Images in Cardiovascular Medicine

Inappropriate Implantable Cardioverter-Defibrillator Shocks in a 72-Year-Old Man With Aphasia

Seri Hanayneh, MD¹; Ramil Goel, MD²

¹Section of Cardiology, Tulane School of Medicine, New Orleans, Louisiana
²Department of Medicine, Division of Cardiology, University of Florida, Gainesville, Florida

72-year-old patient with a diagnosis of nonfluent progressive aphasia, who lived independently and was felt to have good judgment, received a biventricular implantable cardioverter-defibrillator (ICD) in the setting of unexplained syncope and cardiomyopathy. He presented to the emergency department 2 months later with complaints of recurrent ICD discharges. His device interrogation revealed more than 60 ICD shocks over the previous week, with rapid irregular electrical activity on the right ventricular (RV) lead, suggestive of atrial fibrillation sensing. The RV lead was not sensing ventricular activity or capturing the ventricle. The ICD pacing and therapies were turned off, and the patient was admitted for further investigation. Chest x-ray (Fig. 1) revealed retraction of the RV lead to the midatrial location and retraction of the atrial lead to the subcutaneous pocket. The leads were also notably entangled and twisted around each other near the device. When confronted with these findings, the patient admitted to repeatedly manipulating the device. He was taken for a lead revision procedure, during which a photograph was obtained (Fig. 2), which shows the appearance of the entangled leads caused by the patient's repeated twisting and turning of the device.

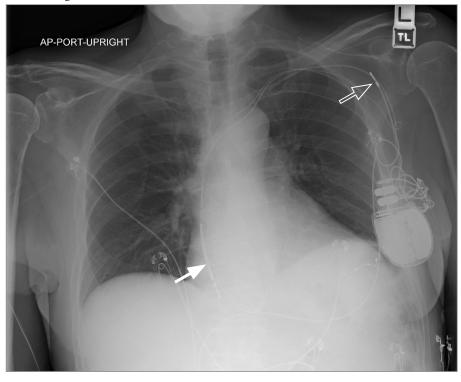


Fig. 1 Chest x-ray obtained on presentation (solid arrow = right ventricular lead in the right atrium; open arrow = right atrial lead).

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Corresponding author:

Seri Hanayneh, MD, Section of Cardiology, Tulane School of Medicine, 1430 Tulane Avenue, New Orleans, LA 70112

E-mail:

shanayneh@tulane.edu

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Fig. 2 Entangled leads postexplanation.

Comment

Twiddler syndrome refers to a patient's recurrent manipulation of the pulse generator, either consciously or subconsciously, which often leads to lead dislodgement and device malfunction.^{1,2} This phenomenon was described soon after patients started receiving pacemakers.³ The loss of contact of the leads with the cardiac tissue resulting from retraction can result in a fatal loss of pacing in pacemaker-dependent patients or recurrent inappropriate device therapies, such as those described in the presenting patient. An excessively mobile device, obesity, pocket discomfort, and psychiatric/cognitive disorders are factors associated with this clinical entity. There are several preventive measures to be considered: careful anchoring of the device and its leads, submuscular device implantation, nonabsorbable antimicrobial device envelopes, and ensuring appropriate pocket size.⁴

Twiddler syndrome should be on the differential whenever lead malfunction is suspected, especially soon after a device implant.

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