

Reversal of thalamic hand syndrome by long-term motor cortex stimulation

Case report

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✓ The authors describe a case of complete recovery from the so-called “thalamic hand” syndrome following chronic motor cortex stimulation in a 64-year-old man suffering from poststroke thalamic central pain. As of the 2-year follow-up examination, the patient’s dystonia and pain are still controlled by electrical stimulation.

It is speculated that a common mechanism in which the thalamocortical circuit loops are rendered out of balance may sustain hand dystonia and central pain in this case of thalamic syndrome. To the authors’ knowledge this is the first reported case of its kind.

KEY WORDS • cortical stimulation • thalamic syndrome • pain • dystonia •
neuronavigation

THE term “thalamic hand” was introduced by Roussy and Cornil⁸ to describe a peculiar abnormal posture of the hand that may develop along with central pain following the emergence of a thalamic lesion.

The pathophysiology and treatment of thalamic central pain have been objects of several recent studies.^{1,3,5,6,10} However, the etiopathogenesis and treatment of the thalamic hand syndrome remain poorly investigated. We describe a case of complete reversal of thalamic hand syndrome following motor cortex stimulation, which was performed to ease the patient’s pain. Such a singular case raises additional questions and hypotheses about this peculiar focal dystonia. To our knowledge no similar case has been reported in the literature.

Case Report

History and Examination. This 64-year-old right-handed man was admitted to our hospital in 1997 complaining of dysesthesias and a burning pain in his right arm and hand. These symptoms had developed gradually a few weeks after the patient had experienced a reversible ischemic neurological deficit (transient right hemiparesis) in 1993. Along with pain, a tonic postural hand attitude had developed, disabling all voluntary movements of the patient’s fingers and wrist (Fig. 1). This peculiar focal dystonia, which was exacerbated by shifts in motion, resulted in the complete loss of use of the affected hand in occupational and personal duties. Neurological examination disclosed a

mild dolorific and tactile sensory deficit with slight hyperesthesia and hyperreflexia in the affected arm; allodynia was also present. Neuropsychological investigations revealed no impairment of superior cognitive functions. No additional neurological signs were noted. A computerized tomography scan revealed an ischemic lesion in the left thalamus; this was more clearly defined on MR images, which demonstrated the involvement of the sensory thalamus and multiple ischemic lesions in the hemispheric white matter on both sides of the brain (Fig. 2).

The patient’s pain was unresponsive to drugs currently used for central nervous system pain treatment (amitripty-

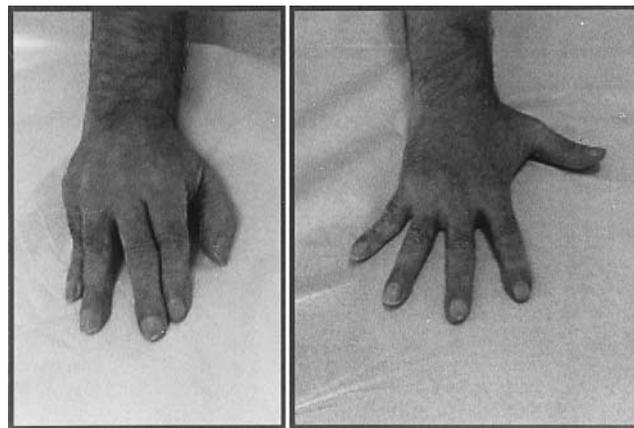


FIG. 1. Photographs showing the patient’s right hand during a period of no stimulation (*left*) and a period of stimulation (*right*).

Abbreviation used in this paper: MR = magnetic resonance.

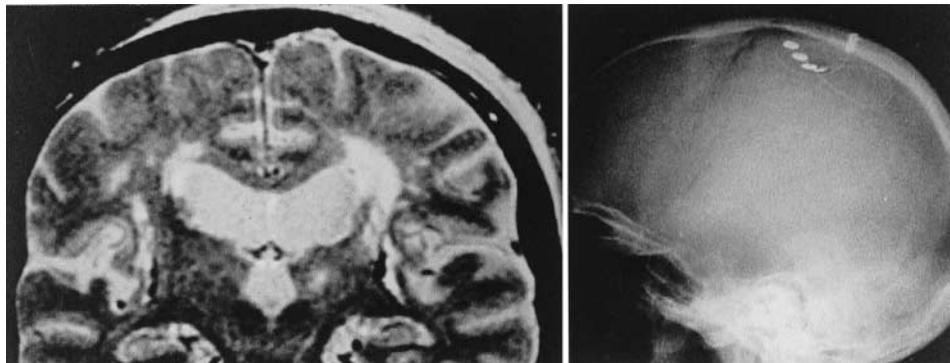


FIG. 2. *Left:* Coronal T₂-weighted MR image revealing ischemic lesions in the left sensory thalamus. *Right:* Radiograph demonstrating the locations of the stimulating electrodes.

line hydrochloride, clonazepam, and carbamazepine) and to orally and intrathecally delivered opioid agents, baclofen, and naloxone hydrochloride. No drug had any effect on the patient's hand dystonia.

Operation. After localized anesthesia had been induced in the patient, a quadripolar electrode strip (Resume; Medtronic, Inc., Minneapolis, MN) was inserted epidurally through an MR image-guided (EasyGuide; Philips Medical Systems, Milan, Italy) single precentral burr hole, which, based on Penfield's homunculus, had been placed over the expected projection of the hand area of the motor cortex (Figs. 2 and 3). Intraoperative stimulation (pulse

duration of 1 msec, 5-Hz trains) at a low frequency (16–20 Hz) and moderately high intensity (10 mA) induced muscle contractions in the painful area, confirming correct placement of the electrode. The extension wire was connected to the electrode, tunneled, and brought out percutaneously for trial stimulations. After 1 week, following the positive effect of a test stimulation on both the patient's pain and his dystonia, the electrode was connected to a subcutaneous subclavicular implanted pacemaker (Itrel II; Medtronic, Inc.) while the patient was in a state of general anesthesia.

Postoperative Course. The patient's postoperative

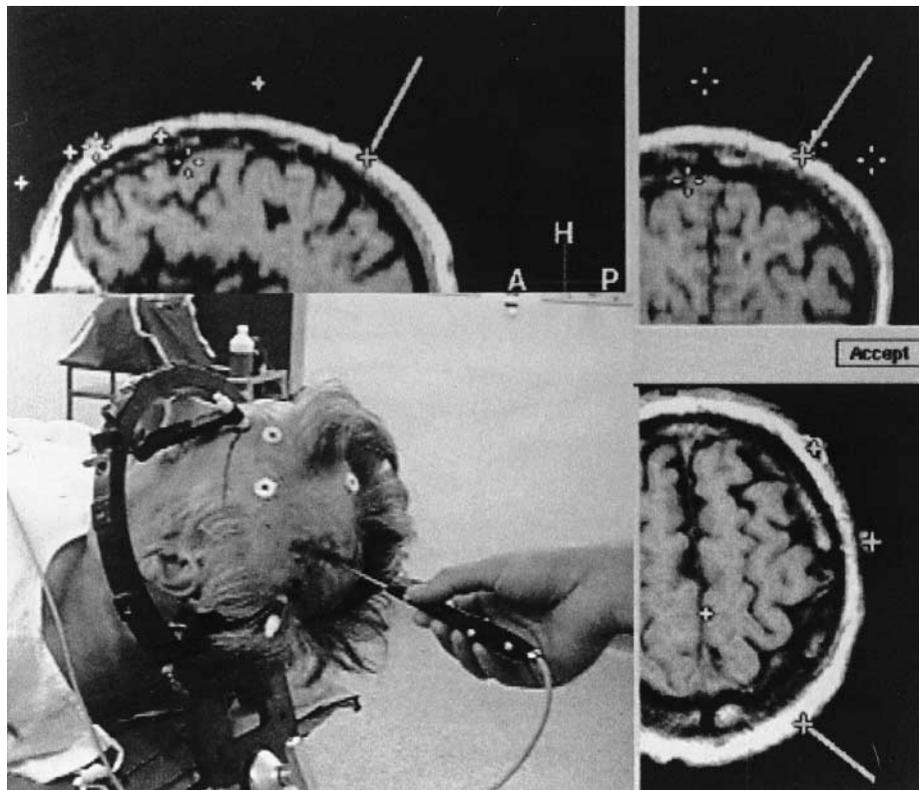


FIG. 3. Photograph and MR images demonstrating intraoperative image-guided localization of the central sulcus. A Cosman-Roberts-Wells ring (Radionics, Inc., Burlington, MA) is used as a head holder.

Thalamic hand syndrome reversal by cortical stimulation

course was uneventful. Analgesia was achieved to a level of approximately 50% by using the following electrical stimulation parameters: 60 Hz, 0.3 msec, 2 V, 0 to 3 setting, and continuous stimulation. The patient's hand dystonia completely disappeared.

At the 1-month follow-up examination, the patient had regained use of his hand for personal and even occupational tasks (bookbinding), with satisfactory pain relief. The effect of the stimulation has been long lasting and is still present as of the 2-year follow-up examination. Discontinuation of stimulation for more than 1 day results in reappearance of the pain and hand dystonia, which are promptly reversed by switching the pacemaker back on.

Discussion

The development of a dystonic hand attitude following the emergence of thalamic lesions has been well documented since the original description of thalamic syndrome by Dejerine and Roussy.² Schuster⁹ identified six patients with thalamic hand syndrome among 27 patients with anatomically verified thalamic syndromes. The dystonic attitude of the fingers can vary from patient to patient and the best way to classify them is to compare them with athetotic movements frozen at a definite moment, as remarkably illustrated by Schuster's photographs.

A dysfunction of the thalamocortical connections has been recently advocated to explain the appearance of dystonia after thalamic stroke.⁴ A mechanism of critical thalamocortical imbalance has also been hypothesized to explain the development of thalamic central pain after a lesion has formed in the sensory thalamus.¹⁰

To our knowledge, this is the first reported case in which the symptoms of thalamic hand syndrome have been reversed by long-term cortical stimulation. The dramatic effect of motor cortex stimulation observed in our case highlights the key role of the rolandic gyrus in sensorimotor corticosubcortical loops involved in the pathogenesis of central pain and dystonia. Brodmann's Area 4 has been reported to be connected to the primary and secondary somatosensory cortices, Brodmann's Area 5a, sensory and motor thalamus (ventral anterior, ventral lateral, ventral posterolateral, and posterior medial nuclei), hypothalamus, periventricular gray matter, and locus ceruleus.⁷ Reciprocal connections between the motor and sensory cortices seem to carry primarily nonnoxious information such as that related to targeted muscles.⁷ Precentral gyrus stimulation likely activates nonnociceptive neurons in the sensory cortex selectively, restoring a possible pain-inhibitory function.¹⁰ The interest of this case relies on the concomitant therapeutic effect of cortical stimulation on both pain and dystonia, showing that appropriate stimulation of this area may induce a modulating effect also on motor pathways.

We suggest that there was a common mechanism by which sensory and motor thalamocortical circuit loops

were rendered out of balance by the thalamic lesion. On the basis of complex motor cortex connections, we hypothesize that appropriate motor cortex stimulation may play a rebalancing role by desynchronization of the abnormal activity induced by the lesion.

Conclusions

Despite all etiopathogenetic speculations, the ultimate mechanisms of thalamic central pain and thalamic hand syndrome are unclear, as is the mechanism of action of motor cortex stimulation. The reported case stresses the well-known importance of extradural cortical stimulation as a modality of pain control and suggests a possible role for this procedure in the control of movement disorders as well. Neuronavigation devices used in localizing the motor cortex make this surgery easier and safer. Our experience illustrates the need to test motor cortex stimulation in patients with central pain and dystonia. The costs associated with the stimulation procedure are balanced by the early recovery of hand dexterity, avoiding the need for long-term rehabilitation.

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