ABSTRACT

Twiddler’s syndrome refers to malfunction of pacemaker or implantable cardioverter-defibrillator (ICD) resulting from deliberate or unintentional twisting of pulse generator in device pocket, resulting in lead dislodgment. We report a case of a 65-year-old woman with multiple ICD shocks and antitachycardia pacing events on device interrogation, while she was unaware of the ICD discharges. Twiddler’s syndrome is rare and mostly occurs in obese or elderly because of loose subcutaneous tissue allowing pulse generator to rotate in its pocket. It can be prevented by patient education against hand manipulating the generator. Smaller device pocket or suturing generator to fascia can also prevent future episodes.

Key words: Implantable cardioverter-defibrillator (ICD), shock, twiddler’s syndrome

INTRODUCTION

Twiddler’s syndrome occurs as a result of pacemaker or implantable cardioverter-defibrillator (ICD) pulse generator twisting, resulting in the loss of device function. Therefore, twiddler’s syndrome should be recognized and corrected early. It may be asymptomatic or may present with a wide spectrum of symptoms, depending on the site of the lead dislodgment.

CASE PRESENTATION

We report a case of a 65-year-old woman with medical history of heart failure with reduced ejection fraction, hypertension, and diabetes mellitus type II who had multiple ICD shocks and anti-tachycardia pacing events on device interrogation, while she was unaware of the ICD discharges. The patient denied shortness of breath or chest pain. She had a history of ventricular fibrillation arrest in the past and had undergone placement of an ICD 7 months ago. Device interrogation revealed lead sensing and captured failure with drop-in shock impedance. Electrogram showed lead noise misinterpreted as ventricular fibrillation by the device, resulting in inappropriate ICD shocks [Figures 1-4]. Chest X-ray revealed that ICD lead had dislodged from the right ventricle (RV), retracted into axillary vein, and coiled around the generator while generator itself was flipped around its axis [Figures 5-7]. She underwent lead revision with extraction of the dislodged lead and insertion of new RV lead.

DISCUSSION

First introduced in 1968, twiddler’s syndrome refers to malfunction of pacemaker or ICD, resulting from deliberate or unintentional twisting of pulse generator in device pocket. It can be prevented by patient education against hand manipulating the generator. Smaller device pocket or suturing generator to fascia can also prevent future episodes.
Jabri, et al.: ICD shock secondary to twiddler syndrome

The incidence of twiddler’s syndrome lies between 0.07% and 7%.\(^\text{[2]}\) Twiddler’s syndrome mostly occurs in obese or elderly because of loose subcutaneous tissue allowing pulse

Figure 1: Electrogram from implantable cardioverter-defibrillator showing noise being interpreted as ventricular fibrillation.

Figure 2: Electrogram from implantable cardioverter-defibrillator showing attempted antitachycardia pacing.

Figure 3: Electrogram from implantable cardioverter-defibrillator showing implantable cardioverter defibrillator shock.
generator to rotate in its pocket. Other risk factors include female gender, weight loss, upper limb exaggerated movements, device manipulation, or large pocket size relative to the defibrillator.\[3-5\]

Most cases occur in the 1st year after implantation of the defibrillator;\[6,7\] however, twiddler’s syndrome can still occur after that.\[8\] The established guidelines for ICD placement follow-up are recommended after 72 h from placement and then after 2–12 weeks. Thereafter, in-person or remote follow-ups in 3–6 months can be performed.\[9\] These follow-up intervals are assigned to identify dysfunctioning ICDs. Twiddler’s syndrome can be asymptomatic\[8\] or represented with various symptoms depending on the final site of the displaced lead. Most symptoms occur due to missed capture, stimulation of nearby structures, or improper exaggerated response to an input.\[10\]

Twiddler’s syndrome can be life-threatening due to ventricular arrhythmias and usually requires rapid intervention with lead revision. A plain chest X-ray can quickly identify lead dislodgment and twiddling of pulse generator. To best aid in diagnosis of twiddler’s syndrome, an electrocardiogram, chest X-ray, and pacemaker interrogation should be obtained as routine workup.\[11\] To restore the ICD function, correction of lead dislodgment, new lead installation, or pulse generator relocation can also be considered.\[10\] Anchoring sutures to the pectoralis fascia can be performed to prevent the re-twisting of the generator or leads. Another approach
described in the literature to prevent the dislodgment of the lead was nonabsorbable antimicrobial pouches.\cite{12} In the study by Osoro et al., the rate of twiddler’s syndrome recurrence with nonabsorbable antimicrobial pouches used in correction of lead dislodgment was compared to other approaches, including sutures to the underlying pectoralis fascia.\cite{12} The results showed a rate of 0% recurrence for the antimicrobial pouches compared to 50% recurrence to other approaches, with a \( P < 0.05 \).\cite{12}

Patient education against hand manipulating the generator is another major determinator for twiddler’s syndrome recurrence, and therefore, concealing should be provided. Patient selection may also play a role in recurrent cases of twiddler’s syndrome. Patients with mental disorders or with high potential for psychoorganic syndrome will tend to disregard the manipulation instructions.\cite{11} Moreover, appropriate follow-up visits should be instructed to patients to aid in diagnosis of missed asymptomatic cases like our case. More frequent device interrogation sessions may be performed in the high-risk population.

CONCLUSION

Twiddler’s syndrome is a vital lethal diagnosis that necessitates early corrective measures. Diagnosis requires high level of suspicion due to the variety of presentations including asymptomatic patients to others with random complaints related to the site of the device dislodgment. Proper device interrogation visits should be advised to patients for better detection.

Declarations of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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