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Case Report

Asymptomatic cable twisting in a patient with impending Twiddler syndrome detected during deep brain stimulation surgery for Parkinson's disease: A case report

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ABSTRACT

Background: Deep brain stimulation (DBS) has consistently demonstrated high efficacy and safety in patients with Parkinson's disease. Twiddler's syndrome is a rare occurrence of hardware failure in patients undergoing neuromodulation. We report here a case of subclinical cable twisting jeopardizing Twiddler's syndrome in a patient with Parkinson's disease who underwent DBS surgery targeting the globus pallidus internus (GPI).

Case Description: A 70-year-old woman with a 7-year history of Parkinson's disease refractory to medication was referred to our department for treatment of involuntary movements of the left hand and leg. She underwent right GPI DBS implantation. Left GPI DBS implantation was subsequently planned to manage resting tremors that developed in the right leg after the first surgery at around one year after the first surgery. During a routine check-up before the second surgery, we incidentally detected Twiddler's syndrome. The patient showed no neurological deficits in the left extremities, the same as before right GPI DBS. We performed left GPI DBS concomitantly with the revision of the implantable pulse generator and extension wire.

Conclusion: Twiddler's syndrome is a rare complication of DBS. Subclinical risk of cable twisting jeopardizing Twiddler's syndrome is rarely detected without clinical indications of hardware failure. Neurosurgeons should be cognizant of and regularly monitor the implanted device in case serious complications occur.

Keywords: Deep brain stimulation, Parkinson's disease, Twiddler's syndrome

INTRODUCTION

Deep brain stimulation (DBS) is the treatment of choice for patients with movement disorders, including Parkinson's disease, generalized dystonia, tremor, and Tourette syndrome. DBS has consistently demonstrated high efficacy and safety for movement disorders.[12] Therefore, it is anticipated that the number of DBS surgeries performed will increase over time alongside an accompanying increase in the incidence of related morbidity.[11] According to a systematic review by Hamani and Lozano [8], after DBS surgery, there is a 9% risk of morbidity associated with the device itself, a 2.8% risk of intracerebral hemorrhage, a 0.7% risk of developing a permanent neurological deficit, and the mortality rate is approximately 0.4%. All complications of DBS can be divided into those associated with (1) the surgical procedure, (2) the device itself, and/or

(3) the electrical stimulation applied. Hardware failure, lead migration, or lead fracture are among the most frequent DBS-related complications.

Twiddler's syndrome is a rare complication that occurs in patients undergoing neuromodulation with an implanted pacemaker or defibrillator[3] and causes hardware failure in about 1% of cases.[10] Twiddler's syndrome was first reported in 1968 to implantable cardiac devices[3] and has since emerged as a known side effect of other implantable devices, such as those used in spinal cord stimulation systems and DBS systems.[4] The syndrome develops as a result of the patient's intentional or unintentional movement of the device, which causes it to rotate in the pocket, create torsion, and dislodge the implanted lead.[10] In this report, we describe a case of subclinical cable twisting jeopardizing Twiddler's syndrome in a patient with Parkinson's disease who was undergoing DBS targeting the right globus pallidus internus (GPI). This report adds to the limited literature on Twiddler's syndrome in patients with Parkinson's disease following DBS implantation.

CASE DESCRIPTION

A 70-year-old woman with a 7-year history of medication-refractory Parkinson's disease was referred to our department for treatment of involuntary movements of the left hand and leg. She had no relevant psychiatric history, such as personal or family history of obsessive-compulsive disorder. She was independent in activities of daily living on medication; however, she was suffering from severe on/off motor fluctuations. Her preoperative neuropsychic evaluation was not significant for dementia or mental disorders. The Mini-Mental State Examination was 27/30, and the Montreal Cognitive Assessment was 27/30. Frontal Assessment Battery was 15/18. In addition, the Geriatric Depression Scale was 1/15, and the State and Trait Anxiety Inventory score was 31/80.

She underwent elective DBS targeting the right GPI. The DBS lead (model B33015, Medtronic, Minneapolis, MS) was implanted in the right GPI under local anesthesia. The implantable pulse generator (IPG; B35200, Medtronic) was implanted in the left subclavicular pouch without suture fixation under pectoral fascia on the same day under general anesthesia. Since the patient is right-handed, she requested to implant the IPG in the left subclavicular. A skull radiograph was performed immediately after surgery to confirm the location of the lead [Figure 1a]. Functional outcomes were favorable, and medical therapy could be markedly reduced. The patient was discharged with no evidence of surgical complications or adverse events. No impairments or neurological deficits were evident on routine follow-up visits.

She had kept having benefits from the first DBS therapy and was then planned to undergo left GPI DBS for the right hemibody symptoms one year later. Around one year after DBS surgery, she developed a resting tremor in the right leg, and DBS implantation in the left GPI was planned. Before the surgery was performed. However, a chest X-ray during a routine check-up incidentally revealed that the extension wire was twisted in the chest cavity, and the IPG was flipped left to right [Figure 2]. Normal impedance measurements suggested that her DBS system was intact and functioning, and she had no recollection of turning or twisting the IPG actively or passively. She did not complain of any tenderness along the extension wire path. We performed left GPI DBS concomitantly with the revision of the implantable pulse generator and extension wire. We also fixed the IPG to the pectoral fascia by sutures. The left GPI DBS was uneventful, and no adverse events have been reported as of the 6-month follow-up.

DISCUSSION

Although DBS of the GPI has been proven to be a safe and effective treatment for Parkinson's disease, there is a risk of hardware, surgical, and/or stimulation complications.[11] Twiddler's syndrome has been reported in 1.3% of patients with movement disorders, and DBS leads were the cause in 1.4% of reported cases.[4,10,13] Twiddler's syndrome has been well described in the cardiac literature for many decades, especially in patients with pacemakers and implantable cardioverter-defibrillators.[3] It occurs when the IPG moves within an overly large pocket. As a result, the IPG can repeatedly twist until the leads are tightly coiled and break or pull away. Additional risk factors have been reported, including psychiatric disorders, weight loss, advanced age, female sex, and looping the lead outside the pocket, as well as loss of large amounts of subcutaneous fat in previously obese women that makes them more susceptible to the IPG movement in the pocket.[7,15]

Our patient with Parkinson's disease showed clinical improvement after right GPI DBS and did not recall having manipulated or moved the IPG intentionally. Identifying a tendency for the IPG to twist within the subcutaneous pocket is difficult, and it is unclear whether the movement disorder in Parkinson's disease plays a contributing role. Neuropsychological and psychiatric profiles may not be able to detect any characteristics of Twiddler's syndrome, which could include anxiety, dementia, depression, obsessive-compulsive behaviors, or paranoia. However, our patient had a risk factor of poorly fixed IPG in the loose subcutaneous space due to obesity.

The development of Twiddler's syndrome is characterized by the recurrence of clinical symptoms and device failure resulting from manipulation or movement. Pain at the IPG site or along the extension wire path may accompany these clinical symptoms.[1,6] The normal impedances found

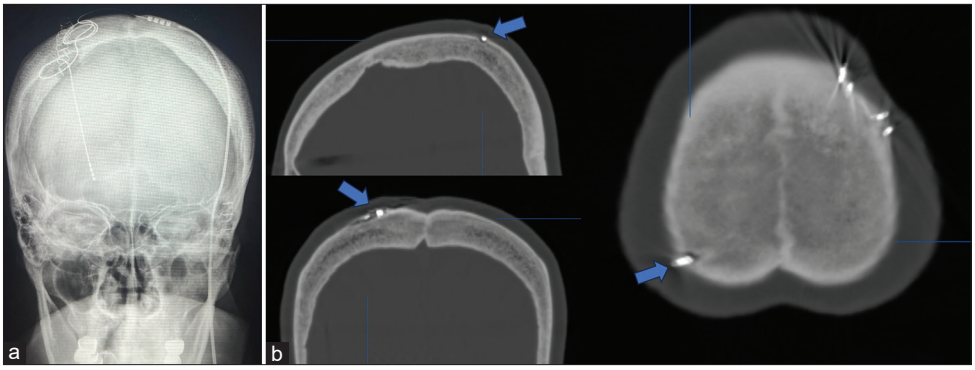

Figure 1: (a) Postoperative skull radiograph image showing brain electrode implantation on the right GPI. (b, arrows) Head computed tomography scans show a small groove made in the parietal region of the skull to countersink each connector during deep brain stimulation surgery.

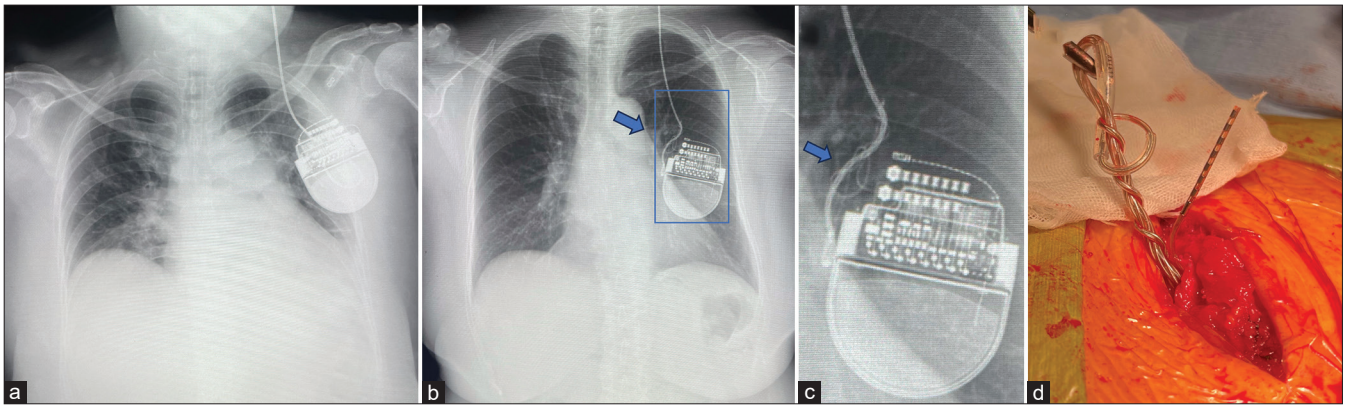

Figure 2: Images of the implantable pulse generator (IPG). (a) Chest radiographs showing the IPG in the left chest wall immediately postoperative and (arrows b and c) coiling of the extension wire around one year after deep brain stimulation surgery targeting the right globus pallidus internus. (c) A magnified image reveals the IPG has flipped left to right. (d) Intraoperative finding showing multiple coils in the extension wire near the IPG.

in this patient suggest that her DBS system was intact and functioning. Nevertheless, the described presentation suggested a potential hardware issue, and surgical intervention likely prevented a further malfunction of the DBS system.[9] When a DBS device fails to relieve symptoms that were previously responsive to stimulation, Twiddler's syndrome should be ruled out as a potential cause of lead or hardware failure by looking at the leads on an X-ray. Diagnosis is usually made using plain radiographs.[2] Given that intracranial electrode retraction may occur in severe cases, brain imaging should be considered.[15] The presenting problem in Twiddler's syndrome is a double-helix or braided pattern in the extension wire [6,9], which is similar to the finding seen on chest X-rays in our case [Figure 2]. Interestingly, this was a case of subclinical cable twisting jeopardizing Twiddler's syndrome. The patient may subclinically manipulate IPG, and this may jeopardize the DBS system. There was no intracranial electrode retraction or lead breakage, and the patient did not show any symptoms resulting from hardware failure. The syndrome was detected only incidentally when planning for left GPI DBS as a second surgery. This condition may be partly due to poorly fixed IPG and loose subcutaneous space, especially because of the patient's age. There was no potential psychiatric disorder due to neuropsychic evaluation. A countersinking procedure during lead externalization can prevent the extension wire from twisting and pulling away from the intracranial electrodes. During lead externalization, in our department, we drill a small groove in the parietal region of the skull to countersink each connector to minimize its prominence and prevent delayed scalp erosion [Figure 1b].

Treatment of Twiddler's syndrome usually involves surgical revision, fixation of the IPG, and the replacement of any damaged hardware. The IPG can be stabilized surgically to prevent it from twirling by fixing the IPG within a tight-fitting subcutaneous pocket using a nonabsorbable silk suture that is passed through the designated IPG hole and fastened to the muscle, fascia, or artificial pouch.[5,11,14] To prevent recurrence, the IPG pocket should be kept as compact as possible, and the device should be

sutured to muscle. However, if leads have been damaged or intracranial electrodes have been pulled away, more extensive revision surgery and replacement of the hardware may be required.[7]

CONCLUSION

Twiddler's syndrome is a rare but serious complication of DBS resulting from IPG manipulations by the patient. Neurosurgeons should be mindful that the patient may subclinically manipulate IPG, and this may jeopardize DBS system dysfunction named Twiddler syndrome. Clinicians should carefully monitor the device in patients treated with DBS and be cognizant that it could be damaged during procedures or surgeries.

Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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