Isolated, Chronic, Epilepsia Partialis Continua in an HIV-Infected Patient

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**Background:** The characteristic clinical feature of epilepsia partialis continua (EPC) is chronic focal myoclonus, usually involving the distal part of one extremity. A variety of pathogenetic factors have been implicated in EPC. In children, the most common cause is Rasmussen encephalitis; in adults, it is vascular disease or tumor involving the sensorimotor cortex. Epileptic seizures are a relatively common manifestation of central nervous system involvement in patients infected with human immunodeficiency virus (HIV), but, to our knowledge, isolated, chronic EPC has not been previously reported.

**Objective:** To describe a case of typical EPC in a patient infected with HIV.

**Design and Setting:** Case report from an epilepsy center.

**Patient:** A 58-year-old man infected with HIV had continuous myoclonus that involved the right arm and was associated with intermittent motor seizures. The electroencephalographic findings were normal at the onset of the symptoms, but left central theta rhythm appeared later. Serial magnetic resonance imaging scans obtained over a 3-month period showed a progressively increasing left rolandic T2-weighted hypersignal. Histologic study of a stereotactic biopsy specimen demonstrated inflammation characterized by perivascular mononuclear cell infiltration. The only detectable cause was HIV infection. Immunochemical tests ruled out JC virus. Neuropsychological testing showed no evidence of cognitive impairment. An electroencephalographic-electromyographic “back-averaging” study showed a reproducible transient left biphasic complex preceding the bursts by about 30 milliseconds on the C3 and F3 electrodes, thus demonstrating that the myoclonus was of cortical origin. High-dose corticosteroid (prednisone, 100 mg/d) and anti–HIV-1 therapy led to marked radiological and clinical improvement. Infection with HIV enhances the risk of seizures, but, to our knowledge, this is the first reported case of “inflammatory” EPC.

**Conclusions:** The present case suggests that the possibility of central nervous system involvement by HIV-1 should be taken into account in the diagnostic workup of patients with EPC. This case also indicates that treatment can be effective.

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a combination of isoniazid and rifampin. The patient, who was right-handed, reported that the twitching, which had begun 5 months earlier, impaired writing ability and was getting progressively worse. He further reported that the myoclonus had become constant during the day within the last month and that it worsened whenever he tried to write or grasp objects.

Examination confirmed the presence of characteristic focal myoclonus with backward flexion of the fingers and adduction of the wrist. Myoclonus was associated with intermittent motor seizures with tonic contraction that progressed from the right hand up into the arm and was followed by clonus and distal motor deficit of the right upper extremity for 15 minutes. Motor seizure was unpredictable but occurred approximately once a day. Secondary generalization was never observed. Closer examination revealed slight hypertonia of the right upper extremity. No other abnormalities were noted. Heart rhythm and fundoscopic findings were normal. The results of standard laboratory tests were normal. The T4 lymphocyte count was 0.007 × 10^9/L. A computed tomogram of the brain and a conventional EEG obtained 1 month earlier showed no abnormalities. In April 1996, a magnetic resonance imaging scan of the brain demonstrated a T1 hyposignal and a T2 hypersignal, indicating a focal parenchymatous lesion in the left rolandic region (Figure 1, A).

In May 1996, a digital EEG (sampling rate, 250 Hz) with polygraphic recording clearly demonstrated rhythmic myoclonus of the right hand varying in frequency from 1 to 4 Hz (Figure 2). The EEG also showed irregular theta activity in the left central region. To confirm the relationship of this finding with myoclonus, back-averaging was performed. Electromyographic (EMG) activity in the extensor and flexor muscles of the right upper limb was recorded. Averaging performed on 80 cycles of 1000 milliseconds of EEG centered on the beginning of myclonia demonstrated a biphasic complex in the left central cortical region preceding EMG bursts by about 30 milliseconds on the C3 and F3 electrodes (Figure 3).

A wide range of tests failed to detect any causes other than HIV infection. The results of the following evaluations were normal: cytochemical analysis and immunoelectrophoresis of cerebrospinal fluid; tests for opportunistic blood and cerebrospinal fluid infection (by polymerase chain reaction), herpes, cytomegalovirus, and JC virus; and serologic tests for toxoplasmosis infection. Herpes simplex virus, varicella-zoster virus, Lyme disease, cytomegalovirus, ricketsiosis, syphilis, cryptococcus, KB, and autoimmune factors, including rheumatoid factor, native anti–DNA antibodies, antinuclear antibodies, circulating immune complexes, and anticardiolipid antibodies, were not detected.

Treatment with carbamazepine (800 mg/d) failed to improve the patient’s myoclonus. In July 1996, his symptoms worsened: he not only had the persistence of myoclonus and somatomotor episodes in the right upper extremity, but he also developed myoclonus in the left hand. Subsequent magnetic resonance imaging of the brain confirmed progression of lesions, with the

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**Figure 1.** T2-weighted magnetic resonance imaging (MRI) scan of the brain showing evolution of lesions. A, April 1996, small hyperintensity in the left rolandic region. A limited area of hypersignal is also seen in the right precentral gyrus. B, July 1996, MRI scan showing an extensive hypersignal in both precentral regions. C, January 1997, MRI scan showing partial regression of lesions.
development of a hypersignal in the right ascending frontal convolution (Figure 1, B). Injection of gadolinium was followed by heterogeneous contrast of the lesions.

Histologic study of a stereotactic biopsy specimen from the left rolandic region demonstrated nonspecific inflammatory perivascular lesions (mononuclear cell infiltration) and slight demyelinization. There was no evidence of any abnormal inclusions in oligodendrocytes (Figure 4). Immunocytochemical tests did not detect JC virus or cytomegalovirus.

Cerebral angiography showed a poorly vascularized area opposite the left rolandic artery. The lumina of the arteries located distally to the devascularized area were greatly narrowed, thus suggesting vasculitis. Prednisone therapy (100 mg/d) combined with treatment with anti-HIV agents (dideoxycytidine [80 mg/d], lamivudine [400 mg/d], and indinavir [2400 mg/d]) and carbamazepine was started in August 1996. A marked improvement was gradually observed, with the disappearance of partial seizures, a significant decrease in the myoclonus in the right upper extremity, and disappearance of the myoclonus in the left hand.

A third magnetic resonance imaging scan of the brain in January 1997 (Figure 1, C) confirmed a sharp regression of lesions. During the first year of follow-up, the only adverse effect of the corticosteroid therapy was an increase in body weight. Intercurrent infection has not been noted, and the last T4 lymphocyte count was 0.025 × 10⁹/L.

The defining clinical feature of EPC is continuous focal myoclonus usually involving the distal part of one extremity. Association of myoclonus with partial simple motor seizures as observed in our case is common.¹,² The anatomical and physiological mechanisms underlying EPC have fueled much controversy. However, available evidence now indicates that EPC is of cortical origin in most cases.³⁻⁵ Indeed, while conventional surface EEG has not generally been successful in determining the origin of EPC,³ back-averaging may reveal a reproducible EEG potential arising from the contralateral motor cortex and preceding the EMG burst by a short interval that is appropriate to conduction through the corticonuclear pathway.⁸ Furthermore, giant evoked somesthetic potentials have usually been measured after stimulation of the zone involved by myoclonus.⁵ In our patient, back-averaging revealed a reproducible EEG potential arising from the contralateral motor cortex that preceded the EMG burst by an interval compatible with conduction through the corticonuclear pathway. The focal nature and short duration of myoclonus were also consistent with cortical myoclonus.

A number of conditions can lead to EPC, including Rasmussen syndrome (type II) or lesions involving the central nervous system (type I).³⁻⁵ To our knowledge, however, this is the first report of EPC attributed to HIV infection. Indeed, in the present case, EPC was the only neurologic manifestation. There was no sign of...
HIV-1 encephalopathy or any other form of diffuse encephalitis. The histologic and angiographic findings in our case were consistent with those of relatively localized vasculitis.

Infection by HIV-1 is associated with an increased incidence of vasculitis. In some patients, vasculitis occurs in association with lymphoma or opportunistic infection (eg, syphilis, aspergillosis, and herpes-zoster virus), but in others, it is due directly to the virus itself and can result in clinical manifestations resembling stroke or diffuse encephalopathy.\textsuperscript{9,10} In our patient, vasculitis was probably linked directly to HIV-1, since there was no evidence of opportunistic infection and since treatment with corticosteroids and anti–HIV-1 agents was successful.

The present case suggests that the possibility of central nervous system involvement by HIV-1 should be taken into account in the diagnostic workup of patients with EPC. It also indicates that treatment can be effective.

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REFERENCES