

LETTERS TO THE EDITOR

Extrapyramidal syndrome with generalised chorea as an atypical presentation of progressive multifocal leukoencephalopathy[☆]



Síndrome extrapiramidal con corea generalizada como forma de presentación atípica de leucoencefalopatía multifocal progresiva

Dear Editor:

Progressive multifocal leukoencephalopathy (PML) is a demyelinating disease of the central nervous system caused by reactivation of the JC virus. The condition usually presents with motor weakness, speech disorders, cognitive deficits, visual impairment, and ataxia,¹ and is associated with neuroimaging findings of subcortical white matter lesions which typically do not show contrast uptake or perilesional oedema.²

We present the case of a 43-year-old woman with stage C3 HIV infection who had discontinued antiretroviral treatment. The last CD4 lymphocyte count revealed 1 cell/mm³ with a viral load of 56 586 copies/mL. The patient visited our centre due to involuntary movements affecting all 4 limbs, with no associated systemic symptoms. Examination revealed generalised chorea predominantly affecting the left side of her body, hypophonia, hypometric saccades and smooth pursuit eye movements, hypomimia, bradykinesia during alternating movements of hands and feet (predominantly affecting the right side of her body), and ataxic gait.

From an anatomical viewpoint, these findings were associated with lesions in the internal capsule, basal ganglia, and thalamus in cranial computed tomography (CT) images (Fig. 1A). A complete blood count revealed no abnormalities, and serology tests for hepatotropic viruses, *Treponema pallidum*, and *Toxoplasma gondii* yielded negative results.

A biochemical analysis of the cerebrospinal fluid (CSF) yielded 8 cells/mm³, no erythrocytes, a glucose level of 56 mg/dL, and a protein level of 19.9 mg/dL. Gram and nigrosin staining was negative, ruling out infection due to *Leishmania*, CMV, Epstein–Barr virus, herpes simplex virus 1 and 2, enterovirus, and varicella-zoster virus. Further tests were performed while we awaited polymerase chain reaction (PCR) results for JC virus.

A nuclear magnetic resonance imaging (MRI) scan revealed patchy lesions to the corona radiata, basal ganglia, and thalamus, associated with oedema and contrast uptake (Fig. 1B).

We started early antiretroviral treatment (abacavir, lamivudine, and raltegravir) plus empirical treatment for cytomegalovirus encephalitis, tuberculosis, toxoplasmosis, and neurosyphilis. Despite this treatment, symptoms progressively worsened and the patient displayed marked bradyphrenia, somnolence, and subsequently right facio-brachiorucral hemiparesis (Fig. 1C). A multidisciplinary team opted for a brain biopsy; the anatomical pathology study of the tissue sample led to a diagnosis of PML (Fig. 1D).

JC virus is a type of polyomavirus which causes PML, a demyelinating disease of the CNS affecting immunocompromised individuals. The condition may present as the initial manifestation of HIV infection.³ A diagnosis of probable PML is made when an immunocompromised patient presents the typical clinical and radiological findings of the disease and tests positive in PCR for JC virus in the CSF. A diagnosis of possible PML is made when presence of the virus is not confirmed.¹

The literature reports some cases of encephalitis due to JC virus infection presenting with atypical neurological manifestations. Tallantyre et al.⁴ describe a case of PML presenting in the form of lower limb muscle weakness, bradykinesia, and bradyphasia. A previous article by Fontoura et al.⁵ reported the case of a patient with HIV and hepatitis C virus infection who developed PML in the form of myoclonic ataxia, progressing to tetraparesis. An MRI scan revealed bilateral lesions to the basal ganglia and primary motor cortex. Gallia et al.⁶ published the case of an HIV-positive patient with tonic-clonic seizures, dysphagia, disorientation to time and place, and fever. Movement disorders are infrequent in patients with PML, with a maximum incidence of 2.6%, and very few cases of movement disorders as the main or only manifestation of PML have been reported in the literature.^{7–9}

In MR images, lesions are predominantly found in the subcortical white matter and the periventricular area; these lesions are hypointense on T1-weighted and hyperintense

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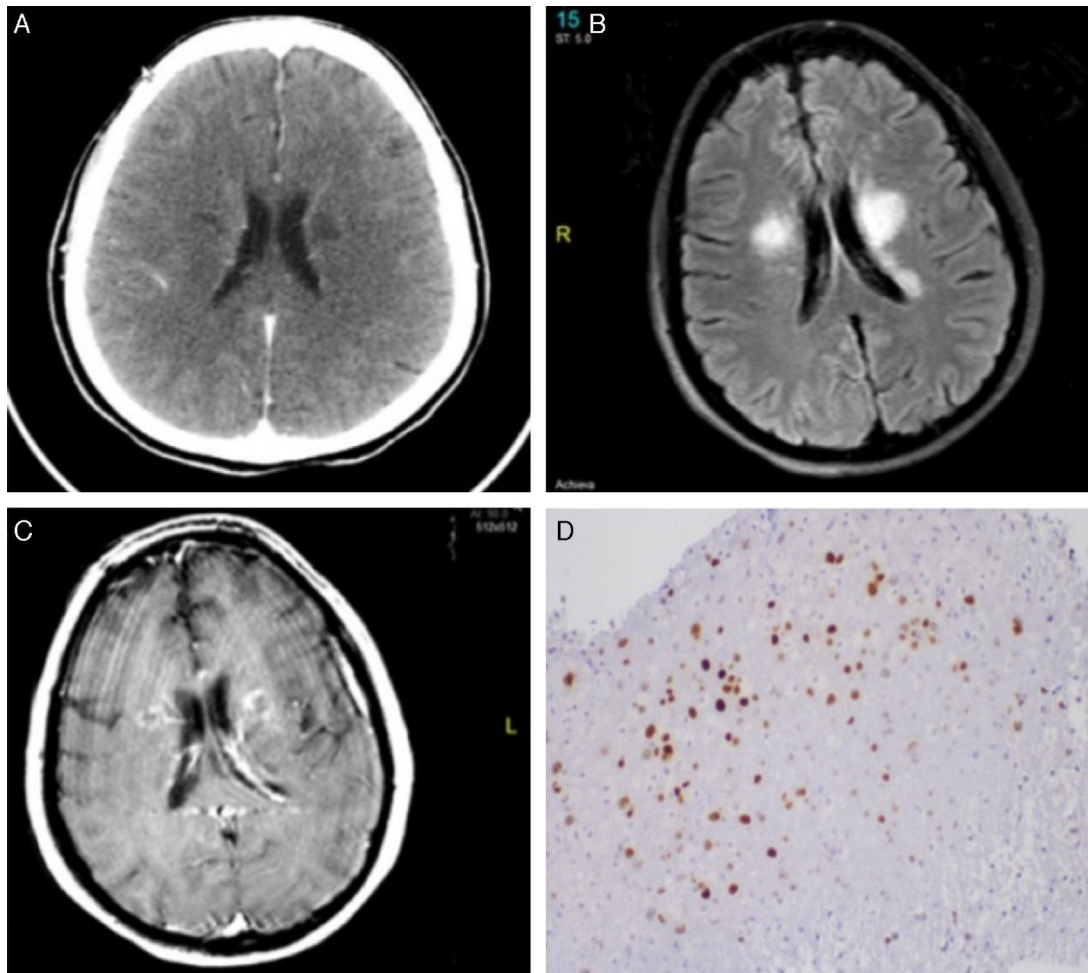


Figure 1 Neuroimaging and anatomical pathology findings. (A) Cranial CT scan showing bilateral focal hypodensities in the area around the ventricles, internal capsule, and thalamus. (B) FLAIR and (C) T1-weighted sequences. A brain MRI scan revealed lesions to the corona radiata, basal ganglia, and thalamus, predominantly in the left hemisphere, as well as slight oedema and contrast uptake. (D) Anatomical pathology study of brain tissue showing JC virus infection (SV40 antibody staining).

in T2-weighted and FLAIR sequences. These lesions rarely involve the thalamus and basal ganglia, have a mass effect, or show contrast uptake.²

Our case is unusual in that PML was associated with atypical clinical and neuroimaging findings, which made it difficult to diagnose the condition: a brain biopsy was necessary in view of the delay in viral diagnosis.

In conclusion, PML is a rare condition that may present with unusual manifestations and should therefore be considered in the differential diagnosis of immunocompromised patients with neurological clinical symptoms.

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Decreased spasticity in primary lateral sclerosis after botulinum toxin injection: A case report[☆]



Mejoría de la espasticidad en esclerosis lateral primaria tras la inyección de toxina botulínica. A propósito de un caso

Dear Editor:

Primary lateral sclerosis (PLS) is a variant of amyotrophic lateral sclerosis (ALS) in which the upper motor neuron and, secondarily, the corticospinal tract degenerate, with no clinical or neurophysiological involvement of the lower motor neuron.¹ Clinically, the disease is characterised by progressive spasticity and poor limb coordination. Prognosis of PLS is significantly better than that of classic ALS, with slower progression and higher survival rates.² Treating spasticity can have a meaningful impact on patients' quality of life, although few studies have addressed this topic. We present the case of a patient with PLS presenting with severe spasticity from onset, whose symptoms improved significantly following treatment with botulinum toxin (BTX).

The patient is a 37-year-old man who began experiencing gait alterations in 2010. Examination revealed hyperreflexia, spasticity, and poor coordination of the lower limbs, with no sensory involvement. Blood testing (including an autoimmune study and monoclonal component detection) and a lumbar puncture yielded normal results. The neurophysiological study ruled out lower motor neuron involvement. A brain magnetic resonance imaging (MRI) scan revealed hyperintensity in the corticospinal tracts on the FLAIR sequence, with an SWI sequence revealing a hypointense rim in the precentral gyrus (Fig. 1). Based on these findings, we diagnosed the patient with suspected PLS and initiated treatment with riluzole. As the disease progressed, the patient presented pyramidal signs in the upper limbs, and spastic dysarthria. An additional electromyography performed 4 years after symptom onset ruled out lower motor neuron involvement; we were therefore able to establish a definitive diagnosis of PLS.¹ Initial treatment with tetrazepam reduced spasticity, but caused

generalised weakness which prevented the patient from standing. In September 2012, the patient displayed spastic gait and was only able to walk short distances. He required 2 canes to walk indoors, with frequent falls, and a wheelchair to move outdoors. He also had poor coordination in the right hand and had a tendency to drop objects. The patient scored 32 on the ALS Functional Rating Scale (ALSFRS-R) (Table 1) and 3-4/5. Deep tendon reflexes were hyperactive in the lower limbs, with persistent clonus and extensor cutaneous plantar reflexes. Spasticity scored 2 on the Ashworth Scale and increased when he began walking, with the patient experiencing freezing. The 10-Metre Walk Test could not be performed, as he was unable to walk 10 metres.

We began treatment with Lioresal[®] in combination with stretches in the therapeutic gymnasium and hydrotherapy. Lioresal[®] was not tolerated, and gait function did not change with the physiotherapy. We therefore proposed attempting to control spasticity with local BTX injections. In February 2013, 50 units of BTX were injected into the left and right solei. The patient continued with the physiotherapy programme, progressively performing stretches, muscle toning, balance control exercises, and gait re-education. One month later, there was a significant improvement in the patient's spasticity, resulting also in a functional improve-

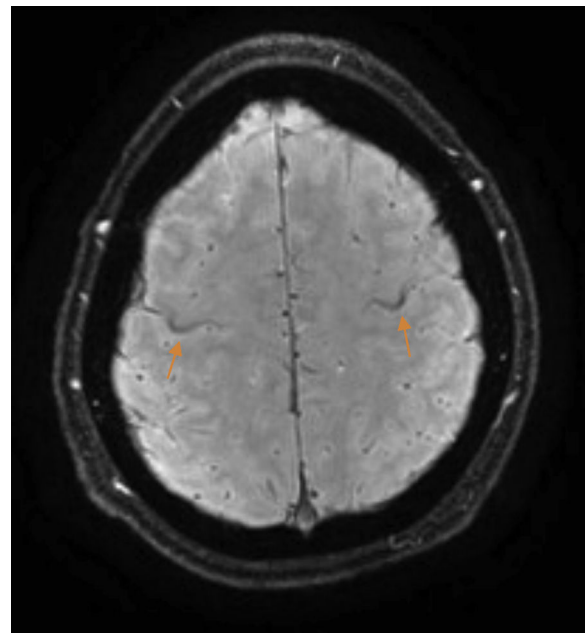


Figure 1 Brain MRI, SWI sequence. A hypointense rim is visible on the hand knob of the precentral gyrus.

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