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Extreme Twiddling: Unusual Presentation of an Old Foe

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Abstract

Characteristic twiddling and lead macro-displacement are well-documented radiological hallmarks of Twiddler's Syndrome. What is less well known is that the different variants of Twiddler's Syndrome can be distinguished by assessing orientation of the pins on X-ray. Illustrations of a change in orientation of the header and pins are rarely reported in cases of pacemaker Twiddler's Syndrome and the phenomenon has not been described in implantable cardioverter-defibrillator recipients. We highlight an important classic finding that is seldom depicted in the literature.

Keywords: ICD; Twiddler's syndrome; Intracardiac twiddling

Abbreviations: ICD: Implantable Cardioverter-Defibrillator; RV: Right ventricular; HV: High voltage; SVC: Superior vena cava; PA: Postero-anterior

Case Report

A 57-year old man underwent ICD implantation for resuscitated ventricular fibrillation in the setting of ischaemic cardiomyopathy in 2006. An active fixation single coil right ventricular [RV] lead [Medtronic 6945-65] was positioned at the RV apex and connected to a Medtronic Secura VR pulse generator before being placed into a sub-pectoral pocket [R wave 14mV, threshold 1.2V, impedance 1278 ohms]. Routine annual device interrogations were uneventful until 2011 when he was found to have rising thresholds with poor sensing [R wave 1.3mV, threshold 3.75V, High Voltage [HV] impedance 42 ohms]. PA chest X-ray did not show evidence of a macroscopic lead displacement or fracture. Lead repositioning was attempted but was unsuccessful due to a complete conductor break; a stylet would not pass beyond the mid portion of the lead, nor would the helix retract. The old lead was extracted and a new dual coil RV lead [Medtronic 6947-65] placed at the RV apex [R wave 12mV, threshold 0.5V, impedance 803 ohms, HV and SVC coil impedance 42 and 61 ohms respectively].

At routine device interrogation 1 month later there was complete failure to sense or capture. Again, the chest X-ray showed satisfactory lead position with no evidence of twisting or coiling and the patient denied any manipulation of the generator. At operation, the old lead was removed and a new dual coil RV lead was positioned at the RV apex [R wave 10.5mV, threshold 0.5 V, impedance 500 ohms, HV and SVC coil impedance 44 and 61 respectively] (Figure 1a).

1 month later HV and SVC impedances, which had always been within normal limits now both measured <20 ohms [R 9.9mV, threshold 2.75V, impedance 437 ohms]. Furthermore, chest X-ray demonstrated an abnormal lead appearance with decreased distance between the proximal and distal coils. The pins and header were orientated in the opposite direction to that seen on the post implantation X-ray (Figure 1b). Lateral chest X-ray confirmed localized intracardiac twisting of the RV lead (Figure 1c). The patient subsequently admitted to manipulating the generator. At operation the RV lead was untwisted, explanted and replaced. On this occasion a non-absorbable suture was placed through the fixation hole of the generator and fixed to the floor of the sub-pectoral pocket to help anchor the device. No further problems have since been encountered.

Discussion

Twiddler's Syndrome is caused by deliberate or inadvertent rotational manipulation of the pulse generator. Radiographically it is characterized by twisting of the lead with associated displacement. Twisting of the proximal portion of the lead is a commonly reported observation, whilst discrete twisting of the intracardiac portion of the lead is a less frequent occurrence. A change in orientation of the pins and header on X-ray is a poorly described entity that is clearly depicted in our case. This radiological observation is seldom reported in the literature and has previously only been described in pacemaker recipients [1].

Since Bayliss first described Twiddler's Syndrome in 1968 [2], additional variants of the condition including Reel and Ratchet Syndrome have been described [3-4]. All are recognized causes of pacemaker, ICD and biventricular ICD malfunction. Whilst classical Twiddler's Syndrome results from rotational manipulation of the generator about its long axis and is characterized by twisting of the leads, Reel Syndrome results from rotation of the generator about its transverse axis and causes coiling of the leads within the pocket. Ratchet

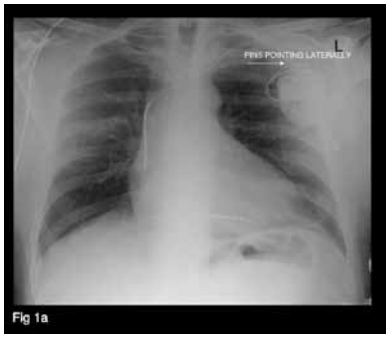

Figure 1a: Post procedure chest X-ray showing clear separation between the proximal and distal coils. The pins are pointing laterally within the header.

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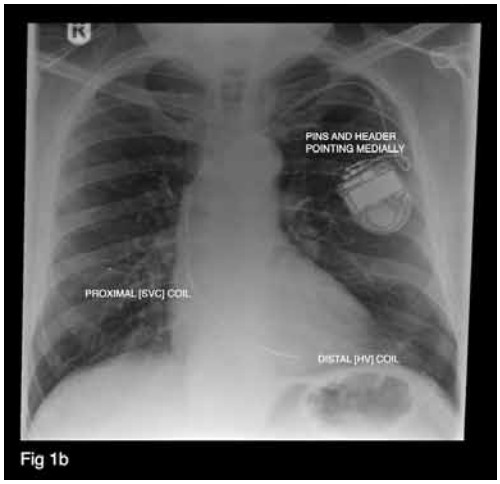

Figure 1b: 1 month later the pins are now pointing medially, the proximal coil has advanced into the heart and there is decreased distance between the 2 coils.

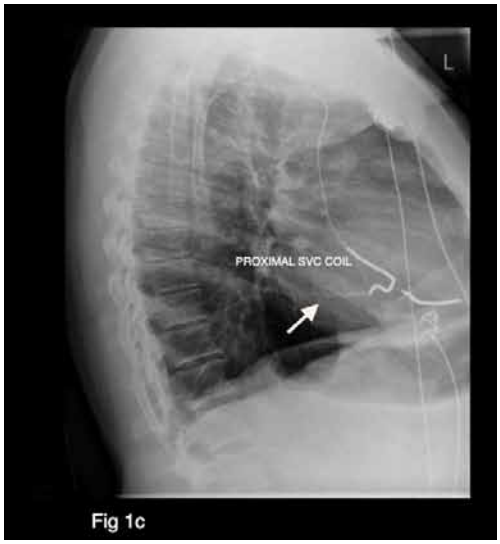

Figure 1c: Lateral chest x-ray showing intracardiac twisting of the RV lead involving the proximal [SVC] coil.

Syndrome stands alone; it is not dependent on rotational manipulation of the device [4]. Rather it results due to inadequate anchoring of the lead within the fixation sleeve, such that movement of the ipsilateral arm causes retraction of the affected lead with subsequent coiling in the pocket. Though the mechanisms differ, all three result in macroscopic lead displacement.

Patient-induced ICD malfunction was initially suspected but the radiographic hallmarks of Twiddler's Syndrome and its variants were not evident. The delay in diagnosing the cause of ICD malfunction was multifactorial. Initial radiographical findings were subtle and did not suggest classical Twiddler's Syndrome. Discernable twisting of the RV lead within the heart was not manifest until the final X-ray. A change in orientation of the header was central to confirming that the mechanism of lead failure was due to rotational manipulation of the pulse generator. Repeated rotation of the generator first one way and then back again may have been sufficient enough to cause lead damage without resulting in overt twisting or displacement of the lead. This may explain the initial 'concealed' presentation. Finally, continuous uni-directional rotation of the generator resulted in twisting of the intracardiac portion of the RV lead. This caused the 2 shock coils to move closer together and subsequent drop in HV and SVC coil impedances, which until that point had always measured within normal limits. These observations all infer that 'fulminant' twiddling did not commence until the later in the course of events when far more convincing signs of twiddling became apparent. Sub-pectoral placement of the pulse generator is described as a potential remedy for Twiddler's Syndrome. However our case demonstrates that this does not prevent a determined patient. Securing the generator with an anchoring stitch and comprehensive directives on refraining from stirring the pocket may be just as influential in reducing the incidence of this patient-induced complication.

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References

1. Rohlfing BM, Hutchinson JC, Webb WR (1977) The flipped pacemaker. Radiographic diagnosis of a cause of malfunction of rechargeable pacemakers. Chest 71: 237-239.
2. Bayliss C, Beanlands D, Baird R (1968) The pacemaker-twiddlers's syndrome: a new complication of implantable transvenous pacemakers. Can Med Assoc J 99: 371-373.
3. Vlay SC (2009) Reverse Twiddler's syndrome. Pacing and Clinical Electrophysiol 32: 146.
4. Von Bergen NH, Atkins DL, Gingerich JC, Law IH (2007) "Ratchet" syndrome, another etiology for pacemaker lead dislodgement: a case report. Heart Rhythm 4: 788-789.