associated with bilateral temporal polyspike and sharp-wave EEG activity were controlled with phenobarbital. His head circumference was at the 75th percentile and disproportionately large. A repeat ultrasound at 7 mos showed increased echogenicity in the midline with normal ventricles, interpreted as blood in the third ventricle with calcification. CT showed a large midline area of decreased density extending into the lateral ventricles, consistent with a lipoma of the corpus callosum and calcifications. At 18 mos the head is large and development is delayed at the 12 mo level. (Imaizumi SO et al. Lesion mistaken for hemorrhage in a premature infant: Lipoma of corpus callosum. Pediatr Neurol Oct 1988;4:313-6).

COMMENT. Blood, fat, and calcium have similar echogenicity by cranial ultrasound, and all three media are hyperechoic when compared to the moderate echogenicity of cerebral white matter, low echogenicity of gray matter, and absent echogenicity of fluid-filled ventricles or cysts. Lipomas are characterized by hyperechoic densities on cranial sonograms.

SPINAL CORD HEMANGIOBLASTOMA

A 6-month-old infant with a spinal cord hemangioblastoma located in the conus medullaris is reported from the University of Washington School of Medicine, Children's Hospital, Seattle, WA. At birth the physical examination was normal except for a lumbosacral dimple. By 1 mo the dimple had deepened and a hemangioma developed at the site. CT and CT metrizamide myelography revealed a posterior filling defect at the level of the conus medullaris. At laminectomy, a congenital dermal sinus tract, cutaneous capillary hemangioma and cord hemangioblastoma were resected. There were no neurologic deficits either postoperatively or following removal of the tumor. (Michaud LJ et al. Hemangioblastoma of the conus medullaris associated with cutaneous hemangioma. Pediatr Neurol Oct 1988;4:309-12).

COMMENT. Spinal cord hemangioblastoma rarely presents in infancy and is usually manifest from the third to the fifth decades. It may be associated with a variety of cutaneous and other lesions, including von Hippel-Lindau and Cobb syndromes. Von Hippel-Lindau disease is an autosomal dominant condition characterized by hemangioblastomas of the cerebellum, medulla and spinal cord; angiomas of the retina, liver, and kidney; pheochromocytomas, adenomas, or cysts of the kidney and epididymis; and pancreatic cysts. In Cobb syndrome, cutaneomeningospinal angiomatosis, spinal cord arteriovenous malformations are associated with cutaneous vascular lesions in corresponding dermatomes. These syndromes were not found in the above case-report.

CHEMOTHERAPY FOR MEDULLOBLASTOMA

The efficacy of adjuvant chemotherapy for patients with

poor-risk medulloblastoma/primitive neuroectodermal tumors (MB/PNET) has been studied at the Children's Hospital of Philadelphia, University of Pennsylvania, PA. Chemotherapy consisted of vincristine during concomitant craniospinal radiation therapy and eight 6-week cycles of vincristine, cis-platinum, and cyclohexylnitrosourea. Twenty five of 26 children (96%) treated remain alive and free of disease at a median of 24 months from diagnosis (range 6-50 mos). Actuarial disease-free survival was statistically significantly better than for control subjects who had received radiation therapy alone during an 8 year period prior to the use of adjuvant chemotherapy. The 2-year disease-free survival was 96% for patients on the protocol of adjuvant chemotherapy as compared to 59% for historical control patients treated with radiotherapy alone. (Packer RJ et al. Efficacy of adjuvant chemotherapy for patients with poor-risk medulloblastoma: a preliminary report. Ann Neurol Oct 1988;24:503-508).

COMMENT. Surgical excision for the treatment of medulloblastoma has less than a 6 month survival rate, and postoperative radiation of the tumor bed alone has minimal benefit. Irradiation of the tumor bed and the entire neuroaxis provides an overall chance of 30% to 50% for 5 year survival. Patients with clinical evidence of arachnoidal seeding or with gross evidence of seeding at surgery do poorly. Survival without evidence of recurrent tumor for a period exceeding the patient's age at diagnosis plus 9 months indicates a probably cure ("Collins' law"). Corticosteroids result in remarkable amelioration of root signs and symptoms and reduction of edema for a brief period. Experience with chemotherapy in the past 25 years has produced better survival rates, and used as an adjuvant these agents are indicated for the poor-risk patients, that is, those less than 5 years of age at diagnosis or with disseminated tumors (Chang stages M1 - M3). (Groover RV. In The Practice of Pediatric Neurology, Eds. Swaiman KF, Wright FS, CV Mosby, St. Louis, 1982.)

MUSCLE DISORDERS

McARDLE'S DISEASE

Muscle biopsy specimens from 48 patients with biochemically proven phosphorylase deficiency (McArdle's disease) have been analyzed by gel electrophoreis (SCS-PAGE), immunoblotting, and immunotitration (ELISA) at Columbia University College of Physicians and Surgeons, New York, NY. The majority had no detectable enzyme protein, 6 had markedly decreased phosphorylase protein, and only 1 had a normal amount of protein. The presence or absence of enzyme protein was not correlated with the clinical presentation or muscle glycogen concentration. In 4 patients tested, messenger RNA was normal in 2, abnormally short in 1, and absent in 1, suggesting