DEGENERATIVE-DEMYELINATING DISEASES

MULTIFOCAL LEUKOENCEPHALOPATHY

A 15-year-old boy with Wiskott-Aldrich syndrome complicated by progressive multifocal leukoencephalopathy (PML) is reported from the University of Miami School of Medicine, FL, and the National Institute of Neurological Disorders and Stroke, Bethesda, MD. At age 4 months he developed thrombocytopenic purpura and with subsequent appearance of eczema and frequent pneumonia and otitis media, the diagnosis of Wiskott-Aldrich syndrome was made at age 7 years. At age 12 years he suffered a retroperitoneal hematoma, liver abscesses, followed by frequent infections, including viral pneumonia, herpes labialis, and Candida septicemia. Neuropsychiatric problems occurred at 14 years of age and progressed, with slurred speech, apathy, and right sided weakness. MRI revealed multiple high-intensity signal lesions in brain stem and cerebrum, including thalamus and basal ganglia. Brain biopsy confirmed PML, characterized by abnormal oligodendroglial nuclei, atypical astocytes, and foamy macrophages. Immunostaining with antibody against papovirus capsid antigen was positive. Bone marrow biopsy and peripheral blood lymphocytes were positive for JC virus DNA. At autopsy the brain showed multiple areas of demyelination. intranuclear inclusions, and a large left frontal hemorrhage. Survival from the time of onset of PML was 10 months. (Katz DA et al. Progressive multifocal leukoencephalopathy complicating Wiskott-Aldrich syndrome. Arch Neurol April 1994;51:422-426). (Reprints: Dr Berger, Dept of Neurology, University of Miami School of Medicine, 1501 NW Ninth Ave, Miami, FL 33136).

COMMENT. Wiscott-Aldrich syndrome is an inherited X-linked recessive immunodeficiency disease characterized by severe eczema, thrombocytopenia, and frequent infections. Patients usually die of infection, hemorrhage, or malignant neoplasm. This case report may be the first described with PML complicating the Wiscott-Aldrich syndrome.

PML is an opportunistic infection of the CNS with JC virus characterized by a rapidly progressive degenerative demyelinating disease. Immunodeficient states associated with PML include AIDS, lymphoma, leukemia, tuberculosis, systemic lupus, organ transplant and rarely, Wiskott-Aldrich syndrome. PML is a disease of adults and is very uncommon in children. An MRI of a 40 year-old man with AIDS and PML is presented by Weiss PJ and DeMarco JK under Images in Clinical Medicine, N Engl J Med April 1994;330:1197. PML changes are best seen on the T2-weighted image as high-intensity abnormalities in the white matter. On T1-weighted image, the lesions are low-intensity and not enhanced by gadolinium, which differentiates them from primary lymphoma of the CNS.

PERINATAL DISEASES

MICROCEPHALY AFTER HI ENCEPHALOPATHY

The development of microcephaly after hypoxic-ischemic cerebral injury in the full-term newborn was studied at the University of British Columbia, Vancouver, Canada. Serial head circumference measurements obtained at 4, 8, and 18 months of age in 54 newborns suffering from acute,