CHILDHOOD BRAIN TUMORS

Brain tumors are the commonest solid tumors of childhood and account for 40 to 50% of all non-leukemic neoplasms in this age group. The reported incidence is 25 per 100,000 per year. In most series, 60% of childhood intracranial neoplasms are situated in the posterior fossa. Cushing’s early report in 1927 of 18 children with cerebral tumors, including a diagnostic base that was dependent upon the symptoms of headache, vomiting, and failing vision in a child found to have a large head, crackpot sign, papilledema and an unsteady gait. The only investigation available was a skull x-ray. Cushing reported an operative mortality of 30%. A dramatic improvement in the diagnosis of brain tumors in children during the 1930's was based on the introduction of ventriculography. Angiography was added in the 1940's and Isotope brain scanning in the 1950's. In the 1970's the computer assisted tomogram increased the accuracy for posterior fossa tumors approaching 98%. The operative mortality has now been reduced to 2% in most specialized centers because of earlier diagnosis, sophisticated surgical techniques, excellent pediatric anesthesia and intensive postoperative care. Secondly, methods of radiotherapy for malignant forms of pediatric brain tumors have improved the quality and length of survival. Medulloblastomas, cerebellar astrocytomas, and brain stem gliomas each accounted for 30% of posterior fossa tumors.

Supratentorial Tumors

1. Parasellar Region.

The parasellar region usually harbors about half the tumors found above the tentorium in children, craniopharyngiomas being the commonest, followed by optic gliomas. While true pituitary tumors are common in adults, they constitute less than 1% of intracranial tumors during childhood. Matson has reported craniopharyngioma to be responsible for 8% of most childhood series. These benign tumors arise as embryonic squamous cell remnants in the region of the pituitary stalk from an incompletely involuted pharyngeal duct. The majority are cystic. Evidence of visual disturbance is present in nearly all patients with craniopharyngioma, the most common visual finding being bitemporal hemianopsia. Features of raised intracranial pressure are present in 25% of patients and endocrine changes are seen in more than half the secondary sex characteristics, obesity, or diabetes insipidus. Skull x-rays show suprasellar calcification in 50 to 80% of cases.

The tumor is reached by a subfrontal or pterional approach and, after aspiration and collapse of the cyst, total surgical resection should be possible in up to 60% of cases. The risk of recurrence after CT-confirmed total resection is less than 10% in 10 years. After subtotal removal and radiotherapy there is a 20% symptomatic recurrence at 10 years. If the tumor has only been biopsied or the cyst aspirated and radiotherapy given, there is a 50% symptomatic recurrence at 10 years. Total removal of the craniopharyngioma almost invariably results in diabetes insipidus within 24 to 48 hours after surgery, but this can now be adequately controlled by nasal administration of antidiuretic hormone substitutes. In most series there is a 10% incidence of fatal delayed coma, probably due to hypothalamic dysfunction.

Optic gliomas represent 4% of all intracranial tumors. Often these are related to neurofibromatosis. Optic gliomas are pilocystic astrocytomas arising in the optic apparatus and anatomically can be described as being prechiasmatic (optic nerve) or chiasmatic. They are very slow growing, usually composed of spindle cells containing Rosenthal fibers. Optic nerve gliomas usually present produce visual symptoms, headache, and hypothalamic disturbances. One third of children with this condition have associated neurofibromatosis. Optic canal views are diagnostic if an optic canal is over 7 mm. in diameter or there is a 2 mm. difference in diameter between the two sides. If the chiasm is involved, the child should be followed by serial CT scans and radiotherapy withheld to allow for normal growth and development unless tumor extension becomes evident. One third of patients will present with a painful third nerve palsy due to tumor involving the cavernous sinus.

Tumors of the pituitary gland itself are extremely uncommon in children.
2. Cerebral Hemisphere Region

Cerebral hemisphere tumors account for 10%. These are usually astrocytomas of low grade.

3. Third Ventricular Region

The third ventricular tumors present because of obstruction to the cerebrospinal fluid pathway and often require shunting for hydrocephalus.

4. Pineal Region

Pineal region tumors are seen in approximately 10% of series. Germinomas and pinealoblastomas were the commonest. Germinomas and astrocytomas are the commonest pineal tumors in North America, accounting for approximately 30% each. They all present as cases of raised intracranial pressure due to hydrocephalus and some have Parinaud’s syndrome. A pineal tumor should be suspected if there is a pineal calcification on plane x-ray in a child under the age of 10 or if the calcification is larger than 1 cm. CT scans are often diagnostic. When a shunt is placed, cerebrospinal fluid should be sent for human chorionic gonadotrophic hormone and cytological examination. A pineal germinoma may give a positive HCG assay and the presence of malignant cells indicates that the lesion is likely to be a germinoma or pinealoblastoma. Both these tumors are difficult to remove surgically, but are extremely radio sensitive. If CSF-screening tests are negative, the tumor should be approached surgically. There are three standard approaches: The supratentorial trans-callosal exposure as described by Dandy, Poppen’s supratentorial occipital approach and the infratentorial supracerebellar approach popularized by Krause and Stein.

Metastatic Brain Lesions

Brain metastases are extremely uncommon in children, where as in adults 20% of intracranial tumors are secondary growths.

Meningeal Tumors

Meningeal tumors in children are rare. When they occur they are often malignant.

Posterior Fossa Tumors

Although the posterior fossa contains less than 1/4 of the intracranial contents, it is the site of over 60% of brain tumors in children. Hydrocephalus is an almost invariable accompaniment and main source of symptoms. These symptoms include headache, vomiting, papilledema, being the classic triad. Headache is usually worse in the morning and vomiting also tends to occur in the morning. Acute visual loss may complicate the already tragic situation.

Medulloblastomas

Medulloblastomas have a slight male predominance. Over 70% are diagnosed by age 10. The tumor arises in the roof of the 4th ventricle and histologically appears as sheets of cells with small dark nuclei in scan cytoplasm. Tumor seeding occurs to the spinal cord and above the tentorium in 10 to 15% of cases and systemic metastases are occasionally seen. In addition to headache, vomiting and papilledema, truncal ataxia without nystagmus or appendicular ataxia may also be seen. CT scan or magnetic resonance image scanning are tests of choice.

The usual plan of action is placement of ventriculoperitoneal shunt. This eliminates the risk of acute loss of
 vision. A possible disadvantage to preoperative shunting is the development of extraneural metastases, but these have been shown to be no more common in cases that have been shunted than those that have not. Another reported hazard of preliminary shunting is upward herniation by the distended posterior fossa contents. This may occur as many as 5% of cases. If it is impending, expeditious removal of the tumor can subsequently be performed. All patients are subjected to postoperative craniospinal radiotherapy. Without this all patients would be dead within one year. Collin's rule is used to predict the period of risk for recurrence, as for any embryonal tumor. This is the age of the child plus 9 months. Chemotherapy is given according to various protocols, usually for 12 months to those children with incomplete removal. There are now reports of 70% five year survival with total macroscopic removal followed by x-ray and chemotherapy.

Ependymoma

Ependymomas account for 11% of childhood tumors. These neoplasms arise from the floor of the fourth ventricle in the region of the obex. Presenting symptoms include vomiting as the earliest symptom because of the tumor's attachment to the posterior aspect of the medulla. CT scan often reveals calcium flecks within the tumor. The usual surgical attack includes preliminary shunting followed 7 to 10 days later by tumor removal and then postoperative irradiation. The prognosis is less good than for medulloblastomas with 5 year survival rate of between 25 and 50%.

Cerebellar Astrocytomas

These tumors account for 10% of pediatric brain tumors and approximately 30% of posterior fossa tumors. The tumors are usually cystic with a mural nodule, although the solid and cystic varieties are said to occur in equal frequency. Over 90% are Grade I to Grade II histologically. After shunting the entire tumor should be removed with at least 80% long term survival. Their survival, after incomplete resection, can be improved with the addition of local radiotherapy.

Brain Stem Gliomas

These are amongst the most frustrating tumors treated by neurosurgeons since the deterioration is relentless and the prognosis hopeless and very little can be done for the unfortunate child. Most die within 2 years after diagnosis; however, 20 to 35% of these patients are reported to survive several years. Hoffman reported that infants with exophytic brain stem gliomas survive substantially longer than children with the usual endophytic tumors. The survival period was significantly reduced in children whose clinical manifestations began with cranial nerve palsies and whose tumor biopsies contained mitoses. Survival times were significantly longer in children with exophytic tumors and in those who received more than 4,000 Rads. The prognosis is also improved if the biopsy specimen contains calcification or Rosenthal fibers. The presence of an early, highly focal neurological deficit may reflect the invasive and destructive nature of a rapidly growing neoplasm. If on CT scan the tumor enhances the prognosis is actually better. The prognosis may also be improved if the boundaries of x-ray therapy are based on magnetic resonance image scanning since the extent of pediatric brain stem gliomas exceeds that visible on CT scan in 50% of cases. Debated in the literature is the worthwhileness of doing a surgical biopsy in this highly critical portion of the nervous system. A single site biopsy is of value for estimating prognosis and planning treatment. Up to 50% of cases present with raised intracranial pressure due to hydrocephalus. As the tumor spreads, bilateral cranial nerve palsies appear and long tract signs can be seen in both legs. Eventually bulbar nerves become involved with difficulty in swallowing and coughing, bronchopneumonia being the usual terminal event.

Metastatic Posterior Fossa Lesions - Rare, including Wilm's Tumor.

SUMMARY: Nationally, 1200 to 1500 new cases per year are expected within the realm of pediatric brain tumors. Astrocytomas comprised the single largest group by far and there are only approximately 600 childhood cases per year nationwide. Medulloblastoma, a more malignant solid tumor in children accounts for
less than 150 new cases per year nationwide. The locations and infiltrative nature of many tumors hinders surgical debulking of the primary lesion. Toxicity associated with high dose of craniospinal irradiation prevents the use of maximally effective radiation doses. The low metabolic and mitotic rate makes many brain tumors unresponsive to both radiation therapy and chemotherapy. Certain tumors develop rapid biologic resistance to therapy. The normal blood/brain barrier limits access of many chemotherapeutic agents to tumor cells. Finally, there is no universally accepted histological nomenclature; histologies may be interpreted differently at different treatment centers, making the choice of type specific treatment difficult.