Imaging of Common Adult and Pediatric Primary Brain Tumors

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The primary goals of brain tumor imaging are lesion detection, localization, delineation of extent, and characterization. This information is used to formulate an appropriate differential diagnosis that is extremely helpful for the referring neuro-surgeons and the neuro-oncologists. In addition, imaging studies play a vital role in therapy planning, such as stereotactic location for surgery or radiotherapy, and assessing response to therapy.

This article discusses the imaging characteristics of several common pediatric and adult primary central nervous system (CNS) tumors. The review will focus on the initial imaging diagnostic workup and will give a useful radiological approach based on age, localization, imaging characteristics, and relative frequency of brain tumors. Advanced magnetic resonance (MR) techniques including spectroscopy and perfusion can provide additional information of brain tumors. These techniques will be discussed separately in this supplement.

Classification

The World Health Organization (WHO) classifies primary brain tumors according to their cellular origin. The major categories include neuroepithelial tumors, tumors of the meninges, lymphoma and hematopoietic neoplasms, germ cell tumors, tumors of the cranial and paraspinal nerves, tumors of the sellar region, metastatic tumors, and cysts and tumor-like lesions. Tumors of neuroepithelial origin comprise a significant number of primary brain tumors, including astrocytomas, oligodendrogliomas, ependymomas, choroid plexus tumors, neuronal and mixed neuronal-glial tumors, pineal lesions, and embryonal tumors.

Grading of brain tumors is according to the WHO classification that assigns a grade of 1-4 from benign to malignant, taking into account the presence of nuclear changes, mitotic activity, endothelial proliferation, and necrosis. The prototypic WHO grade 1 tumor is the pilocytic astrocytoma (PA) that has a 90% 5-year survival after resection whereas glioblastoma multiforme (GBM) represents one of the most common WHO grade 4 tumors with a 4% or less survival at 5 years.

Although the final diagnosis and grading of brain tumors is determined by histologic analysis, imaging can be very helpful in the initial assessment of neoplasms and can, in many cases, help direct surgical biopsy or treatment. Imaging features associated with increasing malignancy include mass effect, vasogenic edema, enhancement, necrosis, and hemorrhage, particularly when astrocytic tumors are compared. Cerebral blood volume (CBV), which can be calculated from MR perfusion studies and choline/creatine (Cho/Cr) ratios based on MR spectroscopy, are relatively elevated in malignant tumors, secondary to increased vascularity and cell proliferation, respectively. However it is important to note that some low-grade tumors can demonstrate “aggressive features” on imaging. In children, PAs may have a malignant appearance on conventional imaging, increased metabolism on positron emission tomographic images, and increased rCBV and Cho/Cr ratios. In adults, oligodendrogliomas often have increased rCBV.

Diagnostic Approach to Intracranial Tumors Based on Frequency, Age, Location, and Imaging Characteristics

The radiologist’s ultimate goal in the initial imaging evaluation of brain tumors should be to provide an appropriate differential diagnosis that will guide future intervention and treatment. Accurate diagnosis is based not only on the radiological appearance of the tumor but also on the patient’s age and the location and relative incidence of the lesion (Table 1). Each of these factors must be taken into account.

The yearly incidence of primary brain tumors is 16.5 cases per 100,000 population or approximately 30,000-35,000 new cases per year. In comparison, intracranial metastases are much more common with as many as 170,000 cases reported each year. The overall incidence of primary tumors...
increases with age. The pediatric population (0-19 years) has a low incidence rate of approximately 4.5 cases per 100,000 population/year while the highest occurrence rate is observed in patients older than 50 years of age, accounting for over 30 cases per 100,000 population/year.

The location and histology of primary brain tumors vary with age. Although the most common location for primary neoplasms in both adults and children is the cerebral hemispheres; brain tumors in children have a special predilection for the posterior fossa. In comparison, primary tumors located in the cerebellum are unusual in adults. Therefore, in the adult population, a solitary lesion in the posterior fossa is most commonly a metastasis.

Differentiating between an intra- and extraxial location of a mass will significantly alter potential diagnoses. Although extraaxial primary tumors, such as meningiomas, are common in adults, they are very uncommon in the pediatric population. Intraventricular tumors have a very specific differential diagnosis, which can vary depending on which ventricle is involved. If a tumor is intraxial, determination of its specific location such as cortex, corticomedullary junction, or white matter lesion will alter possible diagnoses.

### Table 1 Diagnostic Approach of Primary Brain Tumors by Age, Location, and Relative Frequency

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Abbreviations: DNET, dysembryoplastic neuroepithelial tumor; S-PNET, supratentorial primitive neuroectodermal tumor; ATRT, atypical teratoid rhabdoid tumor; SEG, subependymal giant cell astrocytoma; LCH, Langerhan's cell histiocytosis; PMA, pilomyxoid astrocytoma.
Different tumor types occur at very different rates at various ages (Fig. 1). The most common tumor of childhood is the PA, followed by medulloblastoma. However, in adulthood, the most common tumor is meningioma, followed by glioblastoma. Oligodendroglioma, which is common in adults, is very rare in children. Neuronal and mixed neoglial tumors are more common in children than in adults (Table 1). Therefore, correlating the location and characteristics of a tumor with the age of the patient is crucial in determining an appropriate differential diagnosis.

**Tumors More Common in Childhood**

**Pilocytic Astrocytoma (WHO Grade 1)**

PA is the most common primary brain tumor in children. Overall, patients with PA’s have a good prognosis after resection with a 94% survival rate at 10 years. Common locations for PA’s include the cerebellum, optic nerve/chiasm/hypothalamic region, or brain stem. Involvement of the cerebral hemispheres is less frequent.

Pilocytic astrocytomas are typically well-defined lesions that classically present as a cystic mass with an enhancing mural nodule, however solid lesions do occur. Significant vasogenic edema is rare. These are slow growing tumors that often present due to localized mass effect. Lesions occurring in the cerebellum may significantly compress the fourth ventricle, making it difficult to identify the tumor’s parenchymal origin. The main differential diagnosis is between the 2 most common pediatric fourth ventricular tumors, medulloblastoma (MB), and ependymoma (Fig. 2).15,16

PA’s occurring in the optic nerve/chiasm/hypothalamic region have been separated into 3 subsets. In the pediatric population, tumors are divided into 2 types: those associated with neurofibromatosis type 1 (NF1) and those not associated with NF1. PA’s in patients with NF1 are usually bilateral nonenhancing tumors involving the optic nerves and less commonly, the optic chiasm/hypothalamic region. They have an indolent course. PA’s in non-NF1 patients usually involve the chiasm/hypothalamic region. They are typically solid/cystic enhancing tumors, which are often larger than PA’s associated with NF1 and have a less indolent course. The third subset of tumors, which is identified in adults, demonstrates more aggressive behavior.

As mentioned earlier, PA may demonstrate a false “aggressive appearance” on MR spectroscopy with significant elevation of Cho/Cr ratios. Recent studies have shown very low concentrations of Cr in PA’s compared with other pediatric tumors, as well as diminished quantities of total Cho. Therefore, the paradoxical increase in Cho/Cr ratios does not necessarily indicate increased cell proliferation.17

**Medulloblastoma/ PNET-MB (WHO Grade 4)**

MB or PNET-MB (primitive neuroectodermal tumor-medulloblastoma) is a highly cellular, rapidly growing malignant embryonal tumor. It is the second most common tumor in children but the most common posterior fossa lesion. These masses are typically located in the fourth ventricle arising...
from the superior medullary velum. Involvement of the cerebellar hemispheres is rare, usually occurring in older children and adults and typically representing desmoplastic medulloblastomas. MBs have a variable prognosis after resection with a 50% survival rate at 10 years.

MBs are typically isointense on T1 weighted images, mildly hypointense to cortex on T2 weighted images, and homogeneously enhance. The desmoplastic variant that has a better prognosis often demonstrates a different imaging pattern characterized by calcifications and subtotal heterogeneous enhancement. Diffusion restriction is commonly observed due to the tumor’s dense cellularity. Ninety percent of these lesions are dense on computed tomography (CT), which can aid in the radiographic diagnosis (Fig. 2). Approximately one-third of MB’s will have subarachnoid dissemination at the time of presentation; therefore, imaging the entire neuroaxis is extremely important to evaluate the extent of disease.

Supratentorial primitive neuroectodermal tumors (S-PNET) are a subset of tumors genetically distinct from infratentorial PNETs (PNET-MB). Though only 6% of PNETs are supratentorial, they are important hemispheric masses to consider in newborns and infants. S-PNETs are large, complex hemispheric masses with heterogeneous enhancement.
and minimal peritumoral edema.\textsuperscript{21} The differential diagnosis of these lesions includes teratoid or rhabdoid tumors and ependymomas. Unlike PNET-MB, calcifications, hemorrhage, and necrosis are common (Fig. 3). Patients with S-PNET have decreased survival compared to those with PNET-MB.

\textbf{Ependymoma (WHO Grade 2)}

Ependymomas are the fourth most common posterior fossa tumor in the pediatric population.\textsuperscript{22} They arise from ependimal cells lining the ventricles and are most commonly located in the fourth ventricle. Unlike medulloblastomas, which originate superiorly from the medullary velum, ependymomas arise from the floor of the fourth ventricle (Fig. 2). One-third of ependymomas are located supratentorially where they have a characteristic deep parietal white matter location rather than intraventricular. They are slow growing tumors with an overall 60%-70% survival rate at 5 years after resection. Infratentorial ependymomas typically present as heterogeneous enhancing lesions that often extend out through the

\textbf{Figure 3} Supratentorial primitive neuroectodermal tumor in a 1-year-old boy. (A) Axial noncontrast CT image shows a large mass centered in the trigone of the left lateral ventricle extending into the parietal and occipital brain parenchyma. There are multiple calcifications and enlargement of the left lateral ventricle. Note ventricular drainage catheter tip in the right frontal horn. (B) Axial T2 image shows a large mass with significant heterogeneous signal. Areas of dark signal predominantly in the periphery and cortical regions are consistent with calcifications/hemorrhagic staining. (C) Coronal T1 with gadolinium image shows heterogeneously enhancing mass with solid/cystic components involving the ventricle and brain parenchyma as well as extending into the dural/extracranial spaces. Involvement of multiple spaces is an important characteristic of supratentorial primitive neuroectodermal tumor (Images courtesy of Dr. Blaise Jones, MD, Cincinnati Children’s Hospital).
foramina of Luschka and Magendie into the cisterns. Punctate calcifications occur in approximately 50% of cases, therefore CT may be helpful in differentiating this tumor from MB and PA (Fig. 2). Ependymomas may also demonstrate cystic transformation, hemorrhage, necrosis, and edema.

**Choroid Plexus Papilloma/Carcinoma (WHO Grade 1 and 4)**

Choroid plexus papillomas (CPP) are intraventricular papillary neoplasms that are derived from the choroid plexus epithelium. CPP is one of the most common brain tumors in children below the age of 2 years. In the pediatric population they are located in the atrium of the lateral ventricle. In contrast, tumors in adults are typically located in the fourth ventricle and cerebellopontine angles. CPP is a slowly growing tumor with a 5-year survival rate close to 100%.

Imaging features include a frond-like lobulated mass with intense enhancement. Calcifications and hemorrhage are frequently present. Hydrocephalus is very commonly associated with CPP's and can be due to either overproduction or obstruction of cerebrospinal fluid (CSF).

Choroid plexus carcinomas (CPC) are malignant neoplasms that account for approximately 20%-30% of all choroid plexus masses. Most cases occur in children and also typically arise in the lateral ventricles. CPC's have a poor prognosis with a 50% survival rate at 5 years. Imaging findings that can help differentiate CPP from carcinomas include extension of the lesion beyond the ventricle wall, prominent vasogenic edema, and mass effect. Both CPP's and CPC's can seed the CSF, however metastatic spread is more commonly observed with carcinomas.

**Neuronal and Mixed Neural-Glial Tumors (WHO Grade 1-2)**

Dysembryoplastic neuroepithelial tumors (DNET) and gangliogliomas/gangliocytomas are the most common mixed neural-glial tumors in children and young adults. These lesions are cortical tumors often associated with refractory seizures.

DNET is a benign mixed neural-glial tumor frequently associated with a background of cortical dysplasia. They are most commonly found in the temporal lobes; however, they can arise in other areas of the cerebral hemispheres and posterior fossa. On magnetic resonance imaging (MRI), DNETs typically have a “bubbly” or multicystic appearance. Most lesions are well defined with no edema and little mass effect. Enhancement is present in up to 50% of cases and is usually nodular or ringlike in appearance. Calcification may be present. A “FLAIR (fluid attenuated inversion recovery) bright ring” sign has been described with these tumors, which is a complete or incomplete hyperintense rim that may be caused by the presence of loose peripheral neuroglial elements (Fig. 4). This sign may help differentiate these tumors from other cortical lesions.

Gangliogliomas (WHO grade 1 or 2) and gangliocytomas (WHO grade 1) are also cortically based neoplasms. Gangliogliomas are composed of both benign mixed atypical ganglion cells and neoplastic glial cells, whereas gangliocytomas are primarily composed of dysplastic or neoplastic neurons. Although both lesions are usually benign, rare cases of malignant degeneration or metastatic spread have been reported. These lesions can be either solid, cystic, or mixed. Approximately 50% will present as a cystic mass with a mural...

*Figure 4* Dysembryoplastic neuroepithelial tumor in a 10-year-old girl with seizures. (A) Axial fluid attenuated inversion recovery (FLAIR) image shows a cortical lesion in the left temporal lobe, with an incomplete medial rim of hyperintense signal. (B) Coronal T1 image with gadolinium shows subtle enhancement of the cortical lesion. Note the “bubbly appearance” with small rounded areas of hypointensity on FLAIR and T1 near the cortex (Images courtesy of Dr. Blaise Jones, MD, Cincinnati Children’s Hospital).
nodule. Most lesions demonstrate either solid or heterogeneous enhancement. Mass effect and edema are uncommon.

**Brainstem Tumors (WHO Grade 1-4)**

Most brainstem tumors are gliomas, including fibrillary astrocytomas, PAs, and rarely glioblastomas. Eighty percent of all brainstem gliomas occur in the pediatric population. They are categorized according to where they develop (pontine, medullary, and midbrain) and whether they are diffuse or focal. Prognosis varies greatly depending on the location and grade of the lesion.

Pontine gliomas are more often diffuse lesions that may present with cranial nerve palsies and ataxia. These lesions are often high-grade and have a poor long-term prognosis. Imaging features include expansion of the brain stem with abnormal T2 prolongation. Enhancement is usually absent.

Tectal gliomas typically have a distinctly benign behavior compared to other brainstem tumors. Some authors believe that these tumors are hamartomas; however, other series have histologically shown these tumors to be low-grade PAs. Due to their location adjacent to the sylvian aqueduct, these lesions often present with obstructive hydrocephalus. They usually do not require treatment beyond CSF diversion such as third ventriculostomy or shunting.

**Tumors More Common in Adulthood**

**Astrocytomas**

Gliomas are the most common primary intraaxial mass in adult population. Astrocytomas account for over half of all gliomas and can be divided into 2 major categories, infiltrative and noninfiltrative. Infiltrative astrocytomas are much more common, representing 75% of all lesions. The noninfiltrative subset includes PAs, pleomorphic xanthoastrocytomas, subependymal giant cell astrocytomas, and desmoplastic cerebral astrocytomas of childhood. This section will primarily focus on the infiltrative type.

Infiltrative astrocytomas range from low-grade lesions to highly aggressive malignant neoplasms. Grading these tumors is based on several histopathologic features including cellularity, nuclear atypia, mitotic activity, endothelial proliferation, and necrosis. As described previously several imaging features including mass effect, edema, enhancement, and necrosis can help predict tumor grade.

Low grade astrocytomas (WHO grade 2) comprise approximately 25% of all gliomas. They tend to occur in younger adults, often in the third and fourth decades of life. Low grade gliomas are best visualized on MRI and typically present as nonenhancing well demarcated or ill-defined lesions in the cerebral hemispheres. Edema is usually absent. Typically there is absent or minimal mass effect, a finding which can be striking given the size of some lesions.

Anaplastic astrocytomas (AA) (WHO grade 3) occur in a slightly older population and confer a worse prognosis. The median survival rate for these patients is 2-3 years. Most AA's result from dedifferentiation of low grade gliomas, however some arise de novo. Histologically, AA contains gemistocytes and protoplasmic elements but no necrosis. Dissemination occurs through the white matter tracts. AA's typically demonstrate edema and mass effect that helps to differentiate them from low grade gliomas. Approximately two-thirds of AA's will partially enhance.

GBM (WHO grade 4) is the most aggressive and malignant form of astrocytoma and is characterized histologically by necrosis and neovascularity. It is the second most common tumor in adults after meningioma and usually occurs in patients older than 50 years of age. Involvement of patients

![Figure 5](image-url) Low-grade astrocytoma in a 35-year-old woman who presented with seizures. (A) Axial T2 weighted image demonstrates a moderate sized area of hyperintense signal involving the white matter and cortex of the left frontal lobe. (B) Axial T1-weighted post gadolinium image demonstrates subtle hypointensity within the lesion. No abnormal enhancement is observed. Note the relative lack of mass effect despite the size of the lesion.
less than 30 years is rare. These tumors have the worst prognosis among primary brain tumors. The median postoperative survival time is 8 months, and the overall 10-year survival rate in patients older than 45 of age is less than 1.8%.

GBM’s are most commonly located in the supratentorial white matter. Involvement of the brainstem and cerebellum is rare; however, these locations are more common in children than adults (Fig. 7). On MRI, GBM’s are usually identified as large heterogeneously enhancing masses with significant necrosis, mass effect, and vasogenic edema (Fig. 8).

Hemorrhage of differing stages is often observed. Involvement of the bilateral hemispheres through the corpus callosum (“butterfly glioma”) is a classic presentation.

Surgical resection of GBM’s is nearly always incomplete due to the highly infiltrative nature of the tumor. Biopsies taken from around the margins of resection cavities have demonstrated tumor cells in the surrounding “edema” and even in normal appearing white matter. Surgical resection and radiation are frequently targeted to the enhancing (higher grade) portion of the tumor, however the presence

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**Figure 6** Anaplastic astrocytoma in a 48-year-old man with visual abnormalities and headaches. (A) Axial T2-weighted image shows a large lesion in the right medial occipitoparietal lobes. Surrounding abnormal T2 signal extends into the right parietal white matter and across the splenium of the corpus callosum. (B) Axial T1-weighted post gadolinium image demonstrates a nonenhancing, hypointense mass. Note the mass effect and midline shift that helps to differentiate this lesion from a low-grade astrocytoma. One-third of anaplastic astrocytomas will not enhance.

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**Figure 7** Glioblastoma multiforme in a 5-year-old boy with abnormal gait and vomiting. (A) Axial T2 image shows an intraaxial mass centered in the right brachium pontis with a central area of necrosis and thick peripheral solid component. (B) Axial T1 post gadolinium image shows minimal enhancement of the thick peripheral component. Cerebellar and brainstem glioblastomas are rare, however they are more common in children than adults.
of tumor cells in the surrounding tissue limits curative therapy.

**Oligodendroglioma (WHO Grade 2)**

Oligodendrogliomas are infiltrative gliomas that originate from oligodendrogial cells. They occur in middle-aged adults and often present with seizures. Pediatric involvement is rare. Oligodendrogliomas may be low grade (WHO grade 2) or anaplastic (WHO grade 3). Similar to astrocytomas, higher grade oligodendrogliomas demonstrate increased cellularity, mitotic activity, endothelial hyperplasia, and necrosis. Mixed oligodendrogliomas contain both features of astrocytomas and oligodendrogliomas, and can be classified as grade 2 or 3.

Many oligodendrogliomas demonstrate 1p and/or 19q chromosomal deletions. The presence of these chromosomal losses increases the tumor's response to radiation and chemotherapy and improves prognosis. Patients with both 1p and 19q deletions have a median survival time of 8-10 years whereas patients without the deletions have a survival time of 3-4 years.

Differentiating oligodendrogliomas from astrocytomas on conventional imaging is often not possible. However, several imaging features favor the diagnosis of oligodendroglioma, including cortical involvement, heterogeneous signal, intratumoral cysts, patchy enhancement, and the presence of calcifications (Fig. 9). Calcifications, which can be seen in

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**Figure 8** Glioblastoma multiforme in a 59-year-old man who presented with seizures and neurologic deficit. (A) Axial T2-weighted image demonstrates a mildly hyperintense mass in the lateral right frontal lobe. Note the prominent surrounding abnormal T2 signal in the adjacent white matter that represents edema and infiltrative tumor. (B) Axial T1-weighted post gadolinium image shows an irregularly enhancing mass with areas of necrosis.

**Figure 9** Oligodendroglioma in a 30-year-old man who presents with seizures. (A) Axial T2 FLAIR image shows a large, mildly heterogeneous left frontal mass involving cortex. Focal areas of hypointense signal represent calcifications that were confirmed on CT. The lesion is well-defined with no edema and mild mass effect. (B) Axial T1-weighted post gadolinium image shows patchy enhancement within the lesion.
80% of all oligodendrogliomas, are best demonstrated on CT; however, gradient imaging can increase MR’s sensitivity. Oligodendrogliomas tend to have higher relative CBVs on MR perfusion studies compared to other low-grade glial tumors that is related to a dense capillary network rather than a higher grade of tumor.\(^\text{13}\)

**Pleomorphic Xanthoastrocytoma (WHO Grade II)**

Pleomorphic xanthoastrocytoma (PXA) is a distinct type of circumscribed astrocytic tumor noted for cellular pleomorphism and xanthomatous change. Although usually classified as WHO grade 2 tumors, they can undergo malignant transformation. PXAs are slow growing lesions that typically present in the first 2 decades of life. They are usually located in the cerebral hemispheres with a higher incidence in the temporal lobes. Recurrence after resection is uncommon and the survival rate at 10 years is 70%.

Imaging features of PXAs typically include a supratentorial cortical cystic mass with an enhancing mural node. PXAs are superficial lesions that involve the leptomeninges as well as the brain parenchyma. As a result, a “dural tail” may be present that can help differentiate these neoplasms from other cortical tumors, such as DNET or ganglioglioma.\(^\text{36}\)

**Primary CNS Lymphoma**

The vast majority of intracranial lymphomas are primary lesions that involve the brain parenchyma. More than 90% are non-Hodgkin B-cell type. Secondary CNS lymphoma is rare

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**Figure 10** Primary central nervous system lymphoma in a 62-years-old immunocompetent man. (A) Axial T2-weighted, (B) T2 FLAIR, and (C) post gadolinium images demonstrate an irregular, homogeneously enhancing mass in the anterior genu of the corpus callosum extending into the frontal lobes. An additional lesion is noted in the splenium of the corpus callosum. The lesions are mildly hyperintense on T2 weighted images and are associated with moderate edema. The multiplicity of lesions, periventricular location, and homogeneous enhancement support the diagnosis of lymphoma.
and often presents with dural or leptomeningeal involvement.

Imaging features and clinical prognosis of primary CNS lymphoma vary with the patient’s immune status. In immunocompetent patients primary lymphoma typically presents as solitary or multiple predominantly solid masses in the basal ganglia and white matter, often periventricular in location. These are highly cellular tumors that give them a characteristic hyperdensity on CT and homogeneous hypointensity on T2 with strong homogeneous enhancement (Fig. 10). Diffusion restriction may be present due to the dense cellularity of the lesion. The administration of corticosteroids can modify or annul tumor enhancement, and therefore should not be administered before initial CT and MR imaging unless clinically necessary.

In immunodeficient patients, CNS lymphoma tends to present as multifocal heterogeneous/peripheral enhancing lesions with central necrosis. These lesions may be indistinguishable from infectious etiologies including toxoplasmosis. However, as in immunocompetent patients, lymphoma in patients with acquired immunodeficiency syndrome (AIDS) has a characteristic tendency to involve the ependymal surfaces and periventricular white matter.

The incidence of primary CNS lymphoma has tripled over the past 2 decades, largely due to the increase in patients with AIDS. However, the incidence of lymphoma is also rising in the immunocompetent population. No environmental or behavioral factors have been identified to account for this rate increase. In general the prognosis for CNS lymphoma is poor due to recurrent disease, however it is moderately better in immunocompetent patients. The median survival time for AIDS patients with intracranial lymphoma is only 2-6 months.

Hemangioblastoma (WHO Grade 1)

Hemangioblastomas are benign tumors of vascular etiology that represent approximately 1%-2% of all primary CNS neoplasms. Although rare, hemangioblastomas are the most common primary adult intraxial posterior fossa tumor. Over 85% of hemangioblastomas occur in the cerebellar hemispheres. Less commonly they involve the vermis, medulla, and spinal cord; rare supratentorial lesions have been reported. Males are more commonly affected than women, and most patients present within the third to fifth decades of life. Pediatric involvement is rare.

Most hemangioblastomas occur sporadically, however approximately 25% are associated with von Hippel-Lindau syndrome, an autosomal dominant disease which is also associated with retinal angiomatosis and visceral tumors involving the kidneys and adrenal glands. Surgical resection is usually curative, however patients with von Hippel-Lindau syndrome often have multiple lesions.

The most common imaging presentation of hemangioblastomas is a well-defined cyst of variable size with an intensely enhancing mural nodule (Fig. 11). Approximately 25% of lesions will present as a solid mass without a defined cyst. Prominent flow voids may be present within the solid portions of the tumor due to the vascular nature of the tumor. Uncommonly, hemangioblastomas can present as a cyst without evidence of an enhancing nodule or wall.

Meningioma (WHO Grade 1-3)

Meningiomas arise from arachnoid meningotheelial cells and are the most common benign intracranial neoplasm in adults. In general, these lesions are slow growing tumors (WHO grade 1) with a good prognosis, however atypical and anaplastic meningiomas (WHO grade 2–3) do occur. Aggressive meningiomas can invade the brain parenchyma and will rarely metastasize to distal sites.

The incidence of meningiomas is highest in middle-aged women. Meningiomas may occur in multiples, particularly when associated with neurofibromatosis type 2, and have been associated with a history of radiation treatment. The

Figure 11 Hemangioblastoma in a 43-year-old man who presented with headaches. (A, B) Axial T2-weighted and post gadolinium T1-weighted images demonstrate a large cystic mass with an enhancing mural nodule in the right cerebel-

lum. Marked compression of the fourth ventricle is present.
most common location for meningiomas is in the cerebral convexities. Less common locations include the sphenoid ridge, olfactory groove, parasellar region, cerebellopontine angle, and optic nerves. Intraventricular lesions in the adult population are rare but have a typical presentation in the atria of the lateral ventricles (Fig. 12).

Meningiomas are most commonly present as extraaxial masses with prominent homogeneous enhancement and broad dural attachments. Although the MR signal characteristics of meningiomas on precontrast sequences can vary, typically they are isointense to brain on T1 weighted images and isointense to mildly hyperintense on T2 weighted images. This is due to the homogeneous cellularity of the tumor. Tumoral calcifications and hyperostosis of the underlying skull bone are frequent. Edema is commonly observed in the adjacent brain parenchyma particularly with masses over the cerebral hemispheres. The cause of the edema is not fully understood and may be related to mechanical compression or secretions from the tumor, however there is no direct correlation between the presence of edema and the aggressiveness of the lesion or brain invasion. Lesions that can mimic meningiomas include dural metastases, lymphoma, and hemangiopericytomas.

Meningiomas in the pediatric population are rare, however when they occur they often have distinct characteristics. Childhood meningiomas have a male predilection, are more commonly intraventricular in location compared to adults.
Central Neurocytoma (WHO Grade 2)

Central neurocytoma is an intraventricular neuronal cell tumor that typically arises from the septum pellucidum or ventricular wall. Most lesions are present in the lateral ventricles, however third ventricular involvement does occur. Histologically, central neurocytomas resemble oligodendrogliomas, and until recently were considered “intraventricular oligodendrogliomas.” Neuronal characteristics observed on electron microscopy led to the reclassification of this lesion in 1982.47

Central neurocytomas occur in young adults and often present with signs of increased intracranial pressure or hydrocephalus due to their intraventricular location. The lesion typically has a benign course, however more aggressive behavior including invasion of the brain and seeding of the CSF can occur. The survival rate is approximately 81% at 5 years.

On imaging, central neurocytomas are present as heterogeneous masses often attached to the septum pellucidum (Fig. 13). Calcifications and cystic changes are common.48 Solid portions of the tumors demonstrate variable and irregular enhancement.

Subependymoma (WHO Grade 1)

Subependymoma is an uncommon, benign intraventricular tumor that typically occurs in middle-aged to older adults. The most common location for this tumor is the inferior fourth ventricle followed by the frontal horns of the lateral ventricles attached to the septum pellucidum. Subependymomas are usually asymptomatic, however obstructive hydrocephalus can occur due to the lesion’s intraventricular location.49 Patients with subependymomas have an excellent prognosis; surgical resection is curative in most cases.

Imaging features of subependymomas on CT include a hypodense lobular mass within the fourth or lateral ventricles. On MR these lesions are usually hypo-to isointense on T1 weighted images and hyperintense on T2 weighted sequences. Enhancement is typically absent (Fig. 14).

![Figure 13](image.png) Central neurocytoma in a 39-year-old man with signs of increased intracranial pressure. (A) Noncontrast CT scan demonstrates a large heterogeneous, mildly hyperdense intraventricular mass with involvement of the septum pellucidum. (B) T2-weighted sagittal image shows prominent heterogeneity of the lesion with multiple small cysts. (C) T1-weighted post gadolinium coronal image demonstrates patchy enhancement. Note the lateral ventricular dilatation caused by obstruction at the foramen of Monro.
Conclusion

Imaging is a vital step in the initial work-up of brain tumors. An appropriate differential diagnosis should be based not only on the radiographic characteristics of the tumor but also on the age of the patient, location of the lesion, and the tumor’s relative frequency. Advanced imaging techniques often can provide complementary information; however, conventional CT and MRI remain the primary tools for neuro-oncological diagnosis and early detection.

References


Figure 14 Subependymoma in an asymptomatic 33-year-old woman. (A) Axial T2 FLAIR image shows a small, well-defined homogeneous intraventricular mass arising from the septum pellucidum. (B) T1-weighted post gadolinium coronal image demonstrates no enhancement. The location and lack of enhancement are typical for this benign lesion.