A 72-year-old man was implanted with a dual-chamber cardioverter-defibrillator (Lumax 540-HFT, Biotronik, Germany, Berlin) in secondary prevention. An atrial active fixation lead (Biotronik Solia S53) and a ventricular active fixation lead (Biotronik Linox S65) were used. PredischARGE an X-ray showed a good position of both leads (Panel A) and the internal cardioverter defibrillator was checked and functioned correctly. The patient was followed on remote monitoring (Home Monitoring network, Biotronik SE & Co. KG). Before the first planned in-clinic follow up, we received an alert due to the delivery of a shock for a ventricular arrhythmia and an episode of atrial fibrillation. All atrial and ventricular leads parameters seemed stable. Analysing the electrogram (EGM) (Panel B), we recognized an episode of atrial fibrillation on the atrial EGM. The ventricular EGM showed intermittent irregular rapid activity, with obvious similarity to the atrial rhythm. This could be explained by either intermittent 1:1 conduction of AF or by intermittent cross-talk with oversensing of the atrial signal on the ventricular lead. The patient was admitted and a chest X-ray confirmed a right atrial displacement due to Twiddler syndrome (Panel C). The patient denied conscious manipulation of the device pocket, but reported ‘spontaneous’ rotation of the device. A second intervention was consequently performed, electrodes were completely twisted (Panels D and E) and the ventricular lead showed signs of insulation break. Both leads were extracted and replaced. The generator was fixed in the pocket with a ligature.

Twiddler syndrome causing an inappropriate implantable cardioverter-defibrillator shock

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