Twiddler’s Syndrome: A Rare Cause of Pacemaker Failure

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Twiddler’s syndrome was first described by Bayliss et al. in 1968 [1]. This condition is a rare cause of pacemaker failure, resulting from the dislodging of pacing electrodes by manipulation, often unconsciously, of the implanted pacemaker [2]. This report describes a rare condition of pacemaker failure in a patient with twiddler’s syndrome resulting from unintentional rotation of the pacemaker causing atrial lead dislodgement from the endocardium and malfunction of the device.

Patient Description

A 69 year old diabetic and hypertensive woman was hospitalized for weakness. Complete atrioventricular block was diagnosed on electrocardiogram and a permanent AV sequential pacemaker (DDDR, Biotronik, Germany) was implanted with a screw in Selox ST 60 electrode in the right atrium and a passive-tined electrode in the right ventricular apex. Pre-discharge chest X-ray revealed well-positioned electrodes [Figure A] and ECG showed appropriate function of the pacemaker. Four days later a heart rate of 60 beats per minute was measured, associated with abdominal tingling, namely, a phrenic nerve stimulation (the patient described this feeling as a sensation of fetal movements during pregnancy). The patient mentioned that the external monitor electrode located above the pacemaker generator caused irritation.

The patient was referred to hospital where an ECG showed pacemaker undersensing and non-capture of the atrial electrode and normal ventricular pacing at 60 beats per minute – the lower rate of the pacemaker programming. A chest X-ray revealed a dual-chamber pacemaker with displaced electrodes of right ventricular and atrium, with coiling around the pacemaker [Figure B]. Re-implantation of the electrode was performed successfully using the same electrodes. The patient was discharged 2 days later after evaluation of the pacemaker function and appropriate placement of the electrode on X-ray.

Comment

Twiddler’s syndrome is a rare cause of pacemaker failure, resulting from the dislodging of pacing electrodes by manipulation, often unconsciously, of the implanted pacemaker [2]. Sometimes, when the leads are displaced and loop around the pacemaker body, ipsilateral phrenic nerve or brachial plexus stimulation may occur with resultant cycling contractions of the abdomen [3] or arm muscles [4]. Risk factors associated with twiddler’s syndrome are female gender, obesity, older age and dementia [1,2,4], although it appears that dementia is by far the greatest risk factor [5]. Chest X-ray is the most important and simplest diagnostic tool. The majority of patients with twiddler syndrome are diagnosed within the first year of implant.

Twiddler syndrome may lead to a series of complications such as syncopal attack and lethal cardiac dysrhythmias, especially in pacemaker-dependent patients, as well as catastrophic consequence when the implantable cardioverter defibrillator is involved.

As the general population grows older, it is reasonable to expect an increase in the incidence of implantation of various cardiostimulator devices. A substantial proportion of this elderly population will also have an increased prevalence of dementia, thus the incidence of the syndrome may increase in the near future.

Preventive measures such as patient
education, use of a smaller pocket for the pacemaker, and fixation of the device by sewing it to the fascia in the surgical pocket will reduce the risk of development of this syndrome.

References

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~200 kilobases upstream of the Xic – the X-pairing-region (Xpr) – is sufficient in a single copy to allow a transient interaction between the two Xics at a time before the beginning of X inactivation. This pairing is cell cycle dependent, can occur from an ectopic location, and may activate the expression of Xist, a non-coding RNA that coats the inactive X chromosome.

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Homeostasis, the ability to maintain a steady state in the face of stresses, is a fundamental part of life for cells and for organisms. Wilson and co-authors (J. Neurosci. 2007;27:13581) analyzed homeostatic changes at the level of synaptic connections in hippocampal neurons seeded onto a microfabricated surface. Imprinting the surface with a template of squares of increasing sizes created a series of micrometer-scale islands hosting neurons at identical densities but with an increasing number of potential partners. As the number of neurons on a square increased, the number of synaptic connections increased, but, surprisingly, the functional activity of the neurons as a population (measured in voltage clamp and current clamp experiments) did not. This scaling was mediated by a change in the kinds of connections the neurons made. As network size increased, the proportion of connections between excitatory and inhibitory neurons increased, in other words, neurons made weaker connections. Changes in neuronal connectivity occur as a consequence of development, aging, and disease (such as Alzheimer's disease and autism), and analyses of this kind may contribute to our understanding of the ability of the brain to respond to changes and the pathologies that occur when it cannot.

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CASE COMMUNICATIONS

If you don’t read the newspaper you are uninformed; if you do read the newspaper you are misinformed

Mark Twain (1835-1920), American humanist, humorist, satirist, lecturer and writer, most famous for his novels Adventures of Huckleberry Finn, which has since been called the Great American Novel, and The Adventures of Tom Sawyer. During his lifetime, Twain became a friend to presidents, artists, leading industrialists and European royalty.

Capsule

Chromosome X Inactivation

One of the two X chromosomes in mammalian females is randomly inactivated early in development to match the single active X chromosome of males. This process is regulated through the X-inactivation center (Xic). The two Xics interact in trans at the beginning of X-inactivation, presumably to allow reciprocal activation/inactivation. So far, single copies of elements from the Xic have not been able to recapitulate X inactivation, suggesting additional elements are necessary. Augui et al. found that a region

Capsule

Neuron connections

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