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INTRACRANIAL NEOPLASMS AND A-V MALFORMATIONS

MENINGIOMAS IN CHILDHOOD

The clinical presentation and pathological characteristics of 18 meningiomas among 240 surgically verified intracranial space-occupying lesions in children are reported from the Depts of Neurosurgery and Pathology, National Inst of Mental Health, Bangalore and Nizam's Inst of Med Sciences, Panjagutta, Hyderabad, India. The tumor location was supratentorial in 15, infratentorial in 2, and intraorbital in 1 patient. The majority presented between 11-15 years of age and the sexes were equally affected. The most common presenting symptoms were headache in 11 and vomiting in 4 patients; hemiparesis, deteriorating vision and seizures were early manifestations in 3, 3 and 2 patients, respectively. The duration of symptoms before diagnosis was less than 1 month in 50% patients. The meningiomas were large, 4 showed sarcomatous change, 6 were cystic, and one recurred, requiring 3 operations. Two patients died postoperatively. (Kolluri VRS, Reddy DR et al. Meningiomas in childhood. Child's Nerv Syst 1987;3(5):271-273).

COMMENT. Meningioma is an uncommon intracranial tumor of childhood, accounting for less than 5% in previous reports and 7.5% in the above study. In some larger series, the incidence is quoted at 0.4-1.5%. Contrast CT is usually superior to MRI in radiologic diagnosis (Zimmerman RD et al. AJNR 1985;6:149). CT can distinguish orbital meningioma from optic nerve glioma in about 75% of cases. In the remainder, angiography shows a tumor blush with meningioma, a finding that is absent with optic nerve glioma (Jakobiec FA et al. Ophthalmology 1984;91:137).

Proptosis was an early presenting sign of orbital meningioma in the present study, leading to prompt diagnosis, whereas this manifestation was late in appearance in the following case-report of multiple meningiomas.

MULTIPLE MENINGIOMAS IN A CHILD

The unusual case of a 4-yr, 5-mo-old boy with multiple meningiomas without neurofibromatosis is reported from the Children's Memorial Hospital and Northwestern University Medical School, Chicago, Illinois. The boy had presented with an external deviation, severe loss of vision and retinal "scar"

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in the right eye at 2 yrs of age and a history of progressive painless proptosis of the same eye for one year. At the time of diagnosis, a complete oculomotor nerve palsy with dilated pupil unreactive to direct and consensual light stimulation, ptosis, and a visual acuity of 20/300 were present on the right. Intracranial, intraorbital, and asymptomatic epidural cervical spine meningiomas were demonstrated by CT and MRI scans and by angiography. All tumors were surgically excised successfully. (Tomita T et al. Multiple meningiomas in a child. Surg Neurol Feb 1988;29:131-6).

COMMENT. Proptosis was a late sign of the intraorbital meningioma in this case, preceded by external deviation of the eye and loss of vision for a period of 18 mos. Meningiomas may involve the nasal wall of the orbit and invade the olfactory groove as in the present case, or may originate at the sphenoidal ridge and cause the classical syndrome of unilateral anosmia, optic atrophy or papilledema, and exophthalmos. With extension of the tumor to the cribriform plate, the anosmia can be bilateral and more readily detected even in children.

The young child is unlikely to react to an oil of wintergreen or toothpaste odor test. However, taste is partially perceived through the olfactory system, and the mother may have noticed a loss of appetite sufficient to excite an index of suspicion of involvement of the first cranial nerve, that often neglected part of the neurologic and eye examinations. Early diagnosis and treatment of meningiomas involving the orbit and presenting with loss of vision led to improved visual acuity in 9 of 10 patients with tumors less than 3 cm diameter and in 56% of a total of 85 patients followed postoperatively. (Kadis GN et al. Surg Neurol 1979;12:367).

CHIASMATIC/HYPOTHALAMIC GLIOMAS

Twenty-four children with progressive chiasmatic/hypothalamic glioma (CHG) have been treated with actinomycin D and vincristine combination chemotherapy without radiotherapy and followed for a median of 4.3 yrs at the Children's Hospital of Philadelphia, PA. Diagnosis and treatment required a positive CT or MRI, histological confirmation in 15 without optic nerve involvement, and progressive neurological or visual deterioration. Fifteen (62.5%) are free of progression with normal IQ's, 9 showing tumor regression, and 9 have radiographic or clinical progression of the tumor within 2-6 yrs after initiation of therapy. The authors allude to intellectual and endocrinological sequelae of radiotherapy for CHG in young children and conclude that chemotherapy may significantly delay the need for radiotherapy. (Packer RJ et al. Treatment of chiasmatic/hypothalamic gliomas of childhood with chemotherapy: an update. Ann Neurol Jan 1988;23:79-85).

COMMENT. Both radiotherapy and chemotherapy carry the risk of potentially serious adverse effects. Histological confirmation of the tumor type before initiation of therapy may now be possible since thin-needle biopsy with CT guidance is an accepted low risk procedure for neurosurgeons (Szeiniak et al. Cancer 1984;54:2385).

SPINAL A-V MALFORMATION IN A NEONATE

A newborn male infant with flaccid paraplegia, without contractures, deformities or atrophy, caused by an arteriovenous malformation (AVM) of the dorsal spinal cord, is reported from the Depts Neurosurgery and Pediatrics, Hospital Infantil Nino Jesus and Dept Neuroradiology, Hospital Infantil La Paz, Madrid, Spain. Diagnosis was by lumbar puncture showing blood in the