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Tremor and Other Hyperkinetic Movements

Case Reports

Position-Dependent Dysfunction of Deep Brain Stimulation in Tourette Syndrome: Diagnostic Clues

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Abstract

Background: Detection of defective deep brain stimulation (DBS) contacts/electrodes is sometimes challenging.

Case Report: We report a patient with Tourette syndrome (TS), who presented with abrupt tic increase and mild generalized headache 9 years after DBS implantation. On the suspicion of a hardware defect, a fracture of the DBS electrode and extension lead was ruled out by radiography and standard implantable pulse generator readouts. Further investigation revealed position-dependent modifiable therapeutic impedances, suggesting an impaired contact of the extension lead/adaptor. After replacement normal impedances were recorded, and the patient fully recovered.

Discussion: In DBS dysfunction with inconspicuous hardware check, position-dependent defects might be suspected.

Keywords: Deep brain stimulation, hardware complications, headache, impedance, Tourette syndrome

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Ethics Statement: This study was performed in accordance with the ethical standards detailed in the Declaration of Helsinki. All patients have provided written informed consent.

Introduction

Deep brain stimulation (DBS) in patients with Tourette syndrome (TS) results in a tic reduction of about 40%.[1] Most often used targets are the globus pallidus internus (GPi) and thalamic nuclei (centromedian parafascicular/ventral oral internus nucleus [CMPF/Voi]).[1] The overall reported DBS hardware complication rate in patients with movement disorders is about 8%, mainly due to infections, lead migration, and fractures typically manifested with elevated therapeutic impedances.[2,3] In contrast, in patients with TS hardware complications and particularly infections seem to occur more often.[4]

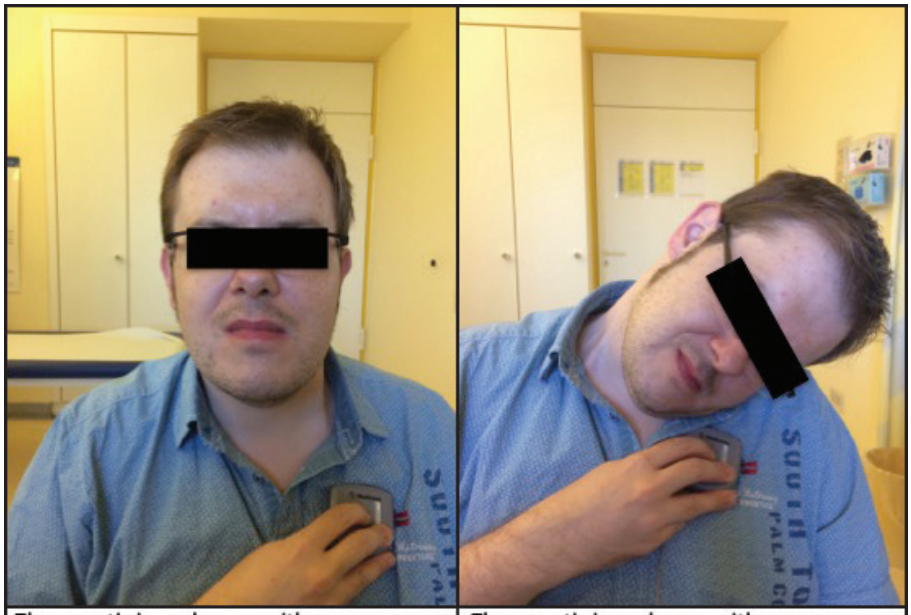
Case report

We report on a male patient with TS, who suffered from severe motor and vocal tics as well as obsessive-compulsive disorder and therefore underwent bilateral DBS electrode implantation (model 3387; Medtronic Inc., Minneapolis, MN) using computed tomography (CT)-stereotactic

surgery guided by magnetic resonance imaging and microelectrode recording in both the GPi and the CMPF/Voi at the age of 29 years. Electrodes were connected to an implantable pulse generator (IPG) (Soletra® Model 7426; Medtronic) on each side using a Y-junction. Stereotactic CT demonstrated appropriate placement of the DBS electrodes. After several adaptations of the stimulation settings, best clinical results were achieved with bilateral DBS of the CMPF/Voi. The patient was followed every 3–6 months in the long-term course. Forty months postoperatively, IPG replacement with switch to a dual-channel rechargeable IPG (Activa® RC; Medtronic) was performed. A specific adapter was used as an interface to allow connection of the lead extensions with the new IPG. The patient received a patient programmer to adapt stimulation intensities within a predefined range. Overall disease course was stable with marked improvement of tics and compulsions compared with the preoperative situation (last DBS settings: left: monopolar stimulation 6-/C+, 2.1mA, 60 μs, 130 Hz; right: monopolar stimulation 3-/C+, 1.5 mA, 60 μs, 130 Hz).

Nine years after surgery, the patient asked for an urgent appointment and reported an abrupt and marked deterioration of his tics since about 1 week. In parallel, he had developed an unspecific general headache, while his programmer indicated a DBS dysfunction. Clinical examination showed aggravated tics; however, the IPG check in supine position showed unremarkable therapeutic and single-contact electrode impedances. System check with the patient programmer neither notified any dysfunction. In addition, DBS electrode or extension lead fracture was excluded by radiography. Therefore, spontaneous fluctuations of tics were assumed and a slight augmentation of stimulation intensities was performed.

During reassessment 1 week later, the patient still reported worsened tics. Again, his programmer had displayed DBS dysfunction, which the patient reported to be possibly motion dependent. Therefore, we checked the DBS system once more and found out that therapeutic impedance of the left electrode was not measurable, when the patient held his head in upright position. Furthermore, the active monopolar contact 6 showed elevated impedances (>40,000 Ohm). Remarkably, therapeutic and electrode impedances were found to be perfectly normal (contact 6: 896 Ohm), when the patient inclined his head to the left (see Figure 1). To ensure movement-dependent variations in



Therapeutic impedances with upright head position			**Therapeutic impedances with inclined head to the left**		
	Left	Right		Left	Right
Programmed current flow	1.1 mA	0.8 mA	Programmed current flow	1.1 mA	0.8 mA
Resistance	**„High“**	1258 Ω	Resistance	**652 Ω**	1268 Ω
Voltage	9,309 V	1,047 V	Voltage	0,781 V	1,055 V

Figure 1. **Assessment of Therapeutic Impedances in Position-Dependent Manner.** Improved values with head inclination to the left are illustrated.

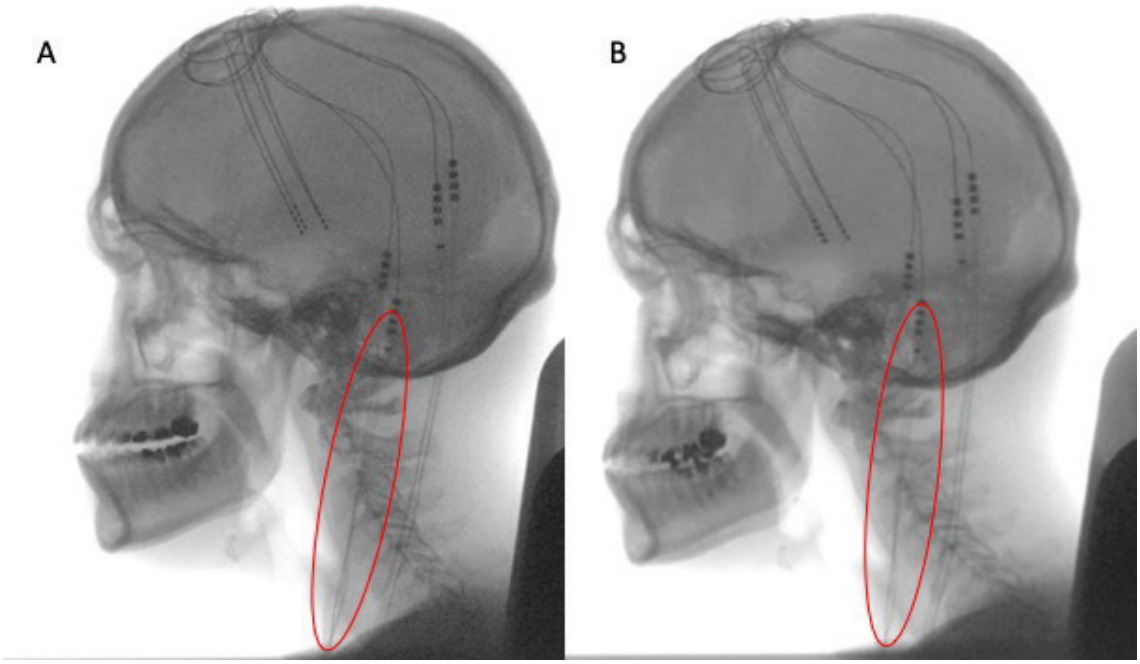


Figure 2. **Radiography in (A) Upright and (B) Inclined Head Position.** A mild traction of the cable (red circle) in the upright position might be observed.

impedances, the described procedure was repeated twice, resulting in the same effect, indicating an impaired contact of the left cable extension. This assumption was further supported by additional dynamic radiography, which showed a traction of the left extension lead, improving with inclination of the head to the left (Figure 2). As the inclined head position could not be hold on in daily practice for longer intervals, a formal clinical assessment of tic frequency could unfortunately not be performed.

We decided for replacement of the left extension lead and adaptors because an impaired contact seemed to be the most obvious cause. Postoperative IPG check showed regular impedances followed by a rapid improvement of the tics to previous levels and relief of headache. The further course was unremarkable with no episodes of dysfunction of the neurostimulation system.

Discussion

This case is instructive for two reasons. First, hardware defects are well-known complications in DBS, which are always considered when acute clinical worsening occurs. Yet, despite extensive hardware check and radiography to exclude fractures of the DBS electrode or the extension lead, the underlying cause and diagnosis was not established at first glance. Normal radiography does not exclude a hardware dysfunction and relevant changes such as a traction of the cable might only manifest very discreetly. In case of clinical DBS dysfunction without obvious evidence for elevated therapeutic or electrode impedances, a defective contact with fluctuating current flow might be considered and uncovered by investigating the IPG function in various head positions. Noteworthy, the patient reported in parallel to the tic increase newly developed generalized headache that relieved after restoration of continuous DBS. It can be speculated that the patient's headache might have been caused by recurrent on–off effects with fluctuating movement-dependent current flow. Even though unspecific, generalized mild headache might be considered as a clinical hint for DBS dysfunction caused by recurrent changes between on and off mode.

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