NATURAL HISTORY OF RASMUSSEN'S SYNDROME

Seizure frequency, degree of hemiparesis and cerebral hemiatrophy are analysed in 13 patients with histopathologically proven Rasmussen's encephalitis (RE) examined at the University of Bonn, Germany. The age at first seizure was in childhood in 10 cases (range, 1.6 - 15.7 years) and in adulthood in 3 (22.1 - 40.9 years). An initial prodromal stage (Montreal Neurological Institute Stage 1), characterized by low seizure frequency, had a median duration of 7.1 months (range 0 months to 8.1 years), shorter in children than adults. An acute (MNI stage 2) phase was recognized by a rapid increase in seizure frequency (to >10 simple partial motor seizures per day) accompanied by development or deterioration of hemiparesis and loss of hemispheric volume. Initial MRI scans showed inflammatory lesions, with multifocal onset between Rolandic and temporomedial areas, which spread across the ipsilateral hemisphere. The median duration of stage 2 was 8 months (range 4-8 months). Residual phase (MNI stage 3) was marked by a relatively stable permanent hemiparesis and diminished seizure frequency.

Patients were divided into two groups, according to age, duration of prodromal stage and outcome. Type 1 patients (n=7), all children, had a more rapid and severe disease than type 2 (adolescents and adults). Type 1 cases had a median age of 5.3 years (range 1.6-6.1 years) at onset of seizures, the prodromal stage was absent or short, and the acute phase lasted 8 months (range 4-8 months). Type 2 patients (n=6) had a median age of 18.9 years (range 6.4-40.9 years) at onset, a long prodromal stage (median duration 3.2 years, range 1.3-8.1 years), focal epilepsy with complex partial or secondarily generalized tonic-clonic seizures, rare occurrence of hemiparesis, of lesser frequency and degree than type 1 patients, only 2 requiring hemispherectomy, and an acute phase lasting 7.5 months (range 7-8 months). Histopathologically, the 2 types were similar, with chronic inflammatory changes in all patients. Most of the brain damage had occurred in the first 8-12 months. (Bien CG, Widman G, Urbach H et al. The natural history of Rasmussen's encephalitis. Brain July 2002;125:1751-1759). (Respond: Dr CG Bien, Department of Epileptology, University of Bonn, Sigmund-Freud-Strasse 25, D-53105 Bonn, Germany).

COMMENT. The authors recommend that future therapeutic interventions should focus on the 8 month period of the acute disease phase, when the most extensive brain damage occurs. In children, this acute phase with increased frequency of seizures and development of hemiparesis occurs early and soon after the onset of seizures, whereas in adolescents and adults, the acute phase is delayed and follows a long prodromal stage. In the later residual stage, the
majority of brain atrophy will have taken place, and the measurement of improvement following immunosuppressive treatment will not be possible. Estimation of hemispheric volume loss may be used as a parameter of the destructive disease process during the acute phase. An autoimmune response mediated by cytotoxic T lymphocytes is proposed as a key mechanism in the etiology of Rasmussen's encephalitis (Bauer J, Bien CG, Lassmann H. see Ped Neur Briefs April 2002;16:27-28).

ANTI-BASAL GANGLIA ANTIBODIES IN SYDENHAM'S CHOREA

The sensitivity and specificity of methods to detect anti-basal ganglia antibodies (ABGA) in Sydenham's chorea were determined in a study at the Institute of Neurology, Queen Square; Great Ormond Street Hospital for Children; Royal Free and University College Hospital Medical School, London, UK; and Federal University of Minas Gerais, Brazil. Samples from 20 patients with acute SC, 16 with persistent SC, control samples from 16 with rheumatic fever (RF), and 11 healthy pediatric volunteers were tested with ELISA and Western immunoblotting (WB) methods to detect ABGA and compared these assays to immunofluorescent (IF) methods. In acute SC, a sensitivity of 95% and specificity of 93% were obtained with ABGA ELISA, while WB and IF had a sensitivity of 100% and specificity of 93%. In persistent SC, ABGA sensitivity was 69% using WB and 63% with IF. WB identified 3 common basal ganglia antigens (40 kDa, 45 kDa, and 60 kDa) in both acute and persistent SC cases. Antibody reactivity to cerebellum, cerebral cortex, or myelin antigen was absent in all groups. WB and IF are recommended for detecting ABGA in acute and persistent cases of SC. The results support an autoantibody-mediated mechanism in SC. (Church AJ, Cardoso F, Dale RC et al. Anti-basal ganglia antibodies in acute and persistent Sydenham's chorea. Neurology July (2 of 2) 2002;59:227-231). (Reprints: Mr Andrew Church, Neuroimaging Unit, Room 917, Institute of Neurology, Queen Square, London WC1N 3BG, UK).

COMMENT. Anti-basal ganglia antibodies are reported in Sydenham's chorea and pediatric autoimmune neuropsychiatric disorder associated with streptococcus (PANDAS). Pathological studies in SC show basal ganglia involvement. An antibody-mediated mechanism is suggested by the latency of the basal ganglia syndromes that may develop after streptococcal infection. WB and indirect IF are the best methods for detecting ABGA, whereas ELISA may be used to monitor ABGA levels over time and to assess treatment response.

DEVELOPMENTAL DISORDERS

SIMPLIFIED CLASSIFICATION OF FOCAL CORTICAL DYSPLASIA

Sections of cortex from 52 of 224 (23%) patients with cortical dysplasia, operated on for drug-resistant partial epilepsy, were retrospectively re-examined histologically at Niguarda Hospital, and Istituto Nazionale Neurologico 'C. Besta', Milan, Italy. Three subgroups were identified as follows: 1) architectural dysplasia (31 patients) with abnormal cortical lamination and ectopic neurons in white matter; 2) cytoarchitectural dysplasia (6 patients) with altered cortical lamination and giant neurofilament-enriched neurons; and 3) Taylor-type cortical dysplasia (15 patients) with cortical laminar disruption and giant dysmorphic neurons and balloon cells. Group 1 architectural dysplasia cases had a temporal lobe location for the epileptogenic zone, with focal hypoplasia, and a significantly lower seizure frequency than groups 2 and 3. Group 3 Taylor-type dysplasia patients had an extratemporal epileptogenic zone, ictalalal distinctive stereo-EEG with high