

Twiddler's Syndrome: a Rare Complication of Pacemakers

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Abstract

Twiddler's syndrome is a rare severe entity first described in 1968 related to transvenous pacemaker placement, involving the pulse generator in a subcutaneous pocket, and the coiling, displacement, or fracture of the leads causing stimulation failure and symptoms recurrence needing immediate revision. Major risk factors for the loss of pacemaker function include bad generator fixation, manipulation, psychiatric disorder, obesity, advanced age, and an oversized pocket. The objective is to emphasize the recent case studies published in this Journal.

Keywords: deep brain stimulation, movement disorders, Twiddler's syndrome.

Dear Journal of Clinical Medicine of Kazakhstan Editors,

Twiddler's syndrome (TS) is a rare severe complication of transvenous pacemaker placement first described in 1968, involving the implanted pulse generator (IPG) in the subcutaneous pocket, with excessive coiling, displacement, or fracture of the leads [1-6]. The stimulation failure, and symptoms recurrence, often demand surgical revision. Risk factors for partial or total loss of pacemaker function include bad generator fixation, manipulation, psychiatric disorder, obesity, advanced age, and an oversized pocket [1-6]. This letter aims to emphasize the two recent case studies published in this Journal by Mammadinova I and colleagues describing the TS related to deep brain stimulations [3]. A 73-year-old man with Parkinson's disease and a 55-year-old woman with multifocal dystonia were longstanding well-controlled by bilateral deep brain stimulation with a left-sided dual-channel implanted pulse generator; but presented worsening motor symptoms. Their evaluations showed abnormal impedance readings, and extensive lead coiling that were surgically corrected with the lead replacement and pulse generator repositioning [3]. The

authors highlighted the TS critical clinical complications of deep brain stimulation, emphasizing the early diagnosis by imaging and impedance monitoring, besides the role of prevention by adequately implanted pulse generator fixation, and the pocket's size [3].

In this setting, some short comments on new literature data seem to be opportune. A 59-year-old man had discomfort at the site of an implantable cardioverter defibrillator; the heart rhythm and pulse were normal, the ultrasound showed a reduced left ventricular ejection fraction, the atrial lead was not visualized, and the shock coil was misplaced [1]. As the X-ray confirmed lead dislodgement and significant entanglement in the pocket, the diagnosis of TS was established, and the patient underwent a surgical revision; the authors stressed the single-session extraction and re-implantation of cardiac implantable electronic device that was successfully performed with a multidisciplinary approach [1]. A 91-year-old woman with chronic renal disease, hypertension, hypercholesterolemia, and obesity grade I, had implantation of a single-chamber pacemaker through the left subclavian vein due to a complete atrioventricular block causing recurrent syncope [2]. Three weeks later, she presented with dizziness and chest discomfort, an

unpleasant rhythmic sensation in the implant site, and involuntary movements of the chest left side, the electrocardiogram showed no ventricle record, suggesting a pacemaker malfunction, while the chest X-ray revealed that the lead was fully retracted into the device pocket [2]. The authors called attention to the diagnosis of Reel syndrome, which is an uncommon variant of TS characterized by the device rotated on its transverse axis; the electrodes coil near the generator and retract away from the heart, causing pacemaker malfunction [2]. Worthy of note, they emphasized the tip of the lead fully retracted into the pouch of the device they estimated to be a phenomenon not yet described in any previous reports [2]. A 54-year-old woman with antecedents of dilated cardiomyopathy, atrial fibrillation, mechanical valve prosthesis, and single-chamber pacemaker presented with chest pain and involuntary contractions of the left pectoral and left upper limb had TS diagnosis [4]. On cardiac monitoring the pacemaker spikes were not correlated with QRS complexes; the electrocardiogram had a rhythm compatible with atrial fibrillation and pacemaker spikes without any associated QRS complexes, revealing a ventricular capture failure; the chest X-ray showed a TS, with the electrode displacement wrapped in the generator [4]. The authors stressed the suspicion of TS based on involuntary contractions of pectoral and shoulder muscles, and no ventricular capture of the pacemaker by monitor or ECG; and the role of an early diagnosis to perform the prompt and adequate management [4]. A 65-year-old woman with previous heart failure and reduced ejection fraction (HFrEF) status post the implantable cardioverter-defibrillator (Barostim) presented with decompensated heart failure 8 months later, and the chest images showed a fractured lead with the proximal extremity twisted and retracted into the pocket confirming a TS [5]. She was treated for HFrEF exacerbation and scheduled for reimplantation of Barostim; the authors commented on the benefits of the device use considering that lead design reduces the risk of carotid

injury when tension is put on the lead with manipulation, and the complication may develop with a delayed presentation and less tension on artery [5]. A 65-year-old woman under cardiac resynchronization therapy-defibrillator had reduced amplitude and occasional detection failure in the atrial lead and increased pacing threshold in the right ventricular lead 13 months post-implant, and a chest X-ray showed twisted dislodged leads, and pulse generator rotation of the TS and Reel syndrome [6].

The authors highlighted the lack of guideline recommendations in the cases of risk factors, for increased follow-up, reimplantation procedures, and routine imaging studies to detect the lead dislodgements, in addition to the electrical testing procedures; moreover, the lead revision is definitively recommended in cases of their dysfunction [6].

Higher suspicion index and practical protocols for TS management can contribute to the outcome improvement among patients with cardiac implantable electronic devices.

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