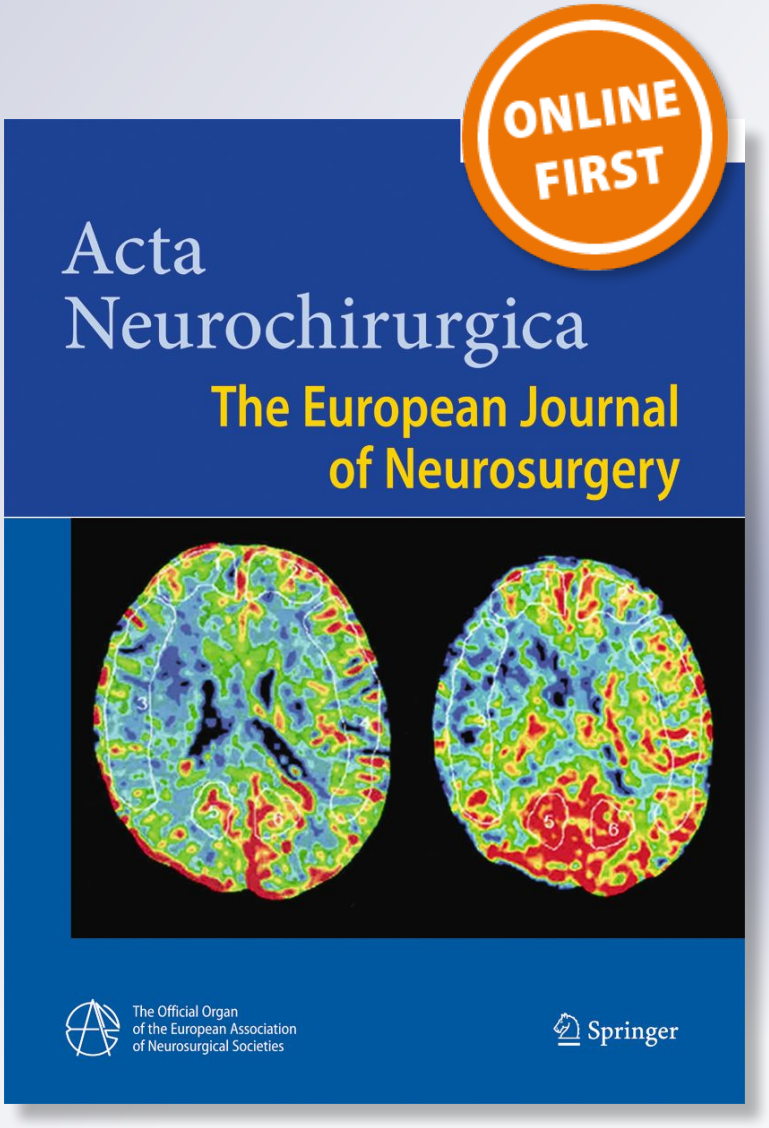
An unusual surgical indication for cerebral tuberculosis: status dystonicus. Case report

Andrea Franzini, Angelo Franzini, Vincenzo Levi, Roberto Cordella & Giuseppe Messina

Acta Neurochirurgica
The European Journal of Neurosurgery

ISSN 0001-6268

Acta Neurochir
DOI 10.1007/s00701-018-3561-5



Springer

Acta Neurochirurgica
https://doi.org/10.1007/s00701-018-3561-5

CASE REPORT - FUNCTIONAL NEUROSURGERY - MOVEMENT DISORDERS

An unusual surgical indication for cerebral tuberculosis: status dystonicus. Case report

Andrea Franzini[1] · Angelo Franzini[1] · Vincenzo Levi[1] · Roberto Cordella[1] · Giuseppe Messina[1]

Received: 29 January 2018 / Accepted: 3 May 2018

Abstract

Actual indications for surgery in tuberculosis are limited to obtaining a diagnosis, acquiring tissue for culture studies, treating hydrocephalus, aspiring a brain abscess, and reducing intracranial pressure in patients with multiple tuberculomas. Tuberculosis-related movement disorders are usually treated pharmacologically. We report on a child affected by post-tubercular generalized dystonia, who progressed to status dystonicus (SD) and underwent stereotactic bilateral pallidotomy. After surgery, SD resolved, and drugs were rapidly tapered. The successful reversal of SD and the motor improvement observed in our patient demonstrate the safety, feasibility, and clinical efficacy of pallidotomy in post-tuberculous-meningoencephalitis dystonia and SD.

Keywords Tuberculosis · Ablative surgery · Status dystonicus · Pallidotomy · Globus pallidus internus

Abbreviations

SD	Status dystonicus
DBS	Deep brain stimulation
Gpi	Globus pallidus internus
FR	Firing rate
EMG	Electromyography
MRI	Magnetic resonance imaging
BFMDRS	The Burke-Fahn-Marsden dystonia rating scale

Introduction

Dystonia is a movement disorder characterized by sustained or intermittent muscle contractions causing abnormal, often repetitive, movements, postures, or both [11]. Patients with dystonia occasionally develop severe episodes of generalized dystonic spasms and rigidity (status dystonicus (SD)) which may be refractory to standard drug therapy [7]. SD's spasms can interfere with respiratory and neurovegetative functions, thus causing metabolic imbalances, respiratory insufficiency, and acute renal failure secondary to rhabdomyolysis. As a consequence, SD is considered a life-threatening condition and urgent treatment is necessary. A gradual stepwise approach, from enteral pharmacological therapy to intensive care unit procedures and surgery, is required [9]. SD more often occurs in the context of acquired (secondary) dystonia, being often precipitated by events such as fever, trauma, surgery drugs, and infections [8]. Among infections, tuberculosis is a known cause of movement disorders such as tremor, chorea, or dystonia. These abnormal movements are the clinical expression of the pathological changes affecting the basal ganglia network during the disease [1]. In this article, we present a rare case of a young patient affected by post-tubercular dystonia, who developed SD and was successfully treated with bilateral pallidotomy.

Case report

This patient is a 6-year-old female child. After her mother's normal pregnancy, she was delivered without any perinatal complications. She developed generalized dystonia following a tubercular meningoencephalitis in July 2014 at age 4. At that time, she was given antituberculous treatment and a ventriculo-peritoneal shunt to address post-meningitis hydrocephalus. Before having tuberculosis, she had no history of abnormal movements or any cognitive impairment. In August 2014, when she was discharged from the hospital, she had developed severe mental and language impairments, spastic quadriparesis, almost complete blindness, and significant

✉ Andrea Franzini
andrea.franzini1@hotmail.it

[1] Department of Neurosurgery, Fondazione IRCCS Istituto Neurologico Carlo Besta, Via Celoria 11, 20133 Milan, Italy

Published online: 15 May 2018

hearing loss as the sequelae of the infection. In addition, abnormal postures and rapid dystonic movements of trunk and inferior and superior limbs were observed, most notably in the right side of the body. The dystonic movements gradually worsened and extended to the oro-facial segment resulting in dysphagia and requiring the positioning of a naso-gastric tube and subsequently of a percutaneous enteral gastrostomy because of malnutrition. Despite maximum pharmacological treatment and the implantation of an intrathecal baclofen pump, in July 2017, she still experienced severe dystonic movements, which progressed to SD after a period of recurrent febrile episodes, probably caused by recurrent urinary tract infections. At that stage, she was admitted to our hospital and required the administration of a high dose of intrathecal baclofene (600 μg per day) and of intravenous midazolam to control the dystonic spasms. Although there was a mild improvement of SD, the condition of good therapeutic control was achieved despite excessive sedation and episodes of apnea (the Burke-Fahn-Marsden dystonia rating scale (BFMDRS) was 110 severity score, 30 disability score). Surgical stereotactic therapy was then considered. Because of malnutrition and the patient's poor skin and subcutaneous status, it was decided to avoid implant devices for chronic deep brain stimulation (DBS) and, as an alternative, to perform bilateral pallidotomy.

A pre-operative brain MRI disclosed a crescentic collection over the right frontal convexity, compatible with a subdural hygroma due to cerebrospinal fluid overdrainage via the ventriculo-peritoneal shunt, diffuse cortical and subcortical atrophy, and a moderate dilatation of the supratentorial ventricular system. Moreover, several T2 ischemic alterations were spread within the internal capsule and the caudatus nucleus bilaterally and in the left putaminal nucleus.

Surgery was performed under general anesthesia with the Maranello frame (Maranello Frame, Italy). Computerized tomography performed under stereotactic conditions was merged with magnetic resonance imaging (MRI) through a Stealth Station (Medtronic Inc.). The definitive stereotactic target was selected in the globus pallidus internus (Gpi). Due to the alteration of the basal ganglia anatomy related to tuberculosis and ischemic lesions, Gpi could not be derived solely using an indirect method based on ventricular landmarks, as in our usual technique [4]. Therefore, we refined the lesion site in the Gpi with careful attention to MR imaging, particularly with the T2 sequence. Hydrocephalus and the altered brain anatomy obstacled the determination of stereotactic coordinates referred to the commissural system and make such values not applicable to other patients. Anyway, the definitive coordinates referred to the commissural system midpoint were 16 mm lateral (*X*), 4 mm posterior (*Y*), and 2 mm above the commissural plane (*Z*) in the left side and 15 mm lateral (*X*), 6 mm posterior (*Y*), and 1.5 mm below the commissural plane (*Z*). Intraoperative continuous limbs' electromyography (EMG) was used during the procedure to assess undesired stimulation of internal capsule. The regimen of general anesthesia used at our institute did not influence spontaneous muscle contractions, which were recorded in most districts during EMG monitoring.

Microelectrode recording was performed on a single track for each side. Very scarce neuronal activity was recorded along the two tracks. The right Gpi activity was characterized by the presence of sporadic action potentials with the exception of two-unit firing at relatively higher frequency. Specifically, one at 6.0 mm above the MRI-based target showed a less than 5-Hz firing rate (FR). The other unit, recorded at the target, had a FR at around 15 Hz (Fig. 1). Both showed an irregular tonic firing pattern. The left Gpi microelectrode exploration was characterized by the absence of neuronal activity with the exception of sporadic action potentials recorded between 10 and 8 mm above the target. A monopolar stimulation (with the following electrical parameters: 50 Hz, 100 μs, up to 7 V) was then performed with the stylet electrode (Radionics, Integra) bilaterally to rule out motor responses due to eventual capsular involvement. The left lesion was moved 3 mm laterally from the planned target position, since electrical stimulation evoked capsular motor responses at the planned site.

The radiofrequency monopolar lesion was performed bilaterally trough a CSK-TC20 electrode (Cosman Medical, MA, USA, 4-mm exposed uninsulated tip, 1 mm in diameter) with a 60-s train of 40 V through the radiofrequency generator, reaching a temperature of 70 °C. The heat generated during the lesioning process was monitored by a thermistor recording the temperature at the probe tip. As a consequence of the radiofrequency lesion, intraoperative EMG showed immediate reduction of spontaneous muscle activity in both hemibodies.

MRI scan performed 1 week after surgery showed circular signal alterations surrounded by T2 hyperintense cerebral edema (Fig. 2) [4]. SD disappeared with a dramatic reduction of dystonic movements on the second day after surgery. No further worsening of pyramidal tract signs was observed. While mild cervical and oromandibular dystonic movements induced by discomfort and pain remained, they were promptly relieved by oral midazolam. Intravenous midazolam and sedatives were tapered until withdrawal on post-operative day three. Baclofen dose was reduced from 600 to 300 μg per day, although the pump for baclofen delivery was not removed.

She recovered environmental contact and the ability to sit in a wheelchair. At 3-month follow-up, the post-operative results are stable (BFMDRS: 50 severity score, 30 disability score).

Discussion

Actual indications for surgery in cerebral tuberculosis are scarce, as shown by a recent review of the literature, being

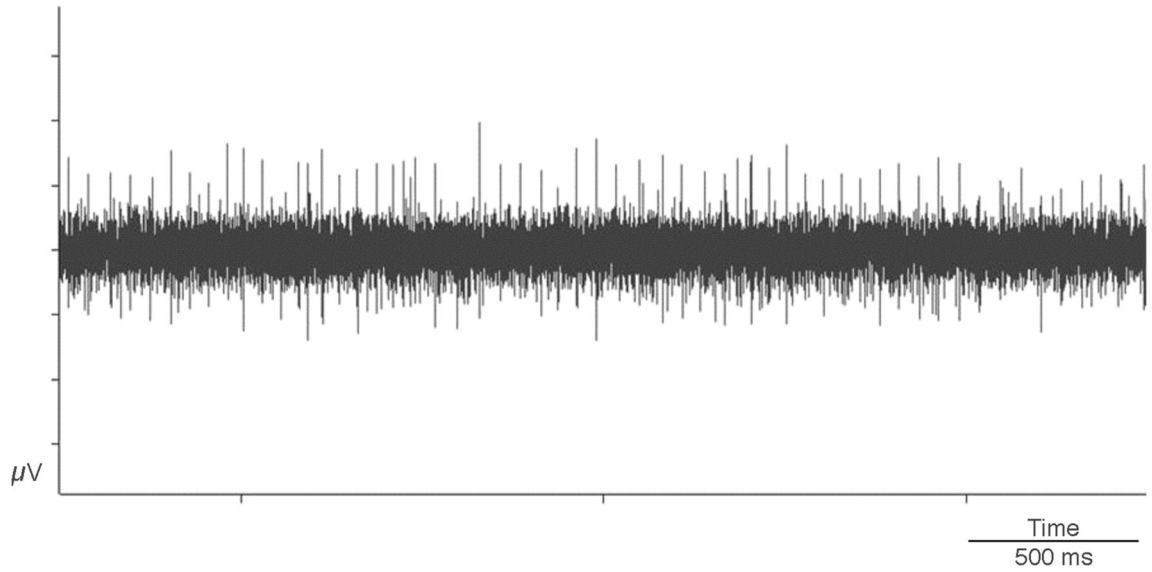


Fig. 1 Raw electrophysiological trace for the unit sampled in the right Gpi. This unit is the only one with a good signal to noise ratio and a firing discharge at around 15 Hz

limited to the following: acquisition of tissue for culture studies, treatment of hydrocephalus, aspiration of brain abscess, and control of intracranial pressure in patients with multiple tuberculomas [10].

In our paper, we describe an unusual indication for brain surgery in a patient who developed dystonia and SD following a tuberculous meningoencephalitis.

Post-tubercular SD is an extremely rare condition, and data about the management of this condition are lacking in the literature, since only limited experience with regard to the treatment of movement disorders associated with cerebral tuberculosis exists. Tremor, chorea-ballismus, dystonia, myoclonus, and Parkinsonism have been described to complicate the course of tuberculous meningitis with a reported frequency of up to 18.6% of cases [1]. Several mechanisms have been addressed in the pathogenesis of such disorders. Infarct of the basal ganglia as a consequence of vasculitis is thought to be involved, although intracranial tuberculomas, arachnoiditis, bacterial toxins, and secondary hydrocephalus are considered to play a role too [1]. These pathological processes determine a dysfunction in the basal ganglia or in their connections with the thalamus, the cortex, or the cerebellum [1], although an exact clinical-pathological correlation cannot always be determined. Referring to dystonia, it is deemed that dysfunction of the striatopallidal complex, the thalamic-subthalamic area, and the brainstem plays an important role in the etiology of the disease. Lesions in these structures, indeed, have been correlated to the generation of the dystonic movements [6]. In our patient, the pre-operative MRI disclosed several T2 abnormalities compatible with ischemic lacunae within the basal ganglia and the internal capsule bilaterally, which

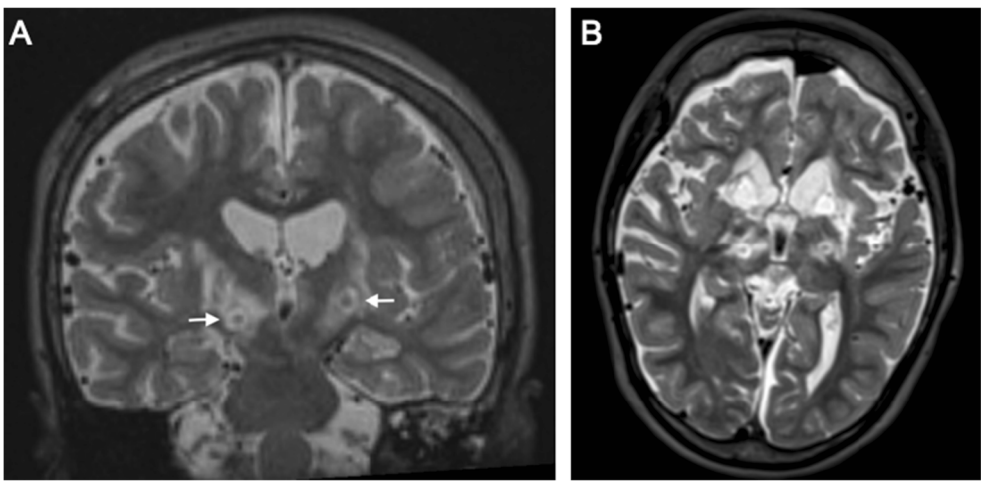


Fig. 2 Coronal (**a**) and axial (**b**) T2-weighted MR images taken on post-operative day 7. Note the circular signal alterations surrounded by hyperintense cerebral edema at the sites of the stereotactic lesions. The left lesion is more lateral than the right one, since electrical stimulation evoked motor responses at the level of the planned target

have probably determined the neurological picture of spastic quadriparesis and drug refractory SD.

The optimal treatment strategy of SD involves the administration of multiple drugs in different combinations (anticholinergics, typical neuroleptics, tetrabenazina, baclofen, benzodiazepines), deep sedation, muscle paralysis, intubation, and mechanical ventilation. This treatment should be conducted in an intensive care setting, due to the risk of life-threatening complications.

Second-line strategies for refractory cases include surgical techniques, such as DBS of the GPi and stereotactic pallidotomy [2]. The latter is preferred when DBS is not available and when the patient exhibits poor nutritional status or demonstrates hardware conflicting with the positioning of a DBS system. [3] Cases of both primary and secondary dystonia of various etiologies, with the SD mostly manifested in childhood, have been described to show a significant clinical improvement after stereotactic pallidotomy [4, 5, 12].

The critical condition and lack of response to oral and intrathecal therapies of our patient required an urgent rescue therapy to avoid the life-threatening neurovegetative and metabolic complications of SD. Gpi DBS was ruled out to avoid the hardware-related problems and infections, which are accentuated in pediatric patients with poor skin and subcutaneous tissue status [8].

To our knowledge, this case is the first description of a bilateral pallidotomy in a patient affected by post-tubercular meningitis SD. The successful reversal of SD and the motor improvement demonstrate the safety, feasibility, and efficacy of this procedure in a subset of patients, whose cerebral anatomy is often distorted by multiple infarcts, atrophy, and chronic hydrocephalus, which may hamper the stereotactic targeting of basal ganglia and their related structures. In this case, intraoperative neurophysiology with microelectrode recording and stimulation proved a valuable adjunct in localizing the correct site for lesioning and for avoiding unwanted post-operative neurological deficits.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Consent The patient's legal guardian has consented to submission of this case report to the journal.

References

1. Alarcon F, Duenas G, Cevallos N, Lees AJ (2000) Movement disorders in 30 patients with tuberculosis meningitis. Mov Disord 15(3):561–569
2. Ben-Haim S, Flatow V, Cheung T, Cho C, Tagliati M, Alterman RL (2016) Deep brain stimulation for status dystonicus: a case series and review of the literature. Stereotact Funct Neurosurg 94:207–215
3. Fasano A, Ricciardi L, Bentivoglio AR, Canavese C, Zorzi G, Petrovic I, Kresojevic N, Kostić VS, Svetel M, Kovacs N, Balas I, Roubertie A, Mishra D, Mariotti P, Temudo T, Nardocci N (2012) Status dystonicus: predictors of outcome and progression patterns of underlying disease. Mov Disord 27(6):783–788
4. Franzini A, Levi V, Franzini A, Dones I, Messina G (2017) Staged pallidotomy: MRI and clinical follow-up in status dystonicus. Br J Neurosurg 28:1–4
5. Jech R, Bares M, Urgosik D, Cerna O, Klement P, Adamovicova M, Ruzicka E, Prihodova I, Oslejskova H (2009) Deep brain stimulation in acute management of status dystonycus. Mov Disord 24(15):2291–2292
6. Lehericy S, Tijssen MAJ, Vidailhet M, Kaji R, Meunier S (2013) The anatomical basis of dystonia: current view using neuroimaging. Mov Disord 28(7):944–957
7. Manji H, Howard RS, Miller DH et al (1998) Status dystonicus: the syndrome and its management. Brain 121:243–252
8. Mariotti P, Fasano A, Contarino MF, Della Marca G, Piastra M, Genovese O, Pulitanò S, Chiaretti A, Bentivoglio AR (2007) Management of status dystonicus: our experience and review of the literature. Mov Disord 22(7):963–968
9. Marras CE, Rizzi M, Cantonetti L, Rebessi E, De Benedictis A, Portaluri F, Randi F, Savioli A, Castelli E, Vigevano F (2014) Pallidotomy for medically refractory status dystonicus in childhood. Dev Med Child Neurol 56:649–656
10. Rajshekhar V (2015) Surgery for brain tuberculosis: a review. Acta Neurochir 157:1665–1678
11. Sanger TD, Chen D, Fehlings DL, Hallett M, Lang AE, Mink JW, Singer HS, Alter K, Ben-Pazi H, Butler EE, Chen R, Collins A, Dayanidhi S, Forssberg H, Fowler E, Gilbert DL, Gorman SL, Gormley ME Jr, Jinnah HA, Kornblau B, Krosschell KJ, Lehman RK, MacKinnon C, Malanga CJ, Mesterman R, Michaels MB, Pearson TS, Rose J, Russman BS, Sternad D, Swoboda KJ, Valero-Cuevas F (2010) Definition and classification of hyperkinetic movements in childhood. Mov Disord 25(11):1538–1549
12. Zorzi G, Marras C, Nardocci N, Franzini A, Estienne M, Conti C, Dones I, Broggi G (2005) Stimulation of the globus pallidus internus for childhood-onset dystonia. Mov Disord 20:1194–1200

Comments

This is a nice and timely paper showing the benefit of pallidotomy in treatment of status dystonicus. In this case, it was tuberculous meningoencephalitis that provoked the dystonias.

This report is especially important since lesional surgery is more and more forgotten in this era of frenetic DBS. Pallidotomy for status dystonicus can be life-saving and much less expensive than DBS, but the skills to perform it properly are becoming forgotten and the young generation of neurosurgeons is not being trained in performing stereotactic lesions.

Marwan Hariz
London, UK