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Case Report

Twiddle me this? Twiddled (Twiddler) Syndrome: A Case Report

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Abstract

Background: Twiddler syndrome is a rare but known complication after pacemaker (PM) placement. Causes of Twiddler syndrome have been described in case reports throughout the last fifty years, which has further elucidated our understanding of risk factors and mechanisms of PM dysfunction

Summary: We describe a case of Twiddler syndrome caused by migrating PM due to a lack of security to the pectoral fascial layer.

Conclusion: Due to the increasing number of iatrogenic causes of PM dysfunction, we propose differentiation from Pacemaker Twiddler Syndrome with known manipulation and Pacemaker Twiddled Syndrome, which includes iatrogenic causes of PM dysfunction. Additionally, the case highlights the importance of a diagnosis in the emergency department, cardiology consultation, and prompt pacemaker replacement.

Keywords: cardiology; ecg; aicd; case report; twiddler syndrome

Introduction

Twiddler syndrome is a rare but known complication after pacemaker (PM) placement. Bayliss et al. first documented this complication of implantable pacemakers in 1968. Since then, Twiddler syndrome has been documented though case series and case reports highlighting the different causes of PM malfunction with a reported incidence of less than 1%.

We report the case a 65-year-old female with iatrogenic Twiddler syndrome, as there was no reported manual manipulation, as the name may implies, with documented incomplete anchoring of pacemaker to the pectoral facial layer. Diagnoses was made in the emergency department by a combination of electrocardiography and x-ray and admitted to inpatient services for subsequent definitive care. The emergency medicine clinician must recognize this syndrome as misdiagnoses may lead to fatal consequences if gone unrecognized.

Case report:

A 65-year-old female with a history of hypertension, diabetes mellitus type 2, and sick sinus syndrome with dual-chamber pacer placed in 2003 presented to the emergency department with fatigue, dyspnea, and weakness over the past month. During her evaluation, she was noted to have profound orthostatic hypotension, bradycardic to the 50s and a new systolic murmur.

An ECG (Image 1) was obtained in the emergency department and compared to a prior ECG (Image 2) 5 years prior, revealing a ventricular paced rhythm with independent non-paced atrial depolarizations.

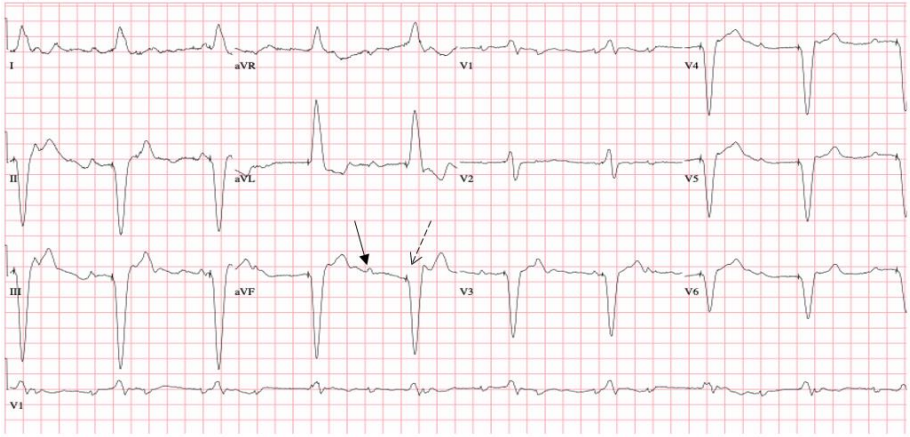

Image 1: ECG on presentation, p waves (solid black arrow) and ventricle pacing spike (dotted black arrow). ECG: AV dissociation with ventricular paced rhythm

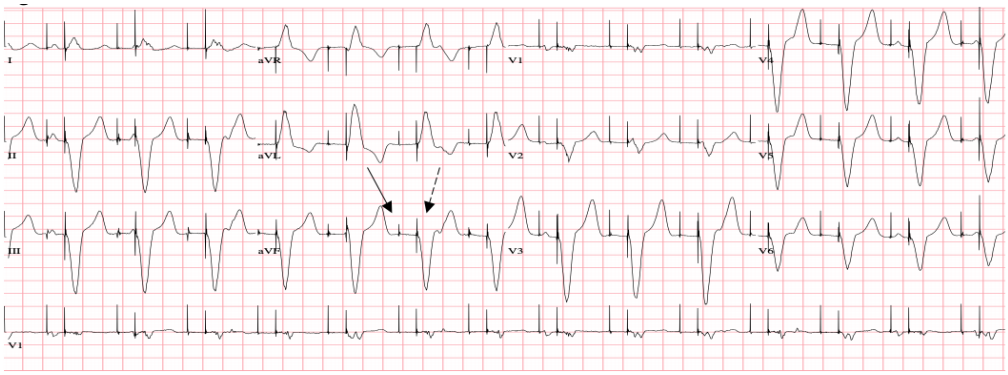

Image 2: ECG 5 years prior to presentation; atrial pace spike (Solid black arrow) and ventricle pacing spike (Dotted black arrow). ECG: Dual atrial-ventricle pacing.

The patient had a chest x-ray (Image 3) which revealed proximal pacemaker leads coiling.

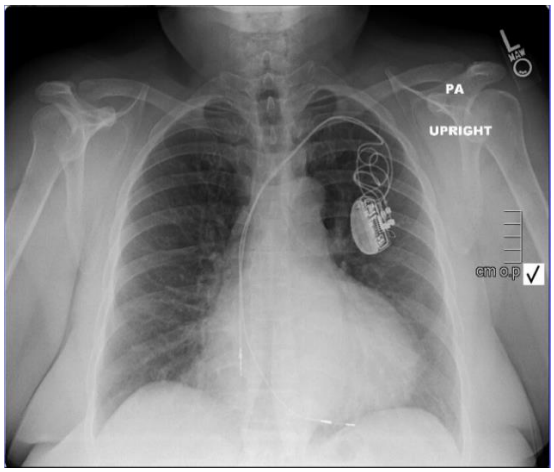

Image 3: Chest X-ray on presentation. Twisted pacer wires (white arrow) causing malfuction of patient's pacemaker

The patient was admitted for a pacemaker replacement. The patient consistently denied manual manipulation of her pacemaker to the admitting medicine and cardiology team, although she did note that she felt the pacemaker move at times while sleeping.

During the pacemaker replacement procedure, the interventional cardiologist noted multiple twists of the pacemaker leads with right atrial lead dislodgement. The pacemaker generator did not appear to be tacked down with sutures and was displaced in the subcutaneous fat layer instead of the pectoralis fascial layer. Both the pacemaker and the right atrial lead were extracted and replaced. The patient was discharged the next day. Subsequent ECGs showed normal sinus rhythm with dual-chamber pacing.

Discussion:

We present a case of iatrogenic Pacemaker Twiddlers Syndrome (PTS) discovered on radiographic and electrophysiologic testing obtained in the emergency department.

Bayliss et al. first described this complication of implantable pacemakers in 1968.1 Pacemaker Twiddlers Syndrome (PTS) was initially defined as the rotation of the pacemaker pulse generator (PMPG) within the pectoral pocket, occurring with and without manual manipulation. Once the pocket became too capacious, pacemaker (PM) leads twisted around the PMPG, a la spaghetti on a fork.1 Although initially described as the now known reeling variant, many case reports have described other variants such as the ratcheting and coiling variant of the pacemaker leads.1-4 These variants may result in lead retraction and displacement or lead fractures.

The reported incidence of PTS is close to <1%, documented by Fahraeus et al. following up on a series of 17,000 pacemaker placements.5 Risk factors for PTS include obesity, existing psychiatric illness, excessive movements of the upper limbs, active manipulation of the generator, and large-sized pockets.2,6 Although initially thought to occur with the first-year post-implementation, there have been more published case reports of 'late onset' Twiddler Syndrome in which PM malfunction has occurred many years after implementation.5,7

In this present case, patient's PM was functioning for at least 15 years per PM interrogation but became symptomatic 16 years after implantation.

To date, three case reports have described iatrogenic PTS. 6,8,9 Examples include:

- Incompletely anchored sutures
- Physical and occupational therapy involving the upper extremity causing PM manipulation.
- Use of orthopedic corset brace over the PM.

The case presented is another example of lack or loss of suture anchoring, highlighting the importance of securing the PMPG to avoid future complications. Additionally, this case exemplifies that PTS can occur without manual manipulation of a PM as the name implies.

Conclusion:

Although the original definition of PTS includes malrotation with or without manual manipulation, the name "twiddler" may insinuate a culprit causing the twiddling, i.e., the patient.

We propose distinguishing between manual manipulation (twiddler) vs. iatrogenic (twiddled) causes of pacemaker and lead malrotation by differentiating these two different processes as Pacemaker Twiddler Syndrome vs. Pacemaker Twiddled Syndrome. Although less significant clinically, this distinction will hopefully broaden clinicians' differential of causes of PM and lead malfunction, leading to less patient blame and stigma.

Nevertheless, pacemaker malfunction from dislodgement or fracture of pacemaker leads has potentially fatal consequences and should be recognized by the emergency medicine physician.

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