

vomiting, hypotonia, dystonia, and aphasia. Deep tendon reflexes were exaggerated and plantar responses extensor. The abnormal neurologic signs and basal ganglia lesions gradually improved with corticosteroid treatment and almost disappeared after one year. The right optic fundus showed chorioretinitis. A stereotactic brain biopsy performed to exclude a neoplasm revealed proliferation of astrocytes of undetermined origin, either reactive or low-grade astrocytoma. The dystonia subsided after one year but peculiar mouthing behavior persisted. A CNS primary lymphoma could not be excluded. (Hirabayashi S et al. Basal ganglia mass lesions in juvenile rheumatoid arthritis. Pediatr Neurol March/April 1991; 7:141-3).

COMMENT. A chronic inflammatory process involving cerebral vessels was suspected in this patient but angiography failed to demonstrate a cerebral vasculitis.

Other rheumatic diseases with CNS complications include lupus erythematosus, polyarteritis nodosa and rheumatic fever. Seizures are a common presenting sign of lupus erythematosus.

SEIZURE DISORDERS

BENIGN FAMILIAL NEONATAL CONVULSIONS

Linkage studies with the chromosome 20 markers D20S19 and D20S20 were performed in two families with benign familial neonatal convulsions at the Department of Pediatrics, The University of Texas Health Science Center, San Antonio, TX. In the first family with 14 affected, none had seizures after two months of age. In the second family with 13 affected, seizures did not remit until 6 to 24 months; febrile convulsions occurred in two, and one had refractory epilepsy until late adolescence. In family one, the odds were greater than 20,000:1 against linkage at 10% recombination; whereas the data from family two favored linkage with a maximum odds ratio of 45:1 at 6% recombination. It was concluded that this autosomal dominant primary epilepsy of infancy is clinically and genetically heterogeneous. (Ryan SG et al. Benign familial neonatal convulsions: Evidence for clinical and genetic heterogeneity. Ann Neurol May 1991; 29:469-473).

COMMENT. These data based on large family pedigrees suggest two distinct genetic loci for benign familial neonatal convulsions. The subtype linked to chromosome 20q may be associated with delayed remission and a higher risk for the development of epilepsy. The authors suggest that absence and benign rolandic epilepsy might also show genetic heterogeneity.

EPIDEMIOLOGY OF ABSENCE EPILEPSY

A population based electroencephalographic study of absence epilepsy in 97 children is reported from the Departments of Neurophysiology and Pediatrics, Goteborg University, Sweden. All patients had regular bilaterally synchronous and symmetrical 2-4 Hz spike-and-slow wave discharges and absences with or without generalized tonic-clonic