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Deep Brain Stimulation in a Patient with Generalized Dystonia Painful Rebel to the Medical Therapy: A Case Report (Clinical Note)

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ABSTRACT

Cervical dystonia is a common malaise in the doctor's office. It is a movement disorder characterized by sustained involuntary muscle contractions and abnormal postures: the patient exhibits involuntary left head and neck turning. It can recognize various more or less severe conditions as etiological agents and still remains a difficult disorder to treat. We reviewed a clinical case, analyzing both the moments of differential diagnostics and the therapeutic choices, with particular interest in cerebral electrostimulation.

Introduction

Cervical dystonia is characterized by involuntary tonic contractions or intermittent spasms of the neck muscles. As for etiology, in most cases, it is idiopathic, while some patients have a family history, and in some of these, a genetic cause has been identified. Some of these patients have other dystonias (e.g., eyelids, face, jaw, hand). Cervical dystonia can be classified as congenital or secondary to other conditions such as brain stem lesions or basal ganglia or taking certain drugs (for example, haloperidol). Deep brain stimula-

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tion appears today as a safe and effective therapy for dystonia, creating a two-way closed circuit.

Case Presentation

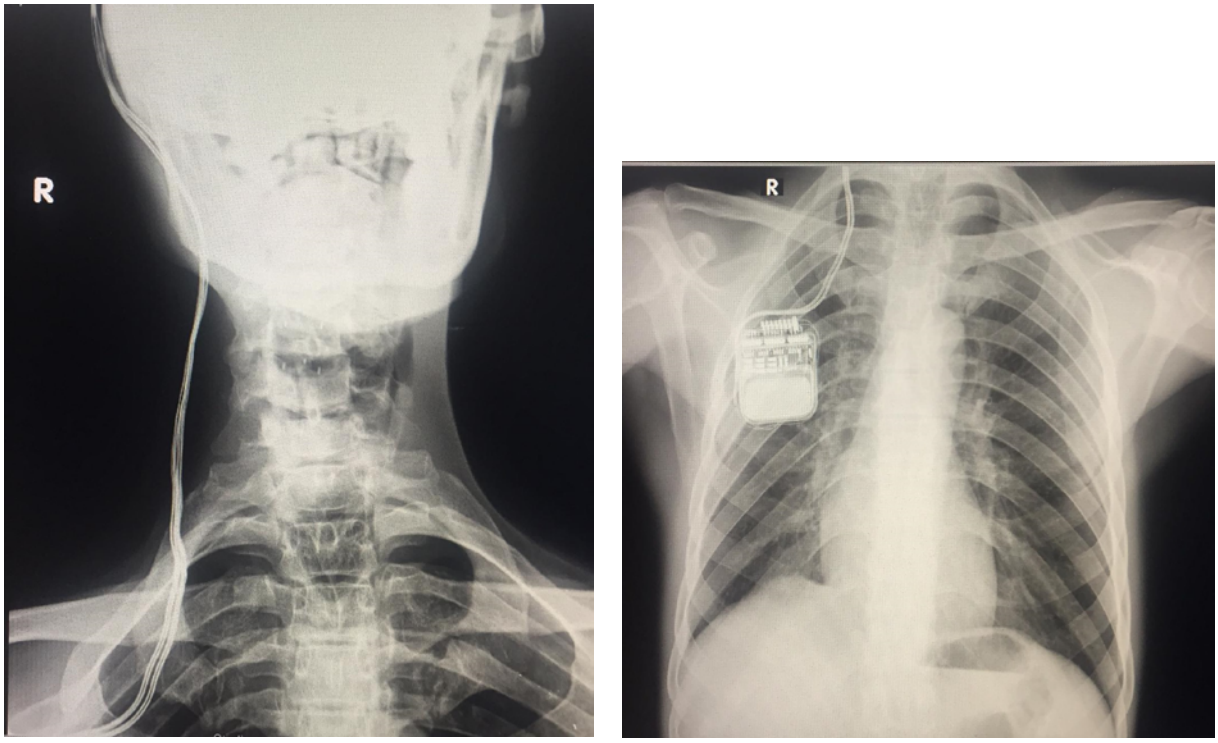
A 42-year-old male patient, suffering from bilateral cervical brachialgia for about two years, came to our observation. Severe algia was detected (Visual Analogic Scale [VAS] and Number Rating Scale [NRS] 10/10) with intense contracture of the neck muscles. The patient was sent to a neurosurgical consultation, which indicated a cervical discectomy.

There was no significant improvement in painful syndrome syndrome (VAS 8-9/10 and NRS 9/10). Indeed, involuntary movements of the neck appeared with associated blepharospasm and oromandibular spasms that forced the patient to several hospitalizations. The patient practiced several drug therapies (muscle relaxants, opioids, benzodiazepines). Daily medical therapy consisted of baclofen 5 mg, trihexyphenidyl hydrochloride 16 mg, clonazepam 8 mg, oxycodone/naloxone 5 mg/10 mg.

Management and results

The patient appeared to be characterized by difficult walking and marching disturbed by contractions and generalized choreoathetotic movements with associated oromandibular spasms. There was also widespread hypertonicity and rigidity. The remaining general physical examination appeared negative. Laboratory investigations seemed to be normal. With partial symptomatic control, infiltrative therapy with botulinum toxin was also attempted during our supervision. In the meantime, we were trying a diagnostic investigation in the suspicion of painful dystonia on a genetic basis. Still, the studies carried out (molecular analyses for mutations in the DYT-1 and DYT-6 genes) did not show anomalies confirming these hypotheses.

Therefore, it was decided to initiate the patient for surgery with bilateral lead implantation in the internal globus pallidus (framed procedure, after recording the action potentials and control stimulation) and a dual-channel device implant capable of providing bilateral



Figures 1. X-rays showing an implanted deep brain stimulation system

X-ray inspection in the post-operative period. It highlights the correct positioning of the electrodes in the region of the nuclei of the base and the pulse generator in the subcutaneous seat of the right subclavicular, ensuring the goodness of the surgical procedure for positioning the electrostimulation system in the absence of complications. Collaterally, the images also show the outcomes of cervical vertebroplasty, the reversal of lordosis, and the consequences of prosthetic disc grafting (at the level of the cervical vertebrae C3-C4, C4-C5, and C5-C6). Partial ventral synostrosis of the interbody bridges.

stimulation with a single device. (Activa PC Neurostimulator®, MEDTRONIC) with a connection to previously implanted leads. Computed tomography (CT) and control radiography showed the correct positioning of the device in the right subclavian subclavicular area (Figures 1). The patient was then discharged with the diagnosis of generalized painful dystonia and entrusted to outpatient clinical monitoring for follow-up.

Discussion

We took care of a patient with the typical movement disorder characterized by abnormal muscle contractions, supported by agonist and antagonist muscle groups together, which cause abnormal postures or repetitive movements, pain, with other signs and symptoms present here (such as tremors). We have tried to classify the disorder based on the age of onset, anatomical distribution, and cause [1].

Primary dystonias are often idiopathic and characterized by a lack of identifiable cause or underlying neurological anomaly. Frequently, but not always, there are associated known genetic mutations, such as the DYT1 mutation (associated with early-onset torsional dystonia) or DYT6 (autosomal dominant dystonia, often craniocervical dystonia), which we have researched and excluded [2, 3]. Deep Brain Stimulation (DBS) represents a form of high frequency (biological) electrical stimulation of subcortical target structures that is based on the use of stimulating electrodes positioned through stereotaxic surgery and a subcutaneous stimulator [4].

Various experiences are described in which the device recorded good results in other situations in which dystonia coexisted with well-defined pathologies. For example, some studies on cohorts of Parkinson's disease patients with various types of movement disorders (tremor, bradykinesia, and dystonia), using an implanted clinical neurostimulator, have obtained good results [5, 6], as well as treating some affected patients from severe and non-responsive epilepsy to common drug treatments [7], head injuries [8] or even neuropsychiatric syndromes [9]. In particular, most of the evidence for DBS regarding dystonia shows promising efficacy in primary generalized dystonia and cervical dystonia, so our patient seemed eligible for treatment and with a better chance of success [10-12] after botulinum toxin treatment failure [13].

Conclusions

There is also growing evidence of DBS in patients with dystonia, a condition known to be difficult to treat medically. Secondary dystonias tend to benefit less from DBS, perhaps due to the wide range of etiologies and potentially affected areas of risk. The clinical case described is an example of a multi-step treatment protocol in which this method can find valid applications to improve the health and quality of life of patients affected by this disorder.

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this research.

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Conflict of interest

The Authors declared no conflict of interest.

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