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,

hypoxic-ischemic encephalopathy showed that a decrease in head circumference ratios of >3.1% between birth and 4 months of age was highly predictive of the development of microcephaly before 18 months. Head circumference ratios were actual head circumference/mean head circumference x 100%. They were correlated with severity of neonatal HIE and outcome at 18 months. (Cordes I et al. Early prediction of the development of microcephaly after hypoxic-ischemic encephalopathy in the full-term newborn. Pediatrics May 1994;93:703-707). (Reprints: Dr Alan Hill, Division of Neurology, British Columbia's Children's Hospital, 4480 Oak St, Vancouver, BC, Canada V6H 3V4).

COMMENT. A decreased rate of head growth in the first 4 months after acute, intrapartum HIE correlates with the later development of microcephaly and neurological sequelae in the full-term neonate as well as very low birth weight infants.

VASCULAR MALFORMATIONS

TREATMENT OF CVA AND VEIN OF GALEN ECTASIA

Optimal methods of evaluation and treatment of newborns with cerebral arteriovenous malformation associated with ectasia of the vein of Galen are outlined along with a case report of an inoperable malformation from the Dept of Pediatrics, Malarsjukhuset, Eskilstuna, and Dept of Neuroradiology, Karolinska sjukhuset, Stockholm, Sweden, and Centre Hospitalier Universitaire de Bicetre, France. The infant developed a general seizure at 10 min of age. She had a loud bruit over the skull and neck and congestive heart failure. Neurosonography showed a mass in the midline of the brain. CT confirmed a A-V malformation and ectasia of the vein of Galen, complicated by in utero encephalomalacia and more recent ischemic injury. Endovascular occlusive treatment was not appropriate and the child died at 15 days of age. Transarterial embolization of feeding arteries using bucrylate (isobutyl cyanoacrylate) is now considered the treatment of choice, in the absence of contraindications such as brain damage, prenatal cardiomegaly, or evidence of multi-organ failure. A free interval between birth and development of cardiac failure are factors of favorable prognostic value. (Swanstrom S et al. Conditions for treatment of cerebral arteriovenous malformation associated with ectasia of the vein of Galen in the newborn. Acta Pediatr March 1994;83:255-7). (Respond: Dr S Swanstrom, Dept of Pediatrics, Malarsjukhuset, S-631 88 Eskilstuna, Sweden).

COMMENT. Rapid clinical deterioration due to cardiac failure is the common presenting feature of this syndrome. Transarterial endovascular occlusive treatment is available only in specialized centers. The rapid evaluation of patients suited for intervention is essential for successful outcome. The authors caution against the use of contrast material for CT and advise against any form of angiography unless therapy is contemplated at the same time.