Therapy-Resistant Opsoclonus-Myoclonus Syndrome Secondary to HIV-1 Infection

To the Editor—Chaotic eye movements and myoclonus are key features of the opsoclonus-myoclonus syndrome, also termed “dancing eye” syndrome [1]. The etiology of opsoclonus-myoclonus syndrome may be inflammatory and can occur in many contexts, most notably paraneoplastic, in association with autoimmune diseases, drug toxicity, or infections [1]. Human immunodeficiency virus (HIV)—associated opsoclonus-myoclonus syndrome has been described [2–4] and is known to respond well to combination antiretroviral therapy (cART). Here, we describe a unique case of therapy-resistant opsoclonus-myoclonus syndrome presenting as a first symptom of HIV-1 infection.

In December 2008, a 27-year-old previously healthy woman presented with a 3-week history of uncontrolled eye movements with disabling oscillopsia (see Video 1). This was constantly present: at rest, after fixation, and when the eyes were closed. The complaints started after a short influenza-like illness. Her vision was normal. There were mild complaints of unsteadiness and vertigo, which caused her to refrain from work. She had no other complaints and did not use medication or recreational drugs. Neurological examination showed opsoclonus in addition to discrete myoclonus and bilateral hand tremor. Registration of eye movements showed an elliptic pendular nystagmus. The complete blood count and blood chemistry; computed tomographic imaging of the thorax, abdomen, and pelvis; mammography; and cerebrospinal fluid analysis did not show any abnormalities. The patient initially received a diagnosis of idiopathic opsoclonus for which she was treated with intravenous immunoglobulins without any effect. Nine months after her original presentation, an HIV-1 test was performed, and the result was positive (plasma HIV-1 RNA load, 9971 copies/mL; CD4 cell count, $50 \times 10^6$ cells/L; CD8 cell count, $620 \times 10^6$ cells/L). Other bacterial and viral serologic test results remained negative. We made the diagnosis of HIV-associated opsoclonus-myoclonus syndrome and started cART, consisting of emtricitabine-tenofovir and efavirenz [5]. In September 2009, the patient’s plasma HIV-1 RNA level was undetectable and the CD4 cell count had increased to $810 \times 10^6$ cells/L. However, the opsoclonus persisted. An 8-week course of prednisolone (starting dose, 50 mg once daily) also did not influence her symptoms. Last, the antiepileptic $\gamma$-aminobutyric acid agonist gabapentin was initiated in increasing doses up to 900 mg 3 times daily for 6 months with no success. At the end of 2010, our patient became pregnant and antiretroviral therapy was switched to lamivudine-zidovudine and lopinavir-ritonavir; her pregnancy proceeded in an otherwise uneventful way. Three years after her original presentation, the uncontrolled jerky movements of the eyes are still present in lasting and constant intensity despite treatment.

In conclusion, we present a patient with opsoclonus myoclonus syndrome as a rare and first manifestation of HIV-1 infection that did not respond to suggested treatment with immunoglobulins [1], cART [3, 4], steroids [3], or gabapentin [6]. The pathogenesis of this disease remains a mystery [3, 4]. The absence of clinical improvement after initiation of therapies that target the underlying disorder (cART) or the immune system (immunoglobulins, steroids, or gabapentin) has never been described. New insights are urgently needed for this debilitating condition.

Note

Potential conflicts of interest. All authors: No reported conflicts.

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