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Progressive multifocal leukoencephalopathy in the AIDS era

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As a consequence of the AIDS pandemic, PML has ceased to be a rare disease. Approximately 5% of the HIV-infected population will ultimately develop PML. Explanations for the disproportionately high incidence of PML in HIV relative to other immunosuppressive conditions remain undetermined, but may be related to the nature and degree of immunodeficiency, interactions between HIV and JCV, or unique aspects of the brain microenvironment engendered by HIV infection. In approximately 20% of patients, PML is the heralding manifestation of HIV. The clinical manifestations of PML are diverse and differ in some respects from those observed with PML associated with other underlying conditions. Language disturbances, seizures, and headache appear to occur with increased frequency in the former group. Magnetic resonance imaging is very sensitive for detecting lesion location. while lesions characteristically occur in the hemispheric white matter, lesions occur in unusual locations, including the basal ganglia, brainstem, and cerebellum. Routine CSF studies are non-diagnostic. The establishment of the diagnosis is dependent on brain biopsy or a clinical picture consistent with disorder, the exclusion of other illnesses, and a positive CSF JCV PCR. To date, no treatment has been established as effective, however, 10% of patients experience prolonged (>12 months) survival following diagnosis. Highly active antiretroviral therapy when coupled with immune recovery appears to contribute to survival.