INFECTIOUS DISORDERS

ALICE IN WONDERLAND SYNDROME WITH MONONUCLEOSIS

Five children with "Alice in Wonderland" syndrome associated with infectious mononucleosis are reported from Assaf Harofeh Medical Center, Zerifin, and Sheba Medical Center, Tel Aviv University, Israel. Visual evoked potentials (VEP) were studied during and after symptoms which included episodes of visual illusions - distortion of form, size, position, movement, or color - associated with headache, nausea, and dizziness. The episodes lasted 5 to 20 minutes and occurred 6 to 23 times daily. During the disease, VEPs showed statistically high amplitudes of P100-N145 compared to the control group, similar to findings in migraine. EEGs showed occipital high-voltage sharp or slow waves in 3 patients, between episodes. After recovery, repeat studies were normal. MRI or CT were normal. A focal decrease in cerebral perfusion is postulated as the pathophysiologic abnormality. (Lahat E, Berkovitch M, Barr J, Paret G, Barzilai A. Abnormal visual evoked potentials in children with "Alice in Wonderland" syndrome due to infectious mononucleosis. J Child Neurol 1999;14:732-735). (Respond: Dr E Lahat, Head, Pediatric Neurology Unit, Assaf Harofeh Medical Center, Zerifin 70300, Israel).

COMMENT. Alice in Wonderland syndrome, or metamorphopsia, associated with abnormal visual evoked potentials, is reported in infectious mononucleosis. It is also described in patients with migraine and epilepsy, and during acute febrile states. Cerebral perfusion PET studies have demonstrated decreased perfusion in regions near the visual tract and visual cortex in 4 children with this syndrome (Kuo YT et al. Pediatr Neurol 1998;19:105-108).

Polymerase chain reaction in CNS infections. The clinical use of cerebrospinal fluid PCR in the diagnosis of infectious neurologic viral diseases is reviewed from the University of Colorado Health Sciences Center, Denver (DeBiasi RL, Tyler KL. Arch Neurol 1999;56:1215-1219). PCR and detection of minute amounts of DNA or RNA in various tissues or fluids may help to identify infectious causes of diseases of previously undetermined etiology. It may also differentiate recurrent viral infection (PCR-positive) from post-infectious immune-mediated disease (PCR-negative).

BENIGN ACUTE MYOSITIS AND INFLUENZA VIRUS

Thirty-eight children (32 boys, 6 girls) with 41 episodes of myositis between 1978 and 1997 are reported from the Royal Children's Hospital, University of Melbourne, Australia. Viral studies during 24 of the episodes were positive in 10 (42%), with influenza B isolated in 5 (50%).

A typical case was a 7-year-old boy admitted with calf pains and difficulty in walking. Fever, cough, and lethargy had preceded the muscle pain by 5 days. He walked on his toes, the calf muscles were tender, and passive ankle dorsiflexion exacerbated the pain. Muscle tone, power, tendon reflexes, and sensation were normal in all extremities. Abnormal laboratory studies included an elevated creatine kinase (CK) of 4762 U/L (normal, 40-240 U/L), sedimentation rate (ESR) of 12 mm/h (normal, <6), and leukopenia (3.0×10^9). Parainfluenza 3 was isolated from a nasopharyngeal aspirate. Pain resolved within 24 hours after admission, and recovery was complete at 6 day-follow-up.

Mean age at onset was 8.1 years. Children were ambulant in 75% of episodes, but gaits were abnormal, on toes or wide-based stiff-legged, with the

trunk flexed at the hips and both knees extended. Muscle tenderness was restricted to the gastrocnemius-soleus muscles, and CK was elevated during all episodes. Muscle biopsy in one case revealed active degeneration with necrosis, myophagia, and ghost fiber formation, findings consistent with a viral infection. Symptoms resolved within 1 week, and only 3 had recurrent episodes. Benign acute myositis occurs mainly in boys, almost always in mid-childhood, and in response to a viral infection, especially influenza. (Mackay MT, Kornberg AJ, Shield LK, Dennett X. Benign acute childhood myositis. Laboratory and clinical features. Neurology December (1 of 1) 1999;53:2127-2131). (Respond: Dr Andrew J Kornberg, Department of Neurology, Royal Children's Hospital, Flemington Rd, Parkville, Victoria, Australia).

COMMENT. This acute childhood muscle disorder, first described by Lundberg A (Myalgia cruris epidemica. Acta Paediatr 1957;46:18-31), may initially mimic a more serious cause of limb pain and refusal to walk - Guillain-Barre syndrome. Benign acute childhood myositis (BACM) is differentiated by the normal reflexes, normal muscle power, and elevated CK. The disease is epidemiologically associated with viral outbreaks, especially influenza. Could BACM sometimes explain the idiopathic myositis of childhood referred to as "growing pains?"

Myositis, or inflammatory disease of muscle, may be acute, subacute or chronic and occurs in two main forms: 1) caused by an identified virus (eg influenza), parasite (eg trichinosis), or pyogenic bacterium (eg *staphylococcus, streptococcus)* and 2) idiopathic but presumed to be inflammatory because of histopathologic changes in the muscle (eg polymyositis, dermatomyositis). Polymyositis is closely related to the rheumatic or connective tissue diseases (rheumatic fever, lupus erythematosus, and polyarteritis nodosa). The muscle biopsy in true myositis is characterized by exudation, infiltration by neutrophilic leukocytes, and by degeneration of parenchymal and interstitial cells. (Adams RD et al. Diseases of Muscle 2nd ed. New York, Harper and Row, 1962). The PCR may now be used to define a viral etiology in cases where the diagnosis is in doubt. In children presenting with BACM, a muscle biopsy is rarely required since the symptoms are acute and remission rapid.

NEUROMUSCULAR DISORDER

CORTICOSTEROIDS IN BELL'S PALSY

The effects of corticosteroids on the early and late outcome of 42 children (21 boys, 21 girls) with Bell's palsy (acute idiopathic facial nerve paralysis) were evaluated in a prospective randomized study at the University of Istanbul, Turkey. Patients were examined in the first 3 days after onset and at 4, 6, and 12 months follow-up. In the group (n=21) that received methylpredisolone (1 mg/kg daily for 10 days orally), complete recovery occurred in 86% and 100% at 4 and 6 months follow-up, respectively. In the control untreated group (n=21), 72% and 86% had recovered completely at 4 and 6 months, and all patients had recovered by 12 months. No significant difference was found in the two groups, and the steroid group exhibited no serious side effects. (Unuvar E, Oguz F, Sidal M, Kilic A. Corticosteroid treatment of childhood Bell's palsy. Pediatr Neurol 1999;21:814-816). (Respond: Dr Emin Unuvar, Munif Pasa Sok, 65:4; TR-34300 Haseki, Istanbul, Turkey).

COMMENT. In this large group of children with Bell's palsy, steroid therapy begun within 3 days of onset had no significant effect on the outcome.