PEDIATRIC NEUROLOGY BRIEFS
A MONTHLY JOURNAL REVIEW

J. GORDON MILLICHAP, M.D., F.R.C.P., EDITOR

Vol. 15, No. 4 April 2001

AUTOIMMUNE DISORDERS

NEUROLOGIC COMPLICATIONS OF LUPUS ERYTHEMATOSUS

Neuropsychiatric symptoms of 10 children (8 girls and 2 boys, ages 7.5 to 17 years) with systemic lupus erythematosus were studied at the Children's Hospital Los Angeles, CA. Psychiatric symptoms at presentation included withdrawal (2), confusion (4), disorganized behavior (3), and sleep disorder (4). Psychosis in 7 was manifested by auditory, visual, and tactile hallucinations, and persecutory or bizarre delusions. Mood was depressed (6) or apathetic, irritable, anxious, or manic. Affect was constricted or blunted (5), labile or flat. One-half the patients were suicidal, and all exhibited poor attention and concentration. Neurologic symptoms in 8 patients included seizures (7) with status epilepticus (1), optic neuritis (2) with transverse myelitis (1), peripheral neuropathy (2), and headache (2). CSF was normal except for increased pressure in 2. EEGs recorded in 8 were normal. Serum anti-neuronal antibodies were elevated at onset and declined with resolution of symptoms. Anti-phospholipid antibodies were positive in 4. MRI showed a pontine infarct in one of 9 studied. SPECT was abnormal in all 10 patients, showing multiple small cortical defects with reduced cerebral perfusion. Neuropsychiatric symptoms improved within 6 months, but chronic sequelae including neuropathy and cognitive problems were frequent. Psychosis and depression recurred after intervals of 2 and 5 years in 2 patients. (Turkel SB, Miller JH, Reiff A. Case series: Neuropsychiatric symptoms with pediatric systemic lupus erythematosus. J Am Acad Child Adolesc Psychiatry April 2001;40:482-485). (Respond: Dr Turkel, Children's Hospital Los Angeles, 4650 Sunset Blvd #82, Los Angeles, CA 90027).

COMMENT. Neuropsychiatric disorder (seizures or psychosis) is included among the diagnostic criteria for systemic lupus erythematosus (American College of Rheumatology 1997). Children with an acute onset of delirium, psychosis, confusion, depression, or mania, with or without seizures or other neurologic symptoms, should be investigated for systemic lupus erythematosus. Diagnosis may be confirmed by serum anti-neuronal antibody (ANAB) testing, and resolution of symptoms is correlated with reduction in ANAB levels. In the

PEDIATRIC NEUROLOGY BRIEFS (ISSN 1043-3155) © 2001 covers selected articles from the world literature and is published monthly. Send subscription requests ($63 US; $65 Canada; $73 airmail outside N America) to **Pediatric Neurology Briefs - J. Gordon Millichap, M.D., F.R.C.P.-Editor,** P.O. Box 11391, Chicago, Illinois, 60611, USA.
The editor is Pediatric Neurologist at Children's Memorial Hospital and Northwestern University Medical School, Chicago, Illinois. PNB is a continuing education service designed to expedite and facilitate review of current scientific information for physicians and other health professionals. Fax: 312-943-0123.

above study, SPECT was abnormal in all patients and remained abnormal at follow-up.

Cerebral vein thrombosis as a complication of SLE was reported in 3 girls admitted with persistent headache at the Hospital for Sick Children, Toronto (Uziel Y et al. J Pediatr 1995;126:722-727; see Progress in Pediatric Neurology III, PNB Publ, 1997;173-4).

Autoimmune-mediated, childhood onset obsessive-compulsive disorder and tics are the subject of a review (Hamilton CS, Swedo SE. Clinical Neuroscience Research Jan 2001;1:61-68) from the National Institute of Mental Health, Bethesda, MD. In this subgroup of PANDAS there is a temporal association between Group A Beta Hemolytic Streptococcal infection and onset or exacerbation of symptoms. If a causal link can be confirmed, antibiotic treatment may be justified as an adjunct therapy of OCD and tics.

SPECT findings in early-onset OCD. Regional cerebral blood flow (rCBF) SPECT was measured in 13 early-onset (<10 years) and 13 late-onset (>12 years) adult OCD subjects and in 22 healthy controls, in a study at the University of Sao Paulo Medical School, Brazil. Early-onset OCD subjects showed decreased rCBF in the right thalamus, left anterior cingulate, and both inferior prefrontal cortices relative to late-onset cases ($p<.0005$), and severity of OCD symptoms corresponded with left orbitofrontal rCBF. The mechanisms involved in OCD may differ according to age at onset of symptoms. (Busatto GF et al. J Am Acad Child Adolesc Psychiatry March 2001;40:347-354).

ISOLATED ANGIITIS OF THE CNS

The clinical features, pathology, and outcome of isolated (primary, idiopathic) angiitis of the CNS (IACNS) in two new and 8 previously reported cases are analysed at the Hospital for Sick Children, Toronto, Canada. Cases were grouped according to the size of arteries affected - 1) small or 2) large and medium:

Group 1. IACNS affected small vessels in 5 cases, including the 2 new cases. Onset of symptoms was gradual, with persistent headaches in 3, multifocal neurologic deficits (2), cognitive impairment (1), mood disorder (1), and focal seizures (3). Brain MRI was abnormal in all 5, showing a single tumor-like mass in 2. Angiography was abnormal in only 1, showing stenosis of a quaternary branch of the middle cerebral. Diagnosis was confirmed by CNS biopsy, showing a nongranulomatous (lymphocytic) vasculitis. Immunosuppression treatment with prednisone was successful in 4 who are alive without recurrence; 1 died at 18 months after presentation, despite the addition of cyclophosphamide in thgerapy.

Group 2. Large artery IACNS in 5 cases presented with ischemic stroke in 2, transient ischemic attacks (1), and subarachnoid hemorrhage (2). Four died within 10 days, and the fifth child had cerebral hematomas secondary to recurrent ruptured aneurysms. The wall of one resected aneurysm showed inflammatory infiltration, and this child died 7 years after presentation. All 5 at autopsy showed granulomatous infiltration and necrosis of large and medium vessels. Elevated ESR, inflammatory CSF, and abnormal angiograms are more frequent in group 2 patients. Clinical evaluation excluded other causes, including drug exposures, systemic infection, thromboembolism, and rheumatic disease. Patients with poor outcome had involvement of large and medium-sized arteries, presentation with acute stroke, granulomatous angiitis on brain biopsy, and