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Authors
Baram, TZ
Mitchell, WG
Snead, OC

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Prognostic Significance of Acute Epilepsia Partialis Continua

Tallie Z. Baram, MD, PhD, Wendy G. Mitchell, MD, and O. Carter Snead, III, MD

We present 3 patients in whom epilepsy partialis continua was the presenting sign of an acute, rapidly evolving and catastrophic neurologic illness. Initial seizures were partial simple (i.e., eye deviation in one, finger twitching in one) which progressed to multifocal partial seizures. The course of the epilepsy partialis continua was 36-41 days. Prognosis was uniformly poor (i.e., death in 2, vegetative state in 1); therefore, epilepsy partialis continua in the context of an acute neurologic illness may herald a grim outcome.


Introduction

Epilepsia partialis continua (EPC) denotes partial simple or partial complex seizures continuing for a prolonged period [1,2]. Described in both children and adults, EPC has diverse etiologies and outcomes [1]. The majority of reported pediatric EPC cases have been associated with chronic, progressive encephalitis of the Rasmussen type. Rasmussen encephalitis is characterized by EPC and progressive hemiparesis usually evolving over many months to years, eventually resulting in hemiplegia and loss of function of the involved hemisphere [3-7].

We present 3 patients in whom EPC was an early and major component of an acute, rapidly evolving illness, clinically and etiologically distinct from Rasmussen encephalitis. The course was rapid; maximal neurologic dysfunction was evident within days. Causative factors varied, but the outcome was uniformly poor. EPC may present in the context of acute, catastrophic neurologic disease, resulting in a dismal outcome.

Case Reports

Patient 1. A 7-year-old Salvadoran boy developed fever (40.5°C), pharyngitis, and gastroenteritis. A generalized seizure occurred on the first day, followed by very frequent partial seizures and stupor. Seizures originated in either arm or in the face, sometimes occurring simultaneously bilaterally. Electroencephalography (EEG) disclosed nearly continuous partial onset of electrographic seizures, correlating closely with observed motor activity. Epileptic activity originated in either hemisphere, sometimes with bilateral asynchronous independent discharges. Antiepileptic drugs (AEDs) that failed to alter the seizures included carbamazepine, phenytoin, lorazepam, and clonazepam. Very high-dose phenobarbital eliminated clinical seizures only when an electrographic burst-suppression pattern was achieved. Over a period of several weeks, a continuously increasing dose of phenobarbital was required to maintain seizure control [8]. The course was progressively downhill; death occurred on the 39th day. Cerebrospinal fluid (CSF) obtained on the fourth day of illness harbored an enterovirus with the growth properties of Coxsackie A. Autopsy revealed diffuse neuronal loss and gliosis.

Patient 2. A 5-year-old Hispanic girl presented with fever, gastroenteritis, and intermittent eye deviation to the left. On the first 3 days of illness she was alert and coherent during episodes of persistent leftward eye deviation with irregular left jerk nystagmus. On the fourth day, one of these events progressed to a generalized tonic-clonic seizure, followed by altered sensorium. She continued to have almost constant partial seizures with an electrographic seizure discharge localized to the right parietal-occipital region which correlated with the eye movements (Fig 1). Subsequently, bilateral but independent clinical and electrographic seizures developed. Acutely, the seizures were refractory to conventional doses of AEDs and abated only when very large doses of barbiturates (i.e., phenobarbital, pentobarbital) induced a burst-suppression pattern on EEG. During the second week of illness, high-dose carbamazepine transiently decreased the frequency and amplitude of motor phenomena. CSF findings are listed in Table 1.

Computed tomography (CT) and magnetic resonance imaging (MRI) revealed a cerebellar infarct and bitemporal abnormal signal. Cerebral angiography did not reveal a vasculitis and a brain biopsy from the involved cerebellum demonstrated inflammation and nonspecific gliosis. Corticosteroids and acyclovir were administered with no change in mental status or seizures. EPC abated 36 days after onset: however, 6 months later, the child is able to open her eyes but has no purposeful movements and little response to the environment. She has frequent focal motor seizures, mainly of the right arm.

Patient 3. A 27-year-old man with hemophilia and transfusion-related AIDS presented with focal motor seizures involving the left hand which occurred 6 months after an episode of severe measles. (The measles virus was cultured from the CSF.) Four days later, continuous left thumb twitching developed. Electrographic discharges at the right midparietal region coincided with the thumb movements. Clinical seizures progressed to involve the left arm and, eventually, the left side of the face. MRI revealed a small lesion involving a single gyrus in the right mid-parietal cortex (Fig 2) The seizures were refractory to high levels of AEDs; independent partial seizure activity of the right arm and face developed, as well as palatal and laryngeal involvement. The patient died 41 days after the onset of EPC.

CSF results are listed in Table 1. Brain biopsy and autopsy revealed cortical and white matter gliosis and microglial nodules, without multi-nucleated giant cells. Cowdry A inclusions were found on the 2 brain specimens, but neither measles nor human immunodeficiency viral particles were found. Progressive measles encephalitis was diagnosed on...
Discussion

EPC first described at the end of the nineteenth century, is considered to be a manifestation of focal cortical dysfunction, involving the motor strip [1,12]. The clinical entities associated with EPC have included infection (granuloma, abscess, encephalitis), autoimmune disease, infarction, trauma, metabolic encephalopathy, and others [1,3,11-14].

Although EPC is commonly associated with epileptic discharges limited to one cortical region, Thomas et al. emphasized that in a significant number of patients more than one focus is found and the patients may exhibit “concurrent though independent twitching” [1]. Thus, as in our patients, it may be difficult to distinguish EPC from multifocal epilepsy.

The majority of patients in series of EPC have had an indolent course. The mean duration of EPC in the 32 patients reported by Thomas et al. was 25 months [1]. In pediatric patients, a well-delineated entity of slowly progressive unilateral hemispheric dysfunction associated with EPC was described by many authors [3-7]. In reviewing 48 patients with “chronic encephalitis” and epilepsy, Rasmussen and Andermann found about 50% with EPC [6]. The course was indolently progressive, with “insidious and gradual” neurologic deterioration.

The patients we encountered had a distinctly different course: explosive onset of focal or generalized seizures progressed rapidly to EPC, followed by altered sensorium leading to coma and death or a vegetative state. Their ages of onset and course are also quite distinct from that of the hemiconvulsion-hemiparesis syndrome [15]. In the series reported by Thomas et al., 5 of 12 patients with EPC of less than 1 week’s duration died [1]. Their ages were not recorded. Mikati et al. described a boy with an abrupt onset of EPC leading, over 1 month, to coma [16]. The patient recovered and his neurologic examination 1 year later was normal. The etiology of his EPC was not elucidated. EPC

<table>
<thead>
<tr>
<th>Patient No./Age</th>
<th>CSF Chemistry (mg/dl)</th>
<th>CSF Cells</th>
<th>CSF Culture</th>
<th>Brain Pathology Biopsy/Autopsy</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/7/M</td>
<td>Protein 32, Glucose 91</td>
<td>5 white, 6 red</td>
<td>Enterovirus</td>
<td>Meningitis, gliosis, neuronal loss</td>
</tr>
<tr>
<td>2/5/F</td>
<td>Protein 27, Glucose 83</td>
<td>15 white, 1 red</td>
<td>—</td>
<td>Cerebellum-gliosis</td>
</tr>
<tr>
<td>3/27/M</td>
<td>Protein 33, Glucose 109</td>
<td>2 white, 0 red</td>
<td>Measles virus</td>
<td>Gliosis, microglial nodules</td>
</tr>
</tbody>
</table>
Figure 2. MRI scan performed on Patient 3 demonstrating a small lesion in the right midparietal region (arrow). This MRI was obtained on a 0.5 T Technicare instrument. T₂-weighted image: TR 3,000 msec, TE 96 msec.

has been described with progressive measles encephalitis in 4 immunocompromised children [9]. Their courses, as in Patient 3, were short (9 days to 2 months) and fatal.

Our patients had in common a fulminant illness, probably resulting from viral cerebritis. CSF studies in all were abnormal and a putative causative organism was found in two. Thus, EPC may herald an acute encephalitis whose etiology should be aggressively sought and treated because otherwise the outcome may be poor.

References