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Case Report **Journal of MAR Cardiology (Volume 4 Issue 1)**

Twiddler's Syndrome

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Introduction:

Twiddler's syndrome was first described by Bayliss, et al. in 1968 when a patient manipulated and rotated the pulse generator in the pocket (ref1). In a pacemaker-dependent patient, it can be a lethal complication. The manipulation results in the rotation of the pacemaker generator on its long axis followed by the coiling of lead resulting in lead retraction, lead damage, lead fracture and/or insulation leakage. The treatment involves readmission, repositioning of the dislodged leads and suture fixation of the lead and pulse generator within its pocket. The syndrome was originally described with pacemakers but the condition has also been reported with the implantable cardioverter-defibrillators. The condition typically presents with device malfunction and occurs when the patient either consciously or unconsciously twists and rotates the implanted device in its pocket, resulting in torsion and dislodgement of the implanted lead. We report such a case of an elderly female with device rotation in the pocket and active fixation lead twined on its own and retracted in the pocket.

Case Report:

80 years old female patient, a known case of coronary artery disease, with angioplasty to LAD with stent implantation done in 2012. She presented as a survivor of sudden cardiac arrest (SCD) and was referred for further treatment. Her check angiography showed a patent stent and no significant disease. Her 2D echocardiography showed Severe tricuspid regurgitation with LVEF 35%. Her Holter study done for 24 hours showed Non-Sustained Supraventricular Tachycardia (NSVT). She underwent single chamber Automatic Implantable Cardioverter Defibrillator (AICD) implantation on 19/09/2019 for secondary prevention of SCD.

Soon after discharge, she had restlessness, inability to sleep, and urinary retention because of a urinary tract infection for which she was treated with intravenous antibiotics according to the culture and sensitivity of the bacteria by the home care. She had hyponatremia with disorientation due to low oral intake and improved after Tolvaptin was added to her regimen.

In June 2020, during the Covid-19 lockdown, the relative made a call, stating that the patient felt the device has moved from its original position and dislodged, but the patient didn't do a physical visit into the device clinic due to Covid-19 scare. The patient eventually came to the device clinic in Oct 2020 and the device was rotated in the pocket. The right ventricle (RV) defibrillator lead sensing and pacing parameters were deranged. The fluoroscopy image revealed the findings consistent with Twiddler syndrome (fig1).

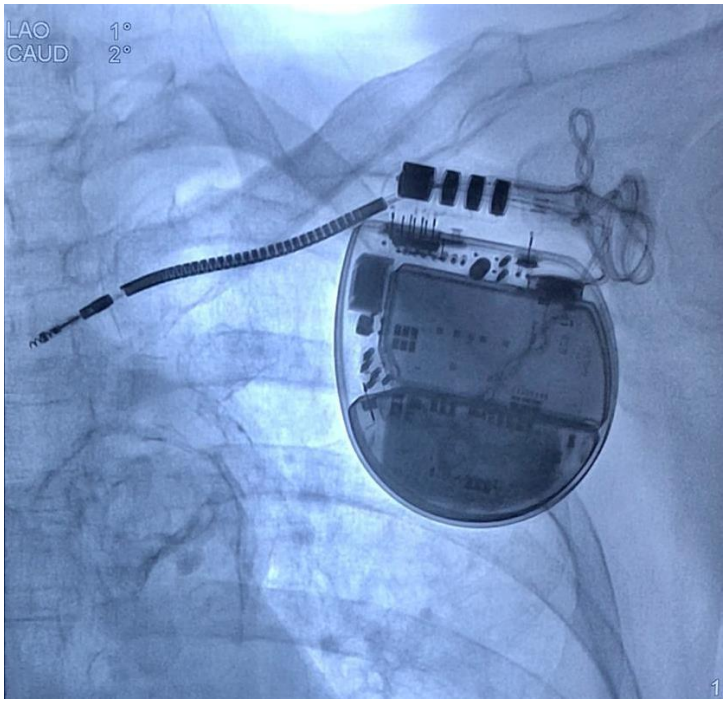

Fig 1: Fluoroscopy image showing RV lead coiled in the pocket

Fig 2: Active fixation RV defibrillator lead knotted and twisted hence discarded

(a) **(b)**

Fig 3 (a & b): AICD extraction demonstrated significant coiling of lead

During the AICD revision in the catheterization lab, the device was rotated in the pocket, the stay suture on the device was broken. Significant knotting of active fixation RV defibrillator lead was apparent and it was discarded (fig2&3). The patient underwent a successful new AICD implantation procedure. To avoid recurrence, the device was placed deeper in the pectoralis major muscle, and the stay suture was placed to the generator and the muscle.

A detailed discussion with the relatives revealed the constant meddling with the device by the patient during restlessness and disorientation. The relatives were apprised of the problem and counseled to avoid such a situation in the future. Ongoing follow-up has been satisfactory.

Discussion:

Twiddler's syndrome is a rare complication after pacemaker implantation and is caused by the conscious or unconscious manipulation at the implantation site by the patient resulting in device malfunction. The reported incidence is around 0.07-7% and usually occurs within the first year following implantation with the earliest case reported at 17 hours after implantation. In a pacemaker-dependent patient, it can be a lethal complication. Furthermore, in patients with a defibrillator, it is dangerous because of inappropriate shocks due to loss of adequate sensing and capture and possible false treatment of malignant ventricular arrhythmias (ref2).

The patient-related factors like pediatric age group, elderly, obese, female gender, psychiatric illness, cognitive dysfunction, as well as the operator-related factors like making large pocket size relative to the device size, are the various risk factors that increase the propensity of this syndrome. Various mechanisms have been proposed with the primary focus being upon the scratching that leads to rotation of the pacemaker generator causing stretching and dislodgement of leads which probably rewind on their own. The patients usually present with syncope due to underlying disease of bradyarrhythmias, diaphragmatic pacing due to phrenic nerve stimulation, rhythmic arm twitching due to stimulation of brachial plexus, or inappropriate shocks due to malsensing of defibrillators (ref3).

The chest X-ray and/or fluoroscopy are simple and easily available investigations for diagnosis. The treatment includes urgent temporary pacemaker implantation if the patient is pacemaker dependant, replacing the lead, minimizing the pocket size, suture fixation of the pulse generator, and use of a Dacron patch, if need be, to promote tissue growth around the device. It is equally important to counsel the patient and educate the family about living with a pacemaker and devices.

Conclusion:

Twiddler's syndrome should always be considered as a cause of pacemaker or device failure when the patient presents within one year of implantation. The easily available first-line investigations for its diagnosis are a simple ECG and chest X-ray/ fluoroscopy. The readmission for pocket revision and in most cases new lead placement increases the cost of the patient. Proper patient education and counseling to prevent further manipulation are very important in long-term management.

References

1.Jackson J Liang, Eric R Fenstad ; Twiddler's syndrome; thelancet.com Vol 382 December 14, 2013

http://dx.doi.org/10.1016/ S0140-6736(13)61419-1.

2.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: 195-196

3.Andre Briosa e Gala,Anthony Dimarco; Twiddler Syndrome: An Unusual Cause of Implantable Cardioverter-Defibrillator Failure Mayo Clin Proc. July 2019;94(7):1296-1297 https://doi.org/10.1016/j.mayocp.2019.02.032