

The twiddler's plus syndrome – a case report

Skrajna postać *twiddler's syndrome* – opis przypadku

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Abstract

We present a case of a 43-year-old male with an extreme form of the twiddler's syndrome – almost complete lead dislodgement associated with their fracture.

Key words: pacemaker, twiddler's syndrome

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Introduction

First described in 1968, pacemaker twiddler's syndrome refers to permanent malfunction of a pacemaker due to the patient's manipulation of the pulse generator. The sequence of symptoms begins with the patient's deliberate or subconscious spinning of the pacemaker's pulse generator in a capacious pocket. The leads are dislodged, and ventricular pacing ceases. Subsequently, with continual reeling of the leads around the generator, the ipsilateral phrenic nerve is stimulated, resulting in diaphragmatic pacing and the sensation of abdominal pulsations. As the leads are further wrapped around the generator, rhythmic arm twitching occurs when the brachial plexus is paced.

Case report

A 43-year-old man, a football player by occupation, was brought to our emergency room with recurrent episodes of syncope. He had been implanted with a permanent pacemaker fifteen years ago with a diagnosis of bifascicular block with intermittent complete heart block with one episode of syncope. The patient was in regular follow-up until two years ago. He developed some sort of behavioural abnormalities eight years after pacemaker implantation which were diagnosed as schizophrenia and he was doing well on antipsychotic drugs. Since two years ago the patient was lost to both cardiological as well as psychiatric follow-up. He was actively participating in

football tournaments. During one month before admission, the patient had a reeling sensation when walking fast or climbing the stairs, which he neglected until he developed syncope and was brought to the hospital by his wife.

On arrival in the emergency room he had a pulse of 36/min regular and cannon wave visible in JVP. The patient started having a Stokes Adam attack while ECG was being taken. The patient was resuscitated and was moved to the cath lab where temporary pacing was started. The permanent pacemaker and lead were examined. It was found (Figure 1) that all but the terminal 3 cm of the lead had been coiled around the generator in the pocket. The terminal 3 cm of the lead was in the subclavian vein and fixed. Hence the patient was diagnosed as a case of twiddler's syndrome with complete lead dissection and separated segments of lead ('twiddler's plus' syndrome).

He underwent permanent pacemaker implantation on the contralateral side (left side) and the pulse generator was tied with a nonabsorbable suture (Ethibond) to the muscle. The previous pacemaker was explanted from its submuscular pocket along with the lead coiled around it (Figure 2). The patient was discharged on the seventh day.

Discussion

Pacemaker twiddler's syndrome was first described by Bayliss et al. [1] in 1968. It is a rare cause of pacemaker failure, resulting from the dislodging of the pacing electrode by manipulation, often unconscious, of the implanted

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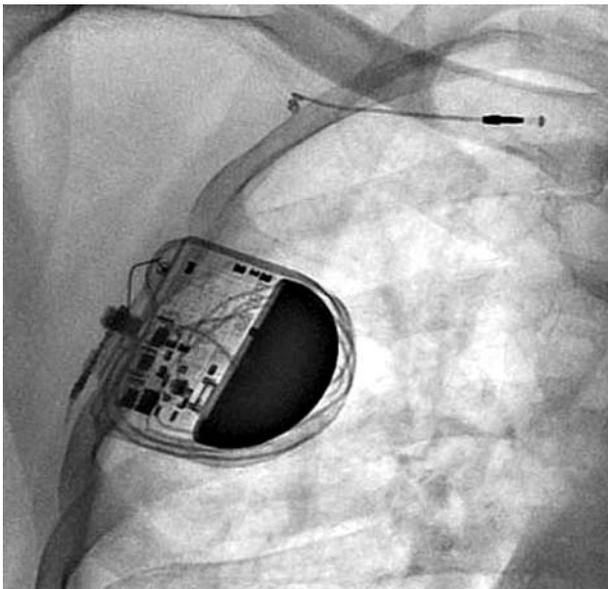


Figure 1. Pulse generator with coiled pacing electrode and terminal segment bisected and lying below the medial third of the clavicle

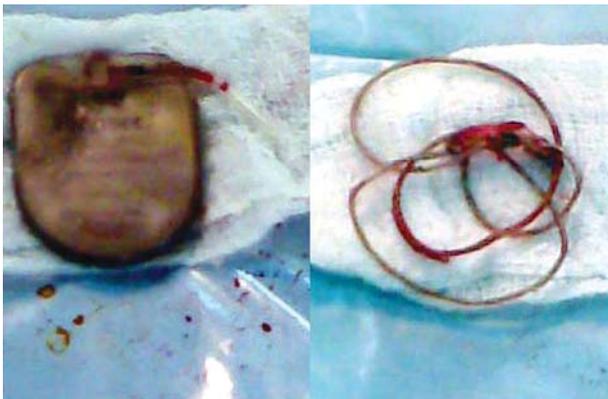


Figure 2. Pulse generator and coiled pacemaker lead after explantation

pacemaker [2]. Risk factors associated with twiddler syndrome are female gender, obesity, older age, and dementia [1–3], although it appears that dementia is by far the greatest risk factor [4]. Although diaphragmatic pacing usually occurs with lead perforation of the right ventricle,

ipsilateral phrenic nerve stimulation due to pacemaker twiddler's syndrome should be considered as a potential cause [5]. Variations of the syndrome have been described with the manipulation of other devices, including implantable cardioverter-defibrillators [6] and chemotherapy infusion pumps [7]. Elderly and obese patients appear to be at increased risk because the presence of loose subcutaneous tissue allows for rotation of the pulse generator in its pocket [8]. Creation of a small surgical pocket and suturing of the device to the fascia can help prevent manipulation of the pulse generator and lead displacement.

Our case has two unique features. The first is the late presentation, as our patient presented fifteen years after pacemaker implantation. The majority of patients with this syndrome are diagnosed within the first year of implantation. Secondly, the pacing lead was not only outside the cardiac chamber but also there was complete bisection of the lead with the terminal three centimetres being widely separated from the coiled up lead around the pacemaker pulse generator. This unique identity of twiddler's syndrome with complete bisection of the lead and widely separated segments has not been reported in the literature previously and has been proposed by us as 'twiddler's plus' syndrome.

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