndrome,⁵ is evident. The stimulation of the motor bundle within the posterior limb of the internal capsule may act through the antidromic stimulation of the primary motor cortex or through the modulation of the descending system that controls voluntary movements at spinal levels.⁴ The posterior limb of the internal capsule harbors a bidirectional network connecting the motor cortex to the final common pathway at the spinal levels. Overactivity of selected pools of motor neurons may be due to abnormal signals traveling on this system and possibly originating from the imbalance between the extrapyramidal and the pyramidal system. High-frequency stimulation (100 Hz) delivered in such a complex system may act as an antidromic inhibition to hyperactive pools of cortical neurons, which allegedly

sustain the abnormal posture.⁸ The same effect may happen in MCS, but it requires a lower frequency of stimulation (50 Hz) directly involving the cortical surface and the hyperactive neuronal pools.

In this patient the disappearance of the foot dystonic posture was soon characterized by recovery of voluntary movements, thereby ruling out the possibility of another dystonic posture simply opposite to the previous one. Previous experiences of internal capsule stimulation to relieve poststroke spasticity and improve gait in hemiparetic patients^{3,4} reinforce these hypotheses, given the successful control of idiopathic fixed dystonia obtained by MCS.¹⁻³ It must also be considered that the active contact of the internal capsule electrode is very close to the GPi and may interact with the fibers of ansa lenticularis and other connection systems traveling through the internal capsule to and from the GPi. The low threshold of motor responses (< 1 V) and the stereotactic coordinates more medial ($X = 18$ mm) and more posterior ($Y = -1$ mm) than the GPi coordinates ($X = 20$ mm, $Y = +2$ mm) rule out the possibility that we were within the GPi nucleus (Fig. 2). Moreover, it should be noted that the result of GPi stimulation was ineffective control of fixed dystonia in our previous report.⁶ In our opinion, neuromodulation of the pyramidal system appears more promising, either at the cortical level for upper limb fixed dystonia⁵ or at the internal capsule level for fixed dystonia of the foot as in this case.

In summary, DBS of the internal capsule motor fibers allows the correction of focal poststroke fixed postures of the foot in the same way that MCS allows the control of fixed postures of the hand, arm, and trunk.⁶ This technique is proposed as an alternative to MCS when the

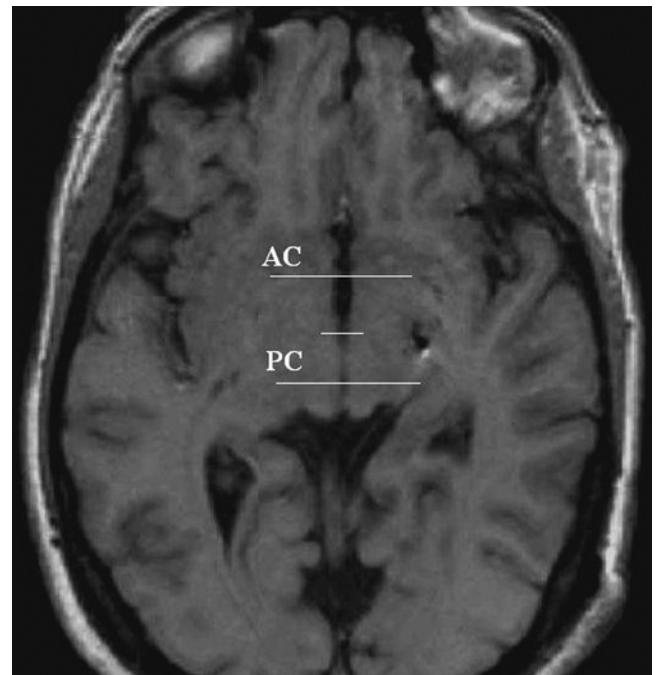


FIG. 2. Postoperative MR image showing the internal capsule electrode. Note the position of the electrode artifact with respect to the superimposed projection of the anterior commissure (AC)–posterior commissure (PC) line.

inferior limb is involved in the disease; the foot cortical representation on the brain mesial surface cannot be easily modulated by implanted strip electrodes. Finally, it is interesting to note that in our experience, therapeutic MCS acts at a stimulation frequency (50 Hz) considerably lower than internal capsule stimulation (100 Hz) delivered in this patient. Only many more cases and double-blind studies will confirm what this case report suggests, which is a new potential indication for DBS in a refractory drug-resistant condition such as poststroke foot fixed dystonia, using an unconventional target for the treatment of dystonic postures.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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