Progressive Multifocal Leukoencephalopathy

National Institute of Neurological Disorders and Stroke (NINDS)

Source

National Institute of Neurological Disorders and Stroke (NINDS). Progressive Multifocal Leukoencephalopathy Information Page.

Progressive multifocal leukoencephalopathy (PML) is a disease of the white matter of the brain, caused by a virus infection that targets cells that make myelin--the material that insulates nerve cells (neurons). Polyomavirus JC (often called JC virus) is carried by a majority of people and is harmless except among those with lowered immune defenses. The disease is rare and occurs in patients undergoing chronic corticosteroid or immunosuppressive therapy for organ transplant, or individuals with cancer (such as Hodgkin's disease or lymphoma). Individuals with autoimmune conditions such as multiple sclerosis, rheumatoid arthritis, and systemic lupus erythematosus -- some of whom are treated with biological therapies that allow JC virus reactivation -- are at risk for PML as well. PML is most common among individuals with HIV-1 infection / acquired immune deficiency syndrome (AIDS). Studies estimate that prior to effective antiretroviral therapy, as many as 5 percent of persons infected with HIV-1 eventually develop PML that is an AIDS-defining illness. However, current HIV therapy using antiretroviral drugs (ART), which effectively restores immune system function, allows as many as half of all HIV-PML patients to survive, although they may sometimes have an inflammatory reaction in the regions of the brain affected by PML. The symptoms of PML are diverse, since they are related to the location and amount of damage in the brain, and may evolve over the course of several weeks to months. The most prominent symptoms are clumsiness; progressive weakness; and visual, speech, and sometimes personality changes. The progression of deficits leads to life-threatening disability and (frequently) death. A diagnosis of PML can be made following brain biopsy or by combining observations of a progressive course of the disease, consistent white matter lesions visible on a magnetic resonance imaging (MRI) scan, and the detection of the JC virus in spinal fluid.