

was known to suffer from tuberous sclerosis and examination of the mother was negative (Millichap J G, Gomez MR Neurofibromatosis II and tuberous sclerosis: Simultaneous occurrence in a 14 year old girl Ped Neur Briefs 1990;4:50-51).

EPIDERMOID TUMORS

Six patients with histologically proven epidermoid tumors are reported from the neurosurgical service, Brigham and Women's Hospital and Children's Hospital, Harvard Medical School, Boston, Mass. All patients were adults between 22 and 52 years of age at the time of diagnosis. Symptoms had been present for more than one to five years and the methods of presentation included seizures in two, ataxia (3), headaches (3), dysmetria (3), left-sided hearing loss (1), visual loss (1) and papilledema (1). The locations of the tumor were variable; cerebellar hemisphere in two, left temporal lobe (2), left cerebral hemisphere (1) and suprasellar cistern (1). Surgery was successful in all patients. None had symptoms related specifically to the tumor in childhood; one had been treated for bulimia for 17 years and another had meningitis at ten years of age. CT demonstrated a hypodense, smoothly contoured, extra axial paramedial mass with a lower density than cerebrospinal fluid. MRI showed an irregularly, but sharply marginated, mass with homogeneous density, variable enhancement with gadolinium, lack of edema in adjacent normal structures, extensive insinuation into cisternal and other cerebrospinal fluid spaces and a high signal intensity on proton-weighted images. Multiplanar magnetic resonance imaging was extremely helpful in showing the full extent of the lesion and its relation to other structures (Panagopoulos K P et al. Intracranial epidermoid tumors. A continuing diagnostic challenge Arch Neurol July 1990; 47:813-816).

COMMENT: Epidermoid, adamantinoma, cholesteatoma, and pearly tumor are terms used interchangeably to refer to this entity. Epidermoid tumors are congenital and arise from misplacement of ectoderm. They are usually benign and slow growing but malignant change may occur. The tumors may be very large at the time of diagnosis, with considerable mass effect but minimal edema. The MRI allows the distinction of an epidermoid from an arachnoid cyst. The MRI or CT is not diagnostic but the MRI is more likely to elucidate the extra axial-nature of the tumor and is preferred. Spinal epidermoid tumors may complicate a lumbar puncture performed 1 to 20 or more years previously. Symptoms are slowly progressive and manifested by back and leg pains progressing to gait difficulties. (Shaywitz BA. J Pediatr 1972; 80:638).

AV MALFORMATIONS: RADIATION THERAPY

Clinical and radiologic follow-up of 86 patients with symptomatic, but surgically inaccessible, cerebral arteriovenous malformations treated with stereotactic heavy charged particle Bragg-peak radiation is reported from the Divisions of Neurosurgery

and Neuradiology, Stanford University School of Medicine, California and the Lawrence Berkeley Laboratory, University of California. Ages at the time of treatment were from 9 to 69 years (mean, 33). Presenting symptoms were hemorrhage (60 patients), neurologic deficits (11), seizures (35), and headaches (40). Three years after radiation treatment the rate of complete obliteration of the lesions as detected angiographically was 100% for smaller lesions and 70% for those larger than 25 cm³. Major neurologic complications occurred in ten patients (12%). Seizures and headaches were less severe in the patients who suffered from these initially. The authors concluded that heavy charged particle radiation is effective for symptomatic surgically inaccessible intracranial AV malformations. Disadvantages of this therapy include the long delay in obliteration of the vascular lesion and a small risk of serious neurologic complications (Steinberg G K et al. N Engl J Med July 12, 1990; 323:96-101).

COMMENT: In an editorial comment by Heros R C and Korosue K of the University of Minnesota, Minneapolis, it is pointed out that the rate of serious morbidity from a hemorrhage from an arteriovenous malformation is about 30% and the mortality rate is about 10%. Not to treat is an unattractive choice for younger patients who will remain at risk for the rest of their lives. In considering the results of radiation therapy for AV malformations, the morbidity and mortality resulting from hemorrhage after treatment must be considered, particularly in comparing irradiation with surgical excision, a treatment method which eliminates the risk of hemorrhage. The delayed adverse effects of radiation on nervous tissue may limit this form of therapy in children. Only patients with previous hemorrhage, severe neurologic deficits, uncontrolled seizures, or disabling headaches were accepted in the author's protocol which included mainly adults.

PERIPHERAL NERVE DISEASES

PLASMAPHERESIS IN CHILDHOOD GUILLAIN-BARRE SYNDROME

The role of plasmapheresis in childhood Guillain-Barre syndrome was examined by retrospective analysis of children admitted to the Children's Hospital of Philadelphia, University of Pennsylvania School of Medicine, Philadelphia, PA. Of 23 patients included in the study nine had been treated with plasmapheresis and 14 served as control subjects. Therapeutic plasma exchanges were performed on an alternate day schedule. The mean age was 8.8 years and the duration of the illness prior to admission was 5.9 days. The plasmapheresis treated group recovered to the stage of independent ambulation significantly faster than the control group, 24 versus 60 days, respectively. By six months after discharge all children in both groups were ambulating independently. Plasmapheresis diminished morbidity by shortening the interval until recovery of independent ambulation, but this treatment