Late twiddler syndrome in a patient with a submuscular implantable cardioverter defibrillator

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DESCRIPTION
A 20-year-old woman with long-QT syndrome received a submuscular Fortify DR-St Jude implantable cardioverter defibrillator (ICD) after an episode of cardiac arrest. She felt an ICD pocket vibration 23 months later due to a high lead impedance and loss of capture. Chest radiography (figure 1A,B) showed both lead displacement and winding of the leads beside the ICD generator. These findings were confirmed during surgery (figure 1C). The patient strongly denied any box manipulation. She was discharged after new leads implantation with no complications.

There is a report of three patients with ICD with submuscular devices in whom a twiddler’s syndrome occurred despite any risk factors were be seen. It is discussed that the cause of twiddling in these cases is the development of a loop in the lead caused by the craniocaudal movement during the abduction of the arm which aggravated like a cogwheel with each movement. Another theory suggests that the device may be twiddled between the muscles by movements.1

Patients with ICD may suffer additional problems as inappropriate shock delivery and the lacking capability for the treatment of life-threatening ventricular arrhythmias by the device. As worst case scenario, an inappropriate ICD therapy may be proarhythmic and may lead to sudden cardiac death.2 Our patient did not have any symptom probably due to Fortify DR-St Jude early detection of the high lead impedance leading a programmed ICD generator vibration.

Learning points
▷ Twiddler syndrome is defined as generator rotation with lead twisting and secondary malfunction of the leads.
▷ The causes of this syndrome are not always clear. However, predisposing factors include the creation of a large subcutaneous pocket, obesity, unconscious repetitive movements of the left arm, manipulation of the pocket or the adoption of repeated poor postural positions.3 Majority of cases were related during the first year of implant, but rarely can happen lately.

REFERENCES