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Case report

Clinical neurophysiological evaluation for simple motor tics

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ABSTRACT

Objective: To demonstrate the usefulness of neurophysiological evaluation to distinguish simple motor tics and functional myoclonus.
Methods: Careful clinical assessments, multichannel surface EMG, and EEG-EMG jerk-locked back-averaging were performed.
Results: Urge to move and ability to voluntarily suppress the movement were reported. EMG bursts showed variable duration and triphasic pattern of the antagonist muscles mimicking voluntary movements. Only the late component of the Bereitschaftspotential (BP2) was present prior to the involuntary movement onset.
Conclusion: Combination of the isolated late BP, premonitory urge, and suppressibility leads to the diagnosis of simple motor tics rather than functional myoclonus.
Significance: The physiological approach in addition to careful clinical assessment is helpful to support the diagnosis of tic.

1. Introduction

Simple motor tics are sudden, rapid, recurrent, nonrhythmic jerk-like movements that are often preceded by premonitory urge (Kwak et al., 2003) and may be voluntarily suppressed for a brief period (Himle and Woods, 2005). Some clinical characteristics of motor tics such as sudden onset, variability of movement distributions, distractibility, suggestibility, temporary remissions and waxing and waning courses may lead to incorrect diagnosis of functional (psychogenic) movement disorders (Jankovic and Kurlan, 2011; Demartini et al., 2015). Careful history taking and electrophysiological study might help to differentiate between these disorders. Here we described a case of motor tics that illustrates the physiological approach. The patient gave informed consent for participating in an IRB approved protocol and for publishing the video.

2. Case description

A 36 year-old right-handed female was referred to our clinic for a second opinion concerning the presence of jerky movements all over the body, randomly in the hands, arms, legs and face, predominantly on the right for 2 months. The referring diagnosis was functional movement disorder. Before the movement, she sometimes had a feeling of something clawing in the certain part of the body and relieved whenever she moved it. She was able to suppress the jerky movement by clenching her fist as well as tensing up that particular muscle for a brief period, but it eventually built up again and she needed to move. The movements occurred more often when she was relaxed whereas they occurred less frequently when she was concentrating on something. She was taking diazepam to control these jerks without any benefit. She had history of similar movements when she was teenager for 5 years and was diagnosed as having a psychogenic movement disorder also at that time. These movements spontaneously disappeared without any treatment. She denied history of obsessive–compulsive symptoms and attention-deficit syndrome during childhood. Neurological examination demonstrated random jerky movements of the right fingers, arm, shoulder and the right leg. The movements were sometimes accompanied with urge to move. She was able to suppress them for a brief period (see video). These movements were sometimes distractible, but not entrainable with tapping maneuver. Magnetic resonance imaging was normal.

The diagnosis of our patient with jerky movements can either be motor tics or functional myoclonus due to variability of her movements. The clawing sensations prior to the movements and

Abbreviations: EEG, electroencephalography; EMG, electromyography; ECR, extensor carpi radialis; FCR, flexor carpi radialis; BP, Bereitschaftspotential.

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the feelings of relief after the movements are consistent with premonitory urge. Urge to move and ability to voluntarily suppress the movements favor tics rather than functional myoclonus. However, distractibility during the exam is suggestive of functional myoclonus. The history of similar movement during teenager further supports the diagnosis of tics that reoccurs in adulthood even if she was previously diagnosed as having a functional movement disorder. While we favored tic, the referring doctor was firm in the functional diagnosis; hence, electrophysiological study was done for further investigation.

2.1. Clinical neurophysiological evaluation

The first step in the evaluation of a hyperkinetic movement is multichannel surface electromyography (EMG) (Hallett, 2003a). The purpose of this investigation is to see the duration of the EMG bursts in the involved muscles and the relationship of the bursts in the different muscles, particularly in antagonist pairs. Here there were EMG bursts with variable duration of 300–1000 ms in the right deltoid, triceps, biceps, extensor carpi radialis (ECR) and flexor carpi radialis (FCR) muscle. The jerking movements demonstrated a triphasic pattern of the agonist and antagonist muscle activity resembling voluntary ballistic movements (Fig. 1).

The second step is to evaluate the relationship of the EEG to the EMG bursting (Hallett, 2012). This is accomplished with EEG–EMG jerk-locked back-averaging. Multichannel EEG is recorded together with an active EMG channel, and the EEG before the EMG onset is averaged and evaluated. In this case, the back-averaging was performed with respect to the onset of EMG activity on the right triceps and averaging EEG in central and frontal leads (Fz, Cz, F3, F4, C3, C4). A steep rising negative potential was seen in the central leads (Cz and C3) beginning at 140 ms prior to the movement onset with amplitude of 8.5 and 5.9 μV consistent with the late Bereitschaftspotential (late BP) (Fig. 2). Then similar back-averaging was done with voluntary movements mimicking the involuntary movements. In this circumstance, a slow rising negative potential was seen in all leads beginning at 1100 ms prior to the movement onset with amplitude of 9.6–14.5 μV (Fig. 3).

The study was interpreted as supporting the suspected diagnosis of chronic motor tics. The reasoning for this interpretation is given below. The patient was sent back to the referring neurologist who now accepted the diagnosis. Subsequently the patient started taking tetrabenazine 25 mg for tic control with significant improvement.

3. Discussion

Resolving the differential diagnosis between tics and functional myoclonus can be difficult (van der Salm et al., 2013). Simple motor tics are sudden, brief, repetitive jerk-like movements that vary in frequency, intensity and distribution (Jankovic and Kurlan, 2011; Hallett, 2015). Some clinical features of motor tics including variability of movements, sudden onset, distractibility, suggestibility, temporary remission and fluctuating courses are also frequently observed in functional myoclonus (Gupta and Lang, 2009; Jankovic and Kurlan, 2011; Baizabal-Carvallo et al., 2014; Demartini et al., 2015). Motor tics often have a premonitory urge and may be temporarily suppressed (Kwak et al., 2003; Himle and Woods, 2005). Lack of premonitory sensation, inability to suppress the movements, lack of typical rostrocaudal tic distribution over time, adult onset, absence of childhood and family history

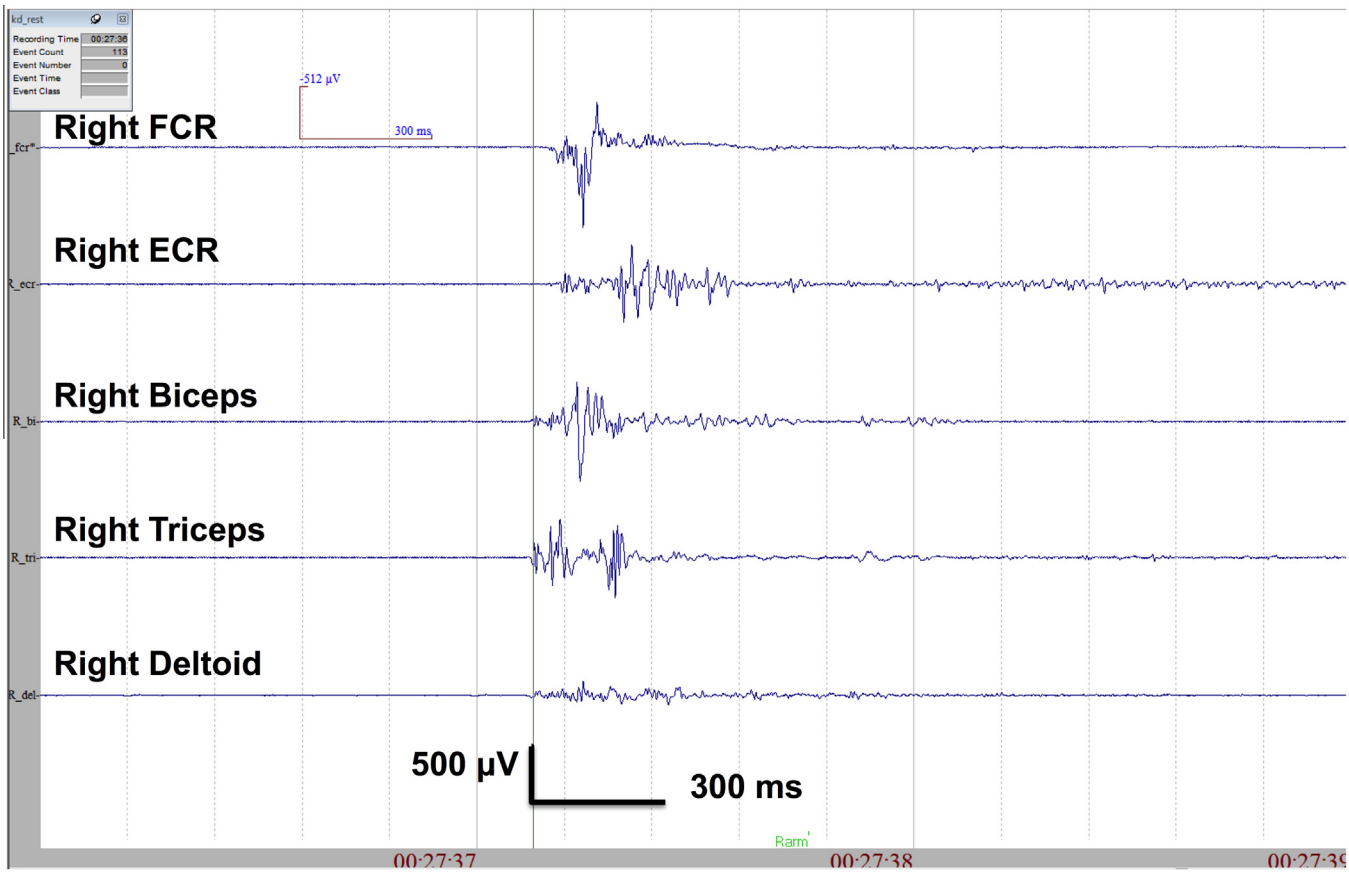

Fig. 1. Multichannel surface EMG showed EMG bursts with variable durations in the triphasic pattern of the agonist and antagonist muscle activity resembling voluntary ballistic movement. FCR is flexor carpi radialis and ECR is extensor carpi radialis.