Parkinsonism and Related Disorders 63 (2019) 232–234

Contents lists available at ScienceDirect

Parkinsonism and Related Disorders

journal homepage: www.elsevier.com/locate/parkreldis

Correspondence

Diaphragmatic myoclonus owing to electrode dislocation of implantable cardioverter defibrillator

ARTICLE INFO

Keywords:
Diaphragmatic myoclonus
Cardioverter defibrillator
Electrode
Diaphragmatic flutter
Myoclonus

ABSTRACT

Myoclonus is a sudden and brief involuntary muscle contraction presenting with jerk-like movements that can occasionally involve the trunk muscles or the diaphragm as in the case of spinal myoclonus[1]. We here present an unusual case with unilateral diaphragmatic myoclonus owing to electrode dislocation of an implantable cardioverter defibrillator.

1. Case summary

A 62-year-old man presented with involuntary movements of his trunk muscles that had started three weeks previously. 11 months prior to that he survived cardiac arrest and subsequently received a combined cardioverter/defibrillator (ICD). He suffered from coronary artery disease with ischemic dilated cardiomyopathy, and he had a history of an acute myeloid leukemia with a confirmed *FAB M2* mutation. He had no family history of movement disorders.

Upon clinical examination, he had episodic bursts of rhythmic contractions of his right upper abdominal wall involving the right pectoralis major muscle. Movements worsened when sitting and lying on his right side. There were no clinical signs of an underlying movement disorder. Surface multichannel electromyography of trunk muscles (at Th8/9) revealed rhythmic, synchronous, 3-phasic potentials lasting less than 10 ms with a maximum amplitude of approximately 2.2 mV registered on the right pectoralis major muscle. Discharges were not synchronous with the patient's palpable pulse nor did they correspond to the ECG artifact. The ECG artifact constantly emerged throughout the recording asynchronously to the rhythmic discharges, apparently arguing against an interconnection with the cardiac activity (see Video 1 and Fig. 1).

Supplementary video related to this article can be found at https://doi.org/10.1016/j.parkreldis.2019.03.004.

As unilateral myoclonus is unusual and the surface EMG found myoclonic activity of atypical morphology with 3-phasic potentials instead of commonly polyphasic myoclonic bursts, we judged our findings as an atypical myoclonus. With the potentials' morphology, the ultra-short duration, and a rise time of below 400 μs, we further considered a non-biological generator the most likely explanation for the phenomenon.

We thus hypothesized that an electrical device generated the discharges. Although a previous check-up of the cardiac device was normal, we decided to admit the patient to the cardiologist. The cardiac device survey test revealed an ”exit block“ of the atrial lead, meaning a loss of sensing resulting in ineffective stimulation. Atrial under-sensing can lead to irregular ICD pacing despite a sufficient heart rhythm. Any time the atrial electrode was externally stimulated, myoclonus was reproducibly induced. Conventional chest X-ray confirmed a malrotation of the ICD with a dislocation of the right atrial electrode lead projecting to the superior vena cava and the right phrenic nerve (see Fig. 1). This phenomenon is known as “twiddler's syndrome“, commonly resulting in failure of ventricular pacing. Thus, capture of the diaphragm is more frequently due to the left ventricular electrode lead. However in this case, singular affection of the atrial lead appears rare. Thus, the diaphragmatic capture was clearly related to the atrial stimulus. The atrial electrode was deactivated and the cardiac device was programmed to the VVI mode, exclusively stimulating the ventricles when pacing. Myoclonus promptly disappeared.

Only a few cases of diaphragmatic myoclonus have been reported to date. It can occur immediately following an incorrect pacemaker emplacement as presented by Onesti et al. [2]. Diaphragmatic stimulation resulting from a wide-reaching electrical field or via direct stimulation of the phrenic nerve due to anatomical proximity of electrode leads commonly occurs immediately after implantation of a cardiac device. In contrast to previous findings, where stimulus induced myoclonus acutely occurred, in our case similar movements started with a delayed onset of 11 months after correct ICD implantation. Lack of synchronicity of the EMG and ECG activity is contradictory to the case presented by Onesti et al. [2] and initially considered to argue against attribution to the device, but this is probably explained by the loss of atrial signal. Thus, a missing EMG/ECG synchronicity not necessarily refutes a “cardiac origin”. Apparently, changes of body position led to a false trigger for the misapplied stimulation. In another case [3] it was assumed that an atrioventricular dyssynchrony provoked diaphragmatic myoclonus via increased right atrial pressure and afferent induced enhancement of vagal activity rather than a direct stimulation of the phrenic nerve. We cannot exclude this possibility in our case.

The most relevant differential diagnoses include segmental spinal myoclonus or peripheral myoclonus [1,4]. While etiology varies, their clinical presentation can be challenging to distinguish, as both involve abdominal and trunk muscles. Notably, segmental spinal myoclonus typically involves multiple muscles of interrelated spinal levels. A dysfunction or loss of inhibitory spinal interneurons or structural reorganization of local neuronal circuits may explain the pathophysiology of spinal generated myoclonus, but there were no cues for these events in our case. Furthermore, the brevity of the EMG activity clearly argued against a spinal origin.

https://doi.org/10.1016/j.parkreldis.2019.03.004
Received 10 August 2018; Received in revised form 27 February 2019; Accepted 6 March 2019

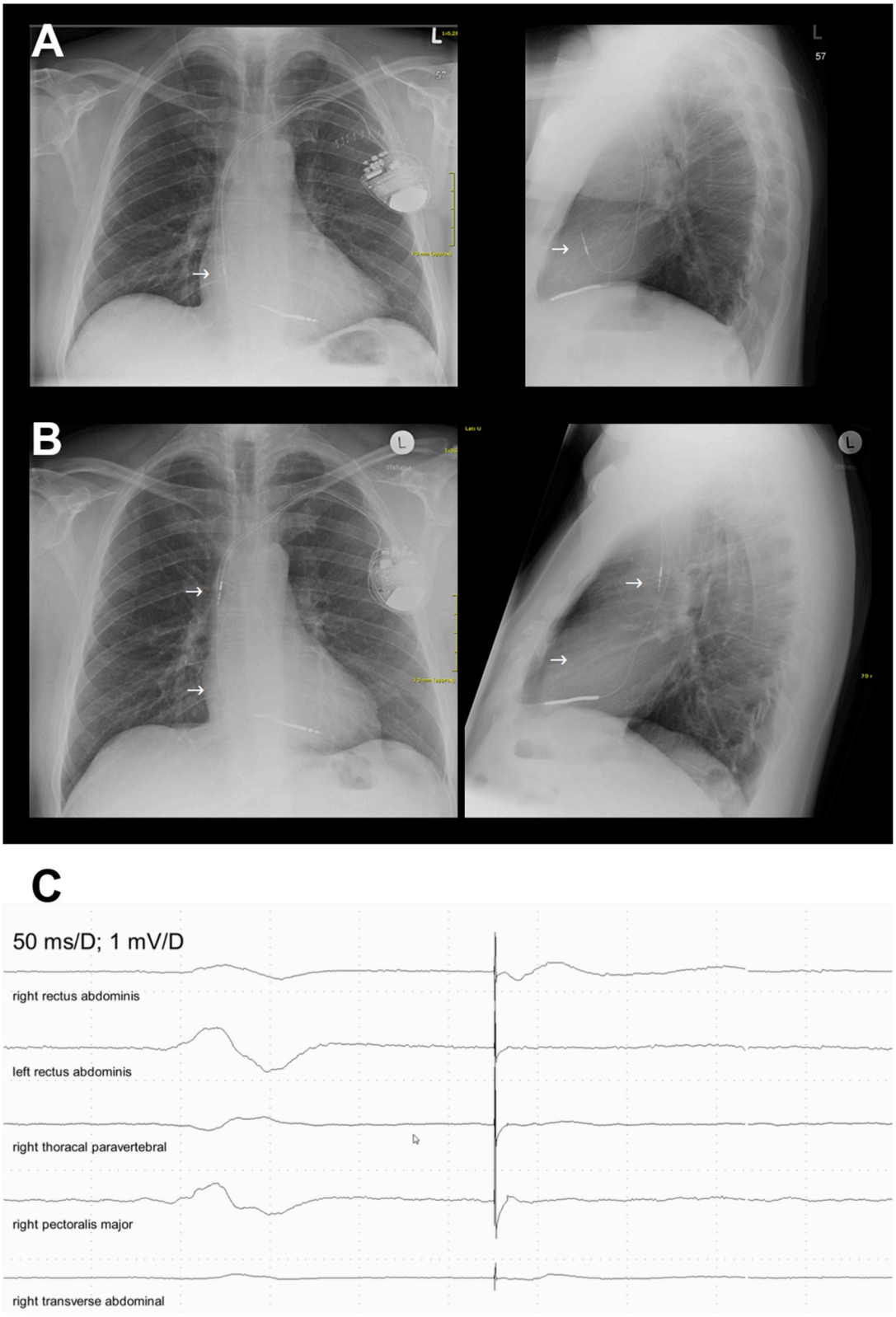

Fig. 1. Imaging and surface EMG.
Chest X-ray imaging with p.a. projection; A, imaging directly after the implantation of the cardiac device shows the correct placement of the electrode leads, and B, a visible malrotation of the device after myoclonus had started. C; Propagation pattern of myoclonus, recorded by surface multichannel EMG of right-sided muscles. Bursts have a duration of max. 10 ms and a maximum amplitude of appr. 2.2 mV. Note that the EMG activity was not synchronous with the ECG artifact nor with the patient's palpable pulse.

In summary, electrophysiological assessment is required to detect whether myoclonus is of cortical, subcortical or spinal/segmental origin, or due to a non-biological stimulus, such as in the case presented here. Our case highlights the importance of a careful interpretation of the duration, distribution and stimulus sensitivity of muscle jerks, and particularly of the configuration of electrophysiological data, demonstrating the possibility of a non-neurogenic origin of abdominal or diaphragmatic myoclonus.

Author contributions

Bedarf J R, conception and design of the study, acquisition, analysis, and interpretation of data, drafting of the manuscript, final approval of the submitted version. Nitsch L, acquisition, analysis, and interpretation of data, revising the manuscript for intellectual content, finals approval of the submitted version. Malotki R, acquisition, analysis, and interpretation of data, revising the manuscript for intellectual content, final approval of the submitted version. Andrié R revising the manuscript for intellectual content, final approval of the submitted version. Reimann J, conception and design of the study, acquisition, analysis, and interpretation of data, revising the manuscript for intellectual content, final approval of the submitted version.

Study funding

No funding reported.

Author disclosures

The authors report no disclosures relevant to the manuscript. Dr. Bedarf was supported by research support by the PD Fonds Deutschland gGmbH. She received travel grants and honoraria for talks for Ipsen Pharma and Merz Pharmaceuticals. Dr. Nitsch received travel grants from Merck, Bayer, Sanofi Genzyme, TEVA and travel grants and honoraria for talks from Novartis. Dr. Malotki has nothing to declare. PD Dr. R. Andrié has nothing to declare. Dr. Reimann serves as an associate editor for BMC Neurology. He received a travel grant from Amicus Therapeutics and a speaker's honorarium from.Natus Europe.

Informed consent

Our patient gave written informed consent for the video publication of his data.

Acknowledgements

We thank Okka Kimmich for her valuable contribution to this manuscript.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.parkreldis.2019.03.004.

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