Clinical commentary

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Bilateral synchronous gamma range firing of bulbar motor units during tonic seizures

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ABSTRACT – We present a patient with tonic seizures of the bulbar muscles. Video-EEG recording during the multiple attacks showed no ictal EEG abnormalities. The tonic seizures were clearly visible as EMG artefacts on the EEG recording and consisted of 3-6 symmetric tonic bursts of EMG activity of 2-4 seconds and an interval of 2-6 seconds. Spectral analysis showed a very distinct, bilateral synchronous, dominant gamma peak on the EMG spectra during the seizures with a cross correlation of 0.67 between left and right without a time lag. We discuss whether this peak originates from the firing frequencies of the bulbar motor units, and whether this is consistent with the Piper rhythm described during maximal voluntary contraction in controls and related to firing in the gamma range of the cortical motor neurons. We conclude that the EMG artefacts recorded on EEG during seizures can be informative with regards to the type and pathophysiology of epilepsy.

Key words: tonic, epilepsy, EEG, EMG

In epilepsy, the EEG is important to detect interictal spikes but is also often informative during seizures as ictal EEG patterns are well described (Fisher *et al.*, 2014). However, it is not unusual to find no ictal EEG abnormalities during seizures; generally, this is thought to be related to the small surface area of active cortex. Often, the EEG is then found to be full of artefacts. However, despite the negative connotations associated with the word "artefact" (Tao *et al.*, 2005), we want to draw attention to the possible value of studying such (EMG) artefacts. There is evidence that during tonic seizures, the frequency content of the surface EMG has higher values and a shift of the median frequency to higher frequencies is noticed (Conradson *et al.*, 2011). The following case illustrates the added value of studying EMG artefacts on EEG.

Case study

A 35-year-old man had nocturnal seizures for 12 years that consisted of paresthesias on the right side of the mouth, followed by a burning pain around the mouth and nose on both sides (but more prominent on the right side), which was followed by myoclonia of the right side of the face. This was sometimes also

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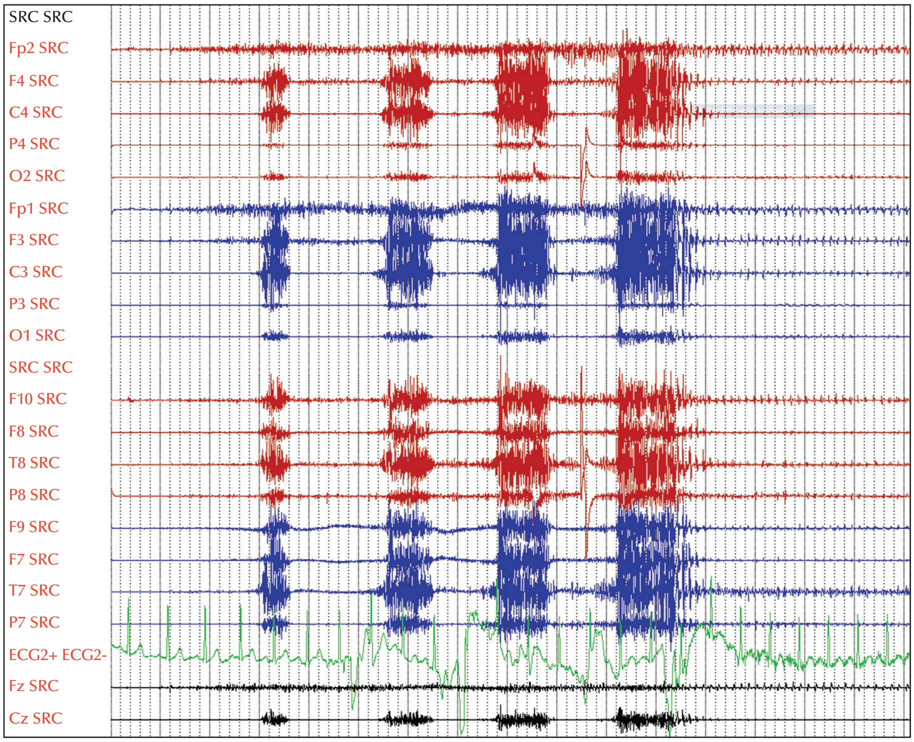

Figure 1. Overview of one tonic seizure with four bursts of bilateral EMG activity followed by single-unit ~12-Hz firing. Source derivation. X axis: each division represents a second; Y axis: 250 mmV/cm.

followed by myoclonic movements of the head on the right side with a total duration of 30 seconds. At the age of 25, the seizures became generalized three times to produce tonic-clonic seizures, and the patient sought medical help.

EEG and MRI showed no abnormalities and levetiracetam was prescribed. The tonic-clonic seizures stopped but the focal aware seizures continued. Addition of gabapentin, substitution of levetiracetam by lamotrigine, and addition of valproate and clonazepam did not change the situation. An increase in seizure frequency to several seizures every night, resulting in loss of employment, led to analysis of the seizures, taking advantage of the high seizure frequency to perform ictal registration. The semiology of the seizures was changed (see below).

During a 24-hour video-EEG recording (Micromed System Plus®; sample frequency: 256 Hz), no interictal abnormalities were identified, as previously reported. Over 24 hours, a total of 23 seizures during the night and three during day time were registered. All seizures consisted of a tonic cramp around the mouth with extension to the other bulbar muscles (facial and masticatory muscles). Consciousness was not impaired.

The ictal EEG did not show epileptic activity either, however, muscle artefacts were present with the following structure: 3-6 symmetrical tonic bursts of a 2-4 seconds with intervals of 2-6 seconds followed by rhythmic single motor unit activity, at about 10 Hz on both sides (*figure 1*). The EMG bursts were maximal in the temporal and fronto-central regions. Frequency analysis of the bipolar derivations showed a very sharp peak of ~53 Hz for the first burst decreasing to 42, 38, and 33 Hz for the second, third, and fourth burst, respectively (*figure 2B*). Bilateral synchronous firing is shown in *figure 2A*. The EMG signals between left and right showed a maximum cross correlation value of 0.67 (Pearson correlation) with no time lag. In order to compare the EMG signals between those associated with the seizures and those with normal contraction, we also analysed the EMG signals during normal chewing (*figure 2C, D*).

Discussion

The EEG during the seizures of this patient showed no ictal activity but according to the EMG artefact, tonic contraction of the faciale and masticatory muscles occurred with a strong peak in the power spectrum at around 53-33 Hz. The determination of the surface EMG power spectrum is based on the power spectrum density function of the underlying motor unit action potentials multiplied by the power spectra of the

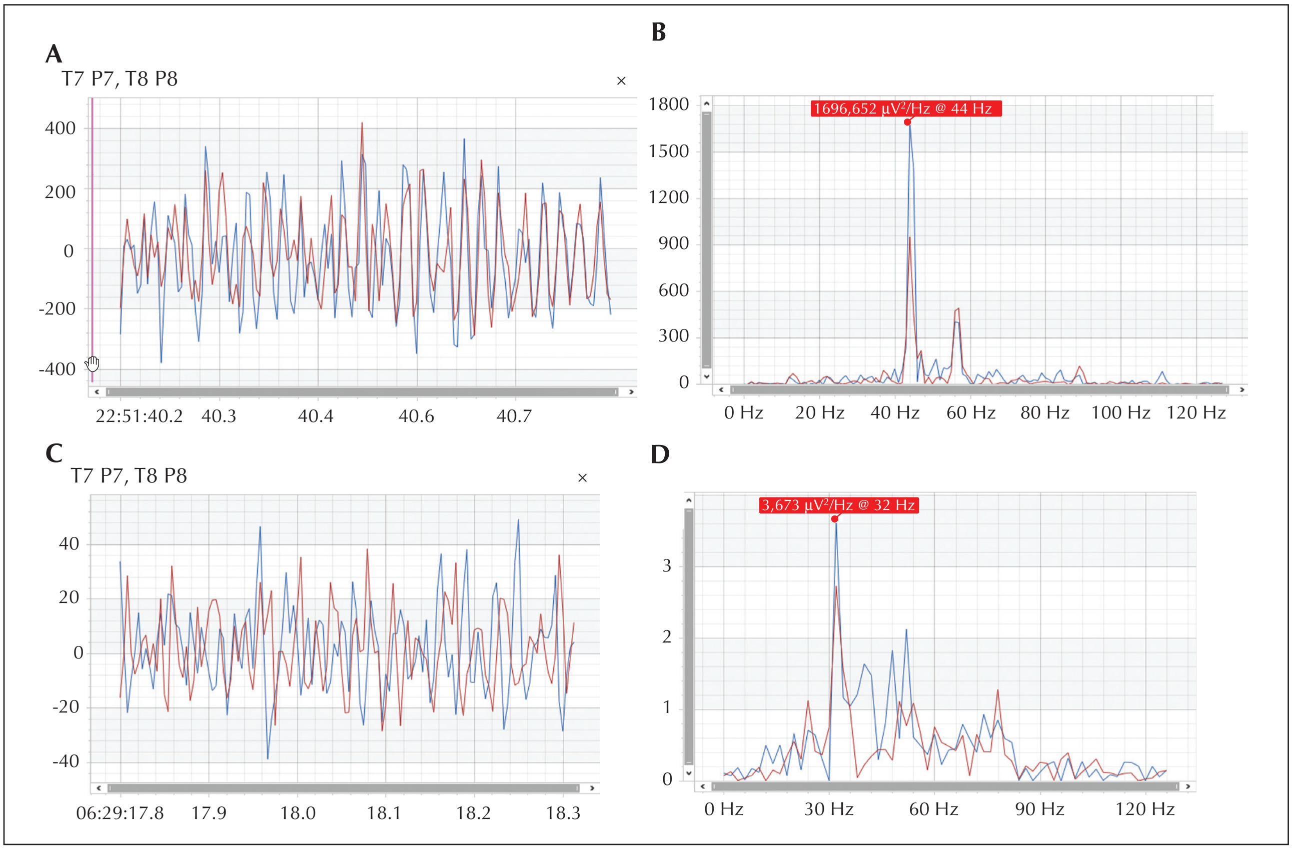

Figure 2. (A) EMG activity during a tonic seizure at T7-P7 and T8-P8. Note the synchronous activity between the right and left side. X axis: seconds; y axis: microvolts. (B) Frequency spectrum of the second tonic burst (*Figure 1*), showing peak firing at 42 Hz. For comparison, the temporal EMG plot (C) and its spectra (D) for the same electrodes during normal activity (chewing) of the patient are presented (also note the different scale of the y axis).

firing processes (Lago and Jones, 1977; van Boxtel and Schomaker, 1983; Hermens *et al*., 1992). During normal contraction, the spectrum of surface EMG has a broad band frequency content from zero to about 200 Hz. In the case of strong synchronization of the firing processes, a clear peak within the spectra emerges. During the tonic phase of seizures, the frequency spectrum of the surface EMG is reported to shift to higher frequencies (about 10-20 Hz), but the global form of the spectrum does not change (Conradsen *et al*., 2011). Thus, the spectra of our patient can be explained by a very prominent and abnormal firing pattern of the motor units consisting of a monorhythmic 53-Hz firing, decreasing to 33 Hz in the final burst. In addition, this firing was bilateral and synchronous. Interestingly, such a prominent peak frequency at around 40 Hz was described during maximal isometric contractions of controls and this is known as the Piper rhythm (Brown *et al*, 1998). However, the frequency spectra of these maximal contractions do not show such a prominent peak, as in our patient. The Piper rhythm of controls has also been shown to strongly correlate with EEG and/or MEG activity in the contralateral motor area and it is reasoned that this is the result of firing within the gamma range of the neurons in the motor cortex. Moreover, the bilateral synchronous firing (as demonstrated by the high level of cross correlation of 0.67) (*figure 2A*) is also very abnormal and is not found during normal maximal contractions in humans. This bilateral synchronous firing can be explained by a single pacemaker that is responsible for the bilateral activation.

In our view, a possible explanation for the phenomenon is that the abnormal epileptic discharge from the left motor cortex or possibly insular/opercular area (left side, in view of the clinical signs) drives the right motor cortex by spreading and subsequently activates the lower motor nuclei innervating the bulbar muscles. In this case, this abnormal firing is highly synchronized and occurs close to the Piper rhythm where the cortex seems to have a preferential firing rate during maximal contractions. Interestingly, the masticatory muscles innervated by the motor nuclei of the trigeminal nerve have been shown to be activated optimally between 40 and 60 Hz, corresponding to the optimal stimulation frequency of the cortical masticatory area to induce mastication (Morquette *et al*., 2012). We could not find other examples in the literature of such an abnormal firing behaviour during tonic seizures, besides the generally higher values of the surface EMG and a shift of the median frequency to higher frequencies in tonic seizures, although without such a distinct peak frequency (Conradson *et al*, 2011). In conclusion, the highly abnormal EMG patterns in the bulbar muscles during the tonic seizures of this patient, as measured on the scalp showing a strong peak in the power spectra at 53-33 Hz and abnormal bilateral hypersynchrony, are an expression of the abnormal epileptic discharge originating in the motor cortex. □

Supplementary data

Summary didactic slides are available on the www.epilepticdisorders.com website.

Disclosures

None of the authors have any conflict of interest to declare.

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TEST YOURSELF

(1) What determines the shape of the power spectrum of surface EMG?

(2) In the literature, the power spectrum of surface EMG during tonic seizures changes. Does the frequency increase or decrease?

(3) What is the Piper rhythm?

Note: Reading the manuscript provides an answer to all questions. Correct answers may be accessed on the website, www.epilepticdisorders.com, under the section "The EpiCentre".