Twiddler’s Syndrome

Ragesh Panikkath MD, DNB, DM, Alejandro Perez-Verdia MD

A 54-year-old obese woman with a history of cardioverter defibrillator implantation (ICD) in 2004 for symptomatic ventricular tachycardia presented with discomfort at the site of the pulse generator and a “feeling of a knot”. She had undergone pulse generator replacement two years ago at which time twisting of the leads was noted and corrected. A pocket revision was performed, and the leads were noted to be intertwined due to counterclockwise rotation causing the “knot” felt by the patient (figures). Sensing and pacing functions of the leads were normal. The leads were untwined, and the pocket size was reduced. The pulse generator was secured to the pocket, and the leads were placed below the pulse generator. Although she had entangled her leads twice, this did not lead to lead displacement or malfunction.

**DISCUSSION**

Twiddler’s syndrome refers to malfunction of a pacemaker due to either deliberate or subconscious spinning of the pacemaker in the pacemaker pocket.¹ This causes reeling of leads around the pulse generator. The leads get dislodged, and sensing and pacing malfunction ensues. These leads can also stimulate the phrenic nerve or brachial plexus, resulting in diaphragmatic pacing and rhythmic twitching of the arm, respectively.² Twiddler’s syndrome is more common in obese, elderly, and mentally subnormal patients. Cases that occur late after implantation have been reported, but the majority of cases occur within a year of pacemaker implantation. Pacemaker malfunction can cause disastrous consequences in patients who are pacemaker dependent. Since newer devices are smaller than older ones, reduction in the size of the pulse generator pocket at the time of pulse generator replacement and fixing the device in the pocket should reduce the incidence of this syndrome. Although this
syndrome was initially described in pacemakers, variants of the syndrome have been described in patients with ICDs (as in our patient) and in patients with infusion pumps.\textsuperscript{3,4} Twiddler’s syndrome can cause malfunction of the ICD system, including inappropriate shocks from the device secondary to lead fractures.\textsuperscript{5}

References