

Twiddler's syndrome presenting as life threatening electrical storm

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Abstract

A 75-year-old man underwent implantation of a single chamber implantable cardioverter defibrillator (ICD) for primary prevention of his underlying severe non-ischaemic cardiomyopathy. Thirteen months later, he presented to the emergency room (ER) with inappropriate ICD shocks as a result of over sensing of the right ventricular lead and double counting of the right atrial signals. The chest X-ray (CXR) revealed a right ventricular ICD lead displaced into the right atrium with coiling in the pocket. The right ventricular shocking coil was noted at the tricuspid annulus. The lead was removed from the pocket and was replaced with a new lead. This case represents the classical Twiddler's syndrome in an ICD with potential lethal consequences.

Keywords: Case report, Twiddler's syndrome, Implantable cardiac defibrillator, Electrical storm, Recurrent shocks.

Introduction

Twiddler's syndrome (TS) is a rare but potentially lethal complication of pacemaker or ICD device placement.¹ It is characterized by device malfunction due to painless dislodgement of cardiac leads resulting from some form of manipulation by the patient. Twiddler's syndrome has never been reported from Pakistan. Here we present a case of Twiddler's syndrome in an elderly man who had an ICD implanted for primary prevention thirteen months prior to the presentation.

Case Report

A seventy five years old man presented to the emergency room (ER) of Aga Khan University Hospital (AKUH), Karachi, in November 2015, with inappropriate ICD shocks. A single chamber ICD (Ellipse St. Jude Medical, CD 1277-36Q) with an active fixation ventricular lead (Durata 7120 Q-58 St. Jude Medical, USA) had been placed in October 2014 for primary prevention of his underlying severe non-ischaemic cardiomyopathy. The ICD pocket was appropriately matched with the device size.

Postoperatively, acceptable lead impedance of 590 Ω , RV lead sensing at 15 mV and RV lead pacing threshold at 0.5V at 0.5ms. The CXR showed satisfactory lead position. (Figure-1A) He had no known psychiatric disease and his follow up visits were uneventful with satisfactory wound healing and evolving device parameters.

His past medical history was significant for hospitalization in 2012, with acute pulmonary oedema and atrial fibrillation with rapid ventricular rate (RVR). An echocardiogram showed an ejection fraction of 20% with global hypokinesia. He underwent coronary angiogram; in view of low EF, which revealed normal epicardial coronaries. Hence was treated as non-ischaemic cardiomyopathy and was optimized on anti-heart failure therapy. He was started on anticoagulation with warfarin, keeping target INR of 2.0 - 2.5, for atrial fibrillation with rapid ventricular rate as his CHADS₂VASC score was 3 while HASBLED score was 1. He had no signs of twiddling with the ICD either through clinical history or on device checkup parameters at three monthly followup visits.

He presented thirteen months later with multiple shocks delivered by the ICD, while doing his routine household chores. Immediately after the shock, he had brief episode of syncope after which he was rushed to the hospital. At presentation to ER, he was severely anxious with stable vital parameters. On interrogation of the device, a series of eight inappropriate shocks and numerous non-sustained episodes of ventricular tachycardia (VT) were noted in the ICDlog. The device was deactivated by application of a magnet. The R-wave sensing had decreased to 0.5 mV and impedance was within the normal range (690 Ω). During the coronary care unit (CCU) stay, he had runs of ventricular tachycardia and was managed on the lines of electrical storm and was started on intravenous amiodarone infusion. CXR showed most part of RV lead was pulled back into the right atrium with significant twisting of lead making a figure of 8 around the pulse generator. The right ventricular shocking coil was pulled towards the tricuspid annulus. (Figure-1B) During interrogation of the device, there was p-wave oversensing during sinus tachycardia by the ICD lead. This double counting was responsible for the inappropriate shocks. The cause of the over sensing was a lead dislodgment due to rotation of the device by manual manipulation by the

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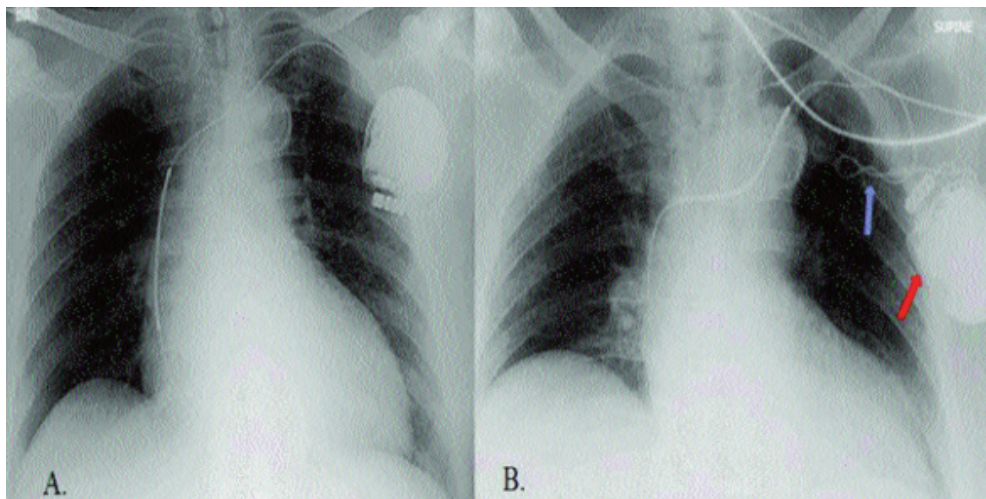


Figure-1: A. Frontal chest X-ray post ICD placement. B. Thirteen months later repeat X-ray shows twiddling of the ICD leads around pulse generator (blue arrow) with RV lead pulled back into right atrium and making a figure of 8 around the pulse generated (red arrow).

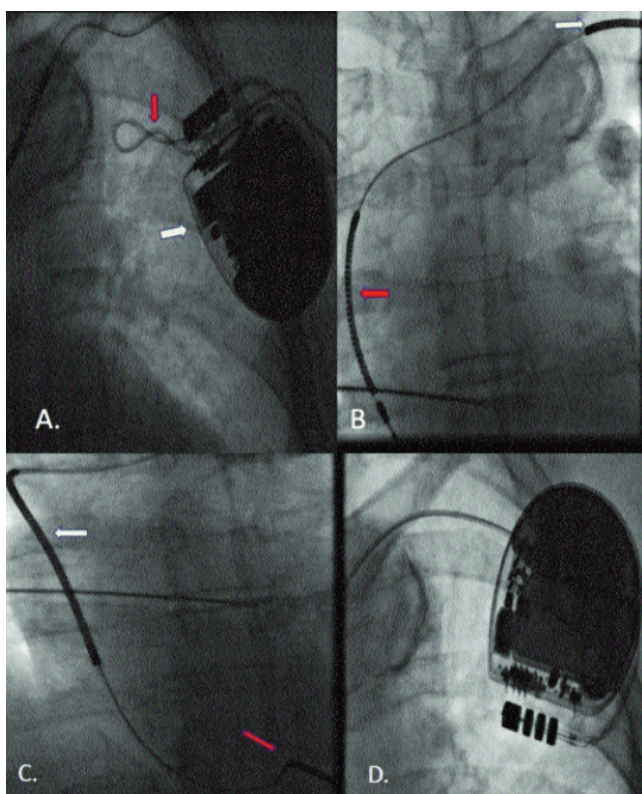


Figure-2: A. Fluoroscopic images shows twiddled lead (red arrow) around the pulse generator, (white arrow) counter clockwise rotation of pulse generator. B. (white arrow) primary shocking coil pulled towards the left subclavian vein, (red arrow) secondary shocking coil pulled into the right atrium. C. New Lead inserted with primary shocking coil (white arrow) at the junction of superior vena cava and right atrium. While a secondary shocking coil (red arrow) is seen at RV apex. D. Final position of pulse generator after lead replacement.

patient. Upon questioning, he admitted to have manipulated the device within its pocket repetitively because he "did not get used to it very well!"

He was shifted to the fluoroscopy suite. A new incision was made through the old scar. The tightly twisted lead (Durata 7120 Q-58 St. Jude Medical) was identified and dissected free. A figure of "8" loop was noted. The lead was unscrewed and removed without any complications. A new lead (Durata 7120 Q-58, St. Jude Medical) was implanted at the RV apex

via the left subclavian approach. (Figure-2) The ICD was placed in the pocket and anchored to the underlying muscle with silk 0 for stability. The pocket was closed with vicryl sutures. Postoperatively, acceptable lead impedance, capture and sensing thresholds were obtained. The CXR showed optimal position of lead. No further complications were detected at one year of follow-up. His heart failure therapy and anticoagulation with warfarin were continued.

Discussion

Twiddler's syndrome (TS) is known to be an uncommon cause of device malfunction caused by excessive twisting of the pacemaker or ICD device in the pocket, resulting in dislodgement of leads and device malfunction.² The prevalence of this syndrome was reported to be 0.07 - 7 % in cases with a pacemaker.^{2,3} It was first reported by Bayliss et al. in 1968.⁴ In ICD patients, the Twiddler's syndrome was most frequently observed in the era of abdominal implantations. Very few publications have been reported in ICD placed in the pectoral region.^[5] Twiddler's syndrome may present as a rare (ICD) malfunction that is potentially fatal.

TS can result from subconscious, inadvertent or deliberate rotation of a pacemaker or an ICD in its subcutaneous pocket. Thus, the device may be turned over and over until the lead is twiddled and may get dislodged by traction. As the tip of the lead is pulled back towards the pocket according to its position, it may produce failure to pace, diaphragmatic contraction by phrenic nerve stimulation, pectoral muscle, or brachial

plexus stimulation resulting in rhythmic arm twitching and may wrap around the pulse generator.⁶ In our patient, there was oversensing and failure to pace.

TS is more serious and potentially life threatening in cases with an ICD because treatment of ventricular arrhythmias would be disabled and inappropriate shocks could be delivered due to oversensing.⁵ As a worst case scenario, an inappropriate ICD therapy may be proarrhythmic and may lead to sudden cardiac death. As in our case, the patient recognized and reported that he consciously twiddled with the device and ultimately came to emergency room with inappropriate shocks which were potentially life threatening. There was also sustained monomorphic VT noted in the CCU while the device was disabled. This could have in reality caused serious consequences with the device failing to recognize true VT.

TS has been described to present in cases where the subcutaneous tissues are lax, the generator is untethered in its pocket, or the size of the pocket exceeds that of the generator. Patients who are most at risk of this condition include middle-aged, obese, women and patients with mental disorders such as dementia.⁷

There are several risk factors for TS, such as obesity, excessive movements of the upper limbs, active manipulation of the generator, large size pocket, or dementia. However, preventive maneuvers like anchoring the generator to the bottom of the pocket or subcutaneous infraclavicular region implants instead of abdominal implants should be considered.⁴ Furthermore, Twiddler's syndrome usually occurs one year or less after implantation of the device. However, cases of late presentation have also been reported in literature.⁸ In our case, the patient was well oriented, had no psychiatric disorder in past and the device was implanted subcutaneously in the left infraclavicular region.

The best approach to TS is prevention.⁹ Careful matching of the size of the device pocket to the implanted device is essential for patient comfort as well as to limit the available room in the pocket for pulse generator rotation. In addition, carefully anchoring the transvenous lead to the pectoral muscle is essential. When TS has been confirmed, the leads are typically reanchored to the pectoral muscle with multiple anchoring sleeves.⁹ In addition, the device is then aggressively anchored to the pectoral muscle which was done in our case as well.

Conclusion

While twiddling is certainly uncommon, it remains a significant clinical problem, as damage to or dislodgement of leads may result in inappropriate shocks with possible injury or death. Furthermore, in patients in whom the device was placed for primary prevention, dislodgement of the ICD lead is frequently asymptomatic as these patients nearly always have the ICD programmed to minimize ventricular pacing. As a result, the only symptom in such patients may be failure to detect or treat potentially lethal ventricular arrhythmias. With more and more ICD implants been undertaken in Pakistan, we have to be cognizant of this life threatening complication.

Informed Consent: Written informed consent was obtained from the patient for their anonymized information to be published in this article.

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Disclaimer: This case report has never been published or presented in a conference previously.

References

1. Boyle NG, Anselme F, Monahan KM, Beswick P, Schuger CD, Zebede J, et al. Twiddler's syndrome variants in ICD patients. *Pacing Clin Electrophysiol* 1998; 21: 2685-7.
2. Konishi H, Tokano T, Nakazato Y, Komatsu S, Suwa S, Komatsu K, et al. Twiddler's syndrome detected by patient's complaint of implantable cardioverter-defibrillator rotation in the subcutaneous pocket. *J Arrhythm* 2012; 28: 239-41.
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. Bayliss CE, Beanlands DS, Baird RJ. The pacemaker-twiddler's syndrome: a new complication of implantable transvenous pacemakers. *Can Med Assoc J* 1968; 99: 371-3.
5. Spencker S, Poppelbaum A, Müller D. An unusual cause of oversensing leading to inappropriate ICD discharges. *Int J Cardiol* 2008; 129: e24-6.
6. Nicholson WJ, Tuohy KA, Tilkemeier P. Twiddler's syndrome. *N Engl J Med* 2003; 348:1726-7.
7. De Buitelir M, Canver CC. Twiddler's syndrome complicating a transvenous defibrillator lead system. *Chest* 1996; 109: 1391-4.
8. Arindam P, Achyut S, Imran A, Sanjeeva CN, Rabin C. Pacemaker twiddler's syndrome: Review through a case report. *J Cardiovasc Dis Res* 2015; 6: 148-51.
9. Parikh V, Barsoum EA, Morcus R, Azab B, Lafferty J, Kohn J. Unique presentation of Twiddler's syndrome. *World J Cardiol* 2013; 5: 207-9.