

Long-Term Outcome of Dentatotomy in a Dystonic Patient

Avaliação a longo prazo de paciente distônico submetido a dentatotomia

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Abstract

Dystonia is characterized by sustained or intermittent muscle contractions leading to abnormal movements and impairment in daily activities. Stereotactic dentatotomy has been a treatment option in cases of spasticity or dystonia, especially in patients with cerebral palsy, but the long-term effect of dentatotomy in dystonia is still unknown. Here we describe a dystonic patient who underwent dentatotomy for symptomatic treatment of refractory dystonia and whose 20-year follow-up showed improvement in symptoms.

Keywords

- ▶ dystonia
- ▶ dentatotomy
- ▶ cerebral palsy

Resumo

A distonia é caracterizada por contrações musculares intermitentes ou sustentadas que levam a movimentos anormais e ao comprometimento das atividades cotidianas. A dentatotomia estereotática tem sido uma opção de tratamento nos casos de espasticidade ou distonia, especialmente em pacientes com paralisia cerebral, mas o efeito a longo prazo da dentatotomia na distonia ainda é desconhecido. Descrevemos aqui um paciente submetido a dentatotomia para tratamento sintomático de distonia refratária e cujo acompanhamento por 20 anos demonstrou melhora nos sintomas.

Palavras-chave

- ▶ distonia
- ▶ dentatotomia
- ▶ paralisia cerebral

Introduction

Dystonia is a neurologic disorder characterized by sustained or intermittent muscle contractions causing abnormal movements and/or postures¹ and involves a heterogeneous group of disorders with many underlying causes, both known and unknown. Etiologic classification relates to whether dystonia is inherited, acquired, or due to identifiable brain

abnormalities.¹ Dystonia can arise secondary to brain damage, including stroke, trauma, or adverse medication effect (tardive dystonia), or as a symptom of other diseases, such as Parkinson disease. Pharmacologic therapies, especially the anticholinergic agents, can provide modest symptomatic improvement but can lead to significant adverse effects. Botulinum toxin injections can provide relief in many patients.^{1,2} However, the injections must be repeated every few

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months, patients can become resistant, and adverse effects, such as weakness, are not uncommon. Surgical interventions have included rhizotomy, for cervical dystonia; ablation of the thalamus (thalamotomy); internal globus pallidus (pallidotomy); dentate nucleus (dentatotomy), and deep brain stimulation for more generalized dystonias.¹⁻³

For many decades, stereotactic dentatotomy has been a treatment option in cases of spasticity⁴ or dystonia, especially in patients with cerebral palsy. Though vastly used in the past, in the last twenty years, published papers on the matter were rare. In this line, the long-term effect of dentatotomy in dystonia is still unknown. Here we describe a dystonic patient who underwent dentatotomy for symptomatic management and was monitored for 20 years.

Case Report

A 39-year-old right-handed man presented with dystonia since early childhood due to hypoxia during birth. His symptoms were mainly distal on four limbs, as well as on larynx, and significantly impaired his work, study and leisure activities. He had no cognitive disability, no motor involvement other than dystonia/dystonic tremor, and no remarkable neuroimaging findings. All the treatment attempts were frustrated, including physical therapy and medical therapy with diazepam, clonazepam, valproic acid and trihexypenidyl. By the time the patient was 20 years-old, in 1995, due to refractory symptoms and with patient's consent, a left dentatotomy was performed. The stereotactic coordinates of the targets were determined based on previously published anatomical studies.^{5,6} The target point was located 10–12 mm behind the fastigial point, 3–5 mm below the fastigial line, and 8–10 mm from the midline. The patient reported good outcome in the motor function, improvement in left side dystonia, action tremor, walking and performing manual tests, such as drinking. Despite the improvement in dystonia, our patient remained symptomatic on the right side, and in the same year, deep brain stimulation (DBS) was placed on the left subthalamic nucleus. Deep brain stimulation was set as bipolar (contact 0 as anode and 2 as cathode), at 2,1 V, 270 μ s and 125 Hz. Twenty years after the first surgery (dentatotomy), the Unified Dystonia Rating Scale (UDRS) was 9,5 (highest score on this scale: 44), with symptoms predominantly in the distal parts of the upper limbs and larynx. He was otherwise asymptomatic.

The patient had the impression that the procedures improved ~ 50% of his dystonia, mainly on his legs, now considered asymptomatic.

Discussion

Dentate lesions result in prolonged reaction time associated with a corresponding increase in the latency of movement and related responses of cortical neurons, and may affect movement programming through the cortico-neocerebellum cortical loop.⁷ It may improve abnormal movements through decreasing in the facilitatory outflow of the dentate

nucleus over the motor cortex.⁸ Basal ganglia or cerebral cortex damage tend to lead the contralateral cortex to hyperexcitability that can be reduced with dentatotomy.^{9,10}

In agreement with this hypothesis, Schneider and Crosby⁹ found that the cerebellar cortex aided abnormal posturing and hypertonia in patients with cerebral palsy.

After those results, several studies showed benefits of dentatotomy in dystonia, choreoathetosis and spasticity in cerebral palsy patients.¹¹⁻¹⁴ Improvement seemed more substantial in the lower limbs,¹⁵ which happened in our patient. A combination of dentatotomy and thalamo-subthalamotomies showed even better result in cases of spasticity than dentatotomy alone.¹⁶ To our knowledge, the case reported is the first to show the association between dentatotomy and subthalamic deep brain stimulation.

Despite the report of good outcomes seen in the literature, the benefit over long periods is unknown. In 1970, Heimburger¹⁷ reported improvement in 50 out of 61 patients submitted to dentatotomy, with benefits lasting from 4 months to 5 years and relapse in 11 cases. Siegfried and Verdie¹⁸ described a reduction in improvement after months or years of the operation, with a failure rate of 10% in 6 months, 20% in 2 years and 24% in 3 years of follow-up. Our patient remained stable after 20 years follow-up.

Finally, although dentatotomy can be a safe treatment of spasticity and dystonia, the current trend is the DBS surgery, which usually brings better results with fewer side effects.¹⁹ Even so, ablative surgery appears to be a good option for a proportion of patients to whom the DBS is contraindicated, as well as for those with social problems,²⁰.

Conclusion

This case report showed sustained benefit of dentatotomy associated with unilateral subthalamic DBS for secondary dystonia. The dentatotomy can be considered as a good option for refractory dystonia, which should be further explored in future studies. Our study protocol was safe; it laid the groundwork for larger studies regarding dentatotomy, with or without DBS, in this patient population.

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