Abstract:
Central nervous system tumors are the most common solid malignancy in childhood and the leading cause of cancer-related mortality in children. With an average of 13 new patients being diagnosed daily with a central nervous system tumor, many of these patients will present to the emergency department for evaluation and management. Furthermore, many children experience complications and/or adverse effects directly from the tumor itself, from multimodality treatment (ie, surgery, radiation therapy, and/or chemotherapy), and/or as long-term survivors. Oncologic emergencies include increased intracranial pressure, spinal cord compression, seizures/status epilepticus, central diabetes insipidus, and adrenal insufficiency. Evaluation and management by emergency medicine practitioners are critically important for these patients in all phases of their diagnosis and treatment and can contribute significantly to an improved prognosis.

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*Department of Pediatrics; †Division of Hematology, Oncology, Neuro-Oncology and Stem Cell Transplantation; ‡Division of Neurology; §Division of Endocrinology;

Pediatric Central Nervous System Tumor Diagnosis, Complications, and Emergencies

Alicia Lenzen, MD*†¶,
Rebecca M. Garcia Sosa, MD*‡¶,
Reema Habiby, MD*§¶,
Arthur J. DiPatri Jr, MD‖¶,
Natasha Pillay Smiley, DO*†¶

Central nervous system (CNS) tumors are the most common solid malignancy and the second most frequent malignancy of childhood.[1] There are approximately 3500 new diagnoses per year in the United States, with the majority of these patients presenting to the emergency department (ED).[2] The clinical presentation of children with CNS tumors depends on tumor location, tumor type, and the age of the child. Signs and symptoms are related to obstruction of cerebrospinal fluid (CSF) leading to the oncologic emergency of increased intracranial pressure (ICP), or caused by focal brain dysfunction.[3] Children undergoing treatment for known CNS tumors present to the ED throughout their treatment; they can present as a result of complications and/or adverse effects from surgery, radiation therapy, and/or chemotherapy. Emergency medicine practitioners are critical in both the diagnosis and management of children with CNS tumors and the numerous complications and/or emergencies they are susceptible to. In this review, we discuss a practical overview of childhood CNS tumors, including diagnosis, complications from both the tumor and the treatment, as well as long-term emergencies in survivors of CNS tumors.

||Division of Neurosurgery; ¶Ann & Robert H. Lurie Children's Hospital of Chicago, Northwestern University Feinberg School of Medicine.
Reprint requests and correspondence: Alicia Lenzen, MD, Ann & Robert H. Lurie Children's Hospital of Chicago, 225 E Chicago Ave, Box 30, Chicago, IL 60611.
alenzen@luriechildrens.org
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ED DIAGNOSTIC EVALUATION

With an average of 13 new patients being diagnosed daily with a CNS tumor, many of these patients will present to the ED for diagnostic workup.[1] At the onset of illness, there are a variety of neurological and systemic symptoms, and there is no single clinical finding that is pathognomonic for the diagnosis of a childhood CNS tumor.[4] Additionally, in young children, the diagnosis is not always straightforward, as the symptoms are similar to those of more common illnesses such as gastrointestinal disorders with nausea and vomiting.[3]

Signs and symptoms may reflect the site of tumor origin, the direct result of tumor infiltration into the adjacent brain/spinal cord, and/or a consequence of CSF obstruction leading to increased ICP (Table 1).[4,5] The most common presentations in the ED include progressively worsening headache, nausea/emesis, signs/symptoms of hydrocephalus, and gait disturbances.[6] Other signs/symptoms include seizure, vision changes, papilledema, ptosis, cranial nerve deficits, behavior/school performance changes, altered mental status, back/neck pain, facial asymmetry, sensory deficits, focal motor weakness, macrocephaly, loss of developmental milestones, and/or anorexia.[3,6]

The evaluation of a patient suspected of having a CNS tumor is an emergency and requires complete history, physical examination, laboratory assessment, and neuroimaging. Key components of a physical examination should include head circumference assessing for macrocephaly; ocular examination assessing for papilledema; skin examination looking for any café au lait macules or neurofibromas associated with neurofibromatosis 1 (NF1); and complete neurological examination assessing mental status, cranial nerves, motor and sensory functions, deep tendon reflexes, coordination, and gait. A formal neurologic examination should not be deferred because the child is considered too weak, in too much pain, or uncooperative.[4] Patients with CNS tumors are at risk for bleeding disturbances and electrolyte abnormalities. Therefore, baseline laboratory evaluation should include complete blood cell count (CBC), coagulation studies (prothrombin time/partial thromboplastin time), and basic metabolic profile. For neuroimaging, the quickest and easiest imaging study obtained in the ED is computed tomography (CT), although, recently, modified rapid magnetic resonance imaging (MRI) techniques offer an excellent alternative with no ionizing radiation. Once a mass is recognized, further evaluation should be done by MRI. Differing MRI signals and sequences accentuate varying properties; T1-weighted signal is used for anatomic imaging and evaluation of blood products, T2-weighted is best to evaluate edema, and fluid-attenuated inversion recovery (FLAIR) sequence improves the conspicuity of edema (Figure 1).[7]

CNS ONCOLOGIC EMERGENCIES

Increased Intracranial Pressure

Many children with CNS tumors present with increased ICP as the tumor blocks the flow of CSF, usually at the level of the third or fourth ventricle (obstructive hydrocephalus), or compresses the cerebellum and brainstem, forcing it through the foramen magnum (Figure 2).[4,8] Signs and symptoms of increased ICP vary with age. In infants and toddlers, patients may present with changes in personality, irritability, emesis, lethargy, head banging, developmental delay or loss of previously acquired developmental skills, failure to thrive, widening of the sutures, or increasing head circumference.[4,8] In older children, presenting symptoms include early morning headache and emesis, lethargy, vision changes (blurry vision, diplopia, intermittent loss of vision), and/or altered mental status.[8] In extreme cases, severely increased ICP can lead to Cushing triad (abnormal respirations, bradycardia, and hypertension).

If increased ICP is suspected in a clinically stable patient, initial management includes neuroimaging (noncontrast CT or rapid MRI) and notifying a neurosurgeon to assist in further management. If there are signs of markedly increased ICP (unequal pupils, bradycardia, hypertension, deteriorating levels of consciousness), it is critical to use all available modalities as quickly as possible (Table 2). These patients should be intubated immediately and hyperventilated to maintain a pCO_2 of 30-35 mm Hg, pO_2 80-100, and pH 7.3-7.5.[8] If a mass lesion is suspected,