SEIZURE DISORDERS

GELASTIC EPILEPSY AND HYPOTHALAMIC HAMARTOMA

Three patients with hypothalamic hamartoma and gelastic seizures were studied using ictal single-photon emission computed tomography and EEG recordings with depth electrodes at the University of Alabama at Birmingham Epilepsy Center. Precocious puberty developed by age 7, and the patients were large for their age, above the 95th percentile. Cognitive and behavioral abnormalities were revealed on neurologic examination, with impulsivity, irritability, and aggressive tendencies. Laughing seizures began in the newborn period in 2 patients and before 6 years in a third. The frequency of attacks ranged from 1 to 5 minor episodes daily to 10 complex partial seizures each month. Interictal EEGs showed rare sharp waves from various areas. Ictal EEGs revealed diffuse theta. MRIs showed hypothalamic hamartomas, with displacement of the floor of the third ventricle. Ictal SPECT showed focal hyperperfusion of the hamartoma and thalamus. Depth electrode EEG within the hamartoma recorded focal spikes. Stereotactic radiofrequency surgical treatment of the hamartoma of one patient resulted in seizure remission. (Kuzniecky R, Guthrie B, Mountz J et al. Ann Neurol July 1997;42:60-67). (Respond: Dr Kuzniecky, Department of Neurology, University Station, Birmingham, AL 35294).

COMMENT. Gelastic (*'gelos'* Gr for mirth) seizures in children with precocious puberty and hypothalamic hamartoma originate from the hamartoma and involve adjacent diencephalic areas. Seizures occur more commonly when the tumor displaces structures adjacent to the hypothalamus. See Progress in Pediatric Neurology II, PNB Publ, 1994;p41, for further articles on gelastic epilepsy. The anterior cingulate region may also be involved in some cases.

NONCONVULSIVE STATUS AND RING CHROMOSOME 20

Six cases of epilepsy and ring chromosome 20 are reported and 20 additional cases in the literature are reviewed from the National Epilepsy Center, Shizuoka Higashi Hospital, Japan. Seizures consisted of a prolonged confusional state, and the ictal EEG showed bilateral high-voltage slow waves with occasional spikes. Neurological exam and MRI were usually normal. Mental retardation or bordeline IQ was present in 5 of 6 cases. Seizures were refractory to AEDs. (Inoue Y, Fujiwara T, Matsuda K et al. Ring chromosome 20 and nonconvulsive status epilepticus. A new epileptic syndrome. Brain June 1997;120:939-953). (Respond: Dr Yushi Inoue, National Epilepsy Center, Shizuoka Higashi Hospital, Urushiyama 886, Shizuoka 420, Japan).

COMMNENT. Chromosome analysis may be indicated in children with drug refractory epilepsy and learning disabilities, despite the absence of dysmorphic features. Ring chromosome 20 in a child with gelastic and partial complex seizures was included in a section on mental retardation syndromes in Progress in Pediatric Neurology III, 1997;p384.

GENETICS OF EPILEPSIES WITH FOCAL SHARP WAVES

The clinical manifestations and genetic aspects in 147 children with benign focal sharp waves were evaluated at the Epilepsy Center, Raisdorf, Germany. Seizures in 134 patients included febrile convulsions (26%), rolandic