BRAIN TUMORS

ADVERSE EFFECTS OF RADIATION AND CHEMOTHERAPY

The long-term consequences of radiation and chemotherapy on intellectual and endocrine function in children with brain tumors is reviewed from the Departments of Neurology and Pediatrics, State University of New York, Buffalo, NY. The young age at time of radiation, high doses of radiation, site of the tumor, and use of methotrexate are risk factors for the development of both endocrinopathies and intellectual dysfunction. Delay in the use of radiation and the substitution of post-operative chemotherapy in patients younger than 3 years of age may delay development of intellectual dysfunction in surviving children. The omission of methotrexate from treatment programs may avoid the complications of leukoencephalopathy and dementia, although the influence of other chemotherapeutic agents on intellectual functioning is as yet unknown. All Pediatric Oncology Group protocols currently include base-line neuropsychologic evaluations as well as yearly follow-up exams throughout the course of study. Improved quality of life is a primary goal of treatment regimens. (Duffner PK, Cohen ME. Long-term consequences of CNS treatment for childhood cancer, part II: clinical consequences. Pediatr Neurol July/Aug 1991; 7:237-242).

COMMENT. The increase in survival rate of children with brain tumors over the past 20 years has been accompanied by a decrease in intellectual functioning and other serious complications. The recognition of risk factors for long-term adverse consequences of radiation and chemotherapy should lead to modifications of treatment regimens and improvement in academic potential.

In a study at the Children's Hospital and Harvard Medical School, Boston, MA, comparing the effectiveness of thallium-201 SPECT imaging with MRI in tumor assessment, SPECT allowed a more precise assessment of the functional state of the tumor but was less reliable than the MRI in detecting possible radiation injury in some patients (O'Tuama LA et al. SPECT imaging of treated childhood brain tumors. Pediatr Neurol 1991; 7:249-257).

Green DM et al, at the Roswell Park Cancer Institute, Buffalo, NY, report that the frequency of congenital anomalies was not increased in children of men or women who had received chemotherapy for cancer during childhood and adolescence, with the possible exception of those treated with dactinomycin, at increased risk for cardiac defects (N Engl J Med July 18, 1991; 325:141-6).

PROGNOSIS OF BRAIN TUMOURS WITH EPILEPSY

The prognosis of 560 patients with a clinical and CT diagnosis of intrinsic supratentorial brain tumour was examined retrospectively at the Department of Neurosciences, Walton Hospital, Liverpool, England. Epilepsy was the first symptom in 164 patients. Patients presenting with epilepsy had

a median survival of 37 months compared to 6 months in those presenting with other symptoms; they were more likely to have a normal clinical examination, a non-enhancing low density lesion on CT scan and a low grade tumour. Significant independent variables which adversely affected prognosis were increasing age, focal neurological signs, an enhancing CT lesion at diagnosis, surgical biopsy, and male sex. Primary intracerebral tumours presenting with epilepsy were relatively benign. They were less likely to receive radiotherapy or biopsy, but more likely to undergo resective surgery. No beneficial effects from either early resective surgery or radiotherapy were demonstrated. The average duration of symptoms before CT diagnosis was 28 months (range 2 days - 232 months in those presenting with epilepsy) and 4 months (range 1 day - 175 months in those with other presentations). Seizures were refractory to anti-epileptic drugs with only 11 of 164 patients ever achieving a 1 year remission (Smith DF et al. The prognosis of primary intracerebral tumours presenting with epilepsy: the outcome of medical and surgical management. J Neurol Neurosurg and Psych Oct 1991; 54:915-920).

COMMENT. Backus RE and Millichap JG analyzed the records of 291 consecutive children treated for intracranial tumor at the Mayo Clinic from 1950-1959 with particular attention to patients with seizures (Pediatrics June 1962; 29:978-984). Seizures occurred in 17% of the total group - in 25% of patients with supratentorial tumors and in 12% of those with infratentorial tumors. They were the initial symptoms in 15% of patients with supratentorial tumors and in 1% of those with infratentorial tumors. The average age at onset was 4.9 years in the supratentorial group and 4.8 years in the infratentorial group. The diagnosis of supratentorial tumors was delayed for an average of 2 years, whereas infratentorial tumors were diagnosed within an average time of 3 months of the initial seizure. The electroencephalogram was of localizing value in 92% of tumors that involved the cerebral cortex. The histologic type of tumors was verified in 46 of the 50 patients who had seizures. Seizures were more common in patients with slowly growing astrocytomas grades 1 and 2 (67% incidence) than in those with rapidly expanding astrocytomas grades 3 and 4 (10% incidence). Symptoms of increased intracranial pressure were present at the time of the first seizure in 20% of patients with supratentorial tumors and in 79% with infratentorial tumors. A delta pattern in the EEG indicative of an expanding lesion occurred in 50% of patients and spike, sharp wave, or spike and wave seizure discharges compatible with a static lesion were recorded in 32% of patients with supratentorial tumors (Millichap JG et al. The electroencephalogram in children with intracranial tumors and seizures. Neurology May 1962; 12:329-336).

With the increasing availability of neuroimaging techniques and particularly the MRI, the early diagnosis of tumor in patients with focal epilepsy will be facilitated and may lead to earlier surgical resection. The present study questions the advisability of resective surgery in patients with primary brain tumors presenting with seizures. A

prospective, randomized, study of aggressive versus conservative management is required.

INFECTIOUS DISORDERS

RELAPSING HERPES SIMPLEX ENCEPHALITIS WITH CHOREA

Herpes simplex encephalitis (HSE) followed a biphasic course with relapse in 3 children aged 11 years, 9 months, and 11 months reported from the Hospital for Sick Children, London and South Hampton General Hospital, England. All 3 presented with fever and seizures and relapse was accompanied by generalized or hemi-chorea in 2 patients. All patients received repeat courses of acyclovir, antibiotics and anticonvulsants. Dexamethasone was added in the older patient and the acyclovir course was followed by vidarabine in 2 patients. All patients had residual sequelae at discharge: 1 had a mild right facial weakness and persisting dysphasia, another was severely handicapped with persisting chorea and a third was visually inattentive and had a right hemiplegia and left hemichorea. The cause of the relapse was not definitely determined. A possible resistant organism might indicate a longer initial course of high dose acyclovir or alternatively, a post-infectious neuroallergic process might suggest the substitution of immunomodulatory treatment in place of further antiviral treatment (Pike MG et al, Herpes simplex encephalitis with relapse. Arch Dis Child Oct 1991; 66:1242-4).

COMMENT. Relapsing herpes simplex encephalitis is not uncommon in adults but is rare in children. Chorea in association with HSE relapse is of interest. The authors refer to 7 additional patients reported in the literature.

Autism is another unusual sequel to herpes simplex encephalitis described in a 31 year old adult at the University of Goteborg, Sweden. (Gillberg IC. Autistic syndrome with onset at age 31 years: Herpes encephalitis as a possible model for childhood autism. Dev Med Child Neurol Oct 1991; 33:920-924). Other cases in the literature have occurred in pre-adolescence. Autism is not necessarily a developmental disorder and temporal lobe damage caused by herpes encephalitis may explain some cases. The Kluver-Bucy syndrome, characterized by emotional instability, hypersexuality and hyperorality, has also been described as a complication of herpes encephalitis when both temporal lobes have been involved.

PROGNOSIS OF ENCEPHALITIS

A population based, controlled follow-up study of the general outcome of 73 children followed for 2 to 12 years after the acute phase of childhood encephalitis is reported from the University of Oulu, Finland. Varicella accounted for 23% of cases, mumps 11%, herpes simplex 8%, measles 5%, and the etiology was unknown for 44%. The mean age at onset was 5.9 years