Both suffered febrile tonic seizures and one was hyperkinetic and had stereotypic hand movements. There was generalized muscle hypotonia, deep tendon reflexes were absent and serum creatine kinase activity was elevated. There was nystagmus in all directions of gaze. Biopsies of the biceps and brachia muscles showed selective type I fiber atrophy and mild type I fiber predominance similar to that observed in congenital fiber type disproportion myopathy. (Kohyama J et al. Congenital fiber type disproportion myopathy in Lowe syndrome. Pediatr Neurol Nov-Dec 1989; 5:373-376).

COMMENT. Selective type I muscle atrophy is uncommon and occurs in congenital nonprogressive myopathies, including nemaline myopathy, myotubular myopathy, central cord disease, and congenital fiber type disproportion. In association with type I fiber predominance it is observed in Pompe disease, Krabbe leukodystrophy and multiple sulfatase deficiency in which peripheral nerves are involved. The oculo-cerebro-renal syndrome of Lowe is characterized by mental retardation, glaucoma, congenital cataracts, and renal impairment. The elevated CPK led to the muscle biopsies in the present patients.

INTRACRANIAL TUMORS

HYPOTHALAMIC HAMARTOMA AND SEXUAL PRECOCITY

Four boys with hypothalamic hamartomas associated with sexual precocity are reported from the Departments of Pediatrics and Neurosurgery, University of Pittsburgh, and the Department of Pediatrics, Johns Hopkins University School of Medicine, Baltimore, MD. Two patients were treated surgically by resection using current microsurgical techniques and two received medical management. Precocious puberty caused by hamartomas occurs early in life with enlarged penis and muscular build noted in early infancy. Growth is accelerated and bone age is advanced. Deepening of the voice and the appearance of acne are common. Other clinical findings include mental retardation, behavioral disturbances, and seizures of gelastic, absence, and generalized tonic clonic patterns. In the two patients treated surgically, subsequent growth and development were normal and in two who were diagnosed late in childhood and treated medically, the adult height was not particularly compromised. The authors recommend surgical resection if the hamartoma is pedunculated or in cases where the patient is young and would require years of parenteral medical treatment. The nontreatment option exists because there is no evidence that the tumors will grow or subsequently cause other problems. (Starceski PJ et al. Hypothalamic hamartomas and sexual precocity. Evaluation of treatment options. AJDC Feb 1990; 144:225-228).

COMMENT. The use of MRI with sagittal, coronal, and axial views enable better visualization of hamartomas and earlier diagnosis. Treatment to allow growth and pubertal development to occur at an age appropriate time may avoid considerable

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