

in treatment by controlling the accumulation of toxic metabolites in some peroxisomal disorders, and the addition of glycerol trioleate, a lipid containing unsaturated fatty acid, is a promising new therapy that reduces the synthesis of C22-26 fatty acids. For further information on peroxisomal disorders, refer to a special article by Moser HW Neurology Oct 1988;38:1617. A family with Refsum's disease (heredopathia atactica polyneuritiformis) in whom 4 out of 6 siblings were affected is reported from the Department of Neurology, Westminster Hospital, London (Britton TC, Gibberd FB. JR Soc Med Oct 1988;81:602-3). Retinitis pigmentosa was the presenting diagnostic sign in the index case, and other affected members of the family were detected by screening for raised plasma phytanic acid levels. Early diagnosis is important because dietary treatment will prevent the development of neuropathy, ataxia, cardiac arrhythmias, and ichthyosis. Retinitis pigmentosa, anosmia, and ataxia should suggest the diagnosis.

CNS TUMORS

POSTERIOR FOSSA DERMoids

Three children with dermoid cysts of the posterior fossa are reported from the Assaf Harofeh Medical Center, Zerifen, and Hadassa Medical Center, Jerusalem, Israel. Two presented with acute meningitis at 1 and 2 yrs of age, and the third patient had hydrocephalus treated by ventriculo-peritoneal shunt at 7 mos and complicated by meningitis and cerebellar abscess at 9 mos of age. CT scans with enhancement and bone window setting revealed the midline bony defect and low density lesion with ring enhancement in the posterior fossa. (Starinsky R et al. Dermoids of the posterior fossa. Case reports and review. Clin Pediat Dec 1988;27:579-582).

COMMENT. Recurrent meningitis or brain abscess in an infant or young child should prompt a search for a sinus, fistula, and bone defect in the occipital area. Dermoid cysts of the cerebellum and posterior fossa account for 2% of intracranial tumors in children. Astrocytoma, medulloblastoma, brain stem glioma, and ependymoma of the IVth ventricle are the most frequently encountered IC tumors.

LIPOMA OF CORPUS CALLOSUM

A lipoma of the corpus callosum diagnosed by CT at 7 mos and mistaken for hemorrhage in a premature infant is reported from the Medical College of Pennsylvania, Philadelphia, PA. The Apgar scores were 3 at 1 and 5 min, and the infant had hyaline membrane disease that progressed to bronchopulmonary dysplasia. Cranial ultrasound at 10 hrs demonstrated a subependymal hemorrhage with unchanged appearance at 17 days. Seizures