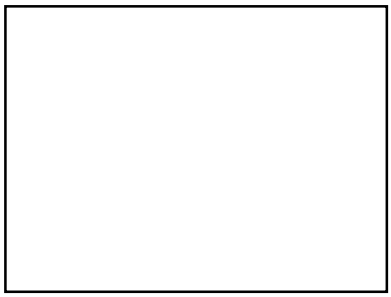
GUEST REVIEW

Scenario of Pediatric CNS Tumors in India

Raj Kumar

Primary central nervous system tumors (CNS) are the most common solid neoplasms in children. The incidence of CNS tumors is 2-5 new cases per 100000 per year, which is stable throughout the World. However; this ratio may be very variable in specific type of tumor. Twenty subtypes of tumors occur in children. Few histeotypes are relatively common, which include primitive neuroectodermal tumor (PNET) (Figure - 1,2), craniopharyngiomas, brain stem gliomas, astrocytomas, mixed gliomas, ganglioglioma, ependymomas and germ cell tumors arising in pineal gland1. In our experience, out of total 277 operated cases during last 13 years, medulloblastomas and primitive neuroectodermal tumor were maximum in numbers 86 (31% these were mainly posterior fossa tumor), gliomas (mainly astrocytomas grade I & II) 80 (29%), craniopharyngiomas 34 (12%), brain stem gliomas 29 (10%), ependymomas 22 (8%), hypothalamic gliomas 14 (5%) and germ cell tumor 12 (4%).

Although infratentorial tumors predominate in children. Overall, supratentorial tumors are more common in the first 2 to 3 years of life. Sixty percent of hemispheric tumors in children are (World Health Organization) grade 1 or 2 astrocytomas, in contrast to the predominantly grade 3 or 4 tumors that occur in adults (1). 68% out of 80 gliomas were benign in our experience. But most of the tumors were larger than 4 cm at presentation, many involved nearly half of the hemisphere. However in children with brain tumors diagnosed in the first year of life 77% to 85% of them are high grade. The most common tumor is astrocytoma accounting for 33.6% (1,2). Ependymoma is the most common nonastrocytic supratentorial neoplasm in children constituting 10% to 15% of such tumors. Supratentorial ependymomas however, account for 30% of intracranial ependymomas (1,2) (Figure - 3,4,5). Primitive neuroectodermal tumor and ganglioglioma each account for 5% to 10% of pediatric hemispheric tumors. Oligodendroglioma accounts for 4% to 8% of supratentorial tumors in children (1,2).



Fig, 1. MRI saggittal cut showing a ring enhancing cystic mass in vermis and cerebellar hemispheres suggestive of astrocytoma

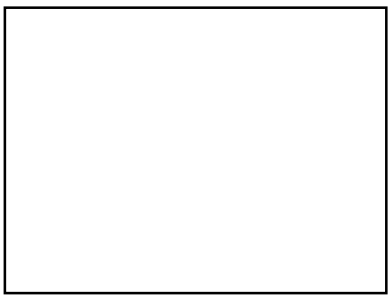


Fig. 2. Intraoperative photograph showing floor of 4th ventricle and aqueduct following total removal of same tumor.

Fig. 3. CT showing no residual tumor in posterior fossa in same child as (Fig. 1)

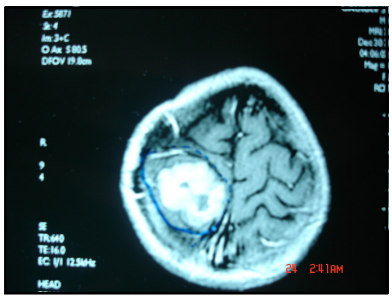


Fig. 4. MRI of a child showing a large, extensive, enhancing hyperintense mass in left temporal lobe (S/o PNET).

From the Deptt. of Neurosurgery, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Luckow (U.P.) India.
Correspondence to : Dr. Raj Kumar, Associate Professor, Department of Neurosurgery, SGPGIMS, Lucknow - 226014, U.P. (INDIA)

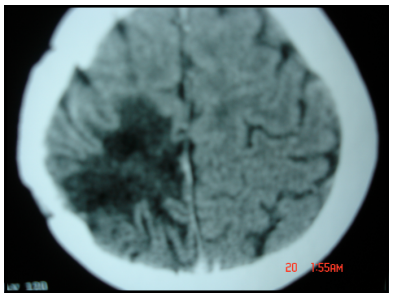
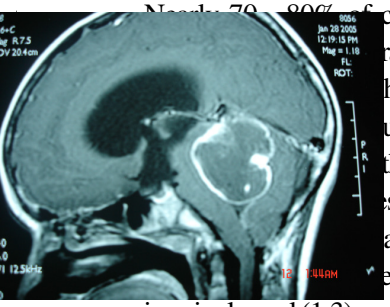


Fig. 5. Collapse of hemisphere after excision of the same tumor (Fig. 5)

Supratentorial cerebral hemispheric tumors frequently cause seizures due to irritation of the cortical gray matter. Overall 30% of patients present with seizures only. The interictal neurological examination is normal in almost all cases. Approximately 80% have a personality change or failure to thrive. Among children who present with seizures, 90% have congnitive change. Other common presenting symptoms include headache, nausea, emesis, visual disturbance and gait difficulty. Focal neurological deficits such as hemiparesis aphasia and hemianesthesia can occur, depending on the location of the tumor. Papilledema may occur. Bulging fontanelle and macrocepahly may be evident in infants (2).



Nearly 70 - 80% of children in our country present with ranial presence (along with neurological her due to delayed referral or because ums, beliefs and poor access to medical hey either present in terminal stage of s inoperable when attend the hospital. are located in supratentorial space and erior fossa. The remaining 5.1% occur in spinal canal (1,3).

Roughly 70% of cerebellar astroctomas are diagnosed in children2. These are benign tumors (1). Clinical findings include headache, vomiting and papilledema. These are usually consistent with increased intracranial pressure from fourth ventricular compression and subsequent hydrocephalus. Gait disturbance and diplopia are also common, whether from increased intracranial pressure or direct compression of the cerebellum and brainstem. These may present dramatically with coma or major brainstem dysfunction with respiratory failure and circulatory collapse. Hydrocephalus is evident at presentation in 60% to 93% of children (1,2,3). In contrast to Western literature we observe hydrocephalus in most of children (about 95%) with posterior fossa astrocytomas, because these tumor attain a large size to obstruct the 4th ventricle at presentation.

Ten year survival rates for patients with gross total resection of tumor ranged from 52% to 100%, survival of more than 88% at 10 years after gross total resection in known (1). Cerebellar astrocytomas presenting to us are significantly large tumor rendering total excision difficult. Total or near total excision is possible in 65% cases only with refined hand at higher centers. In addition to it, an additional CSF diversion also becomes inevitable in form of ventriculoperitoneal shunt in 35% cases. Shunt CSF diversion can be avoided in most of these cases if CSF pathway is re-established following total excision of tumor otherwise. Sometimes they even fill more than 2/3 of posterior fossa with brain stem invasion (rare otherwise) and gross hydrocephalus in our country. Total excision becomes a difficult task in such cases even at many good centers (Fig. 6).

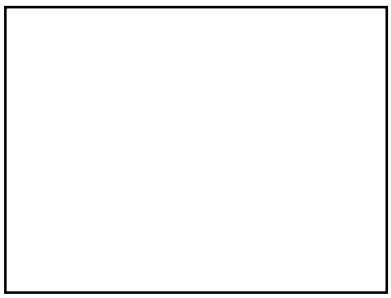


Fig. 6. CECT of a child with cystic right frontoparietal mass showing ring enhancement (s/o ependymomas).

Medulloblastoma is one of the most common and highly malignant tumors of the posterior fossa constituting 20% to 25% of all pediatric brain tumors (4). Which remains a common tumor of posterior fossa in our experience also (4). Medulloblastomas are relatively well differentiated from the surrounding brain. Gross total resection of tumor has high incidence of prolong survival. There was a marked difference in survival rates between the good and poor risk groups, with 5 year survivals of 60% to 85% (1,2,4). Poor risk group includes, (a) an age below 5 years (b) dissemination of disease (c) incomplete excision of tumor. The 5 year survival in poor risk range from 35% to 50%. About 40% of our children present either

with brain stem invasion or with a very large size tumor and total excision becomes impossible with un-experienced hands at many centers. Three year survival in our country hence does not exceed 50% with all available treatment modalities (in contrary to 60% at 5 years in Western literature). Few children present with subfrontal masses and spinal dissemination at follow-up, following surgical treatment and radiotherapy (4).

Craniopharyngiomas are the most common tumors of disordered embryogenesis. They are benign histologically but difficult to remove because of their location and their adhesion to vital vascular and neural structures (2). Almost 93% of children have signs of growth failure. Children often, present with high degree of visual loss, half of patients present with headache, which is often related with an associated hydrocephalus (2). Six of 24 children of craniopharyngiomas presented with either total blindness or near blindness because of ignorance or delayed referral due to miss diagnosis in our experience. While 13 of 24 were so large tumors to indent the floor of 3rd ventricle causing obstructive hydrocephalus. Total removal of craniopharyngiomas is the most desirable operative outcome and that it offers the best chance of cure (2). Only 11 of 24 cases could be excised in toto on account of giant sized tumor becoming inoperable in our experience.

Optic pathway gliomas represent 3% to 6% of all pediatric brain tumors and occur with equal frequency in males and females (5). The association between these tumors and neurofibromatosis type 1 is well known 20% to 50% of patients with optic pathway gliomas have neurofibromatosis (5). Long-term survival and cure following excision of a unilateral optic nerve glioma approach 100%. In case with involvement of the chiasm and hypothalamus, 10 year progression free survival has been poor, however, despite a high incidence of recurrent disease, actuarial survival is greater than 85% (5). Nearly half of our children had indolent course at average follow-up of 3 years following surgery.

Brainstem Gliomas (BSGs) occur in the midbrain, pons and medulla. Approximately 75% of BSGs occur in children (2). Diffuse Brainstem Gliomas are highly malignant tumors and cause cerebellar dysfunction in (87%), cranial nerve palsies (77%) and paresis (53%) because of tumor infiltration into nuclei and tracts (2).

Focal tumors: Focal tumors are of limited size, well circumscribed, they may be partially cystic. Dorsally exophytic tumors appear as intra fourth ventricle tumors (2). Focal tumors are relatively benign in comparison to diffuse brain stem gliomas.

Seventy seven percentage of all brain stem gliomas operated by us were pilocytic astrocytomas (benign histology), while rest were malignant. 33% of all brain stem gliomas were exophytic in our experience. Diffuse gliomas were only 3.7%, though these tumors are non-operable, hence reflecting inadequate and low incidence than actual higher incidence reported in literature. The mean duration of follow-up of our cases of brain stem glioma remained 27 months.

Other than these tumors, tuberculomas and neurocysticercosis are frequently common to present with tumor like features in children of developing countries like ours.

In conclusion primitive neuroectodermal tumor (including meduloblastoma) and gliomas are the most common tumors in our children. The children presenting to us come very late in course of their illness, when tumors attain very large size to become inoperable at times. Long term survival is relatively less in our children in comparison to Western World, despite successful treatment.

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