Life-Threatening Effect of Twiddler's Syndrome with a Subpectorally Implanted Cardioverter Defibrillator

M. ZACHER, D. MARTINOVIC, A. DIEGELER
Kardiochirurgie, Herz- und Gefäß-Klinik GmbH, Bad Neustadt/Saale, Germany

Summary

Twiddler's syndrome occurs only rarely with subpectorally implanted transvenous cardioverter defibrillators (ICDs). This article discusses a case in which adequate shocks delivered to treat ventricular fibrillation failed to terminate an arrhythmia that resulted from Twiddler's syndrome. There are no absolutely safe techniques to prevent Twiddler's syndrome. The known risk factors for Twiddler's syndrome are not applicable to the patient discussed in this case report. The pathological mechanism for such cases still needs to be clarified.

Key Words

Twiddler's syndrome, Implantable cardioverter defibrillator (ICD)

Introduction

Twiddler's syndrome has been known for over 30 years as a rare complication after pacemaker implantation [1]. The often unintended manipulation of the pacemaker by patients causes the device to rotate in the pacemaker pocket, which may result in lead defects or dislocations. The same mechanism exists for transvenously implanted defibrillators and has been described as a rare complication for abdominally implanted devices [2,3]. Very few case reports exist for implantations in the pectoral region [4-6]. In all cases, the observed Twiddler's syndrome resulted in malfunction of the ICD system. Malfunctions are life-threatening for the affected patients due to arrhythmias that cannot be treated or the triggering of inadequate shocks. In one case, the affected patient died [7].
This case report discusses a patient in whom sudden cardiac death could only be prevented with external defibrillation.

Case Report

The 49-year-old patient experienced a posterior wall infarction in April 1994. The invasive diagnosis performed at that time showed the occlusion of a small left marginal branch. In February 2001, the patient was admitted to the hospital due to acute angina pectoris with pain spreading to both forearms. At the hospital, the patient underwent resuscitation due to ventricular fibrillation. Coronary angiography showed no progress of the coronary heart disease, as well as proper left ventricular function despite hypertensive heart disease with low-grade aortic stenosis. No ventricular tachycardias could be induced during an electrophysiological examination. Due to the patient's resuscitation and documented ventricular fibrillation, there was a Class I indication [8] for the implantation of an ICD. The criteria of the AVID study were also fulfilled [9].
In March 2001, the patient received a Microphylax Plus ICD (Biotronik, Germany) in a subpectoral position. An incision was made over the sulcus deltopectoideus, and the intermediate cephalic vein was actively dissected. A quadrupolar Kainox 75/16 lead (Biotronik, Germany) was then advanced through the intermediate cephalic vein and placed in the right ventricular apex. In order to prevent lead dislocation, the lead was fixated in the fascia of the deltoid muscle. Within anatomic structures, a pocket was created between the major and minor pectoral muscle using blunt dissection. After the lead was connected to the pulse generator, the latter was inserted in the pocket using

Figure 1. Device "in situ."

Figure 2. Device after explantation: 11.5-fold clockwise rotation.

slight pressure. The device had a tight fit in the pocket of the muscular and slightly adipose patient. An additional fixation suture was not performed. All measured values and the defibrillation threshold were normal. A 3-hour postoperative bed rest was imposed on the patient, during which a sandbag was placed on the wound. The subsequent out-patient ICD follow-up results were normal. No adverse reactions were noted. In July 2002, the patient again experienced symptoms of angina pectoris. Coronary angiogram showed a progression of the coronary heart disease with an occlusion of the right posterolateral branch and high-grade stenosis of the diagonal branch of the right coronary artery. During angiography of the right coronary artery, ventricular fibrillation was triggered. This was detected by the ICD, but the automatic defibrillation shocks failed to terminate the arrhythmia. The patient was resuscitated and externally defibrillated three times. Subsequently, the implanted device paced appropriately in VVI mode. Based on ICD interrogation, a lead defect was suspected in the high-energy circuit. Exploratory surgery conducted in July 2003 indicated that the cause of the lead break was a 11.5-fold rotation of the device around the longitudinal axis. The lead was removed and replaced by a new lead of the same type. The device was reimplanted in the same pocket. The position of the pulse generator was fixated by the suture to the fascia of the minor pectoral muscle. The subsequent 14-month observational period (to date) showed no further adverse events and normal ICD parameters.

Discussion

Twiddler's syndrome is rare and can occur after abdominal and pectoral implantation. For the pectoral region, it has been reported to occur both after subcutaneous [10] as well as after subpectoral [11] placement. In the majority of publications, a fixation suture is recommended to obviate Twiddler's syndrome. In one case, this technique caused a transection of the lead insulation after only two rotations of the device [12]. The second most common recommendation is the use of a Dacron pouch, with which the pulse generator is implanted and fixated. There are two reported cases in which this technique was used during the revision operation after the occurrence of Twiddler's syndrome. Despite the Dacron pouch, these two patients' devices still rotated around the longitudinal axis [7,13]. None of the recommended methods can completely exclude the possibility of Twiddler's syndrome. Subpectoral implantation with fixation sutures appears to decrease the probability of occurrence. Including the case described above, we have subpectorally implanted 500 ICDs and exchanged 70 devices at this site. Fixation of the device with non-absorbable thread occurred only in rare exceptions.

Based on the previous individual case descriptions, the following risk factors were identified: elderly female patients with weak subcutaneous tissue [11,14, excess weight with subsequent weight loss [5], subcutaneous pocket [11], pockets that were too large [10], and patients who already experienced Twiddler's syndrome. Our patient is male and muscular. He weighed 80 kg at the first implantation as well as at the revision

and has not lost any weight in the meantime. The device was implanted submuscularly, and the pocket was not significantly larger than the pulse generator even after the revision. The patient denied that he had manipulated the ICD, there were no complaints, and the wound looked outwardly normal. It can thus be maintained that Twiddler's syndrome can occur even when none of the accepted risk factors are present. The implanted pulse generator was hardly palpable, and a habitual manipulation of the device by the patient seems improbable. The patient is a mason by trade and at least sporadically resumed his professional duties after the implantation. The question is whether – for people who are physically active and are also engaged in manual work that is above head level – it is possible to rotate the device by simply engaging in muscle-intensive work. For these patients at least, the exact pathologic mechanism of Twiddler's syndrome remains to be clarified.

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Contact
Dr. M. Zacher
Herz- und Gefäß-Klinik GmbH
Abteilung Kardiochirurgie
Salzburger Leite 1
D-97616 Bad Neustadt/Saale
Germany
Phone: +49 09771 66 0
E-mail: meddok.zacher@herzchirurgie.de