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Seminars in
ULTRASOUND
CT and MRI

Intraventricular Tumors

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Intraventricular tumors represent a unique group of intracranial neoplasm separate from the classic division as intra- vs extra-axial masses. Intraventricular tumors are unique because of the diverse pathologic spectrum, including the entire gamut of neuroepithelial and non-neuroepithelial tumors. Most of these tumors are clinically benign presenting with headaches or signs and symptoms of hydrocephalus. Computed tomography and magnetic resonance imaging play a pivotal role in diagnosis and neurosurgical guidance. Though, practically any intracranial tumor can have an intraventricular location, we would be discussing the common 8 tumors, which together constitute more than 90% of intraventricular masses. Demographics, clinical and imaging findings, are together very useful in narrowing down the differential.
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Introduction

Intraventricular tumors are rare lesions that make up 0.8%-1.6% of all intracranial tumors, with the vast majority of them being benign. They are however more common in children and comprise around 16% of childhood and adolescent intracranial tumors.[1,2] These are a histologically heterogeneous group of tumors that can be divided into primary and secondary intraventricular tumors. "Primary tumors" are neoplasms that originate from the ependymal or subependymal lining, septum pellucidum, choroid plexus, and the supporting arachnoid tissue. This includes a wide variety of tumors unique to the ventricles (eg, choroid plexus papilloma [CPP]) and tumors frequently seen elsewhere (eg, meningioma).[3,4] The ventricular margins are composed of ependymal cells with subependymal glial plate giving rise to ependymomas, subependymomas, and subependymal giant cell astrocytomas (SEGAs). The septum pellucidum consists of 2 layers of both white and gray matter, giving rise to central neurocytomas (CNs), a glial neuronal tumor that is unique to the ventricular system. Choroid plexus is the most vascular structure in the ventricular system and gives rise to CPP and carcinoma, which are cerebrospinal fluid (CSf)-secreting tumors, frequently causing hydrocephalus. Highly vascular tumors such as meningiomas and metastases also generally arise from the choroid plexus secondary to the inherent high vascularity of the structure.[5] "Secondary or paraventricular tumors" is a term used when these neoplasms originate from adjacent brain substance and demonstrate more than two-thirds exophytic growth within the ventricle.[3,4]

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Diagnosis and Management

Clinical signs and symptoms of intraventricular tumor vary according to age and tumor location. Most clinical findings are secondary to CSf obstruction (hydrocephalus) and subsequent increase in intracranial pressure. Infants usually present with macrocephaly, loss of appetite, and irritability. Older children and adults present with headache and vomiting with papilledema commonly seen on examination.[6] Seizures and visual disturbances are uncommon manifestation of these tumors. Tumors of the posterior fossa have a higher tendency to be symptomatic and cause hydrocephalus with signs of cerebellar dysfunction including ataxia and dysmetria. Some intraventricular tumors are asymptomatic and may be incidentally discovered on brain imaging. The best example of this would be a subependymoma seen within the lateral ventricles on a trauma head computed tomography (CT) image.

Imaging studies are the key component in the diagnosis of intraventricular tumors. Magnetic resonance imaging (MRI) is the preferred modality of choice for imaging evaluation, although CT and angiogram may often be helpful in tumor characterization. Cranial ultrasound and Doppler are useful in infants with limited role in adult population. Besides conventional MR imaging, newer sequences like susceptibility-weighted imaging and advanced techniques including perfusion imaging and proton (H^1) spectroscopy are increasingly being utilized for tumor characterization. This is especially important if the mass is benign and conservative follow-up could be the optimal treatment.[7]

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Table The Demographic and Major Imaging Features of the Common Intraventricular Tumors

Tumor	Patient Age	Location	Imaging Hallmark
Choroid plexus tumors (CPP and CPC)	CPPs are more common in children usually <10 y. Carcinomas occur exclusively in children and infants.	50% Lateral ventricles, 40% in fourth ventricle	Hyperdense on CT, avidly enhancing, lobulated, frond-like appearance
Meningioma	40-60 Y, with female predilection (similar to dural meningiomas)	Atria of lateral ventricle	Hyperdense on CT with calcification in 50% and avid enhancement
Ependymoma	Both adults and children, one-third of all brain tumors in children <3 y	Fourth ventricle in > 60% cases	Tumor heterogeneity, hemorrhage, necrosis, cyst, and calcification
Subependymoma	40-60 Y	Fourth ventricle (50%-60%) and lateral ventricular margins or septum (30%-40%)	Poorly enhancing, calcification in 30%
SEGA	First 2 decades of life	Virtually always at the foramen of Monroe	Robust enhancing lesion seen exclusively in patients with tuberous sclerosis
Central neurocytoma	Predominantly in the age group of 20-40 y	Inferior septum pellucidum and the anterior lateral ventricle are the most common locations	Lobulated enhancing mass with "bubbly" appearance due to intratumoral cyst. Glycine peak (3.5 ppm)

A systemic approach taking into consideration patient age, location of tumor, and imaging findings can substantially narrow down the differential, and in many cases suggest a single most probably diagnosis. Patient's age and location are the 2 key concepts for the imaging diagnosis of intraventricular tumors.[8] Tumors such as CN and SEGAs are predominantly located in the anterior portion of the lateral ventricles, whereas ependymomas and subependymomas are more common in the fourth ventricle. Vascular lesions such as meningiomas and metastasis tend to occur within the atria of the lateral ventricles as the choroid plexus is most prominent in this region. Ependymomas and choroid plexus tumors are usually seen in the pediatric age group, whereas meningiomas and CN are usually seen in middle-aged adults (30-40 years).[9] The demographic and major imaging features of the common intraventricular tumors are highlighted in the Table. Microsurgical resection is the gold standard of intraventricular tumor resection. Neuroendoscopic surgery has however become the first-line modality for obtaining tumor samples from most intraventricular tumors along with simultaneous treatment of hydrocephalus, thereby avoiding aggressive conventional surgery in few cases. Endoscopy may also be used for complete excision of intraventricular tumors in selected cases.[10]

Neoplasm of the Choroid Plexus

Choroid Plexus Tumors (Papilloma and Carcinomas)

Choroid plexus tumors constitute about 0.5% of adult brain tumors and 1%-2% of pediatric brain tumors, with majority (80%) being diagnosed as CPP as compared to carcinomas (20%). The frequency of these tumors is much higher in the infant age group, ranging from 15%-20% of intracranial tumors in the first year of life. Most common locations are the atria of lateral ventricle (50%) and the fourth ventricle

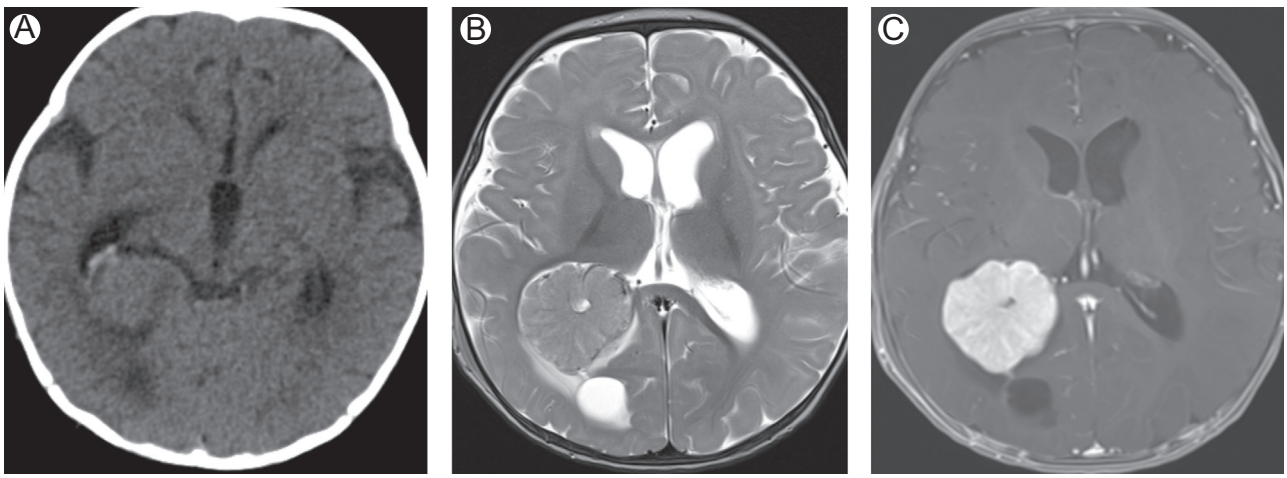

Figure 1 Choroid plexus papilloma. (A) Axial noncontrast CT image shows a circumscribed lobulated hyperdense mass with calcific foci in the atrium of the right lateral ventricle. (B) Axial T2WI and contrast-enhanced T1WI (C) shows "frond-like" appearance and avid enhancing mass with ventricular dilatation.