

Neuropathology. Baltimore, Williams & Wilkins, 1963). Yakovlev and Wadsworth (1946) described schizencephalic porencephalies of developmental origin and encephaloclastic porencephalies caused by destruction of cerebral tissue. A wide variety of etiological factors and clinical findings are reported (Mednick JP, Jerva MJ, Millichap JG. *Trans Am Neur Assoc* 1966;301-303). Hemorrhagic infarction following a germinal matrix hemorrhage in a preterm infant is a frequent destructive cause. Other causes include trauma, vasculopathy secondary to maternal cocaine abuse or congenital infections such as cytomegalic inclusion disease, bleeding disorders, including factor V Leiden and collagen IV A1 mutation with microangiopathy, as in familial autosomal dominant porencephaly, described in the above report.

VASCULAR ABNORMALITIES IN ALTERNATING HEMIPLEGIA

Skin and/or muscle biopsies in 4 patients, ages 18 months, 8, 9, and 16 years, with alternating hemiplegia of childhood (AHC) were examined by electron microscopy and compared with healthy controls in a study at University Hospital, Lille, France. Vascular abnormalities present in both skin and muscle small vessels included endothelial vacuoles, intracytoplasmic vacuoles in vascular smooth muscle cells (VSMCs) in the tunica media, apoptotic nuclei, and isolation of VSMCs from neighboring cells. A primary or secondary vascular pathophysiology is suggested for AHC. (Auvin S, Joriot-Chekaf S, Cuvelier JC, et al. Small vessel abnormalities in alternating hemiplegia of childhood. *Neurology* February (2 of 2) 2006;66:499-504). (Reprints: Dr S Auvin, Service de Neurologie pediatrique, Hopital Roger Salengro, Boulevard du Pr J Leclercq, 59037 Lille Cedex, France).

COMMENT. AHC is characterized by 3 phases; 1) abnormal eye movements and dystonic episodes; 2) episodic hemiplegia and developmental delay; and 3) persistent neurologic deficits. Onset is before 18 months of age, and symptoms resolve during sleep. Formerly ascribed to a migraine equivalent, an epilepsy, or movement disorder, the present evidence suggests a neurovascular mechanism, similar to the cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL).

INTRACRANIAL NEOPLASMS

SUPRASELLAR GERMINOMA AND GROWTH RETARDATION

Three patients, ages 9, 12, and 13 years (1 boy and 2 girls), diagnosed with intracranial germinoma are reported from University Children's Hospital Homburg, Germany. Presenting symptoms and signs prompting an MRI were nausea, vomiting, strabismus, and intermittent headache for 1 year in case 1, migraine in case 2, and a 3-year history of growth retardation in case 3. Patients 1 and 2 also had a history of growth retardation for 2 years. MRI showed tumors in the pituitary region in cases 1 and 2, and a thickened pituitary stalk in case 3. Clinical and laboratory findings showed panhypopituitarism, subclinical diabetes insipidus and hypothyroidism, and growth hormone deficiency in cases 1, 2, and 3, respectively. CSF levels of Beta-human chorionic gonadotrophins (B-hCG) were not elevated, and diagnosis of germinoma was confirmed by

biopsy. Treatment by chemotherapy and/or irradiation was successful. At follow-up of 12 to 18 months, patients were in remission. Patients 1 and 2 were dependent on thyroxine, hydrocortisone, and growth hormone, while patient 3 needed no further hormone substitution. (Gottschling S, Graf N, Meyer S, et al. Intracranial germinoma: a rare but important differential diagnosis in children with growth retardation. *Acta Paediatrica* March 2006;95:302-305). (Respond: Dr Sven Gottschling, University Children's Hospital, Kirrbergerstr, 66421 Homburg/Saar, Germany).

COMMENT. The incidence of pediatric intracranial germ cell tumors varies worldwide, the average being 3.6%, and higher in Asia than the Western hemisphere. Germinomas arise in the pineal or suprasellar regions and if diagnosed early, have a good prognosis. The present small series demonstrates the value of secondary growth retardation as an early sign of suprasellar germinoma. Diagnosis is made by MRI, and a CSF B-hCG level above 50 U/l (under 7 U/l is normal). Biopsy may be necessary when the hormone levels are not elevated.

A larger series of 26 children with germ cell tumors (17 germinomas and 9 teratomas), not cited above, was reported from the University Hospital Hamburg (Haupt C et al. *Eur J Pediatr* 1996;155:230-236; *Ped Neur Briefs* May 1996). Tumor location was the pineal region in 69% and suprasellar/hypothalamic in 31%. Presenting symptoms were increased intracranial pressure, Parinaud's syndrome, and endocrine deficits. Long-term survival rate was 88% for germinomas and 43% for malignant teratomas.

ENCEPHALOPATHIES

ACUTE INFANTILE FRONTAL LOBE ENCEPHALOPATHY

The clinical and radiologic features of nine infants with acute encephalopathy involving the frontal lobes are reported from Dokkyo University School of Medicine and Jichi Medical School, Tochigi; and University of Tokyo, Japan. The prenatal and perinatal histories were uneventful, and milestones of development were normal. Symptoms presented at 7 months to 3 years of age with 1) convulsive status epilepticus with hyperpyrexia and prolonged impairment of consciousness, 2) mental and motor regression after recovery of consciousness, 3) CSF leukocyte count of 8 cells/mm³, 4) MRI showing atrophy of both frontal lobes. The infection was influenza type A in 3 patients, exanthem subitum in 2, measles in 1, and upper respiratory viral illness nonspecified in 3. Regression of behavior on recovery of consciousness involved verbal function and language, stereotypic movements in 4, and cataplexy in 1. On 12-month follow-up, motor function recovered while speech remained retarded. Laboratory tests revealed increased lactate dehydrogenase and creatine kinase, normal CSF protein in all except one patient, and elevated interleukin-6 in 2 of 3 tested. EEG at time of onset showed high-amplitude delta predominantly in both frontal areas, and at 1 year, focal spikes or spike-and-wave discharge in 3 patients. One patient developed localization-related epilepsy. On serial MRI performed between the 20th and 42nd day from onset, atrophic changes were revealed in both frontal lobes of all patients. PET studies in the same period showed decreased perfusion in both frontal lobes, with normalized perfusion at the 7th to 38th month after onset. (Yamanouchi H, Kawaguchi N, Mori M, et al. Acute infantile encephalopathy predominantly affecting the frontal lobes. *Pediatr Neurol*