psychosocial stress and prevent premature skeletal maturation and a shorter adult stature. Medical treatment with long-acting gonadotrophin releasing hormone must be administered through childhood and surgery is sometimes preferred.

INTRACRANIAL LYMPHOMA AND GRADENIGO SYNDROME

A 13 year old black male patient with a T-cell lymphoma who presented with Gradenigo syndrome is reported from the Department of Pediatrics, Tulane University School of Medicine, New Orleans, LA. There was a seven day history of headache described as right-sided, throbbing pain, posterior to the eye and associated with right-sided facial numbness. After five days he developed double vision on right lateral gaze and marked decrease of pinprick sensation of the entire right face and loss of the right corneal reflex. Red glass testing confirmed diplopia from a weakness of the right lateral rectus muscle. CT revealed dural enhancement in the medial aspect of the right middle cranial fossa adjacent to the sella turica. MRI showed the right internal carotid artery compressed and encased by the mass and the Meckel's cave segment of the trigeminal nerve was obliterated. A transphenoidal biopsy provided the diagnosis of T-cell lymphoma, Lennert type, and further evaluation revealed diffuse involvement of bone marrow, spleen, kidneys, and testes. Immunologic workup showed hypogammaglobulinemia. Chemotherapy and radiotherapy resulted in completed resolution of all symptoms and signs. (Norwood VF, Haller JS. Gradenigo syndrome as presenting sign of T-cell lymphoma. Pediatr Neurol Nov-Dec 1989; 5:377-380).

COMMENT. Gradenigo syndrome consists of cranial nerve VI palsy and abnormalities of the sensory component of ipsilateral cranial nerve V. Gradenigo described the symptom complex with middle ear infections and it has been reported as a result of tumors, most commonly neurofibroma. The primary neurologic presentation of lymphoma was unique and a new etiology for Gradenigo syndrome.

RADIOTHERAPY IN BRAINSTEM GLIOMAS

The results of a multiinstitutional phase I/phase II trial, using 100 cGy of radiation therapy twice daily to a total dose of 7,200 cGy in 31 children with high risk brainstem gliomas are reported from the Neuro-Oncology Program, The Children's Hospital of Philadelphia; the University of Pennsylvania, Philadelphia; New York University Medical Center; University of Minnesota; Children's Memorial Hospital, Chicago; and the Robert Wood Johnson Medical School, New Brunswick, NJ. Of the 35 patients evaluated, 24 (69%) had developed progressive disease and 11 (31%) remained in remission at the completion of the three year study period. Survival rate at 20 months was 32%. Patients relapsed at a median of eight months after diagnosis. Those in remission had been followed for a median of 18 months. No patient died as a result of treatment. Glucocorticoid therapy was tapered and discontinued during or soon after completion of treatment. In comparison to control patients and those treated in a previous trial using smaller doses of

hyperfractionated radiotherapy, there was a statistically significant improvement in progression-free survival rate. Further studies seemed warranted. (Packer RJ et al. Hyperfractionated radiotherapy for children with brainstem gliomas: A pilot study using 7,200 cGy. Ann Neurol Feb 1990; 27:167-173).

COMMENT. Brainstem gliomas account for approximately 10% of all childhood central nervous system tumors and are the most resistant to therapy. High risk patients with tumors involving the brain stem diffusely rarely survive after conventional doses of radiotherapy. Hyperfractionated radiation therapy offers greater potential benefit.

DEVELOPMENTAL DISORDERS

CHIARI II MALFORMATION

The theories of the basic embryological defect that lead to the Chiari II malformation are reviewed and a unified theory proposed from the Division of Pediatric Neurosurgery, The Laboratory for Oculo-Cerebrospinal Investigation, The Children's Memorial Medical Center and Northwestern University Medical School, Chicago, IL. Chiari II malformation is almost invariably associated with myelomeningocele and a progressive hydrocephalus. Associated anomalies include small posterior fossa, Luckenschadel of the skull, caudal displacement of pons, medulla, and basilar artery, and upward herniation of the superior cerebellum. Syringomyelia occurs in 50%, and aqueduct stenosis, polymicrogyria, cortical heterotopia, and agenesis of the corpus callosum occur occasionally. Previous theories include the following: 1) Herniation of the posterior fossa contents resulting from supratentorial hydrocephalus with leakage of cerebrospinal fluid into the amnion and herniation of the hindbrain; 2) Traction theory suggesting that the caudal spinal cord may pull the cerebellum and medulla into the lower cervical canal because of tethering; 3) Dysgenesis of the hindbrain and developmental arrest; 4) Small posterior fossa due to mesodermal insufficiency and overgrowth of neuroepithelium causing a neural tube defect. The unified theory proposed by the authors incorporates the previous observation of Padget that leakage of cerebrospinal fluid is one factor in the cause of a small posterior fossa and emphasizes the role of distention of the embryonic and fetal ventricular system in normal cerebral development. The neural tube defect and defective occlusion are the developmental factors that cause the Chiari II malformation and the interrelated cerebral and skull anomalies. Altered inductive pressure on the surrounding mesenchyme is the cause of the Chiari II malformation and Luckenschadel whereas the lack of distention of the developing telencephalic ventricles results in cerebral anomalies, e.g., dysgenesis of the corpus callosum, cortical heterotopia, and polymicrogyria. Chiari II malformation is the result of a series of interrelated time dependent defects in the development of the ventricular system leading to multiple anomalies in brain development,