Neuro-oncological Disorders of the Cerebellum

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KEYWORDS

• Brain tumors • Magnetic resonance imaging (MRI) • Neuro-oncology • Cerebellum • Neuroimaging • Paraneoplastic syndrome

KEY POINTS

- Nonspecific symptoms of cerebellar disease, such as dystaxia, vertigo, nausea, headaches, and loss of developmental milestones, should prompt the neurologist to screen for posterior fossa tumors, especially in children.
- The most common pediatric cerebellar neoplasms are pilocytic astrocytomas, medulloblastomas, or ependymomas, whereas in adults, metastatic disease is the most common.
- The role of conventional neuroimaging in the diagnosis of brain tumors is essential regarding localization, characterization, and establishing a treatment plan.
- Newer magnetic resonance imaging techniques, such as susceptibility-weighted imaging, diffusion-weighted imaging/apparent diffusion coefficient, diffusion tensor imaging, and perfusion-weighted imaging provide additional presurgically important information.

INTRODUCTION

After leukemia, central nervous system tumors are the second most common neoplasm in children, making up the most common solid neoplasms in children. They account for the greatest mortality from cancer in children.[1] These tumors occur with an annual incidence of approximately 3 per 100,000. Supratentorial tumors are more common in infants and children up to the age of 3 years and after the age of 10 years; from 4 to 10 years of age, infratentorial tumors predominate.[2] In contrast to children, intra-axial posterior fossa tumors in adults are rare, and most commonly consist of metastasis from an extracranial site. Most posterior fossa tumors in adult patients are extra-axial and include schwannomas, meningiomas, epidermoid, and metastatic leptomeningeal disease. This article provides an overview of the intra-axial tumors that affect the cerebellum, which can be categorized by location and age (**Table 1**). For each tumor, we review conventional neuroimaging findings and discuss the value of

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Table 1
Cerebellar tumors based on age

Tumor	Age
Ependymoma	2–4 y
Classic medulloblastoma	Usually <10 y of age
Choroid plexus papilloma	18–25 y for 4th ventricle choroid plexus papilloma
Central nervous system lymphoma	Immunocompromised <40 Immunocompetent >60
Pilocytic astrocytoma	1st 2 decades of life
Hemangioblastoma	Sporadic: 5th and 6th decades of life Associated with von Hippel-Lindau disease: 3rd or 4th decade of life
Glioblastoma	Primarily in elderly patients
Lhermitte-Duclos disease	3rd and 4th decades of life
Metastases	Usually >50 y

more advanced neuroimaging techniques. Current management strategies are also briefly discussed. Finally, cerebellar paraneoplastic disorders, are discussed.

The clinical presentation is often dictated by the age of the patient and the location of the tumor. Brain tumors in infants often have relatively nonspecific symptoms initially, which may include irritability, listlessness, vomiting, failure to thrive, loss of developmental milestones, and progressive macrocephaly.[3,4] Although older children are more likely to manifest localizing neurologic findings, these are by no means uniformly present. Instead, many patients have recurrent bouts of headache, nausea, and vomiting, often occurring early in the morning, along with insidiously progressive ataxia. This symptom complex suggests the presence of increased intracranial pressure and is particularly common for posterior fossa tumors.[5–7] In some benign tumors, such as cerebellar pilocytic astrocytomas (PA), ataxia may be the primary symptom, and parents will sometimes comment that the child has been "clumsy" for years before the initial detection of a brain tumor.[6]

The current standard of neuroimaging for brain tumor evaluation is anatomic magnetic resonance imaging (MRI) with gadolinium-based intravenous contrast agent, providing highly sensitive tumor detection and characterization far superior to any other imaging modality. MRI has been found to be more sensitive than computed tomography (CT) in the detection of asymptomatic progression of disease. However, conventional MRI suffers from nonspecificity with respect to vastly different pathologic processes that appear to be similar on imaging. In the past decade, the development and application of various functional imaging techniques have increased, such as proton MR spectroscopy, diffusion-weighted (**Fig. 1**) and perfusion-weighted MRI (DWI/PWI). These advances, along with refinements in surgical techniques and adjuvant management approaches, have contributed to marked improvements in the outcomes of brain tumors. **Table 2** summarizes the characteristic findings of the most common cerebellar tumors.[8]

MEDULLOBLASTOMA

Medulloblastoma (MB) is the most common malignant central nervous system (CNS) tumor in children. All MBs, regardless of the subtype, are designated as World Health Organization (WHO) grade IV neoplasms. MB usually occurs more frequently in male individuals and usually before 10 years of age. Although uncommon, it can occur in