Pectoral muscle contractions as a consequence of Twiddler's syndrome

Des contractions du muscle pectoral révélant un syndrome de Twiddler

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Résumé

Le syndrome de Twiddler constitue une cause rare de dysfonction de défibrillateurs automatiques implantables (DAI). Nous rapportons le cas d'un homme âgé de 51 ans ayant une cardiomyopathie dilatée ischémique à fonction systolique du ventricule gauche altérée nécessitant l'implantation d'un DAI. Quatre mois après la procédure, il consulte pour des contractions régulières du muscle pectoral gauche.
La radiographie du thorax objective un enroulement excessif de la sonde ventriculaire droite autour du boîtier ramenant son bout distal en sous-claviculaire gauche, témoignant d'une rotation du boîtier autour de son axe longitudinal. Le diagnostic de syndrome de Twiddler a été alors posé.

Mots-clés
Défibrillateur automatique implantable – syndrome de Twiddler

Summary

Twiddler's syndrome is a rare but potentially serious cause of implantable cardioverter defibrillators (ICD) malfunction.
We report the case of a 51 year old male with a history of ischemic dilated cardiomyopathy with impaired left ventricle systolic function requiring implantation of an ICD. Four months after the implantation, he was admitted to the emergency department for repeated and regular pectoral muscles contractions.
Fluoroscopy revealed rotation of the generator and dislodgement of the right ventricular lead and its distal tip was in left subclavian vein. Diagnosis of Twiddler's syndrome was made.

Keywords
Implantable cardioverter defibrillator- Twiddler's syndrome

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Twiddler's syndrome is a rare but potentially serious cause of implantable cardioverter defibrillators (ICD) malfunction. It is characterized by deliberate or spontaneous rotation of the pulse generator leading to lead malfunction. It was first described with pacemakers by Bayliss in 1968 (1). In this case we describe dislodgement of ICD's lead due to Twiddler's syndrome.

CASE REPORT

A 51 year old male with a history of ischemic dilated cardiomyopathy with severly impaired left ventricle systolic ejection fraction was implanted with an ICD four months ago . He was admitted to the emergency department for repeated and regular left pectoral muscle contractions.

A chest X-ray was performed and showed that the generator had rotated and the right ventricular lead was excessively twisted around the device and its distal tip was situated in the left subclavian vein.

The right ventricular lead didn't sense ventricular activity which was interpreted as a cardiac pause by the ICD. Thereby, the device was delivering inappropriate stimulations causing pectoral contractions.

The device was rapidly turned off and then we underwent a replacement of the right ventricle lead which was not damaged.

Later, he explained that he manipulated the pocket's device because it was a little bit painful and he was anxious.

DISCUSSION

Twiddler's syndrome is a rare cause of macro-dislodgement and lead failures, which usually occurs one year or less after implantation of a device [1].

It is defined as a deliberate or unconscious manipulation of the device in a too large pocket, winding thereby the electrode around device's longitudinal axis [2].

This usually results in dislocation or fracture of the lead(s), which causes loss of capture and abnormal sensing. The reported frequency of Twiddler's syndrome is around 0.07-7% [2, 4].

Although it was originally described by Bayliss in 1968 with pacemakers [1], it has also been reported with implantable cardioverter-defibrillators, in which case, it is more serious and potentially life threatening because treatment of ventricular arrhythmias would be disabled and inappropriate shocks could be delivered due to over sensing [3].

A widely accepted explanation for the Twiddler's syndrome is that new types of devices (pacemakers and defibrillators) are smaller and therefore easier to twist in their pocket.

This syndrome is mostly observed in elderly women aged between 60 and 85 years. Obesity is also a common finding. This coincidence may be related to feminine anatomy at this age with accumulation of pre-pectoral subcutaneous adipose tissue and eventually pendant breast [3].

Other risk factors for this condition include psychiatric illness, mental disorders such as dementia, lax subcutaneous tissue, excessive movements of the upper limbs, active manipulation of the generator, and the small size of the implanted device relative to its pocket [2, 4].

Possible complications due to ICD Twiddler's syndrome are inappropriate shocks and lack of appropriate shocks. Furthermore, patients having pacemakers may present syncope and near syncopal symptoms caused by loss of pacing capture and bradycardia. Otherwise device lead displacement may produce diaphragmatic contraction by phrenic nerve stimulation and twitching of arm accordingly to the brachial plexus or pectoral muscle stimulation like for our patient [1, 2, 3].

In case of Twiddler's syndrome, the most common behavior is to untwist the leads after making sure they are not fractured, and fix them actively. In some cases, when the leads are damaged and no longer functional, they must be take out and replace them.

Figure 1 : Different severe VHD involvement in the study population

Figure 2 : after right lead replacement

CONCLUSION

Twiddler's syndrome is a rare complication of ICD implantation with potential catastrophic consequences. The chest-X- ray is the simplest diagnostic tool. Fixation of the pulse generator with a ligature during the implantation may prevent twiddling of the device. The use of screw in leads as well as subpectoral implantation has been described as possible preventive strategies, but these are not always successful. Active fixation of leads should always be performed to prevent this situation along with other preventive measures such as minimizing the pocket size. [1] However, patient education remains the single most important means of avoiding pacemaker manipulation.

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