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Case report

Twiddler syndrome in a patient with tremor dominant Parkinson's disease. A case report and literature review

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ABSTRACT

Twiddler syndrome is described as a spontaneous rotation or intentional external manipulation of implanted cardiac or occasionally deep brain stimulation (DBS) devices. We report this hardware related complication in a patient with tremor dominant Parkinson's disease (PD), who underwent unilateral subthalamic nucleus (STN) DBS and subsequently developed twiddler syndrome. The clinical course of twiddler syndrome in this patient is described. Some surgical nuances which may prevent its occurrence are suggested. Our case report indicates that twiddler syndrome occurs in DBS patients. Impedance check of DBS hardware, plain chest X-ray, or palpation for a knobbly extension lead through the skin above the IPG allows the correct diagnosis and subsequently a prompt surgical revision. Our subsequent literature review revealed only 10 patients with twiddler syndrome in DBS patient population worldwide. This number may suggest that this syndrome may be unrecognized or underreported, given the number of patients with movement disorders implanted with DBS hardware worldwide.

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1. Introduction

Twiddler syndrome describes spontaneous rotation or intentional manipulation by its bearer of the implanted internal

pulse generator (IPG) in its subcutaneous or subpectoral pocket causing subsequent hardware malfunction. Twiddler syndrome was first presented in 1968 by Bayliss et al. [1] in a patient with an implanted cardiac pacemaker producing loss of function. The same complication causing hardware

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malfunction was further reported in patients with phrenic and vagal nerve stimulators, subcutaneous ports for chemotherapy or baclofen pumps.

Twiddler syndrome in a movement disorder patient was first described by Geissinger and Neal [2] in 2006. Until today, only a few case reports describing this phenomenon in patients implanted with DBS hardware have been presented [2-8]. Twiddler syndrome may cause fracturing of not only the extension lead but also the distal part of the electrode, or even its upward withdrawal from brain parenchyma [3,6,7]. This complication requires implantation of a new DBS electrode, with some risk of intracerebral hemorrhage. To prevent this complication twiddler syndrome should be properly recognized and prompt surgical revision should follow.

In this report we describe the case of twiddler syndrome in a patient with an implanted single-channel St. Jude IPG for treatment of tremor dominant Parkinson's disease. Clinical presentation, recognition, and surgical technique preventing electrode fracture are discussed. Moreover, our case report has inspired us to perform a literature search of twiddler syndrome in movement disorders patients.

2. Case report

A 67 year old female with 11 years' history of tremor dominant PD was scheduled for bilateral staged STN DBS procedure. The PD symptoms were more pronounced on the right side than on the left. The most incapacitating rest and postural right-sided tremor was poorly controlled by pharmacotherapy. After expressing a written informed consent the patient underwent successful left STN DBS procedure with implantation of a single-channel IPG (St. Jude USA) in the left subclavicular area. Initiation of the stimulation at the second postoperative day eliminated the rest and postural components of the right-sided tremor. The patient gained independence in performing activities of daily living with subsequent postponing of the planned right-sided STN DBS procedure. There was good tremor control over an 8 months' follow-up period. The patient suddenly experienced rapid reappearance of her right-sided tremor, which was even more pronounced than before the DBS procedure. An examination of the DBS hardware revealed high impedance of the DBS system (over 30 units in the case of an implanted Libra St. Jude IPG). This high impedance suggested a fracture of the implanted DBS hardware. Closer examination of the implanted IPG side revealed knobby structures of the extension cable palpable though the skin. Moreover the IPG could be easily moved in the subcutaneous pocket but not turned around. Plain X-ray exams of the patient's chest, neck, and head showed hardware complication. Chest A-P X-ray revealed an extremely coiled extension cable, particularly above the IPG. Neck X-ray showed complete extension cable fracture just above the patient's left ear. There was no evidence of the slippage of the connector placed in the left parietal region or the withdrawal of the electrode from the brain parenchyma (Fig. 1A and B).

Our patient denied any manipulation of her IPG. She also did not complain of pain, tightness along the connection cable

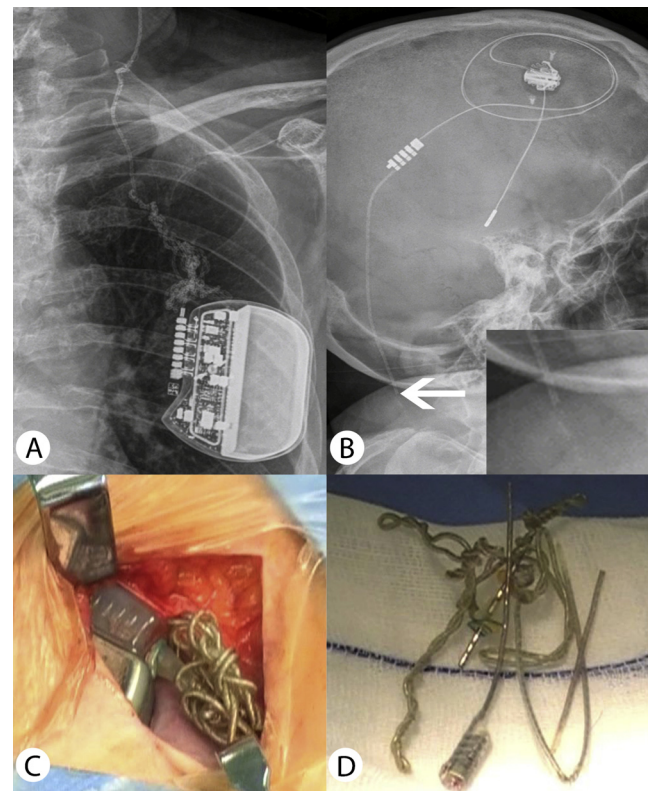


Fig. 1 – (A) Chest X-ray showing the placement of the internal pulse generator in the chest wall with extremely coiled connection cable just above the generator. (B) The lateral skull X-ray demonstrating the DBS electrode in lace with connector placed at high parietal region. The magnification window and the white arrow indicate the point of fracture of the connection cable. (C) The intraoperative view of the extremely coiled connection cable just above the single-channel internal pulse generator (Libra St. Jude). (D) The braid and removed connection cable.

or itching sensation in her infraclavicular incision. The patient's history was negative for any psychiatric co-morbidity including obsessive or compulsive behavior.

The hardware problem was explained to the patient and a revision surgery was offered. During the surgery, after preparation of the connection cable evident braiding of the connection cable just to the IPG (Fig. 1C and D) was identified.

The connection cable was thus removed and replaced with a new one. The connector was freed from scar tissue and again attached to the parietalis fascia with two non-absorbable sutures. The pocket size was reduced by stitching its inferior, medial, and lateral aspects using 0 Vicryl sutures. The IPG fitted comfortably in the reduced pocket and was sutured with one silk suture through the IPG's anchor hole to the muscle fascia. It was additionally immobilized by stitching the overlying subcutaneous tissue. The postoperative period was uneventful. The patient made full recovery with regained antitremor benefit on her right side. The follow-up period of the next 6 months has been free of any hardware malfunction in this patient.

3. Discussion

DBS procedures have become a routine management option for patients with incapacitating movement disorders such as PD, essential tremor, and dystonia. This treatment modality has been available for nearly 25 years and has been more broadly applied worldwide in the recent years. It is estimated that since its introduction approximately 100,000 DBS devices have now been implanted for various movement disorders. According to observations of some authors twiddler syndrome – although rare – may affect approximately 1% of patients [9]. This can suggest that this complication has been an unrecognized and underreported phenomenon. Since the first DBS stimulation procedure in 1999 at our department we have encountered this complication in only 1 patient with tremor dominant PD. So far we have implanted 293 DBS electrodes in 182 patients, which means that the single case of twiddler syndrome represent 0.5% of operated patients and 0.3% of implanted DBS electrodes. Burdick et al. [6] among 226 patients implanted with 326 electrodes encountered this phenomenon in 1.3% operated patients and 1.4% implanted electrodes. Most authors report not the exact number of DBS electrodes implantations but mention that this complication was first encountered after roughly over 200 procedures in their series [3,8]. In the literature search we have found only 7 articles reporting 10 patients with the diagnosis of twiddler syndrome requiring surgical revisions in all cases [2–8]. The articles presented in a chronological order describe small case series or case reports and are summarized in Table 1.

The most common presenting symptom of twiddler syndrome was reappearance of PD symptoms or tremor in PD or ET patients. In some cases the tremor was even more pronounced than before the original surgery, thus indicating a rebound phenomenon – just like in the case of our patient. Other symptoms associated with twiddler syndrome reported by the patients include the postauricular pain, pain along connection cables, and a sensation of IPG mobility [2,5,6]. Due to the relatively small number of reported twiddler syndrome cases in DBS patients population it is not easy to find predisposing factors for the syndrome's development. Most authors suggest that predisposing factors may include advanced age with more loose subcutaneous tissue, adipose patients with thick adipose tissue layer precluding proper immobilization of IPG in the pocket [2–8]. Additional factors, such as creating an excessively large pocket or a patient with compulsive-obsessive behavior may also play an important role [9].

In all patients the twiddler syndrome was recognized by performing chest X-ray with typical appearance of coiled connection cable just above the implanted IPG. Some authors have found high impedance mostly above 4000 Ohms, indicating a fracture of Medtronic DBS hardware [3,5,6]. We have encountered the occurrence of twiddler syndrome in a patient with implanted Libra IPG which has only one anchoring dock, like Medtronic Activa SC 37603. This may theoretically constitute a risk factor for twiddling, as an IPG with two anchor docks is better anchored to the surrounding tissue. This is a preliminary observation which can be

supported by further case reports of twiddler syndrome in the future, as the number of DBS implants performed worldwide keeps growing.

To adjust malfunctioning DBS hardware in twiddler syndrome a revision surgery was undertaken in all reported cases [2–8]. This complication can lead not only to fracture of the connection cable but also causes the risk of a fracture of the extracranial part of the electrode or its upward withdrawal from the brain parenchyma [3,6,7]. Braiding of the extension cable results in its shortening and tension on the connector and the distal part of the electrode. Early in our DBS procedures the connector was not additionally sutured to the parietal fascia which caused slippage of the connector to the neck area with subsequent fracture of the extracranial part of the electrode or even its upward migration from the brain parenchyma. To prevent this complication we routinely placed the connector at the high parietal area and anchored it with two non-absorbable sutures to the parietal fascia. This surgical maneuver used also in this patient prevented the connector from slipping down with potential damage to the DBS electrode. Fracture or dislodgment of DBS electrode or electrodes due to twiddler syndrome has been reported in the literature [3,5,7]. In the report by Gelabert-Gonzales et al. [3], 2 patients developed intracranial complications due to twiddler syndrome. 1 patient experienced dislodgment of the right DBS electrode and fracture of the left DBS electrode, and the other patient experienced the fracture of the left DBS electrode. Additionally 1 of 3 patients in the report by Burdick et al. and 1 patient in the case report by Samuelsson and Blomstedt experienced dislodgment of DBS electrodes [6,7]. This case involved a revision surgery with implantation of a new DBS electrode, which was more complicated than simple replacement of the connection cable or surgical revision of the IPG pocket.

Patients with recognized twiddler syndrome require a prompt revision surgery to prevent the above-mentioned serious complications. The revision surgery requires replacement of the malfunctioning hardware and usually surgical reduction of the IPG pocket. Various techniques have been used to prevent the IPG from repeated twiddling, such as stitching the pocket to reduce its size, placing the IPG in a polyester battery pouch [5], or placement of the IPG beneath the pectoralis muscle [6,7]. Proper securing of the IPG or others parts of the DBS hardware are of paramount importance to prevent recurrent twiddling. The incidence of recurrent twiddling among DBS patient is high and was found in 4 of 10 patients with twiddler syndrome [5–7]. This observation may also suggest that implementing proper revision surgery will not deter a determined pathological twiddler from manipulating the implanted DBS hardware. In our reported case during revision surgery we anchored the connector with two silk sutures to parietal fascia immobilizing it at the parietal region and preventing it from twiddling as had been done in the original surgery. In this way we protected the DBS electrode (the most valuable component of DBS hardware) from twiddling. To our knowledge there is only one case report describing a patient with PD and trichotillomania who through compulsive behavior consisting of picking skin over the connector side produced coiling of the distal part of the extracranial electrode around the extension cable connector

Table 1 – Case series or case report of twiddler syndrome in movement disorder patients presented in chronological order.

Authors and publication year	Indication	Target	Etiology of TD	Presenting symptoms	Occurrence of TD after initial surgery in months	Diagnosis	Type of hardware malfunction	Treatment
Geissinger and Neal [2], 2006	ET	Vim	Unconscious twiddling due performance of normal daily activities	Reemergence of tremor, post auricular pain, sensation of IPG rotation	6 ms	Chest X-ray, high impedance	The connection cable fracture	Surgical revision, placing an IPG in polyester battery pouch
Gelabert-Gonzales et al. [3], 2007	PD	STN	Conscious twisting of IPG by the patient in the abdominal pocket	Reemergence of PD symptoms	36 ms	Chest X-ray	Dislodgment of the right DBS electrode	Repositioning of the right DBS electrode and replacement of the left DBS electrode
Patient 2	PD	STN	Conscious twisting of IPG by the patient in the abdominal pocket	Reemergence of PD symptoms	24 ms	Chest X-ray	Fracture of the left DBS electrode Fracture of the left DBS electrode	Replacement of the left DBS electrode
Israel and Spivak [4], 2008	ET	Vim	Unconscious twiddling	Itching sensations at infraclavicular incision	2 ms	Chest X-ray, normal impedance	Twiddling of extensions lead	Revision of connection cables, suturing IPG to clavicular periosteum
Astradsson et al. [5], 2010	Dystonia	GPI	Conscious and active twisting of IPG	Reemergence of dystonic symptoms	30 ms	X-ray, high impedance over 4.000 Ohms	Rupture of connection cables bilaterally	Multiple revisions and rigorous fixation of IPG
Burdick et al. [6], 2010	ET	Vim	Unconscious manipulation	Reemergence of tremor, pain along the cables, age more than 70	Several weeks	X-ray, impedance more than 2.000 Ohms	Rupture of connection cable two times	Replacement of the connector cables, suturing IPG to fascia
Patient 2	ET	l Vim	Conscious manipulation	Tremor recurrence, sensation of IPG mobility	17 ms	X-ray, impedance more than 2.000 Ohms, inability to interrogate the IPG	The fracture of left extension cable. Dislodgement of left DBS lead. Twisting of the right extension cable	Multiple revision of pocket, suturing IPG to fascia
Patient 3	PD	STN	Sensation of IPG mobility	Sensation of IPG mobility	Approximately 6 ms	Patient history	Loose pocket	Revision of the pocket
Samuelsson and Blomstedt [7], 2014	Dystonic tremor		Conscious manipulation of the IPG due to it rotation	Sensation of IPG mobility Postauricular tenderness	5 ms and 7 ms	X-ray	Fracture of the left connection cable, Dislodgement of the right DBS lead	Replacement of the left connection cable, placement of IPG beneath pectoralis muscle
Silva et al. [8], 2014	PD	STN	Unconscious manipulation	Recurrence of PD symptoms	4 ms	X-ray, high impedance	Rapture of connection cables	Replacement of the connection cables

ET, essential tremor; PD, Parkinson's disease; Vim, nucleus intermedius of the thalamus; STN, subthalamic nucleus; IPG, internal pulse generator; ms, months; TD, twiddler syndrome.

[9]. This resulted in wound infection and repeated revisions consequently requiring DBS hardware removal. The connector in this case was placed in high cervical area rather than in parietal region where twiddling is impossible due to smaller amount of subcutaneous tissue.

4. Conclusion

An increasing number of implanted DBS devices will make twiddler syndrome part of hardware-related complications which should be solved properly and timely by DBS multidisciplinary team members. Prompt recognition and proper surgical technique at first DBS implantation (suturing and placing the connector at parietal region with a pocket tailored to IPG dimensions) may substantially reduce this complication in the future.

Conflict of interest

None declared.

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None declared.

Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; Uniform Requirements for manuscripts submitted to Biomedical journals.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at [doi:10.1016/j.pjnns.2015.10.004](https://doi.org/10.1016/j.pjnns.2015.10.004).

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