Twiddler’s syndrome of an implantable cardioverter defibrillator

A 60-year-old Caucasian man underwent implantation of a biventricular implantable cardioverter defibrillator (ICD) for primary prevention of his underlying severe ischaemic cardiomyopathy. One year later, he presented with a problem of left-ventricular lead failure. He also received inappropriate ICD shocks as a result of poor R-wave sensing in the ventricular leads and double counting from the right atrium. A chest x-ray (CXR) (figures 1, 2) showed that the right-ventricular shock lead was displaced to the right atrium, and the right atrial lead was displaced into the superior vena cava. The left-ventricular lead could not be identified in the CXR. During device extraction, the displaced leads and heavily twisted leads around the ICD box were identified via the fluoroscopy. The left-ventricular lead was retracted into the ICD pocket and was completely displaced out of the venous system. There were extensive coiling and retracting of all three ICD leads within the pocket. This was a classical manifestation of Twiddler’s syndrome (figure 3).1,2

Twiddler’s syndrome is a clinical condition that is commonly caused by excessive twisting of the pacemaker or ICD device in the pocket, resulting in dislodgement of leads and device malfunction. Laser-lead extraction was performed successfully in this patient, and a single-chamber ICD was subsequently implanted. The cause of Twiddler’s syndrome in this patient could be due to an improper implantation technique resulting in a pocket that was too large, thus causing the device to twist and subsequent removal of the leads. In addition, the patient had a history of psychiatric disorder; self-inflicted twisting of the ICD device could thus be another cause.

Figure 1 Chest x-ray: anterior-posterior (AP) view.

Figure 2 Chest x-ray: lateral view.

Figure 3 Extracted implantable cardioverter defibrillator showing extensive coiling of the leads.

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