ELSEVIER
SAUNDERS

PEDIATRIC CLINICS
OF NORTH AMERICA

Pediatr Clin N Am 55 (2008) 121–145

Central Nervous System Tumors

Roger J. Packer, MD[a,b,c,d,*],
Tobey MacDonald, MD[c,d,e], Gilbert Vezina, MD[c,d,f]

[a]*Center for Neuroscience and Behavioral Medicine, Children's National Medical Center, 111 Michigan Avenue, Washington, DC 20010, USA*
[b]*Division of Neurology, Children's National Medical Center, 111 Michigan Avenue, Washington, DC 20010, USA*
[c]*The Brain Tumor Institute, Children's National Medical Center, Children's National Medical Center, 111 Michigan Avenue, Washington, DC 20010, USA*
[d]*The George Washington University School of Medicine and Health Sciences, Washington, DC, USA*
[e]*Division of Oncology, Center for Cancer and Blood Disorders, Center for Cancer and Immunology Research, Children's National Medical Center, 111 Michigan Avenue NW, Washington, DC 20010, USA*
[f]*Department of Radiology, Children's National Medical Center, 111 Michigan Avenue NW, Washington, DC 20010, USA*

Central nervous system (CNS) tumors comprise 15% to 20% of all malignancies occurring in childhood and adolescence [1]. Despite being relatively common, they only occur in between 2500 to 3500 children in the United States each year and may present in a myriad of ways, often delaying diagnosis. Symptoms and signs depend on the growth rate of the tumor, its location in the central nervous system (CNS), and the age of the child. Childhood brain tumors demonstrate greater histological variation, are more likely to be disseminated at the time of diagnosis, and more frequently are embryonal than those arising in adults [1].

The etiology for most childhood brain and spinal cord tumors is unknown. Specific syndromes are associated with a higher incidence of tumors [2]. Patients who have neurofibromatosis type 1 (NF-1) have a higher incidence of low-grade gliomas, including visual pathway gliomas and other types of CNS tumors [3]. Children who have tuberous sclerosis are prone to harbor giant-cell astrocytomas [4], and those who have the Li-Fraumeni syndrome have an increased predisposition to various different tumors including

* Corresponding author. Department of Neurology, Children's National Medical Center, 111 Michigan Avenue, NW, Washington, DC 20010.
E-mail address: rpacker@cnmc.org (R.J. Packer).

doi:10.1016/j.pcl.2007.10.010

gliomas [5]. Rarer conditions, such as the autosomally dominant inherited nevoid basal cell carcinoma syndrome (Gorlin syndrome) and the recessively inherited Turcot's syndrome (germ line mutation of the adenomatosis polyposis coli gene) are associated with an increased incidence of medulloblastoma [6,7]. Exposure to radiation therapy has been the only environmental factor consistently related to the development of brain tumors [8].

Presentation

Approximately one-half of all childhood brain tumors arise in the posterior fossa (Table 1) [1]. The five major tumor types that arise subtentorially may present with focal neurologic deficits, but those filling the fourth ventricle are as likely to come to clinical attention because of obstruction of cerebrospinal fluid with associated hydrocephalus. The classical triad associated with increased intracranial pressure of morning headaches, nausea, and vomiting, may occur, but nonspecific headaches are more frequent. In infants, cerebrospinal fluid obstruction with dilatation of the third ventricle and the resultant tectal pressure causes paresis of upgaze may result in downward deviation of the eyes, the setting sun sign.

The suprasellar and pineal regions are relatively frequent sites for supratentorial childhood brain tumors [1,9]. Tumors in the suprasellar region, primarily craniopharyngiomas, visual pathway gliomas, and germinomas, may present with complex visual findings including unilateral or bilateral decreased visual acuity and hard to characterize visual field loss, as well as hormonal dysfunction. In the pineal region, various different tumor types may occur in the pediatric years, including germinomas, mixed germ cell tumors, pineoblastomas, and lower-grade pineocytomas. Pineal region lesions characteristically cause compression or destruction of the tectal region of the brain stem, and result in Parinaud's syndrome, manifested by paralysis or paresis of upgaze, retraction or convergence nystagmus, pupils that react better to accommodation than light, and lid retraction. Most cortical childhood tumors are gliomas, usually low-grade, but they are anaplastic in approximately 20% of cases. Other tumor types (supratentorial primitive neuroectodermal tumors and ependymomas) may occur. Unlike the situation in adulthood, pediatric low-grade gliomas do not mutate frequently to higher-grade gliomas during childhood. In younger children, large benign supratentorial lesions, such as diffuse infantile gangliogliomas/gliomas and dysembryoplastic neuroepithelial tumors may be misdiagnosed as more aggressive lesions.

Diagnosis

The diagnosis of pediatric brain and spinal cord tumors has been simplified by advances in neuroimaging [10]. Because of the speed and availability of CT, it is often the first imaging technique obtained for children with