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Tremor and Other Hyperkinetic Movements

Video Abstracts

Something that Touches your Heart: an Unusual Case of Abdominal Clonic Movements

Valentina Fioravanti[1,2*], Igor Lamberti[3], Nicola Bottoni[4], Francesco Cavallieri[1,5], Franco Valzania[1] & Matteo Pugnaghi[6]

[1] Neuromotor & Rehabilitation Department, Neurology Unit, Azienda USL-IRCCS di Reggio Emilia, Reggio Emilia, IT, [2] Local Health Unit of Reggio Emilia, Neurology Division, Correggio (RE), IT, [3] Local Health Unit of Reggio Emilia, Emergency Department, Correggio (RE), IT, [4] Local Health Unit of Reggio Emilia, Arcispedale S. Maria Nuova-IRCCS, Department of Cardiology, Reggio Emilia, IT, [5] Clinical and Experimental Medicine PhD Program, University of Modena and Reggio Emilia, Modena, IT, [6] Local Health Unit of Reggio Emilia, Neurology Division, Castelnovo né Monti and Scandiano (RE), IT

Abstract

Background: Rarely, cardiac pacemaker implant can lead to the development of involuntary hyperkinetic movement disorders localized to the abdominal wall or the diaphragm.

Phenomenology Shown: We report a case of a 79-year-old female who developed rhythmic continuous clonic right abdominal movements caused by cardiac pacemaker lead dislodgement.

Educational Value: Our case highlights that, in the differential diagnosis of hyperkinetic abdominal movement disorder, the presence and the possible pathogenic role of a cardiac pacemaker should be kept in mind.

Keywords: Abdominal, diaphragmatic, myoclonus, pacemaker, Twiddler's syndrome

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*To whom correspondence should be addressed. E-mail: valentina.fioravanti@gmail.com

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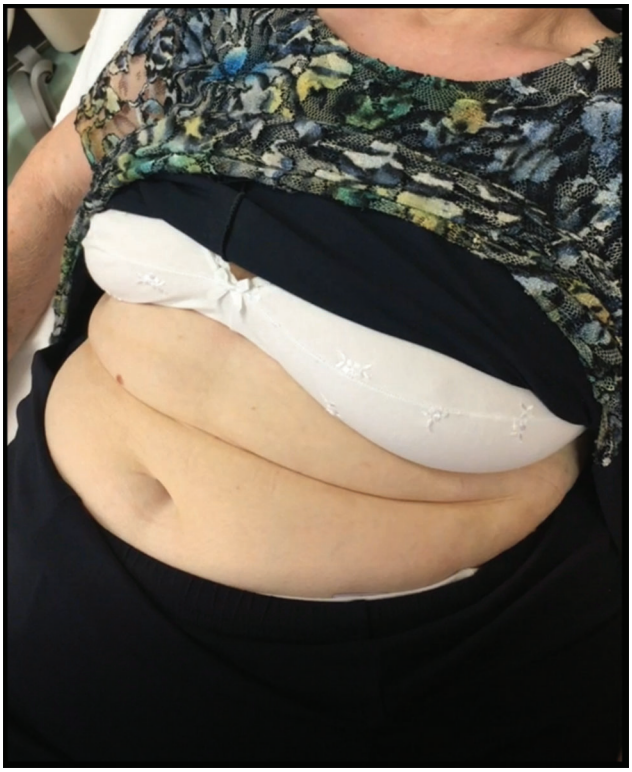
Rarely, cardiac pacemaker implant can lead to the development of involuntary hyperkinetic movement disorders localized to the abdominal wall or the diaphragm. In particular, the pacemaker patient manipulation and the subsequent lead displacement, also known as Twiddler's syndrome, may cause the stimulation of the ipsilateral phrenic nerve or the brachial plexus, respectively resulting in diaphragmatic pacing or ipsilateral rhythmic arm twitching.[1] However, apart from the so-called Twiddler's syndrome, isolated electrode lead misplacement may be associated with abdominal or diaphragmatic involuntary contractions.[2]

A 79-year-old female with a history of hypertensive cardiomyopathy was admitted in the Emergency Room complaining an uncomfortable "right abdominal tremor" onset abruptly and present during wake and sleep. Twenty days before, a dual-chamber cardiac pacemaker was implanted after an episode of syncope due to second-degree 2:1 Mobitz-II atrioventricular block. Neurological examination only showed the presence of rhythmic continuous clonic right abdominal movement, not altered by distraction (Video 1). A brain computed tomography (CT) scan showed the presence of a left frontal meningioma located in the parafalcine region and adjacent to the dorsolateral prefrontal cortex. In suspicion of an epileptic origin of the involuntary movements, an electroencephalogram was performed; however, no epileptiform activity was detected, excluding the possibility of an epilepsia partialis continua syndrome. A chest radiogram was performed that showed the misplacement of the right atrial electrode lead. In the hypothesis of contraction of the right diaphragm related to atrial pacemaker electrode dislodgement, the atrial lead was repositioned 3 days after, with prompt disappearance of symptoms.

During its descent through the chest cavity, the right phrenic nerve interfaces medially with the superior vena cava and the right atrium. At this level, a displaced atrial electrode could stimulate the right phrenic nerve with the subsequent appearance of hemidiaphragmatic



Video 1. **An Unusual Case of Abdominal Clonic Movements.** The video shows continuous rhythmic (approximately 1 Hz, as highlighted by a stopwatch placed in the lower left corner) clonic right abdominal movements because of phrenic nerve stimulation by a dislodged atrial lead of a dual-chamber pacemaker previously implanted for 2:1 Type II A-V block causing relapsing syncopes. Neurological examination was otherwise unremarkable.

twitching. It has also been reported that a correctly positioned atrial lead or a left ventricular electrode placed too close to the course of the left phrenic nerve may cause the appearance of ipsilateral hemidiaphragmatic rhythmic pseudo-myoclonus.[2,3] From a clinical point of view, our case differs from abdominal myoclonus, which is almost always characterized by irregular diaphragmatic contractions of 0.5–15 Hz (usually 2–5 Hz) that disappear during sleep.[4] On the contrary, the abdominal contractions seen in our patient were rhythmic, with a frequency of 1 Hz (exactly like the heart rate) and were present also during sleep. Furthermore, in diaphragmatic myoclonus, breath holding and deep inspiration suppress the movements or decrease their frequency, highlighting the influence of postures and voluntary movements on myoclonic jerks.[5] None of these factors influenced the rate and intensity of involuntary contractions in our patient. Based on clinical examination and CT findings (i.e. the presence of left frontal meningioma), an epileptic origin of the involuntary movements has been hypothesized. However, the absence of electroencephalogram epileptic activity, the temporal correlation with the pacemaker implant, and the misplacement of the atrial electrode lead allow to make the correct diagnosis. In conclusion, our case highlights that, in the differential diagnosis of hyperkinetic abdominal movement disorder, the presence and the possible pathogenic role of a cardiac pacemaker should be kept in mind, even in the absence of Twiddler's syndrome.

References

1. Newland GM, Janz TG. Pacemaker-twiddler's syndrome: a rare cause of lead displacement and pacemaker malfunction. *Ann Emerg Med* 1994;23:136–138. doi: 10.1016/S0196-0644(94)70021-4

2. Onesti E, Ceccanti M, Rubino A, Frasca V, Inghilleri M. Atypical case of diaphragmatic pseudo myoclonus. *Parkinsonism Relat Disord* 2017;43:118–119. doi: 10.1016/j.parkreldis.2017.06.016

3. Khan AA, Nash A, Ring NJ, Marshall AJ. Right hemidiaphragmatic twitching: a complication of bipolar atrial pacing. *Pacing Clin Electrophysiol* 1997;20:1732–1733. doi: 10.1111/j.1540-8159.1997.tb03550.x

4. Llaneza Ramos VF, Considine E, Karp BI, Lungu C, Alter K, Hallett M. Ultrasound as diagnostic tool for diaphragmatic myoclonus. *Mov Disord Clin Pract* 2016;3:282–284. doi: 10.1002/mdc3.12295

5. Aggarwal A, Thompson PD. Unusual focal dyskinesias. *Handb Clin Neurol* 2011;100:617–628. doi: 10.1016/B978-0-444-52014-2.00044-6