PROGRESSIVE MULTIFOCAL LEUCOENCEPHALOPATHY (PML)

WHAT IS PML?

Progressive multifocal leucoencephalopathy is a serious viral infection of the brain.

"Encephalo" means brain. "Pathy" means disease. Encephalopathy is a disease of the brain. "Leuco-" (or "leuko") means pale or white. Leucoencephalopathy is a disease of the white matter of the brain.

"Progressive" means that this disease gets worse in a short time. "Multifocal" means that it shows up in several places at the same time.

Researchers estimate that about 6% of people with AIDS develop PML. Most cases of PML show up in people with CD4 cell counts below 100. The exact rate is hard to know because PML is difficult to diagnose.

Most cases of PML used to be fatal. People diagnosed with PML lived an average of 6 months, and most died within 2 years. However, if people with PML start taking strong antiretroviral medications (ARVs) to control their HIV, they can survive much longer. Now more than half of people with HIV and PML survive for at least two years.

The "JC" virus causes PML. Between 80–85% of all adults are exposed to this virus worldwide. In people with weakened immune systems, JC virus can become active.

HOW CAN PML BE DETECTED?

The first symptoms of PML are weakness or coordination problems in an arm or leg. There may be difficulty thinking or speaking. Vision and memory problems, seizures, and headaches can occur.

These symptoms can also occur with other opportunistic infections, including toxoplasmosis, lymphoma, inner ear infections, or cryptococcal meningitis. It is important to rule out these other diseases.

PML can be diagnosed using a scan of the brain by magnetic resonance imaging (MRI). Another way to test for PML is by checking spinal fluid. The sample is taken by inserting a needle into the spinal canal. This procedure is called a spinal tap.

HOW IS PML TREATED?

A major problem with treating any brain infection is the "blood-brain barrier." The blood vessels around the brain are different from the rest of the body. They are "tightly woven" to protect the brain from toxic substances. Chemicals that dissolve in fat can get through. Those that dissolve in water can't. Unfortunately, this includes most antibiotics and many other medications.

There is currently no proven treatment for PML. Research studies have had conflicting results. Some possible treatments have not been carefully studied. However, PML has slowed down or stopped in some patients taking strong ARVs to fight HIV. Strengthening the immune system is currently the best way to treat PML. This approach might trigger immune reconstitution inflammatory syndrome (IRIS, see fact sheet 483.)

Ara-C (Cytosine arabinoside or cytarabine) has been tried against PML. It was given intravenously, or pumped directly into the brain. It seemed to work in one small study, but not in later ones. Ara-C is very toxic, and damages bone marrow.

High-dose AZT has been tried against PML, because it crosses the blood-brain barrier. Other substances that have been tried with different degrees of success include acyclovir, heparin, peptide-T, beta interferon, dexamethasone, mefloquine, n-acetylcysteine, topotecan and cidofovir.

Because PML can progress rapidly, it is important to begin treatments quickly.

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THE BOTTOM LINE

PML is a viral infection of the brain caused by the JC virus. It is fatal in about 50% of cases. It can be confused with other medical conditions.

There is no approved treatment for PML, although several treatments may be helpful. Strengthening the immune system using antiretroviral therapy is currently the best approach. Any treatment must be started as early as possible. Combination antiretroviral therapy (ART) may slow the progress of PML.

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