CEREBELLOPONTINE ANGLE LIPOMA

A cerebellopontine angle lipoma discovered incidentally in an asymptomatic 17 year old girl during evaluation for trauma is reported from the University of Alabama and Children's Hospital, Birmingham, AL. The CT had shown a low density nonenhancing 1.5 cm mass in the right cerebellopontine angle. The patient denied hearing loss, tinnitus, facial weakness or other neurologic symptoms. On examination she had a left-sided hearing loss, an audiogram showed left conductive hearing loss, and brainstem auditory evoked potentials were abnormal on the left. MRI was consistent with either an epidermoid or a lipoma. At surgical biopsy which confirmed the diagnosis the mass involved the 7th and 8th as well as portions of the 9th, 10th and 11th cranial nerves and was partially adherent to the brainstem. Surgical removal was not possible and postoperatively the patient made an uneventful recovery. (Ashkenasi A et al. Cerebellopontine angle lipoma in a teenager. Pediatr Neurol July/Aug 1990; 6:272-274).

COMMENT. The authors found only one other report of this tumor in a child and 17 in adults. They recommend that surgical removal should be avoided if possible because of postoperative cranial nerve injury. A mass with negative CT density and increased signal intensity on T1 and T2 weighted images on MRI should limit the differential diagnosis to lipomas, dermoids, or cholesterol granulomas, according to reports in the literature.

CEREBRAL MALFORMATIONS

LISSENCEPHALY SYNDROMES

The diagnostic features and clinical signs of 21 patients with lissencephaly type I are reviewed from the Department of Neurology, Westeinde Hospital, The Hague, The Netherlands; the Departments of Child Neurology, Academic Medical Centre, Amsterdam, and Sophia Children's Hospital, Rotterdam. A multicenter study was conducted with the cooperation of all child neurology departments in The Netherlands. Lissencephaly was diagnosed at autopsy in two patients and by CT scan in 19. The criteria of Dobyns WB (1987) were used for the classification of Lissencephaly: Type I Isolated lissencephaly sequence (ILS), Miller-Dieker syndrome (MDS); Type II, Walker-Warburg syndrome (WWS), Fukuyama congenital muscular dystrophy; Rare forms, Neu-Laxova syndrome (NLS), Cerebro-cerebellar syndrome (CCS). Lissencephaly type I is classified according to pathology and radiology as follows: Grade 1, complete agyria; Grade 2, agyria with some sulci; Grade 3, a mixture of about 50% agyria and 50% pachygyria; Grade 4, complete pachygyria. Of 21 patient with lissencephaly type I, 17 had isolated lissencephaly and 4 had the Miller-Dieker syndrome. More severe abnormalities in gross brain morphology occurred in MDS than in ILS. Facial dysmorphism was most frequent in MDS patients and microcephaly in combination with facial abnormalities increases the suspicion of MDS. All children with lissencephaly in this study were severely retarded, 86% developed epilepsy before the age of six months, and one-third had infantile spasms. The somatic signs of 19 patients

with chromosome deletion 17p are described for the present series of patients and those in the literature: malformed fingers, congenital heart-defect, sacral dimple, cryptorchidism, and malformed kidneys. (De Rijk-van Andel JF et al. Diagnostic features and clinical signs of 21 patients with lissencephaly type I. Dev Med Child Neurol Aug 1990; 32:707-717).

COMMENT. The diagnosis of lissencephaly is made by clinical and neuroradiological findings. Cases of isolated lissencephaly may be distinguished from the Miller-Dieker syndrome without the results of chromosome analysis in most cases. The CT signs of lissencephaly include smooth brain surface and no Sylvian fissure with figure 8 appearance, sharp demarcation between gray and white matter, and colpocephaly. Facial dysmorphism may show high forehead, bitemporal hollowing, and micrognathia.

SCHIZENCEPHALY AND BEHAVIORAL CORRELATES

The MRI appearance and neuropsychologic and speech/language evaluations in three patients with schizencephaly are described from the Department of Radiology, Michigan State University, East Lansing, MI. All patients presented with a seizure disorder and they were left-handed without familial sinistrality in first degree relatives. The primary areas of cerebral involvement were the left parasylvian and pararolandic regions with varying degrees of secondary involvement of the right hemisphere. There was mild right-sided limb hypoplasia, and motor dexterity with the dominant left hand was better than with the right hand. Two patients showed significant impairment in finger localization and tactile form recognition particularly with the nondominant right hand. The level of general intellectual functioning related to the amount of brain tissue involved, and neurobehavioral abilities reflected the location of the brain malformation and the prenatal onset of the disorder. The full scale IQ on the WAIS-R ranged from a low of 65 to a high of 87. Only one patient showed a higher verbal IQ than performance IQ and her visuospatial construction abilities and visual memory abilities were significantly impaired. There were varying degrees of linguistic deficit with relatively greater difficulties in syntactical speech than in semantic aspects. (Aniskiewicz AS et al. Magnetic resonance imaging and neurobehavioral correlates in schizencephaly. Arch Neurol August 1990; 47:911-916).

COMMENT. Yakovlev and Wadsworth first described the schizencephalies as congenital defects in the cerebral mantle in 1946. The patients in the present study shared features in common with the pathologic left handedness syndrome of Orsini and Satz; predominantly left-sided cerebral lesion with onset before six years of age and involving speech/language areas of frontotemporal parietal cortex, atypical or right-sided hemispheric speech representation, impaired visuospatial abilities and preserved verbal cognitive abilities, right limb hypoplasia, right hand motor impairment, and absence of familial sinistrality. (Orsini DL, Satz P. A syndrome of pathological