

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 Neuro 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 1, 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