Supratentorial Tumors in Pediatric Patients

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KEYWORDS

• Astrocytoma • Brain tumors • Desmoplastic infantile tumors • Ependymoma • Glioma • Neuroepithelial tumors • Supratentorial • Embryonal tumors

KEY POINTS

- Anaplastic transformation of diffuse astrocytomas is a much less common event in children compared with adults.
- Both subependymal nodules and subependymal giant cell tumors can show contrast enhancement.
- Contrast enhancement and calcifications in pediatric oligodendrogliomas are less common than in adults.
- Almost all gangliogliomas are low grade and present as cystic and solid mass lesions, most frequently arising in the temporal lobes.
- Up to one-third of dysembryoplastic neuroepithelial tumors may show contrast enhancement that may be nodular or ring-like.

INTRODUCTION

Brain and central nervous system (CNS) tumors continue to represent a significant source of morbidity and mortality in the pediatric population. They are the most common solid tumors in children between 0 to 14 years of age, and their incidence is highest during the first year of life. These tumors account for the most cancer-related deaths in the 0 to 14 age group according to the Central Brain Tumor Registry of the United States (CBTRUS).[1] Overall, most brain tumors in children are gliomas, with roughly half of them consisting of pilocytic astrocytomas or other low-grade neoplasms, followed by embryonal tumors. Approximately 21% of all gliomas have a high-grade histology[1] and are associated with an aggressive clinical behavior and a dismal prognosis.[2] When brain stem tumors are excluded, high-grade gliomas are most commonly supratentorial, occurring in the cerebral hemispheres, followed by central gray matter structures.[2]

Fifteen percent of all CNS neoplasms are embryonal tumors,[1] a heterogeneous group of lesions that arise from undifferentiated small round cells, tend to occur in small children, and are associated with a poor prognosis and

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a tendency to disseminate throughout the neuraxis.[3] With the exception of medulloblastomas, embryonal tumors are predominantly supratentorial. Finally, although neuronal and mixed neuronal-glial tumors are not as common, accounting for less than 5% of all neoplasms,[1] they may nonetheless lead to significant morbidity in many patients due to intractable seizures. Many of these lesions share similar clinical and imaging presentations making their prospective diagnosis challenging. This article reviews the neuroimaging characteristics of these entities with particular attention to relevant features that may aid in narrowing the differential diagnosis, including demographics and clinical presentation.

GLIAL CELL TUMORS

Low-Grade Gliomas

World Health Organization (WHO) grade 1 and 2 gliomas roughly account for 60% of all gliomas in children.[1] They are considered benign and usually follow a relatively indolent course with an overall 10-year survival exceeding 80%.[4] However, these tumors may be associated with significant morbidity and even mortality with increasingly recognized leptomeningeal spread in pilocytic astrocytomas and malignant transformation in diffuse astrocytomas, although the latter is less commonly seen than in adults.[4,5]

Pilocytic astrocytoma

Pilocytic astrocytomas account for one-third of all gliomas in children from 0 to 14 years of age and constitute the most common primary brain tumor in this population.[1] Their incidence is relatively evenly distributed across this age group after the first year of life.[1] They are histologically benign (WHO grade I) and demonstrate slow growth over time. Pilocytic astrocytomas have an excellent prognosis, with survival rates as high as 95% at 10 years.[6] They most commonly occur in the cerebellar hemispheres (about two-thirds of lesions in pediatric patients), followed by optic chiasm and nerves and hypothalamus, but they can rarely develop in the cerebral hemispheres (particularly in older children and adults, accounting for half of all tumors in the latter group).[6,7] Most pilocytic astrocytomas are sporadic, but there is a higher incidence in neurofibromatosis type 1, where they occur in up to 20% of patients.[8] Notably, approximately one-third of patients with an optic pathway glioma (the majority of which are pilocytic) have neurofibromatosis type 1.[9] Most pilocytic astrocytomas harbor a BRAF-KIAA1549 fusion gene mutation, which may be associated with improved clinical outcomes.[10,11]

Nearly all pilocytic astrocytomas are well circumscribed on imaging, and approximately two-thirds of those in the cerebellum present with the characteristic appearance of a cystic mass with an avidly enhancing mural nodule.[7,12] The cyst wall rarely enhances. In the cerebral hemispheres, the frequency of this appearance is unknown but appears to be less common than in the posterior fossa. A prior study has shown that approximately 36% of all cerebral astrocytomas present with cystic changes (**Fig. 1**).[13] Pilocytic astrocytomas may also appear as solid enhancing masses (**Fig. 2**). On occasion they may demonstrate an infiltrating pattern in the surrounding tissue and even leptomeningeal spread, which renders their distinction from high-grade tumors challenging (**Fig. 3**).[7] An additional characteristic feature is the lack of significant vasogenic edema in the surrounding parenchyma. When edema does occur, it tends to be limited in relation to the size of the tumor.[12]

Pilocytic astrocytomas are exceptional tumors in that they commonly show avid enhancement despite their benign and relatively indolent biology. They can also show an aggressive profile on magnetic resonance spectroscopy (MRS) that may be mistaken for a high-grade tumor, with increased choline, decreased N-acetylaspartate, and a lipid-lactate peak.[14] However, recent data suggest that pilocytic astrocytomas have higher lipid–lactate/creatine ratios compared with high-grade tumors.[15] The enhancing components of pilocytic astrocytomas tend to have low perfusion with decreased relative cerebral blood volumes (rCBV),[16] although nodules with increased perfusion may at times be encountered. They also show significantly higher apparent diffusion coefficient (ADC) values compared with high-grade tumors by virtue of their low cellularity.[15] Malignant transformation of pilocytic astrocytomas has been described but is an unusually rare event. Some studies suggest that this may be much more common in adults.[17,18]

Diffuse astrocytomas

Diffuse astrocytomas are low-grade tumors (WHO grade II) that are several times less common in children than pilocytic astrocytomas.[4] They can occur anywhere in the CNS, but one-third arise in the frontal or parietal lobes, which represent the most common location.[4] On MR imaging, they have relatively ill-defined margins but are homogeneously hypointense on T1- and hyperintense on T2-weighted sequences, without