

Multifocal Leukoencephalopathy

Published on 22.02.2001

DOI: 10.1594/EURORAD/CASE.874

ISSN: 1563-4086

Section: Neuroradiology

Imaging Technique: CT

Imaging Technique: MR

Case Type: Clinical Cases

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Osteaux

Patient: 32 years, female

Clinical History:

HIV-positive woman presented with progressive ataxia, dysarthria and hyperthermia. Antitoxoplasma therapy was started but had no effect. Subsequently she was admitted to the emergency department with a bipyramidal syndrome. CT and MRI were performed prior to brain biopsy.

Imaging Findings:

A 32-year-old HIV-positive woman presented with progressive ataxia, dysarthria and hyperthermia. Antitoxoplasma therapy was started but had no effect. Subsequently she was admitted to the emergency department with a bipyramidal syndrome. CT and MRI were performed prior to brain biopsy. Histological examination of the tissue sample, obtained by stereotactic biopsy from the right cerebellar hemisphere showed progressive multifocal leukoencephalopathy (PML).

Discussion:

Progressive Multifocal Leukoencephalopathy (PML) is an uncommon demyelinating disease that occurs in immunocompromised patients (lymphoma, leukemia, carcinomatosis, AIDS). The incidence of PML in adult AIDS-patients has been estimated to be up to 4%. PML is caused by central nervous system infection by group B Human Papovaviruses (Jacob-Creutzfeldt virus, simian virus 40), destroying principally oligodendroglia, thus resulting in extensive demyelination. The clinical manifestations are those of multiple, progressively enlarging lesions of the cerebral and cerebellar fiber systems: motor system abnormalities, visual loss, dementia and ultimately death, which occurs four to six months after onset. The typical distribution of PML is bilateral, asymmetric with a predilection for the white matter of the posterior centrum semiovale. The lesions initially tend to be focal and circumscribed; later they become confluent and large. CT scan shows focal, non-enhancing regions of low attenuation without mass effect. MRI, the most sensitive imaging modality for white matter disease, reveals patchy, poorly defined areas of hyperintensity in the white matter on T2-weighted images; hypointense, mostly non-enhancing on T1-weighted images. In AIDS-patients PML increasingly occurs in unusual locations: primary involvement of the cortex, the basal ganglia, the thalamus, the cerebellum and the brainstem is observed in up to 50% of AIDS-patients with proven PML. The lesions may also behave slightly different from the classical pattern, showing discrete contrast enhancement. Hemorrhagic transformation with spontaneous hyperintensity on T1-weighted images is a rare finding. Thus, imaging studies may help to identify and distinguish PML from lymphoma and toxoplasma abscesses, which both appear as focal mass lesions with ring enhancement.

Differential Diagnosis List: Multifocal Leukoencephalopathy

Final Diagnosis: Multifocal Leukoencephalopathy

References:

Mark AS, Atlas SW. Progressive Multifocal Leukoencephalopathy in Patients with AIDS: Appearance on MR Images. Radiology 1989; 173: 517-520. (PMID: [2798883](#))

Hawkins CP, McLaughlin JE, Kendall BE, McDonald WI . Pathological Findings Correlated with MRI in HIV Infection. Neuroradiology 1993; 35: 264-268. (PMID: [8388082](#))

Ng S, Tse VC, Rubinstein J et al. Progressive Multifocal Leukoencephalopathy: Unusual MR Findings. J Comput Assist Tomogr 1995; 19: 302-305. (PMID: [7890860](#))

Enting RH, Portegies P, Algra PR, Valk J, Lange JM. Progressieve Multifocale Leukoencefalopathie bij AIDS. Ned Tijdschr Geneesk 1992; 136. (PMID: [1552955](#))

Figure 1

a



Description: Unenhanced CT scan shows a diffuse hypodense region in the right cerebellar white matter, extending into the cerebellar pedunculus. No significant mass effect is seen. **Origin:**

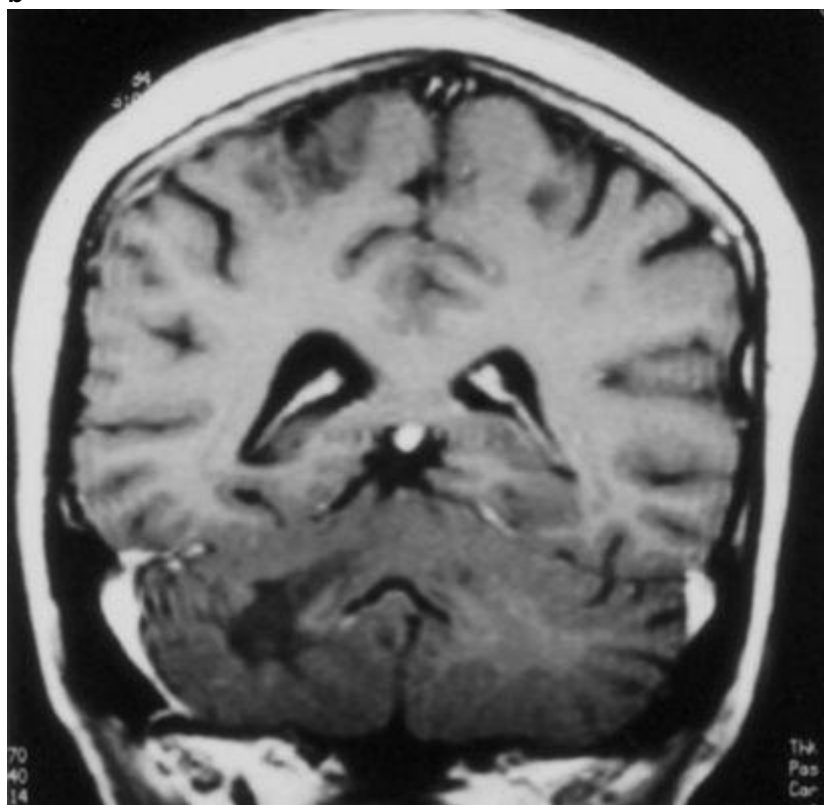
Figure 2

a



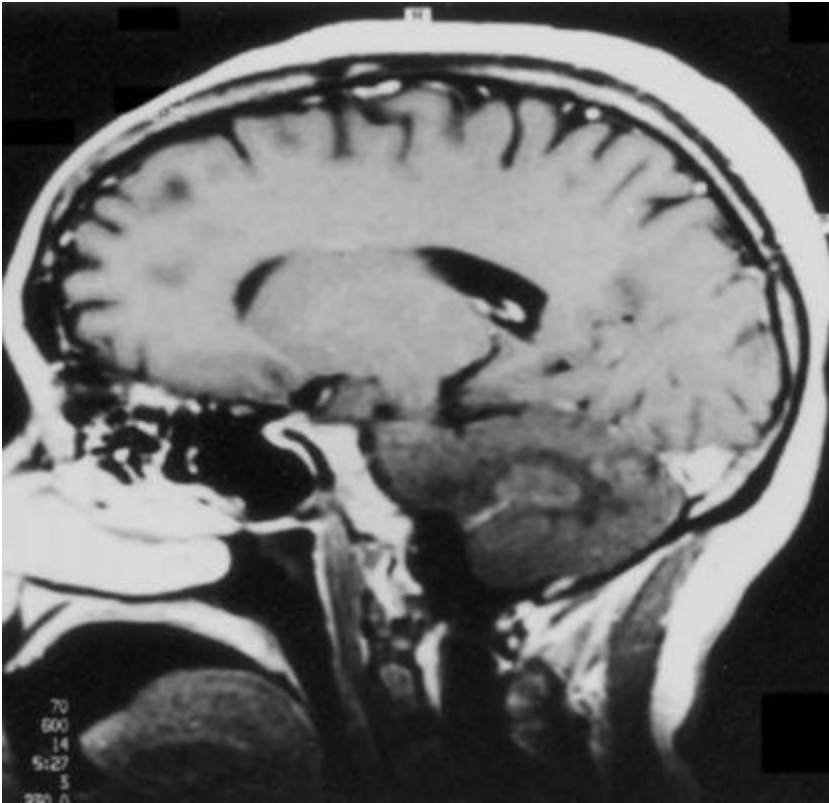
Description: Magnetic Resonance Imaging of the posterior fossa demonstrates, on axial SE T2-weighted image, a diffuse hyperintense region in the central aspect of the cerebellar hemispheres, more prominent on the right side, with extension into the pons. **Origin:**

b



Description: Coronal Gd-enhanced SE T1-weighted image reveals a circumferential hypointense white matter lesion in the right cerebellar hemisphere. **Origin:**

c



Description: Sagittal GD-enhanced SE T1-weighted image demonstrates a small central enhancing area, suggesting local breakdown of the blood-brain barrier. **Origin:**