Reel Syndrome and Pulsatile Liver in a Patient With a Two-chamber Pacemaker

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SUMMARY

Twiddler’s syndrome is characterized by coiling of the pacemaker lead due to the rotation of the pacemaker generator on its long axis. Reel syndrome is another form of Twiddler’s syndrome. It occurs due to the rotation of the pacemaker generator on its transverse axis with subsequent coiling of the pacemaker leads around the pulse generator. In this article we describe a patient with a two-chamber pacemaker who presented with sudden onset of abdominal pulsation and was subsequently diagnosed as Reel syndrome. To the best of our knowledge, this case is the first case of Reel syndrome that developed in a patient with a two-chamber pacemaker. (Jpn Heart J 2004; 45: 1037-1042)

Key words: Twiddler’s syndrome, Reel syndrome, Pulsatile liver

TWIDDLER’S syndrome is characterized by coiling of the pacemaker lead due to the rotation of the pacemaker generator on its long axis. Although a loose subcutaneous pocket is the main reason for the rotation of the pacemaker generator, its size and weight may also play an important role in the development of this syndrome.

Since its first description by Bayliss, et al in 1968, many interesting cases with Twiddler's syndrome have been published.1) It is frequently observed in overweight female patients with loose, fatty subcutaneous tissue.2) Rotation of the generator may occur by the patient’s pathologic manipulations or spontaneously.3) Lead coiling is the manifestation of the rotation of the generator on its long axis. It is usually observed close to the generator and sometimes intracardially.4,5) It may cause lead dislocation or fraction and these in turn may cause life-threatening symptoms in patients with pacemaker dependence.6,7)

Reel syndrome is another form of Twiddler’s syndrome. It occurs due to the rotation of the pacemaker generator on its transverse axis with subsequent coiling of the pacemaker leads around the pulse generator. It was described in a patient with a single-chamber pacemaker, which had been implanted because of the diag-
nosis of presyncope, a low ventricular rate, atrial fibrillation, and previous mitral valve replacement due to rheumatic heart valve disease.\textsuperscript{8}) Similar to Twiddler's syndrome, Reel syndrome may also lead to electrode dislocation or fracture followed by several clinical symptoms such as presyncope, syncope, or even death in patients with absolute pacemaker dependence.

In this report, we describe a patient with a two-chamber pacemaker who presented with sudden onset abdominal pulsation and was subsequently diagnosed as having Reel syndrome.

**CASE REPORT**

A 68-year-old man was admitted to our emergency department because of sudden onset of abdominal pulsation. He had been complaining of fatigue and lightheadness for 10 years. After his evaluation at another hospital he had been diagnosed with “sick sinus syndrome” and a DDDR pacemaker [Medtronic 5594 atrial lead, 5092 ventricular lead, KDR 701VPP pacemaker generator (Medtronic, Inc., Minneapolis, MN, USA)] was implanted. He did well until 1 month after implantation when an abdominal pulsation developed suddenly.

On physical examination his arterial blood pressure was 120/70 mmHg and his pulse rate 75/min. Cardiac auscultation findings were normal. A pulsation in his abdomen was observed on inspection and a regular pulsation in the liver was palpated.

On ECG, pacing spikes at the beginning of the P waves and in the QRS complexes and rare ventricular ectopic beats were observed. P waves were thought to have been developed by pacing and QRS morphology by fusion pacemaker beats or normal ventricular activation-ineffective pacing spikes. Pacemaker interrogation showed effective atrial and ineffective ventricular pacing. Atrial pacing beats were regularly conducted to the ventricle (Figure 1). The pacemaker mode was changed to AAI mode and the patient’s symptoms disappeared.

In the evaluation of bedside chest radiography, which had been obtained just after implantation at the previous hospital, the atrial and ventricular leads and pulse generator were in normal positions (Figure 2). On chest X-rays taken at our university hospital, the atrial lead was straightened, the ventricular lead was in the vena cava superior, and leads were coiled around the pacemaker generator (Figure 3).

A new surgical procedure was performed. The pacemaker pocket was opened; leads were uncoiled, repositioned, and connected to the same generator. The leads and pulse generator were fixed tightly to the fascia (Figure 4).
Figure 1. Pacing spikes at the beginning of the P waves and in the QRS complexes and rare ventricular ectopic beats on electrocardiographic tracing. Pacemaker interrogation shows effective atrial and ineffective ventricular pacing. Atrial pacing beats are conducted to the ventricle.

Figure 2. Bedside antero-posterior chest radiography in previous hospital after implantation. a: Atrial lead, b: Ventricular lead, c: Pulse generator.
Figure 3. On postero-anterior chest radiograph, atrial lead is straightened, ventricular lead is in the vena cava superior, and leads are coiled around the pacemaker generator one month after implantation. a. Ventricular lead, b. Atrial lead, c. Pulse generator.

Figure 4. On postero-anterior chest radiograph, leads are repositioned in the right atrium and in the right ventricular apex. a. Atrial lead; b. Ventricular lead; c. Pulse generator.

DISCUSSION
To the best of our knowledge, the present case is the first case of Reel syndrome that developed in a patient with a two-chamber pacemaker. The patient
had liver pulsations synchronous with heartbeats. The pulsations were thought to be due to stimulation of the right phrenic nerve by the dislocated ventricular lead. We suggest that Reel syndrome, which is another form of Twiddler’s syndrome, should be included among the etiologies of liver pulsation in patients with pacemakers. It is easily diagnosed by chest X-rays with the observation of dislocated leads which are coiled around the pulse generator.

In our case we were not able to find any etiological factor for the pacemaker rotation. The patient was male, not overweight, had thick subcutaneous tissue, and no history of willing or unwilling pathological manipulation. The size of the generator pocket was not large. We suggest that implantation of the atrial lead loosely during the first procedure could be the reason for the straightened and functional lead. The same leads were replaced and connected to the same pacemaker because their functions were normal. Development of new complications was prevented by creating a new generator pocket instead of reducing the pocket’s size and tight fixation of the leads and generator to the fascia. Because of the patient’s thick and strong subcutaneous tissue, we thought subpectoral placement was not necessary.

During follow-up we have not observed any further complications and pacemaker function was normal after 10 months.

Although Twiddler’s syndrome and its other forms are rare conditions, they may result in serious clinical complaints and life-threatening events. To prevent Twiddler’s syndrome, the pulse generator should be implanted between the fascia of the musculus pectoralis major and minor, and fixation to a loose subcutaneous tissue should be avoided.9,10 Although fixation of electrodes with a butterfly and unresorbable suture is not a gold standard, fixation of the generator in at least two points increases the safety of the procedure and decreases the risk of rotation. Other preventive measures include the creation of a small surgical pocket, using a compression band around the upper chest and shoulder, and tightening of the patient’s arm for at least five to seven days.

REFERENCES

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