Case Report

Twiddler’s syndrome: Between mechanical recoil and behavioral troubles✩✩

Mohamed El Minaoui, MD✩, Wassim Beladel, MD

Cardiology Department, University Hospital Agadir, Medical School of Medicine & Pharmacy Ibn Zohr University, Quartier Tilila Bp 7519 Agadir Al Fidia Cp80060, Agadir, Morocco

A R T I C L E   I N F O

Article history:
Received 19 June 2022
Revised 3 July 2022
Accepted 6 July 2022

Keywords:
Twiddler's syndrome
Implantable defibrillator
Behavior troubles

A B S T R A C T

A 72-year-old male with severe dilated cardiomyopathy, benefited of implantable cardiac defibrillator implementation. Device control shows high impedance. On X-ray, electrodes were completely twisted in the generator pocket, they were replaced and the generator was fixed to pectoralis-major fascia. Nurses report patient abnormal movements, scratching implantation area. This was identified as probably the trigger of the complication.

© 2022 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/)

Introduction

Twiddler's syndrome (TS) was first described in 1968 by Bayliss et al. [1] in patients with pacemakers. It consists of the rotation of the generator by its axis, which causes the torsion electrodes. This torque may lead to fracture or displacement of electrodes, causing dysfunction device. It has also been described in patients with implantable defibrillator [2]. We present a case of a patient with this complication in which a precipitating factor could be identified in an old man with behavior troubles.

Case report

This is a 72-year-old man with 6 years history of worsening exercise intolerance compatible with heart failure in relation to a severe dilated cardiomyopathy, who was initially admitted for cardiac decompensation. In the subsequent study, coronary angiography found nonsignificant irregularities in the coronary arteries and left ventricular ejection fraction was at 28%. We decide to set up a double chamber defibrillator for primary prevention of sudden cardiac death (SCD) which was performed without complications. At follow-up, the patient

Abbreviations: TS, Twiddler’s syndrome; DCM, dilated cardiomyopathy; SCD, sudden cardiac death; ICD, implantable cardiac defibrillators; IPG, internal pulse generator.

✩ Competing Interests: All authors declare no conflict of interest.

✩✩ Funding: None.

✩ Corresponding author.

E-mail addresses: elm.mhd@gmail.com (M. El Minaoui), beladelwassim@gmail.com (W. Beladel).

https://doi.org/10.1016/j.radcr.2022.07.035

1930-0433/© 2022 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/)
remained stable without ventricular arrhythmias or inappropriate discharges of the defibrillator. Periodic controls parameters were always correct.

Two months later, he was admitted to emergency department for lipothymia. Also, family reports onset of behavioral disorders. Interrogation of implanted device showed normal functions without recorded tachycardia episodes, however.

Investigations found a recent frontal ischemic stroke. Patient was discharged with a neuropsychiatric follow-up. One month later, the control of the device shows a high impedance of electrode stimulation above 2000 Ω (ohm), suggesting a fracture thereof.

In the chest X-ray, electrodes were completely twisted in the generator bag, with the traction of the intracavitary electrode (Fig. 1).

Electrode extraction was carried out by continuous traction (Fig. 2), and others were replaced, chest X-ray control was done (Fig. 3). Also, the generator was fixed to the fascia of the pectoralis major.

In the days after the intervention, the nurses report repetitive abnormal movements of patient touching and scratching the device implantation area, he unconsciously repeats these stereotyped movements. This movement was identified as probably trigger of the complication, so the patient was advised to avoid this maneuver in the future.

Fig. 1 – Chest X-ray showing the coiling of the device leads around the device in a 72-year-old man.

Fig. 2 – Operative view of the extraction of the twisted leads of the ICD.

Fig. 3 – Chest X-ray control after replacement of new leads.
The patient was integrated to a cardiac rehabilitation program with neuropsychiatric and cardiological follow-up.

Discussion

TS is a rare mechanical cause of permanent pacemaker’s dysfunction. The frequency would vary between 0.07% and 7%.

Firstly described by Bayliss in 1968, it was defined as manipulation of the cardiac implantable device around its central axis within skin pocket which causes the coiling and dislodgment of electrodes [1,3]. This phenomenon usually occurs in the first year following device’s implantation; the earliest case described on the pacemaker had a delay of 17 hours after implantation [4]. More recently, a late Twiddler syndrome has also been described [5].

Furthermore, cases of similar syndrome have been reported with implantable cardioverter-defibrillators and cardiac resynchronization therapy [6,7]. In fact, the first case of Twiddler’s syndrome associated with automatic implantable defibrillators (ICD) was described by Veltri et al. in 1984 [2].

The main risk factors predisposing to the development of TS are: advanced age, female sex, obesity, cognitive, and behavioral disorders. In the elderly, the increased laxity of the subcutaneous tissues favors the displacement of the material, also the creation of too large pockets for internal pulse generator (IPG) facilitates their rotation into the skin pocket most often during physical activities [4,8,9]. In some isolated cases, it has been possible to demonstrate active manipulation of the generator by the patient [10].

Otherwise, a recently published series of cases have suggested that the construction of an implantable pulse generator (IPG) may itself be a predisposing factor: the presence of a single anchoring hole intended for attachment exposes more to displacement of the IPG [11].

Clinically, the displacement of the device is itself painless, the voluntary or unintentional manipulation is often denied by the majority of patient. TS is characterized by the reappearance of neurological or cardiac disorders attributed to the pacemaker dysfunction in dependent patients, which can have dangerous consequences. In our case, the dysfunction is totally asymptomatic but the consequences could be fatal for the patient.

The other clinical manifestations, displacement or fracture of the electrodes can cause minor complications, such as increases in impedance or stimulation threshold [12], or stimulation of the skeletal muscles, but there may also be major complications, such as failures in under or overdrive, as well as increases in the defibrillation threshold [13].

Positive diagnosis of this syndrome is very simple; it’s based on the chest X-ray which shows the displacement of the device with rotation of the conductive wires [7].

Therapeutic procedure is based on the uncoiling of the electrodes, the implantation of new leads with repositioning and fixation of the device [7,9,14]. Some authors encourage the reutilization of the same electrodes.

Active fixations of leads and suturing of the device to the fascia can also help prevent displacement [1,15]; some teams reserve this option for patients with mental disorders and elderly ones with loose subcutaneous tissues [4]. We can also implant it below the pectoral muscle [16], that’s what we did in our patient by the use of a Dacron patch, which promotes tissue growth around the device and ensures better fixation. Also, minimizing the pocket size without redundant space around the generator is essential to prevent the development of this syndrome [4,5].

In addition to optimizing fixation procedures, the education of patients, especially the elderly, is one of the most important means of avoiding manipulation of the device [7].

Radiological follow-up of predisposed patients may also help to make an early diagnosis and thus avoid major complications.

Conclusion

Twiddler’s syndrome is currently recognized as an uncommon mechanical cause of intracardiac device malfunction, especially in the elderly. Its diagnosis is very easy based on chest X-ray. Optimization of the device fixing measures and educating patients to avoid any manipulation of the pulse generator are the most important methods of prevention.

Acknowledgments

The authors thank Dr Thomas Chastre Rhythmologist from Cardiology Department Poissy Saint Germain en Laye Hospital.

Patient consent

Patient consent was obtained from the patient.

References

[1] Bayliss CE, Beanlands DS, Baird RJ. The pacemaker-Twiddler’s syndrome: a new complication of implantable transvenous pacemakers. Can Med Assoc J 1968;99:3713.
[2] Veltri EP, Mower MM, Reid PR. Twiddler’s syndrome: a new twist. PACE 1984;7:1004–9.
[3] Hill PE. Complications of permanent transvenous cardiac pacing: a 14-year review of all transvenous pacemakers inserted at one community hospital. Pacing Clin Electrophysiol 1987;10:564–70.
[4] Fahraeus T, Hijer CJ. Early pacemaker Twiddler syndrome. Europace 2003;5:279–81.
[5] Dursun I, Yesildag O, Soylu K, Yilmaz O, Yasar E, Meric M. Late pacemaker Twiddler syndrome. Clin Res Cardiol 2006;95:547–9.
[6] Sharifi M, Inbar S, Neckels B, Shook H. Twiddling to the extreme: development of Twiddler syndrome in an implanted cardioverter-defibrillator. J Invasive Cardiol 2005;17:195–6.
[7] DeMarco DC, Xuereb RG. 'Twiddling' of the pacemaker resulting in lead dislodgement. Malta Med J 2009;21(3):38–41.
[8] Gupta R, Lin E. Twiddler syndrome. J Emerg Med 2004;26:119–20.
[9] Castillo R, Cavusoglu E. Twiddler's syndrome: an interesting cause of pacemaker failure. Cardiology 2006;105:119–21.
[10] Parsonnet V, Bernstein AD, Neglia D, Omar A. The usefulness of a stretch-polyester pouch to encase implanted pacemakers and defibrillators. PACE 1994;17:2274–8.
[11] Sobstyl MR, Ząbek M, Bruszakiewicz-Kuźmicka G, Pasterski T. Dual anchor internal pulse generator technique may lower risk of Twiddler's syndrome: a case series and literature review. Neuromodulation 2017;20(6):606–12.
[12] Saliba BC, Ghantous AE, Schoenfeld MH, Marieb MA. Twiddler’s syndrome with transvenous defibrillators in the pectoral region. PACE 1999;22:1419–21.
[13] Buitleir M, Canver CC. Twiddler's syndrome complicating a transvenous defibrillator lead system. Chest 1996;109:1391–4.
[14] Nicholson WJ, Tuohy KA, Tilkemeier P. Twiddler’s syndrome. N Engl J Med 2003;348:1726–7.
[15] Mandal M, Pande A, Kahali D. A rare case of very early pacemaker Twiddler's syndrome. Heart Views 2012;13(3):114–15.
[16] Boyle NG, Anselme F, Monahan KM, Beswick P, Schuger CD, Zebede J. Twiddler’s syndrome variants in ICD patients. PACE 1998;21:2685–7.