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## Isolated speech-induced lingual dystonia successfully treated with botulinum neurotoxin



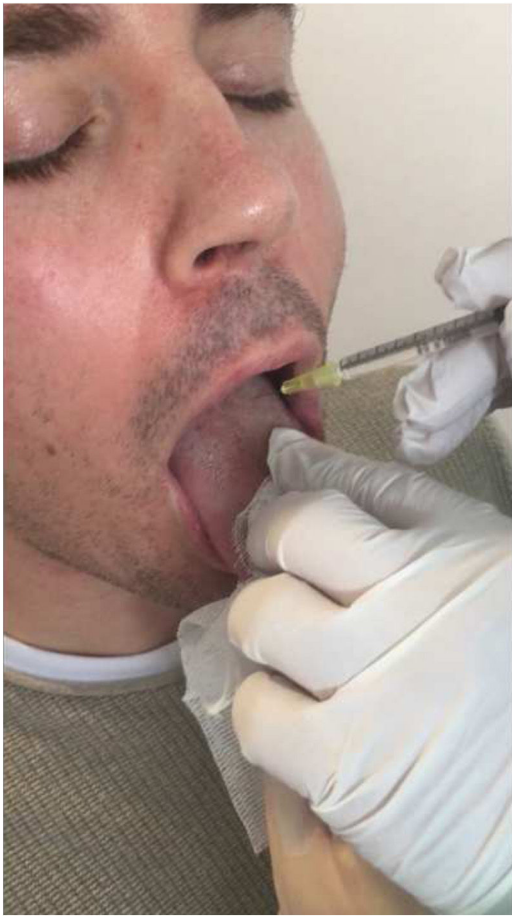
### Distonía lingual inducida por el habla con respuesta favorable a toxina botulínica

Dear Editor:

Dystonia is a hyperkinetic disorder characterized by sustained or intermittent muscle contractions, resulting in abnormal repetitive movements and/or postures.<sup>1</sup> Oro-mandibular dystonia (OMD) is a focal type of dystonia that involves masticatory and/or lingual muscles. Lingual dystonia is one of its subtypes. According to some case series, it might take up to the 4% of all the dystonias assessed in a Movement Disorder Department.<sup>2</sup> Speech-induced lingual dystonia is rare. Mainly, it can be idiopathic and according to literature can be considered as an occupational dystonia in some patients.<sup>3</sup> Despite its low prevalence, it can be very disabling and requires proper identification and management. We report here a case of a young man who presented with an isolated speech-induced lingual dysto-

nia with an excellent response to Botulinum neurotoxin (BoNT).

A 34 year-old man was referred to our department of Neurology with a five month history of slurred speech and progressive difficulty to move the tongue. A second-grade relative suffered from epilepsy, but there was no other family history of interest. He worked as telemarketer and spent up to eight hour-a-day speaking. Due to his symptoms, which continued worsening, he had to stop working and felt embarrassed when holding a conversation. No concomitant treatment or any other precipitating factor were found. His neurological exam revealed speech-induced tongue protrusion associated with mild to moderate dysarthria (Video 1). His symptoms improved when chewing gum (Video 2) or bread, as well as when speaking out. Brain MRI (including T2-sequence), electromyography (EMG), and extensive blood tests (including thyroid function, copper metabolism, folate and vitamin B12 levels) were normal. A diagnosis of idiopathic speech-induced focal dystonia (occupational) was performed and trihexyphenidyl (up to 4.5 mg) was started, obtaining slight improvement without remarkable secondary effects. Six months later BoNT was started. Specifically, 2.5 units of BoNT (Botox®, Allergan Pharmaceuticals) were injected intraoral bilaterally in only 1 point of each genioglossus muscle (Fig. 1). No secondary effects were observed. One week later he noticed slight



**Figure 1** Intraoral injection of BoNT at left genioglossus muscle.

clinical improvement, but only maintained for one month. BoNT injection was repeated within 3 and 6 months, increasing dosage up to 5 units at each genioglossus. The second infiltration led to a clinical benefit that remained for only two months. Five days after the third injection a remarkable and sustained response was referred. The neurological exam was normal four months later (Video 3). Trihexyphenidyl was left progressively without impairment. One year later, the patient was looking for a job and reported a great improvement into his social life.

Speech-induced dystonia is a rare, but potentially really disabling condition. Its pathophysiology is not clear yet. Classical models were based into the idea of an abnormal plasticity regulation.<sup>4</sup> However, some recent publications question this hypothesis, considering the plasticity response variability inter and intra-subject as well as plasticity is not required to develop secondary dystonia.<sup>5,6</sup> Current hypothesis defend that both genetic and environmental factors play an important role in the aetiology of dystonia. Different brain structures (thalamus, cerebellum and sensorimotor cortex) are thought to be involved in its pathophysiology, leading to a network disfunction.<sup>6,7</sup> Due to the low prevalence of lingual dystonia, it has previously been reported on in single case reports<sup>8,9</sup> or case series.<sup>10,11</sup> Recently, Yoshida et al. reported the largest series with 172 patients with lingual dystonia.<sup>2</sup> Lingual protrusion, as in our case, was

the most frequent but retraction, curling and lateropulsion dystonia have been observed as well. Moreover, it can be considered as an occupational dystonia. Indeed, task-specificity was observed in 90.1% of patients reported by Yoshida et al during speaking, chewing, or swallowing. Our patient had no history of exposure to neuroleptic medications, facial or oromandibular injuries, or recent infection. For this reason, the condition was diagnosed as a lingual dystonia specifically induced by speaking after all laboratory examinations had ruled out secondary causes. Isolated lingual dystonia is extremely rare as most reports have described patients with lingual dystonia who also displayed dystonic movements elsewhere.<sup>2</sup> According to this, a case of speech-induced task-specific OMD (involving perioral muscles, the tongue and the jaw) has been reported in a bingo caller<sup>8</sup> as well as in a telemarketer.<sup>9</sup>

A number of treatments are used in oromandibular dystonia such as trihexyphenidyl, baclofen, clonazepam, tiapride, or zolpidem.<sup>11</sup> However, BoNT injection has been demonstrated to be the most effective.<sup>2,8,11–13</sup> The genioglossus makes up the bulk of the tongue's muscle mass and is responsible for tongue protrusion, making it an ideal candidate for injection. We did not use EMG guide for injections. However, its use is recommended as it may help to identify muscles correctly and to reduce the risk of potential complications such as dysphagia or aspiration. In any event, administration should be always done by an experienced professional. BoNT tongue injections can cause some potential life-threatening secondary effects, as dysphagia or aspiration. Therefore, its administration should be done by an experienced professional. In our case, it is worth noting the absence of side effects and the good response with low doses.

As conclusion, speech-induced lingual dystonia is an uncommon movement disorder that can face as an occupational phenomenon. Neurologists and other physicians need to be aware of this condition, even in the absence of other dystonic phenomena. Although a few cases have been reported to date, BoNT seem to be an effective treatment for this condition. Further studies are needed to clarify underlying mechanism and to evaluate the long-term efficacy of BoNT.

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## Author's contribution

Carlos Cores Bartolomé: conception, organization, and execution of the project writing of the first draft of the manuscript.

José Manuel Paz González: review and critique. Treatment of the patient.

María José García Antelo: review and critique. Diagnosis of the patient.

Diego Santos García: review and critique. Diagnosis and treatment of the patient.

## Conflicts of interest

None.

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## Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at [doi:10.1016/j.nrl.2021.05.010](https://doi.org/10.1016/j.nrl.2021.05.010).

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## Germinoma primario del sistema nervioso central como causa infrecuente de afectaciónependimaria difusa en el adulto

### Primary central nervous system germinoma as a rare cause of diffuse ependymal involvement in adults

*Sr. Editor:*

Los tumores de células germinales del SNC son proliferaciones neoplásicas de células germinativas de muy baja incidencia en el mundo occidental, representando menos del 0,5% de los tumores intracraneales primarios. En los países



asiáticos son, en cambio, más frecuentes<sup>1</sup>. Histológicamente se clasifican en diferentes subtipos, siendo el germinoma el más frecuente, representando hasta el 69% de los casos<sup>2</sup>. Suele diagnosticarse en la edad pediátrica, siendo el 90% de los pacientes menores de 25 años<sup>3</sup>. Sus manifestaciones clínicas varían según su localización. Exhiben características radiológicas diversas, presentándose frecuentemente como masas ovoides o lobuladas localizadas en la región pineal (38-57%), supraselar (34-49%) o ambas simultáneamente (5-10%). Únicamente un 3-5% de los casos se presentan en otras zonas incluyendo los ventrículos cerebrales<sup>4,5</sup>. Para su diagnóstico definitivo es necesario el estudio histopatológico. Su tratamiento se basa en radioterapia sola o asociada a quimioterapia, lográndose tasas de curación de hasta el 90%<sup>6</sup>.

Presentamos el caso de un varón de 31 años, natural de España, caucásico, fumador y sin otros antecedentes