Movement Disorders
CLINICAL PRACTICE

CASE REPORT

Emergence of Bruxism after Reducing Left Pallidal Stimulation in a Patient with Huntington's Disease

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Bruxism rarely appears in Huntington's disease (HD)[1] but represents a compromising symptom that puts considerable strain on the patient and family members. We present a 55-year-old female patient who had been subjected to bilateral internal globus pallidus (GPi) deep brain stimulation (DBS) to improve limb hyperkinesia. In the course of disease, the patient developed severe bruxism that appeared only if left GPi-DBS stimulation was reduced.

Case Report

Our patient used to work as a physician up to the age of 42 years with very few patient contacts in the last period of her professional activity. She received the genetically confirmed diagnosis of HD at 44 years of age. According to her friend, the onset of motor symptoms was at 37 years of age. Retrospectively, she developed her first psychiatric symptoms in her mid-twenties, suffering from emotional instability. According to family history, her mother was diagnosed with HD at the age of 40 years.

The patient presented to our inpatient clinic at the age of 45 years for the first time. Prevailing symptoms were psychomotor agitation and anxiety, signs of bruxism were not present. Hyperkinesia of the limbs was present only to a minor extent and disturbances in fine motor skills were mild.

The patient was treated with tiapride to control motor symptoms. Because higher doses of tiapride were not tolerated, an additional treatment with tetrabenazine was established that had only limited effects on hyperkinesia. A medication of sulpiride was prescribed to improve limb hyperkinesia while valproate was occasionally used because of newly developed myoclonus.[2]

In terms of non-motor symptoms, mirtazapine and duloxetine were added and did temporarily improve anxiety and depressive symptoms. Treatments with citalopram and quetiapine were less effective for mood stabilization. Zuclopenthixol and lorazepam were transiently applied to mitigate agitation. Table 1 provides an overview of medication and respective time periods of treatment.

During the disease course, the most compromising symptom of limb hyperkinesia was further progressing. Therefore—at the age of 47 years—the patient was subjected to bilateral GPi-DBS in an external neurosurgery unit. At that time, the patient suffered from mild orofacial dyskinesia but not from bruxism. Directly after DBS implantation, antidopaminergic treatment could be stopped because of a significant improvement of limb hyperkinesia[3] that remained constant over the years. Brain imaging is only available before DBS implantation, because the patient did not want to undergo further imaging procedures after surgery.

Eight years after DBS implantation, the patient was admitted to our inpatient clinic because of overall symptom deterioration. She was bedridden and suffered from severe dysphagia, dementia with mutism, and intermittent mild orofacial dyskinesia. Communication was not possible, and limb hyperkinesia was not present. After reduction of DBS intensity of the left GPi stimulation electrode to evaluate dysphagia, we discovered an immediately appearing severe bruxism that was not the case if we modified stimulation parameter of the right GPi. The reestablishment of left GPi DBS induced a rapid disappearance of bruxism without altering dysphagia. Symptoms such as chorea or dystonia were not affected by changes in DBS settings (Video S1).

Discussion

Bruxism has been reported occasionally in HD[1] and represents a compromising symptom that has not been associated with DBS before. We found that active DBS of the left GPi does adequately suppress bruxism in our patient. Because of the severity of bruxism when reducing stimulation intensity of left GPi, we recognized the risk of possible oromandibular and teeth injury.

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TABLE 1 Duration and daily dose of medication in our patient

Drug	Daily Dose (mg)	Estimated Treatment Duration (mo)
Citalopram	20	5
Duloxetine	60	>132
Lorazepam	0.5–1.5	>132
Mirtazapine	15	>132
Sulpiride	150	2
Tetrabenazine	31.25–87.5	24
Tiapride	50–500	48
Quetiapine	75	5
Valproate	1200	2
Zuclopenthixol	10	2

Therefore, we decided not to turn off DBS or reduce stimulation intensity for longer time periods so that putative long-term effects with DBS cessation such as limb hyperkinesia or dystonia could not be evaluated.

Importantly, our patient was known to suffer from orofacial dyskinesia before DBS implantation but not from bruxism. Bruxism, in general, is characterized by extensive teeth grinding and is considered as involuntary hyperkinetic motor disorder that can be difficult to differentiate from other oromandibular movements such as orofacial dyskinesia.[4] Although the extensive teeth grinding we observed meets the definition criteria of bruxism, this could, to some extent, include features of orofacial dyskinesia. However, the pronounced presentation of teeth grinding without hyperkinetic symptomatology in other body parts is best classified as bruxism in our view. Therefore, the beneficial influence of DBS can best be interpreted as attenuating effect on bruxism.

The etiology of bruxism has been controversially discussed (tardive[5] vs. HD-related[1]). In our patient, there was no history of treatment with classical, highly potent neuroleptic drugs for which tardive complications are commonly described. Atypical neuroleptic drugs, tiapride, and serotonin reuptake inhibitors (citalopram) were used for which bruxism is only an infrequent complication.[6] Therefore, a medication-induced etiology of bruxism in this case is rather unlikely. On the contrary, a causal link between long-term DBS and bruxism has to be considered.

In conclusion, our observation suggests an involvement of left GPi in the pathophysiology of bruxism. A possible association of DBS and bruxism should be considered in DBS-treated HD patients.

Author Roles

(1) Research Project: A. Conception, B. Organization, C. Execution; (2) Manuscript Preparation: A. Writing of the first draft, B. Review and Critique.

D.R.: 1A, 1B, 1C, 2A

C.S.: 1A, 1B, 1C, 2B

L.T.: 1A, 1B, 1C, 2B

All authors read and approved the content of the manuscript.

Disclosures

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Supporting Information

Supporting information may be found in the online version of this article.

Video S1. Appearance of bruxism in different settings of the GPi-DBS in HD. Patient's face appears in close up during different settings of GPi-DBS that are displayed in the lower right corner. The video was captured in consecutive movie sections that closely followed each other. Note that bruxism appeared immediately after reduction only of the left GPi stimulation amplitude.