Cardiac Imaging

Twiddler's Syndrome

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Twiddler's syndrome is a rare mechanical cause of failure of device therapy for dysrhythmia. This syndrome results from manipulation of the implanted device by the patient, leading to the dislodgment of the device leads. Although in the majority of the cases the dislodgment of the leads is painless, the subsequent device failure can be the cause of detrimental consequences.

Case presentation

A 65-year-old man was admitted to our institution with intermittent symptomatic complete heart block. This was a class I indication for pacemaker implantation according to the ACC/AHA/ESC guidelines. A DDDR system was implanted in the left pre-pectoral area and atrial and ventricular bipolar active fixation leads were placed via the left cephalic approach. The 24h post implantation chest X-ray showed satisfactory pacing lead positions (Figure 1) and all the pacing parameters (pacing threshold, sensing and impedance) were satisfactory.

One month later the patient visited the pacemaker clinic for his routine post implantation pacemaker check. He reported that he had been completely asymptomatic and the wound had healed completely. Pacemaker interrogation showed complete loss of pacing and sensing in both the atrial and the ventricular leads. We requested a chest X-ray, which revealed that both the atrial and the ventricular leads had been dislodged. Moreover, the pulse generator had been turned over several times, as the leads were twisted in the pocket and the pacemaker's orientation was opposite to the original (Figure 2). The patient admitted manipulating the pulse generator and we concluded that it was a case of the type of secondary lead dislodgement known as twiddler's syndrome.

We reopened the pacemaker pocket and inspected the segment of the leads in the pocket and the pulse generator. Fortunately, both the insulation and the conductor of the leads were intact. Both leads were repositioned and proved to be functional with good pacing and sensing parameters. The pulse generator was fixed on the pectoral muscle with non-absorbable sutures.

Discussion

Twiddler's syndrome was first described by Bayliss and refers to a permanent malfunction of the pacemaker as a result of rotation of the device causing lead dislodgement.[1] The syndrome occurs more often among elderly, obese and mentally disordered patients.[2] Although the syndrome was described in pacemakers it is

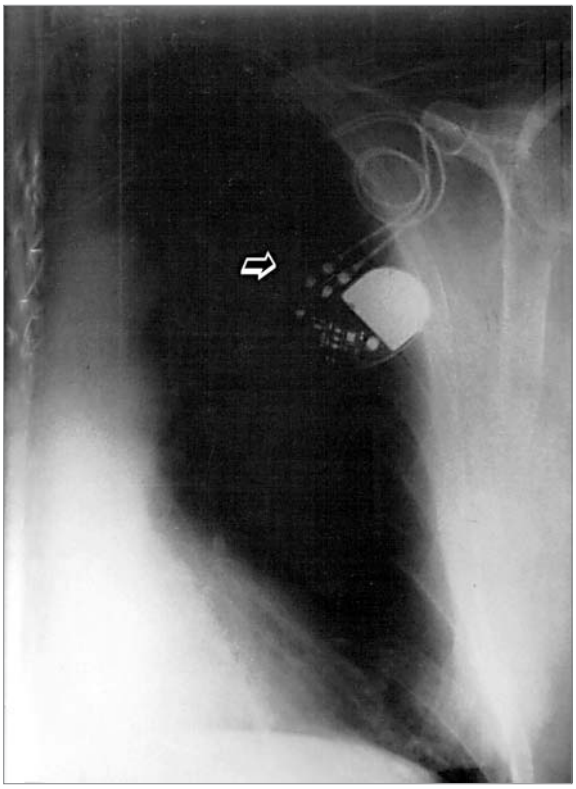

Figure 1. The pacemaker position is shown at 24 hours post implantation. The arrow indicates the clockwise orientation of the header.

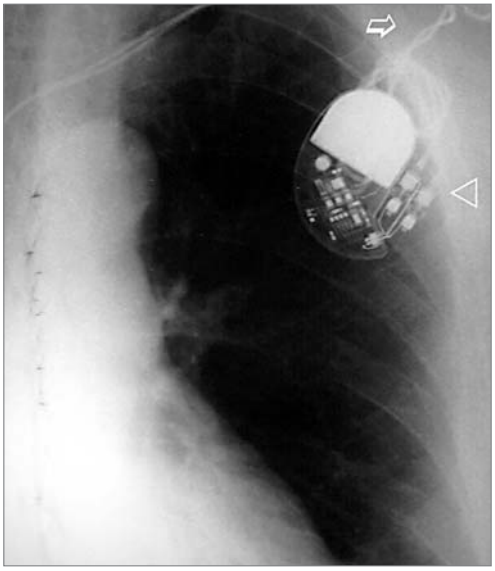

Figure 2. Chest X-ray showing that the pulse generator header has been reorientated counter clockwise (arrowhead) and the leads are twisted (arrow), documenting the diagnosis of twiddler's syndrome.

now recognised as an important cause of implantable cardioverter-defibrillator (ICD) failure.[3] ICD systems implanted in the abdomen are more vulnerable. Although the majority of the cases occur during the first year of implantation,[3] a "late twiddler syndrome" has recently been reported.[4] The miniature size of the new devices permits their rotation during physical activities when positioned in the old pacemaker's pocket. The reduction of the pocket size to fit the smaller device can prevent the development of the late version of the twiddler syndrome.[4]

Although cardiac symptomatology predominates, the stimulation of the phrenic nerve by the dislodged leads can cause diaphragmatic contractions and the stimulation of the brachial plexus can cause rhythmic arm twitching.[5]

Minimising the pocket size and suturing the device will decrease the incidence of this syndrome.[2,5] In addition, suture fixation should be taken into consideration in very lax subcutaneous tissues, especially in elderly people.[6]

Although contemporary pulse generators do not have to be placed in the pocket in a particular way, as they do not have an insulated cover as did older devices, it is worth placing the generators in a pre-specified way. In our institution we implant the devices with the header oriented in a clockwise direction. This can help the physician document the syndrome with a chest X-ray.

In our case, we managed to reinstitute the same leads, and we believe that use of the same leads should be encouraged as they remain intact most of the time.

References

1. Bayliss CE, Beanlands DS, Baird RJ: The pacemaker-twiddler's syndrome: a new complication of implantable transvenous pacemakers. Can Med Assoc J 1968; 99: 371-373.
2. Castillo R, Cavusoglu E: Twiddler's syndrome: an interesting cause of pacemaker failure. Cardiology 2006; 105: 119-121
3. Sharifi M, Inbar S, Neckels B, Shook H. Twiddling to the extreme: development of twiddler syndrome in an implanted cardioverter-defibrilllator. J Invasive Cardiol 2005; 17: 195-196.
4. Dursun I, Yesildag O, Soylu K, Yilmaz O, Yasar E, Meric M: Late pacemaker twiddler syndrome. Clin Res Cardiol 2006; 95: 547-549.
5. Nicholson WJ, Tuohy KA, Tilkemeier P: Twiddler's syndrome. N Engl J Med 2003; 348: 1726-1727.
6. Fahraeus T, Hoijer CJ: Early pacemaker twiddler syndrome. Europace 2003; 5: 279-281.