PEDIATRIC NEUROLOGY BRIEFS
A MONTHLY JOURNAL REVIEW

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

Vol. 14, No. 11 November 2000

BRAIN TUMORS

BRAINSTEM NEUROECTODERMAL TUMORS

Seven histologically confirmed primitive neuroectodermal tumors (PNETs) arising in the brainstem were identified among 146 pediatric brainstem tumors, in a review of the the clinical neuro-oncology database and 1986-1995 files of the Division of Neuropathology at New York University Medical Center. All 7 patients presented with focal cranial nerve pareses (VI in 3, VII in 2, VI & VII in 1, and III in 1), 2 had hemiparesis, and 1 was ataxic. The median age at diagnosis was 2.7 (1.0-8.0) years, and the mean duration of symptoms was 2 (1-6) months. MRIs showed a focal intrinsic exophytic nonenhancing lesion with low T1-weighted and high T2-weighted signals. All patients required a ventriculoperitoneal shunt for hydrocephalus. Meningeal dissemination occurred in 6 patients. Only 2 responded temporarily to therapy, for 4 and 6 months. The median survival was 7 (3-17) months, and all patients died within 17 months of diagnosis. Tests for mutation in the p53 gene were negative. (Zagzag D, Miller DC, Knopp E et al. Primitive neuroectodermal tumors of the brainstem: investigation of seven cases. Pediatrics November 2000;106:1045-1053). (Reprints: David Zagzag MD PhD, Department of Pathology, Division of Neuropathology, New York University School of Medicine, 550 First Ave, New York, NY 10016).

COMMENT. Primitive neuroectodermal tumors (PNETs) account for 5% of pediatric brainstem tumors. Compared to the more common brainstem gliomas, PNETs are encountered at an earlier age, the MRI lesion is localized, nonenhancing rather than diffuse intrinsic, there is a greater predilection for leptomeningeal dissemination, and the prognosis is worse. Brainstem PNETs are more aggressive than cerebellar PNETs. The authors recommend biopsies in cases of brainstem tumor with clinical and radiological characteristics of brainstem PNET. They favor high-dose focal irradiation without biopsy in cases diagnosed clinically as glioma of the pons or medulla and not showing leptomeningeal dissemination or characteristics of PNET. Mutations of the p53 gene, a common finding in those pediatric brainstem gliomas tested, are rare in PNETs, and an added distinguishing characteristic.

PEDIATRIC NEUROLOGY BRIEFS (ISSN 1043-3155) © 2000 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. Visit our web site: www.pnbpublishers.com