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BRAIN NEOPLASMS

PRIMITIVE NEUROECTODERMAL TUMOR PROGNOSIS

Biological factors of prognostic significance in primitive neuroectodermal tumors (PNETs) were evaluated in tumor specimens of 86 children examined at the Children's Hospital of Philadelphia and University of Pennsylvania. Immunochemical evidence of glial differentiation (glial fibrillary acidic protein, GFAP) or neuronal differentiation (neurofilament proteins, NFPs) was associated with a 6.7-fold greater risk of relapse than tumors that did not express GFAP or NFPs. These biological characteristics of PNETs were as significant as tumor location and metastatic stage in predicting rate of relapse. (Janss AJ et al. Glial differentiation predicts poor clinical outcome in primitive neuroectodermal brain tumors. Ann Neurol April 1996;39:481-489). (Respond: Dr Janss, Division of Neurology, Children's Hospital of Philadelphia, 3400 Civic Center Blvd, Philadelphia, PA 19104).

COMMENT. The authors suggest that children with PNETs, especially cerebellar medulloblastoma, that express large amounts of glial fibrillary acidic protein (GFAP) are at increased risk for relapse and should be treated aggressively. Tumors should be classified as low or high risk based on both clinical factors, tumor location and metastatic stage, and biological factors, including GFAP expression.

The Mayo Clinical Update (12;1:1996) notes that malignant brain tumors in children are increasing in incidence at a rate of 1.4% per year. For medulloblastoma, after radical resection followed by cisplatin and craniospinal irradiation, 5-year survival rate is 75%. A protocol involving gene therapy is about to be opened for the treatment of recurrences of some malignant tumors. The tumor is injected with cells that secrete a retroviral vector containing thymidine kinase gene isolated from herpes simplex virus. This therapy should result in killing of tumor cells with sparing of normal brain. New therapies are needed for PNETs since no effective treatment for children with recurrent medulloblastoma has been identified. Intensive therapies presently employed result in complications, including second malignancies, endocrine

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and growth dysfunction, and cognitive impairments. Basal-cell carcinoma, discussed below, is an example of a secondary malignancy induced by spinal irradiation for medulloblastoma. Low risk tumors, identified by the biological method, may be spared unnecessary aggressive and disabling therapy.

NEVOID BASAL-CELL CARCINOMA (GORLIN) SYNDROME

A unique case of a 15-year-old girl with nevoid basal-cell carcinoma (NBCCS), who presented with uncontrolled temporal lobe epilepsy due to neuronal heterotopia, is reported from the Royal Melbourne Hospital, Australia. Seizures began at age 13 months, often precipitated by fever. She had a dermoid cyst removed from her nose, multiple dental cysts, and seborrheic keratosis of the scalp. Growth increased dramatically at age 9 years, and seizures became refractory to treatment. Chest X-ray showed cervical spina bifida occulta and fused ribs. Head CT showed calcification of falx cerebri and tentorium. MRI showed a lesion in the left anterior temporal lobe. EEG recorded seizure activity in the left frontotemporal areas. Temporal lobectomy specimen showed neuronal heterotopias and cortical dysplasia. After surgery, she was seizure free at 6 months follow-up. (Hogan RE et al. Epilepsy in the nevoid basal-cell carcinoma syndrome (Gorlin syndrome): Report of a case due to a focal neuronal heterotopia. Neurology Feb 1996;46:574-576). (Reprints: Dr R Edward Hogan, Department of Neurology, St Louis University, 3635 Vista Ave at Grand Blvd, PO Box 15250, St Louis, MO 63110).

COMMENT. Nevoid basal-cell carcinoma syndrome presents at puberty with multiple basal-cell carcinomata, palmar and plantar pits, odontogenic painful cysts of the mandible, facial paresthesia, macrocephaly, large stature, frontal bossing, broad nasal bridge, narrow shoulders, fused ribs, polydactyly, bone cysts, calcified falx, cervical spina bifida occulta, odontoid agenesis, corpus callosum agenesis, meningioma, and medulloblastoma. Medulloblastoma, occurring in 3-5% of cases, may precede the development of other manifestations of the syndrome, and may have a better prognosis than general. Patients with seizures should be investigated for neuronal heterotopias, which may be treated surgically. The syndrome is autosomal dominant and the gene is localized to chromosome 9q22.

INTRACRANIAL GERM-CELL TUMORS

Prognosis of 26 children with intracranial germ-cell tumors (17 germinomas and 9 teratomas) treated in a 10 year period is reported from the University Hospital Hamburg-Eppendorf, Germany. Median age at diagnosis was 11.5 years (range 1.5-16.8 years). Tumor location was in the pineal region in 69% and suprasellar/hypothalamic in 31%. Symptoms were increased intracranial pressure, Parinaud's syndrome, and endocrine deficits. Surgical resection was attempted in 22 patients, 25 were irradiated, and 8 received additional chemotherapy. Long-term survival rate was 88% for germinomas and 43% for malignant teratomas; 57% had no and 37% had mild neurologic deficits, and 24% had neuro-endocrine dysfunction. Neuropsychological function was normal or only mildly impaired in 53%, and 69% had no change in their level of education. (Haupt C et al. Intracranial germ-cell tumours - treatment results and residuals. Eur J Pediatr March 1996;155:230-236). (Respond: Dr C Haupt, Departments of Paediatrics and Neurosurgery, University Hospital Hamburg-Eppendorf, D-20251 Hamburg, Germany).

COMMENT. In this center, primary intracranial germ-cell tumors (PGCT)