ENCEPHALOPATHIES

ACUTE NECROTIZING ENCEPHALOPATHY

The clinicopathological features of an acute necrotizing encephalopathy are described in a review of 13 consecutive children treated and 28 previously reported cases seen at various institutions in Japan. The onset was preceded by an upper respiratory infection, frequently treated with antipyretics and antibiotics. Aspirin was taken by only 2 patients. Symptoms of brain dysfunction, including impaired consciousness, convulsions, and vomiting, developed rapidly within 1 to 3 days. One third had hematemesis, one half had diarrhea, and all had liver enlargement without clinical jaundice. Hyperpyrexia, hyperventilation, and decorticate or decerebrate posturing occurred at the comatose stage. Twenty eight per cent died. In survivors, recovery of consciousness and neural function beginning after 6 to 10 days was slow, and serious sequelae such as spasticity, mental retardation, and seizures were common. Laboratory findings showed liver dysfunction, uremia, and hypoproteinemia. Liver histology was nonspecific and distinguished from Reye's syndrome. CSF protein was increased. CT and MRI revealed symmetric, multifocal areas of necrosis in the thalamus, white matter, brainstem, and cerebellum. The etiology of this previously unrecognized type of acute encephalopathy was not determined. (Mizuguchi M et al. Acute necrotizing encephalopathy of childhood: a new syndrome presenting with multifocal, symmetric brain lesions. J Neurol Neurosurg Psychiatry May 1995;58:555-561). (Respond: Dr M Mizuguchi, Department of Mental Retardation and Birth Defect Research, National Institute of Neuroscience, NCNP, 4-1-1 Ogawahigashi-cho. Kodaira, 187, Japan).

> COMMENT. In addition to Reye's syndrome, the differential diagnosis included Wernicke's and Leigh's encephalopathies, carbon monoxide poisoning, acute disseminated encephalomyelitis, and acute hemorrhagic leukoencephalitis. The patients reported here appear to show some characteristics that differentiate this form of "acute toxic encephalopathy" from those already recognized. It is of interest that several patients (16%) were retarded, and 16% had congenital anomalies, including ventricular septal defect, radial agenesis, and polydactyly. Evidence of recent viral infections (influenza A and B, coxackie A9, Rotavirus) was detected in 10 patients, and one patient had 3 paternal aunts who had died of Ekiri, a fulminant form of acute encephalopathy secondary to *Shigella dysenteriae* infection, prevalent in Japan up to the 1950s.

AMPHOTERICIN B-INDUCED ENCEPHALOPATHY

Three children with refractory leukemia treated by bone marrow transplantation at the Children's National Medical Center, Washington, DC, developed encephalopathy, leukoencephalopathy, and parkinsonism after receiving high-dose amphotericin B for pulmonary aspergillosis. All 3 had previously been treated with high-dose chemotherapy and total body irradiation. MRIs showed basal ganglia, cerebellar, and cerebral atrophy, and frontal and temporal lobe white matter changes. One died and two recovered after withdrawal of the amphotericin, 1 having intellectual impairment. (Mott SH, Packer RJ et al. Encephalopathy with Parkinsonian features in children following bone marrow transplantations and high-dose amphotericin B. Ann

Neurol June 1995;37:810-814). (Respond: Dr Mott, Department of Neurology, Children's National Medical Center, 111 Michigan Ave, NW, Washington, DC 20010).

COMMENT. Neurologic complications of bone marrow transplantation in children with leukemia are a common occurrence. These have included seizures, infections, and encephalopathies, but Parkinsonian symptoms associated with amphotericin B appear to be unique.

HEMORRHAGIC SHOCK AND INFANTILE ENCEPHALOPATHY

The clinical characteristics, treatment and possible causes of hemorrhagic shock and encephalopathy in infants are described and a 5-month-old patient is reported from the Section of Neurology, The Children's Mercy Hospital, Kansas City, MO. The infant presented with fever and irritability. She developed respiratory distress, requiring endotracheal intubation, followed by cardiorespiratory arrest. Excessive bleeding from puncture sites was associated with a disseminated intravascular coagulopathy. Admission diagnosis was septic shock. Other complications of this encephalopathy are bloody diarrhea and hepatorenal failure. Treatment requires fluids and electrolytes, fresh frozen plasma, and vitamin K. Hyperthermia appeared important in causation. (Chaves-Carballo E. Hemorrhagic shock and encephalopathy: a new neurologic syndrome in infants. Acta Neuropediatr 1995;1:178-184). (Reprints: Dr E Chaves-Carballo, Section of Neurology, Children's Mercy Hospital, 2401 Gillham Road, Kansas City, MO 64108).

COMMENT. The syndrome was first described in Great Britain in 1983 as cited by the author (Levin M et al. Lancet 1983;2:64-67). The differential diagnosis includes septic shock, toxic-shock syndrome, Reye syndrome, and hemolytic-uremic syndrome. Early aggressive therapy was recommended.

BRAIN NEOPLASMS

BRAIN TUMORS AND INFANTILE SPASMS

Two patients, aged 6 and 7 months, with brain tumors who presented with infantile spasms and hypsarrhythmia are reported from Sapporo and Asahikawa Medical Universities, Japan. One had a hypothalamic hamartoma and the other a oligoastrocytoma with calcification in the right temporal lobe. ACTH controlled spasms and EEG seizure discharges. (Asanuma H et al. Brain tumors associated with infantile spasms. Pediatr Neurol May 1995;12:361-364). (Respond: Dr Asanuma, Department of Pediatrics, Sapporo Medical University, School of Medicine, S1 W16, Cho-ku, Sapporo, 060, Japan).

COMMENT. The authors cite 9 additional reports in the literature of brain tumors associated with infantile spasms. Focal brain lesions may underly the origin of infantile spasms.
Brain tumor was not listed as a cause of infantile spasms in an epidemiological study involving 57 patients treated in Sweden for the period 1987-1991. (Sidenvall R, Eeg-Olofsson O. Epilepsia July 1995;36:572-574).

PINEAL AND EPENDYMAL CYSTS AND INFANTILE SPASMS

A 3-month-old infant with infantile spasms and hypsarrhythmia