Epilepsia partialis continua: semiology and differential diagnoses

Christian G. Bien, Christian E. Elger
Dept. of Epileptology, University of Bonn, Germany
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ABSTRACT – Epilepsia partialis continua (EPC) is a rare form of focal status epilepticus. It may have vascular, immune-mediated, neoplastic or metabolic-toxic causes. The origin of EPC has been linked with the motor cortex. This has been solidly supported by sophisticated electrophysiological studies. Here, a series of video sequences from patients with EPC (due to Rasmussen encephalitis, early-stage multiple sclerosis, and steroid responsive encephalopathy with autoimmune thyroiditis), and other cases with repetitive myoclonic jerks or movement disorders (myoclonic epilepsy associated with ragged-red fibers, Jacksonian march, myoclonic seizures in other types of frontal lobe or idiopathic generalized epilepsies, and different types of tremor) is presented.

[Published with video sequences].

Key words: epilepsia partialis continua, semiology, diagnosis

In 1894-1895, Koževnikov (or Koshewnikow) reported in Russian and in German on four patients who suffered from mild to moderate hemiparesis and from what he called “epilepsia corticalis sive partialis continua” in the paretic parts of the body (Koževnikov 1894, Koshewnikow 1895). Today, only the term epilepsy partialis continua (EPC) is still in use. The Russian neurologist conceptualized EPC as a particular form of epilepsy and distinguished it from “common epilepsy” because it did not have the tendency to spread and was often not self-limiting. It was clear to Koževnikov that in the four patients he described, the EPC originated in the hemisphere contralateral to the affected side of the body, close to the primary motor area. He was, however, unable to clarify the nature of the organic brain lesion, which in his opinion was underlying the continuous myoclonic jerks. Based on the slowly progressive nature of the disease in his patients, he hypothesized that a chronic encephalitic process was at work. He was probably right because it can be assumed that the majority of his patients suffered from tick-borne, viral Russian spring summer meningoencephalitis, which was detected only some decades later (Omorokow 1927, Smorodintsev 1958).

In contrast to Koževnikov’s terminology, EPC is today conceptualized as a seizure type rather than a form of epilepsy, and is subsumed under the group of focal status epilepticus (Engel 2001). Following Koževnikov’s publications, subsequent research on EPC has mainly dealt with three – in part interrelated – questions: how should EPC be defined? Which pathological processes cause EPC? Where in the nervous system does this phenomenon arise?

The current, broadly accepted answers to these questions are very briefly
The neurophysiological conceptualization of EPC is complicated – or rather: enriched – by the fact that it has been studied from the viewpoint of both epilepsy and movement disorder research. Depending on their main field of interest, authors may – in addition to the aforementioned clinical features – require the demonstration of epileptiform EEG abnormalities (ideally in a fixed temporal coupling to the muscle jerks), or abnormalities as shown by other electrophysiological studies such as giant somatosensory-evoked potentials (SSEPs) in order to demonstrate the cortical origin of the muscle jerks (Cockerell et al. 1996).

Semiology of EPC

Typical clinical signs of EPC are: combination of the repetitive myoclonic jerks with hemiparesis or – less frequently – with other cortically-generated deficits; monomorphic, simple, brief excursions of the affected limb; regular or irregular occurrence of the jerks; involvement of distal rather than proximal muscle groups; physical exercise, sensitive stimulation or psychic exertion may increase the amplitude and frequency of the myoclonic jerks; more frequent involvement of the upper than the lower half of the body (in 151 patients, the distribution was as follows: head: 16%; head and upper extremity: 14%; upper extremity: 40%; trunk: 5%; lower extremity: 14%; whole side of body: 11% - the sum is larger than 100% because multiple sites may have been affected during the disease course [Löhler and Peters 1974]); EPC more often continues during sleep – sometimes in a milder form – rather than stops (25 versus 5 patients from the literature according to [Löhler and Peters 1974]). The same authors calculated a mean frequency of 90 jerks per minute (i.e. 1.5 Hz; range 3-360 per minute, equaling 0.06-6 Hz) from the literature. There is no preference for one side of the body or the other.

Differential diagnoses

In the second part of the video sequence and the accompanying table (table 1), differential diagnoses for EPC are presented. Tremor has a characteristic alternating agonist-antagonist innervation pattern (patients 13, 15). The cortical myoclonic jerks of patients with myoclonus epilepsies are summarized in the following paragraphs. Thereafter, a description of the semiological features of EPC and possible differential diagnoses are given, with a video sequence illustrating them (see video sequence).
affect the limbs diffusely (patient 14). Sometimes, even extensive investigations do not clarify the origin of the repetitive myoclonic jerks as in patient 12; the long duration of the bursts mitigates against a typical cortical origin. The Jacksonian march (patient 16) is clearly distinguishable because of the sequential involvement of unilateral body parts.

Summary

EPC is a rare type of focal status epilepticus. Characteristic semiological features enable it to be diagnosed and distinguished from other movement disorders or myoclonic symptoms. Additional electrophysiological studies can contribute to an enhanced degree of certainty regarding the cortical origin of the myoclonic jerks. In childhood, the most frequent cause of EPC is Rasmussen encephalitis (Schomer 1993). At older ages, vascular and neoplastic diseases are the most frequent causes. Metabolic, toxic and other immune-mediated causes account for most of the remaining cases. The prognosis depends on the underlying cause and its appropriate treatment. The search for the underlying disease is therefore of paramount importance.

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References


Koževnikov AJ. Osobyj vid kotikal’noj epilepsii. Medicinskoe obozrenie (Moskva) 1894; 42: 94-118.


Legend for video sequence

In the attached video sequence, we present video EEG recordings of patients with repetitive involuntary jerks:

Eleven patients with EPC, defined according to clinical criteria (all patients with the diagnosis of EPC and an adequate video EEG recording from this center during the period September 2000 until March 2007), and 5 patients with other types of myoclonia or movement disorders are included.

Ten of the EPC patients have been diagnosed with immune-mediated disorders; one patient’s EPC was of unknown cause.

In some patients, additional electrophysiological studies were performed. For details, see table 1.

The apparently long duration of the EMG jerks on our recordings are probably best explained by the peculiarities of recordings with an EEG system with a lower sampling rate (here: 200 Hz), and filter settings different from those usually applied in pure EMG recordings.

SSEPs on stimulation of the median nerves were considered as giant potentials if the amplitude of P25-N33 was > 12 μV (Obeso et al. 1985). The overrepresentation of inflammatory conditions and the lack of vascular diseases in this series is due to the patient population from this tertiary center, with its particular interest in immune-mediated syndromes and a (surprisingly) low number of referrals of elderly patients. For recording montages, see figures 1 and 2; in the split-screen videos, red curves represent left hemispheric leads, and blue curves represent right hemisphere leads. EEG channels with epileptiform activity, and EMG channels are marked by red colour.
Figure 1. 23 EEG channels.

Figure 2. 16 EEG channels.

ADM: musculus abductor digiti minimi
AH: musculus abductor hallucis (foot)
APB: musculus abductor pollicis brevis (hand)
centr: central
EPC: epilepsia partialis continua
F: female
FCU: musculus flexor carpi ulnaris
FLE: frontal lobe epilepsy
fr: frontal
hem: hemisphere
Hypoth: hypothenar
IGE: idiopathic generalized epilepsy
L: left
LTG: lamotrigine
M. add. long.: musculus adductor longus
M. grac.: musculus gracilis
M. orb. oc.: musculus orbicularis oculi
M. vast. lat.: musculus vastus lateralis
M: male
MERRF: myoclonic epilepsy-associated with ragged-red fibers
mo: months
MS: multiple sclerosis
Nd: not done
R: right
RE: Rasmussen encephalitis
SREAT: steroid responsive encephalopathy with autoimmune thyroiditis (formerly Hashimoto’s encephalopathy)
SSEPs: somatosensitve-evoked potentials
sync: synchronous jerks in the recorded muscles
temp: temporal
VPA: valproic acid
wks: weeks
yrs: years
Table 1. Patients shown in video with epilepsia partialis continua and with other conditions.

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Age (yrs)</th>
<th>Diagnosis</th>
<th>Time since onset</th>
<th>Body region affected</th>
<th>Epileptiform potentials on EEG</th>
<th>EMG bursts: apparent duration and synchronicity</th>
<th>Amplitude of SSEPs on stimulation of median nerve P25-N33 (µV)</th>
<th>Ipsilateral-contralateral</th>
<th>Additional neurological deficit</th>
</tr>
</thead>
<tbody>
<tr>
<td>1, M</td>
<td>11.3</td>
<td>RE, R hem</td>
<td>3.0 yrs</td>
<td>L face</td>
<td>Yes</td>
<td>Nd</td>
<td>Nd</td>
<td>L hemiparesis</td>
<td></td>
</tr>
<tr>
<td>2, F</td>
<td>11.4</td>
<td>RE, L hem</td>
<td>13 mo</td>
<td>R tongue</td>
<td>Yes</td>
<td>Nd</td>
<td>Normal</td>
<td>R hemiparesis</td>
<td></td>
</tr>
<tr>
<td>3, M</td>
<td>13.2</td>
<td>RE, L hem</td>
<td>4.0 yrs</td>
<td>R face</td>
<td>No (artifacts)</td>
<td>&lt; 200 ms (only one muscle)</td>
<td>30-5</td>
<td>R hemiparesis</td>
<td></td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Same patient, 1.8 yrs later</td>
<td></td>
<td></td>
<td></td>
<td>R face</td>
<td>Yes</td>
<td>Nd</td>
<td>Nd</td>
<td>R hemiparesis</td>
<td></td>
</tr>
<tr>
<td>4, M</td>
<td>18.6</td>
<td>Unclear</td>
<td>3 yrs</td>
<td>Face, pectoral muscle, L arm</td>
<td>No</td>
<td>&lt; 150 ms, sync</td>
<td>12.5-1.8</td>
<td>None</td>
<td></td>
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<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Same patient, 8 mo later</td>
<td></td>
<td></td>
<td></td>
<td>R hand</td>
<td>Yes</td>
<td>Nd</td>
<td>Nd</td>
<td>R hemiparesis</td>
<td></td>
</tr>
<tr>
<td>5, M</td>
<td>10.5</td>
<td>RE, L hem</td>
<td>2.2 yrs</td>
<td>R hand</td>
<td>Yes</td>
<td>&lt; 200 ms, sync</td>
<td>87-7</td>
<td>R hemiparesis</td>
<td></td>
</tr>
<tr>
<td>6, F</td>
<td>23.8</td>
<td>Early-stage MS</td>
<td>Days</td>
<td>L hand</td>
<td>No</td>
<td>&lt; 250 ms, sync</td>
<td>Normal</td>
<td>None</td>
<td></td>
</tr>
<tr>
<td>7, F</td>
<td>62.8</td>
<td>SREAT (Hashimoto’s)</td>
<td>3 wks</td>
<td>L hand</td>
<td>No</td>
<td>Nd</td>
<td>Nd</td>
<td>Dystonia L hand</td>
<td></td>
</tr>
<tr>
<td>8, F</td>
<td>42.1</td>
<td>RE, L hem</td>
<td>26 mo</td>
<td>R hand</td>
<td>No</td>
<td>Nd</td>
<td>Normal</td>
<td>R hemiparesis</td>
<td></td>
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<tr>
<td>Same patient, 1.8 yrs later</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9, M</td>
<td>6.3</td>
<td>RE, L hem</td>
<td>1 mo</td>
<td>R foot and leg</td>
<td>Yes</td>
<td>Nd</td>
<td>Nd</td>
<td>R hemiparesis</td>
<td></td>
</tr>
<tr>
<td>10, F</td>
<td>5.1</td>
<td>RE, L hem</td>
<td>10 mo</td>
<td>R hand and foot</td>
<td>Yes</td>
<td>&lt; 150 ms, sync</td>
<td>Normal</td>
<td>R hemiparesis</td>
<td></td>
</tr>
<tr>
<td>11, F</td>
<td>16.6</td>
<td>RE, R hem</td>
<td>3.3 yrs</td>
<td>L hand and foot</td>
<td>No</td>
<td>&lt; 150 ms, sync</td>
<td>Normal</td>
<td>L hemiparesis</td>
<td></td>
</tr>
</tbody>
</table>

Differential diagnoses

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<tr>
<th>Patient no.</th>
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</thead>
<tbody>
<tr>
<td>12, F</td>
<td>18.9</td>
<td>Myoclonia of unknown origin. FLE or IGE</td>
<td>5.9 yrs</td>
<td>L leg</td>
<td>No</td>
<td>&lt; 600 ms, sync</td>
<td>Nd</td>
<td>None</td>
<td></td>
</tr>
<tr>
<td>13, F</td>
<td>2.8</td>
<td>Tremor ataxia teleangiectasia</td>
<td>2 mo</td>
<td>L arm</td>
<td>No</td>
<td>Nd</td>
<td>Nd</td>
<td>Ataxia</td>
<td></td>
</tr>
<tr>
<td>14, M</td>
<td>23.3</td>
<td>Cortical myoclonia MERRF</td>
<td>14 yrs</td>
<td>Diffuse</td>
<td>No</td>
<td>Nd</td>
<td>Nd</td>
<td>Ataxia, paraparesis</td>
<td></td>
</tr>
<tr>
<td>15, M</td>
<td>26.8</td>
<td>Tremor VPA-LTG overdose</td>
<td>Weeks</td>
<td>Upper extremities</td>
<td>No</td>
<td>Nd</td>
<td>Nd</td>
<td>None</td>
<td></td>
</tr>
<tr>
<td>16, M</td>
<td>21</td>
<td>Jacksonian march. FLE</td>
<td>9 yrs</td>
<td>L hand, spread to rest of hemibody</td>
<td>Late in seizure</td>
<td>Nd</td>
<td>Nd</td>
<td>None</td>
<td></td>
</tr>
</tbody>
</table>