Supratentorial Tumors in Pediatric Patients

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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 histology1 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
a tendency to disseminate throughout the neuraxis. 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, 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. They are considered benign and usually follow a relatively indolent course with an overall 10-year survival exceeding 80%. 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.

**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. Their incidence is relatively evenly distributed across this age group after the first year of life. 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. 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). Most pilocytic astrocytomas are sporadic, but there is a higher incidence in neurofibromatosis type 1, where they occur in up to 20% of patients. Notably, approximately one-third of patients with an optic pathway glioma (the majority of which are pilocytic) have neurofibromatosis type 1. Most pilocytic astrocytomas harbor a BRAF-KIAA1549 fusion gene mutation, which may be associated with improved clinical outcomes.

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. 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). 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). 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.

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. However, recent data suggest that pilocytic astrocytomas have higher lipid-lactate/creatinine ratios compared with high-grade tumors. The enhancing components of pilocytic astrocytomas tend to have low perfusion with decreased relative cerebral blood volumes (rCBV), 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. 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.

**Diffuse astrocytomas**

Diffuse astrocytomas are low-grade tumors (WHO grade II) that are several times less common in children than pilocytic astrocytomas. They can occur anywhere in the CNS, but one-third arise in the frontal or parietal lobes, which represent the most common location. On MR imaging, they have relatively ill-defined margins but are homogeneously hypointense on T1- and hyperintense on T2-weighted sequences, without
Fig. 1. Pilocytic astrocytoma. (A) Postcontrast T1 image demonstrates a partially cystic (arrowhead) mass with an avidly enhancing solid component (arrow) centered in the hypothalamic region and basal ganglia. (B) Coronal T1 image shows marked mass effect on the ventricular system with obstructive hydrocephalus (arrows). (C) Axial FLAIR (fluid attenuated inversion recovery) shows the mass to have minimal surrounding edema (arrowheads). Note periventricular signal due to transependymal flow/interstitial edema (wavy arrow). (D) Corresponding ADC (apparent diffusion coefficient) map shows no evidence of restricted diffusion (arrow).
contrast enhancement or restricted diffusion (Fig. 4).19 Interestingly, while in adults most diffuse low-grade astrocytomas eventually undergo anaplastic transformation, progression to a higher-grade tumor is a rare event in children and accounts for only about 10% of cases.5,20 Diffuse astrocytomas do not show significantly increased rCBV and may show elevated myoinositol on MRS.15,21

**High-Grade Gliomas**

High-grade gliomas are significantly less common in children than in adults, yet they constitute 11% of all CNS neoplasms in the pediatric population, with an estimated incidence of 0.59 per 100,000 person-years.1 Supratentorial high-grade gliomas comprise one-third of all pediatric high-grade gliomas and occur most commonly in adolescents.22 They may be related to prior radiation exposure or occur in the setting of rare syndromes such as Li Fraumeni.23 Most-high grade gliomas in children are purely astrocytic and classified as either anaplastic astrocytomas (WHO grade III) or glioblastomas (WHO grade IV), with other mixed or nonastrocytic types being rare in this population.22 Notably, evidence shows that pediatric high-grade gliomas are genetically and molecularly distinct from their adult counterparts.24

**Anaplastic astrocytoma**

Anaplastic astrocytomas represent close to 2% of all CNS tumors in children.25 They are rapidly growing and infiltrative lesions with poorly circumscribed margins and are most commonly found in the cerebral hemispheres (particularly frontal and temporal lobes), although they can occur in the deep midline structures, brain stem, or cerebellum.19,20 They do not show significant contrast enhancement, hemorrhage, or necrosis, features that are associated with glioblastomas (Fig. 5).19 ADC values of anaplastic astrocytomas are lower than those of pilocytic or diffuse astrocytomas, and they also show increased rCBV compared with lower-grade histologies (Fig. 6).15 On MRS, anaplastic astrocytomas show increased choline and decreased N-acetylaspartate, with decreased myoinositol compared with the spectra of lower-grade gliomas.15,21

**Glioblastoma**

Glioblastomas are rare in children, in whom they constitute about 3% of primary brain tumors.26 Survival is poor but better than that of adult
Fig. 3. Spinal leptomeningeal metastases from pilocytic astrocytoma. (A) Sagittal T2 image of the entire spine demonstrates areas of heterogeneity within the spinal canal and presence of a thoracic syrinx (arrow). (B) Sagittal postcontrast T1 image shows thick leptomeningeal enhancement surrounding the cord (arrows) and numerous enhancing nodules (arrowheads).
glioblastomas. Most glioblastomas occur in the frontotemporal region but can also affect other lobes or the deep gray structures. The imaging hallmark of glioblastoma is that of heterogeneous enhancement with necrosis and marked peritumoral edema (Fig. 7). The solid components show restricted diffusion with low ADC values as well as increased MR imaging perfusion and...
permeability parameters (such as rCBV and Ktrans), which may be helpful in differentiating them from low-grade gliomas or for evaluation of tumor recurrence versus treatment response.28

On MRS, in addition to decreased myoinositol, decreased N-acetylaspartate, and elevated choline, glioblastomas typically have elevated lactate due to anaerobic metabolism and...
elevated lipids due to the presence of necrosis.\textsuperscript{15} Note that most of these studies have been performed in adults due to the rarity of these tumors in children.

Subependymal giant cell tumor
Subependymal giant cell tumors (SGCTs) are slow-growing neoplasms characterized as WHO grade I. They show mixed glioneuronal lineage

Fig. 6. Anaplastic astrocytoma. (A) Axial FLAIR demonstrates a large, diffusely infiltrative and mildly heterogeneous mass involving the entire left temporal lobe (arrows). (B) Axial postcontrast T1 image shows small foci of heterogeneous enhancement within the lesion (arrows). (C) MR rCBV map shows areas of increased perfusion (arrows).
and are not pure astrocytomas. SGCTs are most commonly seen in children and adolescents with tuberous sclerosis complex (TSC), in whom they constitute the most common CNS neoplasm (5%–20% of patients). It is unusual to develop an SGCT after age 21 years if not already present, although tumors that have been diagnosed in childhood can become symptomatic later.

Fig. 7. Glioblastoma. (A) Axial T2 image shows a heterogeneous mass that is predominantly hypointense to cortex and relatively well-circumscribed (arrow). Note moderate peritumoral edema (arrowheads). (B) Coronal postcontrast T1 image heterogeneous but avid enhancement throughout the mass (arrow). (C) Axial susceptibility-weighted image shows areas of increased susceptibility within the tumor due to hemorrhage (arrows). (D) ADC map shows low signal within the lesion in keeping with restricted diffusion (arrows).
Several cases of solitary SGCTs have been described in patients without other manifestations of TSC. However, genetic testing in some isolated SGCTs has demonstrated mutations in the TSC-1 and TSC-2 genes, suggesting that at least some of these tumors may represent a forme fruste of TSC in patients without other clinical manifestations of the disease. They are supratentorial and virtually always located in a lateral ventricle near the foramen of Monro, although they may rarely occur in other locations. SGCTs appear to arise from neoplastic transformation of existing subependymal nodules, but the reason why some nodules grow and others do not is not clear. Enhancement is variable but usually avid and heterogeneous (Fig. 8). However, in and of itself, contrast enhancement is not sufficient for diagnosis, as many subependymal nodules have also been shown to enhance. Both subependymal nodules and SGCTs can calcify and hemorrhage. From a clinical standpoint, the most important factor in the evaluation of a subependymal nodule or SGCT is the development of intracranial hypertension with new papilledema or obstructive hydrocephalus, or growth over serial imaging.

Pleomorphic xanthoastrocytoma

Pleomorphic xanthoastrocytomas (PXAs) are rare tumors that account for less than 1% of all astrocytic neoplasms. They have a wide range of age at presentation, from early infancy to the ninth decade of life, with a median of 20 years at the time of diagnosis. Most are classified as WHO grade II and have a relatively favorable prognosis, with 5- and 10-year survival rates of 75% and 67%, respectively. However, between 10% and 23% display a more aggressive behavior with histologically malignant features, and prognosis seems to be worse in males and with increasing age. Anaplastic pleomorphic xanthoastrocytoma, WHO grade III, has been added to the 2016 CNS WHO as a distinct entity. Patients with such tumors have shorter survival times when compared to those with WHO grade II PXAs. Seventy percent to 80% of patients present with seizures.

The imaging features of PXAs are variable. PXAs occur most commonly in the temporal (39%), followed by the frontal (19%) and parietal (14%) lobes. They are overwhelmingly supratentorial, with only 2 cerebellar tumors out of 213 PXAs in the largest single series published to date. These tumors favor a peripheral location and may scallop the inner table of the calvarium, reflecting their slow growth. Most are heterogeneous, and the solid components show avid enhancement and may characteristically abut the meninges (Fig. 9).

Oligodendrogial tumors

The peak incidence of oligodendrogial tumors is between the fifth and sixths decades of life. They are rare in children, in whom they represent 2% to 4% of brain tumors, with the majority being low grade (WHO grade II). In contrast to their adult counterparts, in whom 1p19q codeletions are common and associated with increased chemosensitivity and improved prognosis, such alteration is rare in pediatric oligodendrogliomas. Other molecular features that have been associated with increased overall and progression-free survival in adult oligodendrogliomas, namely isocitrate dehydrogenase 1 (IDH1) mutations and methylguanine-methyltransferase (MGMT) promoter methylation, appear to have a distinct presentation in the pediatric population. IDH1 mutations, which are frequent in adult oligodendrogliomas, are notoriously rare in children, while MGMT promoter methylation appears to be as common. Oligodendrogial tumors occur most commonly in the frontal lobe, followed by the temporal and parietal lobes. Other locations such as the brain stem, cerebellopontine angle, optic nerve, and spinal cord are rare. The imaging characteristics of oligodendrogliomas are nonspecific, but they tend to be relatively well circumscribed and typically expand the cortex with variable degrees of white matter involvement. Enhancement is less common than in adult oligodendrogliomas and is heterogeneous when present. Susceptibility-weighted imaging (SWI) may be helpful to detect calcification, but this feature also appears to be less common in children (Fig. 10). As opposed to astroglial tumors, where increased perfusion correlates with tumor grade, high rCBV is often found in low-grade oligodendrogliomas. Regardless, perfusion parameters may still have a role in assessing treatment response or disease progression or possibly in predicting malignant transformation.

Ependymoma

Ependymomas constitute 10% of all primary CNS neoplasms in children. Most occur in the posterior fossa, and 40% are supratentorial, half of which are situated within the brain parenchyma. A rare subset of supratentorial ependymomas may selectively involve the cortex and is more commonly associated with seizures. They are
found most commonly in the frontal lobes, followed by the parietal lobes. It is believed that parenchymal ependymomas may arise from embryonic ependymal rests trapped during development of the cerebral hemispheres. Due to their parenchymal location, extraventricular ependymomas tend to be larger at presentation than intraventricular ones, which more commonly

Fig. 8. Subependymal giant cell tumor. (A) Axial FLAIR shows a mildly hyperintense and heterogeneous mass projecting into the left foramen of Monro (arrow). Note obstructive hydrocephalus with transependymal flow/interstitial edema (arrowhead). (B) Axial T2 shows the mass to be slightly hyperintense relative to cortex (arrow). Note small cystic changes (arrowhead). (C) Postcontrast T1 image shows avid enhancement (arrow).
result in obstructive hydrocephalus. On imaging, ependymomas are usually well circumscribed but heterogeneous tumors that show variable degrees of inhomogeneous contrast enhancement. They have a higher incidence of cysts compared with infratentorial ependymomas; about 50% show areas of calcification, and hemorrhage may occur.

Fig. 9. Pleomorphic xanthoastrocytoma. (A) Axial FLAIR shows a large cystic mass (star) in the right temporal lobe with a peripheral solid component abutting the meningeal surface (arrow). (B) Axial T2 image shows the nodule to be only slightly hyperintense relative to the cerebral cortex (arrow). (C) Axial postcontrast T1 image shows avid enhancement of the nodule.
is similar to that of ependymoblastomas and other embryonal tumors, there is suggestion that ependymomas have a higher incidence of cysts that are more often peripherally located and that their enhancement is more commonly inhomogeneous. Similar to their infratentorial counterpart, the solid components of supratentorial ependymomas show low ADC signal due to restricted diffusion.
diffusion. However, ADC values of ependymomas are usually higher than those of embryonal tumors. Perfusion is rarely performed but has been shown to be high with a slow return to baseline. MRS characteristics are nonspecific, with increased choline and reduced N-acetylaspartate, but may be useful for follow-up and determination of tumor recurrence.

Fig. 11. Ependymoma. (A) Axial FLAIR shows a relatively well-circumscribed intra-axial tumor in the right frontal and parietal lobes (arrows). (B) Axial noncontrast T1 image shows large areas of intrinsic hyperintensity related to hemorrhage (arrows). (C) Axial postcontrast T1 demonstrates superimposed enhancement of the lesion.
Angiocentric glioma

Angiocentric gliomas are now recognized as a distinct subset of glial tumors with uncertain histogenesis but with some degree of astrocytic and ependymal differentiation. Two independent case series were first described in 2005, and these lesions were listed as a new entity in the WHO classification of tumors of the CNS in

Fig. 12. Angiocentric glioma. (A) Axial T2 image shows a heterogeneous mass in the left temporal lobe (arrows). (B) Sagittal noncontrast T1 image shows a component of the lesion to be cortically based and to demonstrate intrinsic hyperintensity (arrows). (C) Corresponding postcontrast T1 image shows no evidence of superimposed enhancement.
Angiocentric gliomas are by far tumors of children and less commonly young adults, although a few cases in older patients have also been described. They are relatively indolent and slow growing (WHO grade I), and most come to attention due to longstanding or intractable seizures. Except for 1 tumor that occurred in the midbrain and was characterized pathologically as angiocentric glioma-like, all other reported cases in the literature have been located in the supratentorial brain, most commonly involving the frontal and temporal followed by parietal and occipital lobes. Angiocentric gliomas are superficial nonenhancing cortical lesions, although a few cases showing subtle to mild enhancement have been reported. Some of them may be intrinsically hyperintense on T1-weighted sequences and have a stalk-like extension to the adjacent ventricle on T2-weighted sequences, features thought to be characteristic but inconsistently present.

A recent study using MRS has found a myoinositol and/or glycine peak in an angiocentric glioma, but for the most part their spectral characteristics overlap with those of other low-grade neoplasms. Information on MR imaging diffusion features is scarce in the literature due to the paucity of published cases. However, no significant restricted diffusion should be expected due to the low-grade histology of this lesion, as shown in 1 reported case where DWI (diffusion weighted imaging) showed facilitated diffusion.

NEURONAL AND MIXED NEURONAL–GLIAL TUMORS

Neuronal and mixed neuronal–glial cell tumors are rare, representing nearly 1% of all primary brain tumors in children, with a median age of 9 years at presentation. In the pediatric population, their incidence is highest in the 10 to 14 years age group, among whom they constitute 6.5% of brain tumors. The more relevant neuronal–glial tumors will be discussed, while recognizing that various other entities may be included under the same classification.

Ganglioglioma

Gangliogliomas are composed of neoplastic neuronal elements and astrocytes. Most are relatively indolent and have a natural history that is comparable to that of pilocytic astrocytomas, with an overall survival of 98% at 7.5 years. Almost all of these tumors are

Fig. 13. Ganglioglioma. (A) Coronal T2 image shows an ill-defined hyperintense lesion involving the right insula and temporal stem (arrow). (B) Coronal postcontrast T1 shows a focus of avid enhancement within the lesion (arrow).
<table>
<thead>
<tr>
<th>Tumor</th>
<th>Key Features</th>
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<tbody>
<tr>
<td>Glial cell tumors</td>
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<tr>
<td>Pilocytic astrocytoma</td>
<td>Most common primary tumor in children Excellent prognosis Cystic with enhancing mural nodule or solid mass Lack of significant vasogenic edema</td>
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<td>Diffuse astrocytoma</td>
<td>Much less common in children than in adults Relatively ill defined without contrast enhancement Dedifferentiation rarely seen in children</td>
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<tr>
<td>Anaplastic astrocytoma</td>
<td>Poorly circumscribed margins No hemorrhage or necrosis Usually no contrast enhancement</td>
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<td>Glioblastoma</td>
<td>Rare in children Heterogeneous enhancement Necrosis and marked peritumoral edema</td>
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<td>Subependymal giant cell tumor</td>
<td>Associated with the tuberous sclerosis complex Avid enhancement Virtually always in a lateral ventricle near foramen of Monro</td>
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<tr>
<td>Pleomorphic xanthoastrocytoma</td>
<td>Almost always supratentorial Solid components show avid enhancement Peripheral location abutting meningeal surface</td>
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<tr>
<td>Oligodendroglial tumors</td>
<td>Relatively well circumscribed, expanded cortex Enhancement and calcification less common than in adults High rCBV often found in low-grade tumors</td>
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<tr>
<td>Ependymoma</td>
<td>Half of supratentorial tumors are parenchymal Higher incidence of cysts than infratentorial ones Calcifications, hemorrhage and inhomogeneous enhancement ADC values usually higher than embryonal tumors</td>
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<tr>
<td>Angiocentric glioma</td>
<td>Superficial cortical lesions T1 hyperintensity is a characteristic but infrequent feature Usually no contrast enhancement</td>
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<td>Neuronal and mixed neuronal glial tumors</td>
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<tr>
<td>Ganglioglioma</td>
<td>Most common in temporal lobes Mixed cystic and solid masses with avidly enhancing nodule Calcifications are common</td>
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<td>Desmoplastic infantile tumors</td>
<td>Very rare, typically 18 months of age or younger Predominantly cystic with solid nodules located near cortex Solid components may show low ADC values even if benign</td>
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<td>Dysembryoplastic neuroepithelial tumors</td>
<td>Cortically based, favor temporal lobes 30% associated with cortical dysplasia May have a characteristic bubbly appearance Can rarely have nodular or ring-like enhancement</td>
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<tr>
<td>Embryonal tumors</td>
<td>Usually children &lt;5 y of age Large at presentation with little surrounding edema Intense and heterogeneous contrast enhancement Low ADC values</td>
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low grade: 1 large series with 184 patients classified 93% of gangliogliomas as WHO grade I, 6% as grade II (atypical), and 1% as grade III (anaplastic), with frank glioblastoma documented in a patient who presented with recurrent disease after treatment. They occur most commonly in the temporal lobes, particularly mesial regions (79%), followed by the frontal lobes, and they may be incidentally found after temporal lobectomies in patients with intractable epilepsy (Fig. 13, Table 1). Interestingly, tumors previously diagnosed as other low-grade histologies have been reclassified as gangliogliomas after the 2000 WHO classification system was introduced, and newer and more specific immunohistochemical profiles became available (expression of CD34 and lack of MAP2 expression). Eighty-five percent of these lesions are associated with long-standing seizures.

The conventional imaging characteristics of gangliogliomas are nonspecific. Most of them will present as mixed cystic and solid masses with avidly enhancing tumor components, and calcifications are a common feature (Figs. 14 and 15). As with other lesions, ADC values tend to decrease with higher tumor grades, which may aid in their preoperative evaluation.

**Desmoplastic Infantile Tumors**

Desmoplastic infantile tumors (DITs) are classified as WHO grade I and include desmoplastic infantile astrocytomas and desmoplastic infantile gangliogliomas, with the latter featuring a neuronal component. They are rare and occur most frequently in children 18 months old or younger. Despite their benign classification and usually favorable prognosis, several cases with aggressive pathologic features have been reported.

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**Table 1**

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<table>
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<tr>
<th>Tumor</th>
<th>Key Features</th>
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<td>Atypical teratoid rhabdoid tumor</td>
<td>10% of CNS tumors in children &lt;12 mo of age</td>
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<td>Rare aggressive neoplasms</td>
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<td>Large and predominantly solid with minimal</td>
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<td></td>
<td>edema</td>
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<td></td>
<td>Common calcifications, hemorrhage, and cysts</td>
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<td></td>
<td>Moderate to marked enhancement and low ADC values</td>
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*Fig. 14. Ganglioglioma. (A) Coronal T1 inversion recovery sequence shows a partially solid and partially cystic mass centered in the mesial right temporal lobe (arrows) that results in compression of the brain stem. (B) Corresponding axial postcontrast T1 image shows avid enhancement of the solid components of the tumor (arrow) with a nonenhancing cystic component (arrowhead).*

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features and/or a malignant clinical course have been described. DITs are typically large at presentation and are frequently multilobar, most commonly involving the frontal and parietal lobes. Occasionally they may be located in the suprasellar region. DITs are heterogeneous and tend to be peripherally located, characteristically involving the cortex, leptomeninges, and dura, which may show variable degrees of thickening and enhancement. Most DITs are predominantly cystic with solid nodules that tend to be located near the cortex and which represent tumor with intermixed desmoplasia, although some may be entirely solid. The solid components are hyperdense on computed tomography (CT), iso-to hypointense on T2-weighted MR imaging, and show avid contrast enhancement (Fig. 16). Hemorrhage is rare, and peritumoral edema is variable and in many cases mild or absent. Despite their benign histology, the solid components of DITs may show restricted diffusion.

**Dysembryoplastic Neuroepithelial Tumor**

Dysembryoplastic neuroepithelial tumors (DNETs) are benign glioneuronal neoplasms (WHO grade I) most commonly seen in children and adolescents who present with intractable seizures. They have a peak incidence during the second decade of life and are more common in males. The great majority of DNETs are cortically based lesions and, while they can occur anywhere in the supratentorial brain, they preferentially arise in the temporal lobes. Less common locations include the brainstem, cerebellum, and striatum. DNETs are virtually always solitary, but rare cases of multifocal lesions have been reported. Approximately 30% are associated with adjacent cortical dysplasia. On CT, these lesions are hypodense but may be difficult to visualize unless large. They have minimal if any associated mass effect or edema. On MR imaging, they are well-circumscribed, iso-to hypointense on T1-, and hyperintense on T2-weighted sequences and may show a triangular configuration (Fig. 17). Forty percent of them are cystic, sometimes with a typical bubbly appearance, and the rest are nodular or diffuse (Fig. 18). Calcifications are seen in 40% of cases. Although most DNETs do not enhance, contrast enhancement has been described in up to one-third of cases and may be nodular or ring-like. These tumors do not show restricted diffusion, and therefore ADC values are high. Spontaneous intralesional hemorrhage may occur but is exceedingly rare. DNETs are benign tumors, but presumed malignant transformation has been reported in a few cases. Notably, contrast enhancement can develop over time in some tumors without necessarily indicating malignant transformation. MR imaging perfusion shows low rCBV and rCBF values relative to brain. MRS demonstrates a normal spectrum, but elevated myoinositol has also been reported.

**EMBRYONAL TUMORS**

**Embryonal Tumors Not Otherwise Specified**

One of the major changes in the 2016 CNS WHO classification has been the removal of primitive neuroectodermal tumors (PNETs), which accounted for 15% of all embryonal neoplasms and which are now included within the category of embryonal tumors not otherwise specified (NOS). Embryonal tumors themselves represent 15% of all CNS neoplasms in children. They are usually seen in children less than 5 years of age but can also occur in adults, who appear to have a worse prognosis. Embryonal tumors NOS comprise a heterogeneous group of aggressive malignancies (WHO grade IV) that are biologically distinct from medulloblastomas and histologically include...
neuroblastomas, ganglioneuroblastomas, ependymoblastomas, and medulloepitheliomas. They are typically large at presentation and can arise in the cerebral hemispheres (most commonly), pineal gland, brain stem, or spinal cord. On imaging, they are frequently large, well demarcated, and heterogeneous due to frequent cystic changes and calcifications.

Fig. 16. Desmoplastic infantile ganglioglioma. (A) Axial T2 image shows a large, predominantly cystic left hemispheric mass (arrows) resulting in shift of midline structures and hydrocephalus. Note the minimal amount of edema (arrowhead) for the size of the tumor. (B) Axial T2 image at a lower level shows a hypointense solid component (arrow). (C) Axial postcontrast T1 image shows avid enhancement of the solid components of the lesion (arrows).
Hemorrhage may be present but is relatively rare, and there is typically little surrounding edema (Fig. 20). As is the case with other high-grade malignancies, embryonal tumors also show restricted diffusion with low ADC values and are hyperdense on CT due to high cellularity. Contrast enhancement is generally intense and heterogeneous, and these tumors have a propensity for leptomeningeal spread. One study using dynamic susceptibility contrast MR imaging has described increased relative cerebral blood flow and perfusion parameters, features that are also seen in other high-grade lesions. While the MRS pattern of these tumors is nonspecific and generally follows that of other high grade malignancies, the presence of a taurine peak, deemed to be highly characteristic of medulloblastomas, has also been described in at least 1 supratentorial embryonal tumor previously classified as a PNET.

Atypical Teratoid Rhabdoid Tumor

These are rare but aggressive tumors with a poor prognosis that are most frequently seen in children no more than 3 years of age. They constitute less than 2% of all CNS tumors in the pediatric population but account for 10% of those seen in children under 1 year of age. They are formed by a combination of rhabdoid cells, peripheral neuroepithelial elements, and mesenchymal elements but lack the divergent cellular features of teratomas. Their hallmark genetic feature is alteration of the SMARCB1 tumor suppressor gene. By histopathology they may be confused with other embryonal tumors, including medulloblastomas, a distinction that has clinical relevance, because atypical teratoid rhabdoid tumors (ATRTs) have a much worse prognosis. According to 1 meta-analysis, roughly one-half of reported ATRTs are supratentorial, one-third infratentorial, and 7% occur in the spine. Rare cases of primarily extra-axial ATRTs have also been reported. The appearance of these tumors on imaging is nonspecific. They tend to be large and predominantly solid but heterogeneous due to the presence of hemorrhage, cysts (sometimes peripherally located), and calcifications, which are common. Most ATRTs show inhomogeneous areas of moderate to marked contrast.
enhancement and have an increased tendency for leptomeningeal spread.\textsuperscript{108,109} As opposed to glial and neuroepithelial tumors, transgression of the dura with bone invasion may be a more common phenomenon in ATRTs.\textsuperscript{111,112} As is the case with medulloblastomas and due to their high cellularity, the solid components of ATRTs show low ADC values in keeping with restricted diffusion.\textsuperscript{109,113,114} Tumors are also predominantly hypointense on T2-weighted sequences and hyperdense on CT, presumably on the same basis.\textsuperscript{108,115} Even when large, peritumoral edema in ATRTs may be mild or absent (\textbf{Fig. 21}).\textsuperscript{109} Although the imaging characteristics of medulloblastomas and ATRTs are similar, the latter may more commonly involve the cerebellopontine angle and show intratumoral hemorrhage.\textsuperscript{113} Their MRS features are nonspecific with decreased N-acetyl aspartate, elevated choline, and lipid peaks in some patients.\textsuperscript{109}

\textbf{Fig. 18.} Dysembryoplastic neuroepithelial tumor. (A) Axial T2 image shows a relatively well-demarcated, partially cystic and partially solid mass in the left parietal lobe (arrows). (B) Corresponding axial FLAIR image demonstrates minimal surrounding edema (arrowheads). (C) Axial postcontrast T1 and (D) diffusion tensor images demonstrate no evidence of enhancement or restricted diffusion, respectively.
Fig. 19. Embryonal tumor not otherwise specified. (A) Axial FLAIR shows a lesion that is partially cystic (star) and partially solid (arrow) centered in the left temporoparietal region. Note that the signal of the cyst is higher than that of cerebrospinal fluid. There is little peritumoral edema (arrowhead) for the size of the lesion, as well as obstructive hydrocephalus with transependymal flow/interstitial edema (wavy arrow). (B) Axial postcontrast T1 image demonstrates avid enhancement of the solid component of the tumor (arrow) and enhancing septations (arrowhead) within the cyst (star). (C) ADC map shows low signal from the solid component due to restricted diffusion (arrow).
**Fig. 20.** Embryonal tumor not otherwise specified. (A) Axial postcontrast T1 shows a heterogeneously enhancing mass in the left temporal lobe (arrows). (B) Axial T2 image shows multiple foci of hypointensity within the mass due to hemorrhage (arrowheads). (C) ADC map shows low signal caused by restricted diffusion.
SUMMARY

The breadth of tumors that can arise in the supratentorial brain in children is extensive. With the exception of those that result in seizures and the highly malignant histologies, supratentorial tumors may come to medical attention later as they are less commonly associated with ventricular obstruction. Although there is overlap in the imaging appearance of some of these entities, many have relatively characteristic features that in combination with the patient’s demographics and clinical presentation may aid in narrowing the differential diagnosis.

REFERENCES


Fig. 21. Atypical teratoid rhabdoid tumor. (A) Axial noncontrast CT shows a predominantly hyperdense mass (arrows) with small foci of calcification (arrowhead). (B) Axial FLAIR and (C) axial T2 images show that the mass (arrows) is relatively well circumscribed with large areas of necrosis. There is little peritumoral edema. Note shift of midline structures and obstructive hydrocephalus with transependymal flow/interstitial edema (arrowhead). (D) Corresponding ADC map shows profound hypointensity in keeping with restricted diffusion. (E) Postcontrast T1 image shows heterogeneous enhancement throughout the tumor. (F) Susceptibility-weighted image demonstrates numerous prominent vessels within the tumor due to hypervascularity and scattered puddles of hypointensity related to hemorrhage.


