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Twiddler's Syndrome with A Fatal Outcome

Case Report

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Author Details

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Abstract

Introduction: Pacemaker dislodgement syndromes are exceedingly rare though may be potentially life-threatening. Three syndromes have been described in the literature and different mechanical processes explain the dislodgement and radiographic features

Case report: We describe a case of early Twiddler's syndrome in a pacemaker-dependent patient with a fatal outcome. The chest radiograph demonstrates dramatic braiding of the lead with spontaneous and complete dislodgement.

Discussion: The usually described patient and device risk factors for Twiddler's syndrome are not always present. There are variable radiographic features of the syndrome depending on the underlying generator-lead motion.

Conclusion: Pacemaker lead dislodgement syndromes can be fatal and the usually described risk factors are not necessary for its occurrence.

Introduction

There are three described pacemaker macro-dislodgement syndromes; all of which result in lead displacement with loss of atrial or ventricular stimulation. The differentiating features between the syndromes are based on the underlying abnormal mechanical movement of the generator without its migration, and commonly reported as patient-manipulated, with the subsequent impact on the lead. The first described syndrome in 1968 by Bayliss, et al. is the somewhat stigmatising Twiddler's syndrome likely named after the patient "twiddled" with the generator. The underlying mechanism of lead dislodgement is by the traction created by the rotation of the generator on its long axis. The result is the twisting of the leads outside of the generator pocket creating a braid-like or tangled appearance on X-ray [1]. Carnero-Varo, A et al. in 1999 first described the Reel syndrome as a new variant of Twiddler's syndrome and the proposed and accepted mechanism is the rotational movement of the generator along its short axis. The radiographic features consistent with this are complete or near-complete retraction of the lead with coiling around the generator [2]. The Rachet syndrome, first described by Von Bergen, et al. in 2007, is also associated with complete retraction of the lead and is attributed to the forwards and backwards motion without rotation of the generator which explains why coiling or braiding of the lead is usually not present [3] These complications can occur at any time, although the Reel and Rachet syndromes have been associated most frequently within the first month after implantation and usually some months later in the Twiddler's syndrome.

Idiopathic lead migration is a rare complication of pacemaker or intracardiac device implantation. Ghani, A et al. in 2014 reported 57 (1.4%) lead dislodgements in 1929 implanted devices and 3909 leads [4]. Wang, Y et al. in a 2018 meta-analysis of 18 studies which included 17,321 patients undergoing conventional single or dual-chamber pacemaker implantation reported an incidence of lead dislodgement ranging from 1%–2.69% in individual studies with 1.63% as the mean incidence [5]. In a real-world report by Jorge O. Gomez, et al. in a 2022 study evaluating a five-year retrospective analysis of 1793 patients reported the prevalence of Twiddler's syndrome in 21 (1.2%)

patients [6]. Morales, J et al. in a study in 2017 which included 1472 implanted devices and 78 cases from reports in the literature spanning 1990 to 2012 in a pooled analysis, reported nine (one of Twiddler's syndrome and eight of Reel syndrome) cases of lead dislodgment [7]. The findings of the multicentre FOLLOWPACE study which included 23 hospitals and 1517 patients who were followed for a mean period of 5.8 years reported a 3.3% incidence of lead dislocation or displacement within the first 2 months of implantation [8]. Given the rarity of this potentially life-threatening consequence, most literature is cited around case reports and case series.

Case report

We present a case of a 56-year-old male with conventional vascular risk factors of diabetes mellitus, mixed hyperlipidaemia, hypertension and angiographic evidence of non-occlusive coronary artery disease with no echocardiographic nor ventriculographic evidence of left ventricular dysfunction nor myocardial infarction who attended with a rapidly progressive clinical presentation consistent with heart failure. The index electrocardiogram was consistent with complete atrio-ventricular dissociation with a ventricular rate of 42 beats per minute. A transvenous St Jude Medical, Endurity SR permanent single lead VVIR (ventricular pacing, ventricular sensing, inhibitor mode) pacemaker was implanted with no peri-procedural nor immediate complication [9]. His clinical condition improved, and he was discharged well. Twenty-two days later, he suddenly presented in extremis with an altered level of consciousness, near-complete circulatory failure, a severe anion-gap metabolic acidosis, acute severe hypoxaemic respiratory failure with pulmonary oedema and a KDIGO grade 3 acute kidney injury. The blood pressure on arrival was 70/40mmHg (MAP-50mmHg), his Glasgow coma scale was 8/15 and his pO2/FiO2 ratio was 95mmHg. The sampled arterial blood gas analysis revealed a pH of 7.02; pCO2 of 18.1mmHg; bicarbonate of 4.77mmol/L; base deficit of 26.3mmol/L and a lactate of >20mmol/L.

His creatinine was 1356umol/L in the setting of anuria. The non-contrasted CT scan of the brain performed in the emergency department confirmed left internal capsule watershed infarction and infarction in bilateral posterior limbs of the internal capsules and left head of caudate nucleus. The electrocardiogram confirmed the absence of pacemaker activity and revealed a junctional rhythm with a ventricular escape rate of 40 beats per minute. The bedside focused echocardiogram confirmed reduced left ventricular contractility and the absence of a pacemaker lead in the right ventricle or atrium. Resuscitation measures included emergency endotracheal intubation and positive pressure ventilation and the optimistic administration of atropine and adrenaline as an infusion as a temporising measure while awaiting transvenous pacing as transcutaneous pacing was not imminently available. He rapidly regressed and succumbed to his illness prior to any pacing intervention, likely as a result of the accrued factors of severe metabolic acidosis, specifically severe lactic acidosis; respiratory failure and acute kidney injury with complete circulatory failure supported by evidence of cerebral hypoperfusion, acute tubular necrosis and likely myocardial ischaemia. His chest radiograph demonstrated a complete right ventricular pacemaker lead dislodgement and migration toward the ipsilateral left generator. (Figure 1)

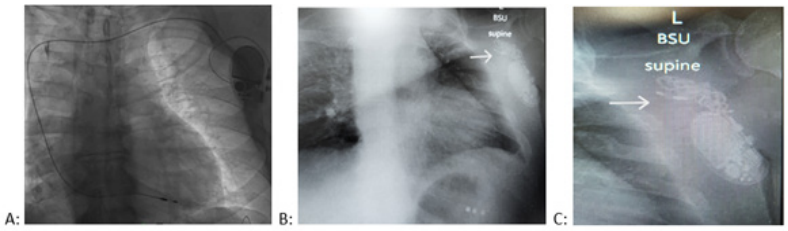

Figure 1:

A. Fluoroscopy image post implantation- demonstrates a single lead pacemaker with the lead tip in the right ventricle.

B. Rotated plain chest radiograph at the time of presentation demonstrates complete migration of the right ventricular pacemaker lead toward although not around the generator (Arrow). The generator appears to be in a consistent orientation as in A (given the rotated plain radiograph), suggesting that no obvious generator rotation occurred.

C. Focused inset of chest radiograph in B- Dramatic and near complete braiding and twisting without coiling of the lead around the generator is appreciated.

Discussion

We report a case of a young male who was pacemaker-dependant with spontaneous and complete lead migration with a fatal outcome. Such dramatic entangling of the completely retracted pacemaker lead close to although with no coiling around the generator was evident on chest radiography and rarely described in previous case reports. Reports of a migrated lead have described phrenic nerve, vagus nerve and brachial plexus stimulation. The absence of expected ipsilateral rhythmic arm-twitching in our case, given the complete migration and positioning of the lead is likely explained by the exceptional entanglement which may have resulted in lead fracture not easily appreciated on plain radiography or simply by lack of contact with the brachial plexus.

There are several patient and device factors which make this case unusual. Firstly; none of the significant risk factors for Twiddlers syndrome cited in several case reports and meta-analyses in the literature; namely obesity, advanced age, female gender and psychiatric illness, specifically obsessive-compulsive disorder, mostly for the inferred risk of patient-incited manipulation are present in our case [6,10-13]. A capacious generator pocket has also been cited as significant risk factor although in our patient, the leads appear outside of the pocket suggesting this is not the case. Secondly, spontaneous lead dislodgement in this case occurred within three weeks of the pacemaker insertion which is unusually early for the classical Twiddlers syndrome. A previous case report described the Twiddler syndrome occurrence as early as within 24 hours of implantation, however, the mechanism in that case was consistent with Reel syndrome rather than the classical Twiddler syndrome [14].

Further, it is apparent that the usual device-related factors significantly associated with Twiddlers syndrome cited in the literature which include a right atrial position of the lead, the device type, the shape of the device and the technique of fixation are not obviously contributory in this case. In the meta-analysis by Ghani, A et al. the right atrial lead is reported to be significantly more frequently dislodged compared with the right ventricular pacemaker lead (p=0.0007); and significantly more lead dislodgments or malfunctions were observed in patients with CRT-D/P devices and dual-chamber devices compared with pa-

tients with a single chamber pacemaker (p=0.006; p=0.002 respectively) [4]. Chauhan A, et al. in their study of 2019 patients also reported more frequent atrial lead dislodgment than ventricular lead within the first 6 weeks as well as a significantly higher complication frequency with dual-chamber implants compared to singe lead pacemaker implantation ($p<0.05$) and the finding was attributed to the higher incidence of atrial lead displacement [15].

Wang Yan, et al. in his metanalysis, reported in patients with conventional pacemaker leads, a higher lead dislodgement rate with atrial as compared to ventricular electrodes and no significant difference in dislodgment risk with the use of active fixation leads compared to that of passive fixation leads [5]. The Pacemaker Selection in the Elderly (PASE) study reported a higher frequency of ventricular lead dislodgement than atrial lead dislodgement although a significantly higher risk of atrial lead dislodgment in patients receiving dual-chamber pacemakers. The PASE study however, evaluated patients older than 65 years and its findings may not necessarily be relevant in this case [16]. The current literature appears to support the lowest device-related risk of complete lead migration in our patient. Lastly, the radiographic appearance suggests a mixed mechanism of lead dislodgment. While the braided and entangled appearance of the lead is consistent with the classical Twiddlers syndrome, most previous case reports describe dislodgment with the lead tip usually remaining intracardiac and with far less dramatic braiding or twisting. Although spontaneous lead displacement with complete retraction has been described in case reports; most cases have been associated with the Reel syndrome where braiding or twisting of the lead is not radiographically appreciated.

The Reel mechanism is thought less likely in our case as the position of the generator appears unchanged in its orientation compared to the immediate post implantation image on fluoroscopy, which suggests the absence of generator rotation and further, the lead does not appear coiled around the generator box. We therefore postulate that the underlying mechanism in this case may have been a combination of the Twiddler mechanism and the Rachet mechanism. The overall extreme presentation and rapid clinical regression in this patient precluded direct visualisation of the generator position in relation to the lead which may have clarified the underlying mechanism. An explanation for this occurrence in this patient may have been the use of passive fixation, which has been cited in some case studies as an important aetiological factor.

Conclusion

Pacemaker lead migration or dislodgement in a pacemaker-dependant patient can certainly be fatal. There is a possibility of a combination of mechanisms for the spontaneous and complete retraction of a pacemaker lead. The usual patient and device risk factors associated with Twiddler's syndrome may not be necessary for its occurrence and idiopathic pacemaker lead migration may indeed be idiopathic.

Author contribution

All authors were involved in the literature search and review and the formulation of this case report.

Author disclosures

None of the authors have any conflict of interest to declare nor declare any competing interests.

Ethical considerations

This article was formulated ensuring patient anonymity.

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