Intraventricular Neoplasms: Radiologic- Pathologic Correlation

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Learning objectives

The purpose of this pictorial review is:

1- To illustrate the MRI characteristics of several types of intraventricular tumors

2- To discuss the clinical presentation and differential diagnoses of these intraventricular lesions

3- To describe the clinical and imaging features of intraventricular neoplasms with emphasis on pathologic correlation

4- To discuss the utility of MRI and to provide the radiologist with an approach to accurately differentiating between these lesions
Background

The differential diagnosis for intraventricular neoplasms can be broad, and many of them have similar patterns of signal intensity and contrast enhancement at imaging. However, the location of the lesion in the ventricular system along with knowledge of the patient’s age, gender, and underlying conditions will help narrow the differential diagnosis.

Embriology:

The cerebral ventricles begin as ependymallined outpouchings from the cranial end of the neural tube, which are called the telencephalic vesicles. The choroid plexus develops from an invagination of primitive pia-arachnoid and vessels into these vesicles, thus creating the choroidal fissures. The epithelial lining of the ventricles is composed of ependymal cells, which are the cell of origin of the ependymoma. Subjacent to the ependymal lining is a layer of subependymal plate composed of glial cells, from which subependymomas are thought to arise.

The septum pellucidum is also lined by glial cells and residual neuronal precursor cells, from which the central neurocytoma may arise. The very vascular choroid plexus produces cerebrospinal fluid (CSF) and may give rise to primary neoplasms of the choroid plexus (choroid plexus papilloma, atypical choroid plexus papilloma, choroid plexus carcinoma); owing to its vascular supply, it may contribute to deposition of metastases in this location. Arachnoidal cap cells, which make up the arachnoid granulations, may become trapped within the choroid plexus during embryologic development; these cells can give rise to meningiomas (Figure 1 and 2).

Ependymoma:

Ependymomas account for 3%-5% of intracranial neoplasms. They are generally well circumscribed
glial tumors with ependymal differentiation that arise from the ependymal cells of the ventricular wall. These lesions can occur either supratentorially (40% of cases) or within the posterior fossa (60%).

Ependymomas can occur in any age group but are more common in younger patients. Those that occur in the posterior fossa are more common in children (mean age, 6 years), whereas the mean age for supratentorial lesions is 18-24 years.

Presenting symptoms depend on the location: Those that occur in the fourth ventricle typically manifest with symptoms of increased intracranial pressure due to obstruction, ataxia, or paresis, whereas supratentorial lesions manifest as headache, focal neurologic deficit, or seizure.

**Pathologic Findings:**

Ependymomas are classified as either World Health Organization (WHO) grade II (lowgrade, well-differentiated) or grade III (anaplastic) neoplasms. At gross inspection, these are soft "plastic" neoplasms. In the region of the fourth ventricle, they may extend through the foramen of Luschka into the cerebellopontine angle cistern or through the foramen magnum. WHO grade II ependymomas are moderately cellular tumors, with rare mitotic figures. The classic histologic findings in ependymomas are perivascular pseudorosettes and true ependymal rosettes. WHO grade III ependymomas demonstrate increased cellularity with brisk mitotic activity, along with ependymal differentiation. Frequently, microvascular proliferation and necrosis are seen.

**Imaging Features:**

Ependymomas frequently demonstrate cystic components and areas of small chunky calcification. Occasionally, intratumoral hemorrhage
may be seen. At computed tomography (CT), the soft-tissue portion is commonly hypo- to isoattenuating.

At magnetic resonance (MR) imaging, they are iso- to hypointense on T1-weighted images and iso- to hyperintense on T2-weighted images. Heterogeneous enhancement is seen on contrast material-enhanced images. Blooming may be seen on T2*-weighted images if calcification or hemorrhage is present. Findings on diffusion-weighted images are variable. Reduced diffusion may be seen in the soft-tissue component of some ependymomas, a finding that most likely reflects higher cellularity in some neoplasms.

Intraparenchymal lesions are typically large at presentation, with up to 94% being over 4 cm in size at the time of diagnosis. Many intraparenchymal supratentorial ependymomas have a large cystic component. They may also have a "cyst and mural nodule" appearance, for which the differential diagnosis includes pilocytic astrocytoma, pleomorphic xanthoastrocytoma, and ganglioglioma; however, they may be completely solid as well.

The cystic component tends to be similar in signal intensity on T1- and T2-weighted images, but it may not be completely suppressed on T2-weighted fluid-attenuated inversion-recovery (FLAIR) images due to proteinaceous content.

Intraventricular lesions may extend into adjacent brain, and there may be vasogenic edema in the adjacent periventricular white matter. Ependymomas involving the fourth ventricle tend to fill the ventricle-like a plaster cast-and may extend through the foramen of Luschka, foramen of Magendie, or foramen magnum. This finding is highly suggestive of an ependymoma but is not pathognomonic, since occasionally medulloblastomas may extend through the foramen magnum (Figure 3,4 and 5).

Subependymoma:

Subependymomas account for 0.2%-0.7% of intracranial neoplasms; however, this may be an
underestimate, since often they are asymptomatic and found incidentally. It is thought that they arise from the subependymal glial layer surrounding the cerebral ventricles, but the exact histogenesis remains uncertain. Most of these lesions occur in the fourth ventricle (50%-60%) and lateral ventricles (30%-40%). Rarely, they may arise from other intraventricular sites or within the central canal of the spinal cord. The majority are less than 2 cm in size, but symptomatic lesions tend to be 4 cm or greater in size.

Subependymomas have a male predominance (male-to-female ratio, 2.3:1), and 82% occur in patients older than 15 years.

**Pathologic Findings:**

Subependymomas are WHO grade I neoplasms with ependymal differentiation. These well-circumscribed lesions are typically attached to the ventricle wall by a narrow pedicle. Histologic analysis reveals a finely fibrillary background and occasional clustering of small, uniform, cytologically bland ependymoma-like nuclei. They often demonstrate numerous small cysts, especially in those lesions that originate from the lateral ventricle. These lesions do not have any significant vascularity, and mitotic activity is low or absent.

**Imaging Features:**

Subependymomas are well-circumscribed lesions that are hypo- to isoattenuating at CT. Cystic degeneration is common, and calcification may be seen in the lesion. Intratumoral hemorrhage may also occur. MR imaging reveals a lesion that is hypo- to isointense relative to white matter and hyperintense on T2-weighted images.

Enhancement is variable, with most lesions demonstrating no or minimal enhancement; less commonly, there is moderate but typically heterogeneous enhancement. Unlike with ependymomas, no invasion into the brain parenchyma occurs, and it is rare for there to be adjacent T2 hyperintensity in the periventricular white matter. No CSF dissemination
occurs. Rarely, multiple subependymomas may be present, but the lack of significant enhancement helps differentiate these lesions from other multifocal processes such as metastases. Angiography reveals an avascular or hypovascular mass, a finding that reflects the relatively avascular nature of these lesions at histologic analysis (Figures 6, 7, 8 and 9).

Central Neurocytoma:

Central neurocytomas account for 0.25%-0.5% of intracranial tumors. The origin of these tumors is unclear, but results of cell-culture investigations suggest they arise from bipotential progenitor cells that are capable of both neuronal and glial differentiation.

Central neurocytomas occur in the lateral ventricle with or without extension into the third ventricle and arise from the septum pellucidum or ventricular wall.

The mean patient age is 29 years, but a wide age range is reported, from 8 days to 67 years; there is no gender predilection. Patients typically present with symptoms of increased intracranial pressure.

Pathologic Findings:

Central neurocytomas are WHO grade II lesions. At gross analysis, central neurocytomas are gray friable lesions often containing calcification or hemorrhage. These neoplasms typically have a benign histologic appearance, featuring solid sheets or large lobules of small round to ovoid neoplastic cells with a delicate vascular network and intervening irregular patches of fibrillary neuropil. Atypical forms demonstrate increased mitoses, atypia, and in some cases microvascular proliferation and necrosis. Various architectural patterns may be observed, including an oligodendroglioma-like appearance and neurocytic rosettes. In fact, these tumors were
mistakenly classified as "intraventricular oligodendroglioma" before the 1980s. The presence of pineocytomatous rosettes allows differentiation of central neurocytoma from oligodendroglioma.

Central neurocytomas usually express immunoreactivity for synaptophysin and neuron-specific enolase, which are both markers of neuronal differentiation, and strong staining for synaptophysin is reported to be the most reliable diagnostic marker.

**Imaging Features:**

Central neurocytomas are well-circumscribed, lobulated masses that frequently have cystlike areas. Up to 50% may contain calcification, and hemorrhage may rarely be seen. At CT, these lesions are hyperattenuating. At MR imaging, central neurocytomas are isointense to gray matter on T1-weighted images and hyperintense on T2-weighted images. These lesions may have a "bubbly" appearance due to the presence of multiple cysts. At contrast-enhanced imaging, the enhancement pattern is variable, but moderate to strong enhancement is typically seen.

Prominent flow voids may be noted, and increased T2 signal intensity may be seen in the adjacent periventricular white matter (Figures 10 and 11).

**Choroid Plexus Neoplasms:**

Choroid plexus tumors account for 2%-4% of pediatric brain tumors, 0.5% of adult brain tumors, and up to 20% of pediatric neoplasms occurring in the 1st year of life. These neoplasms arise anywhere that choroid plexus is located and develop from the choroid plexus epithelium. They most commonly occur in the atrium of the lateral ventricle (50% of cases). Forty percent occur in the fourth ventricle, 10% in the third ventricle, and approximately 5% in more than one location. Fourth ventricle lesions are more common in
males (male-to-female ratio, 3:2), but there is no gender predilection for lateral ventricle lesions.

Lateral ventricle lesions are more common in children (80% of cases), whereas fourth ventricle lesions are evenly distributed among all age groups. Rare cases of extraventricular choroid plexus tumors have been reported.

Pathologic Findings:

CPP is histologically similar to normal choroid plexus, consisting of bland cuboidal to columnar epithelial cells surrounding a delicate fibrovascular stalk. At gross inspection, these are discrete, well-circumscribed, often papillary masses.

Mitotic activity is low; brain invasion and necrosis may occur but are uncommon. Hemorrhage and cyst formation may be seen in all of the choroid plexus neoplasms. Atypical CPPs demonstrate two or more mitoses per 10 randomly selected high-power fields. Up to two of the following features may be present but are not required for diagnosis: increased cellularity, nuclear pleomorphism, blurring of the papillary pattern, and areas of necrosis. CPC is a rare tumor that shows frank signs of malignancy. Diagnosis of CPC requires the presence of at least four of the following features:

- more than five mitoses per high-power field, increased cellular density, nuclear pleomorphism, blurring of the papillary pattern, and necrosis.

Imaging Features:

Imaging alone does not allow distinction between these neoplasms. All of them may demonstrate CSF dissemination; therefore, imaging of the entire neuroaxis is recommended.

Choroid plexus neoplasms are very vascular lesions that demonstrate avid enhancement at
contrast-enhanced imaging. They are iso- to hyperattenuating at CT. CT angiography as well as conventional angiography may reveal an enlarged choroidal artery if the neoplasm is in the atrium of the lateral ventricle. Calcifications and foci of hemorrhage may be seen, and hydrocephalus commonly occurs. CPP and atypical CPP frequently have a papillary or lobulated appearance, which helps differentiate them from other intraventricular neoplasms; conversely, carcinomas tend to have a more irregular contour.

At MR imaging, these lesions are iso- to hypointense on T1-weighted images and iso- to hyperintense on T2-weighted images; flow voids are common. Choroid plexus neoplasms may have a long vascular pedicle, which may twist, leading to tumor infarction. Periventricular vasogenic edema may occur in all choroid plexus tumors, and a suggestion of parenchymal invasion may also be seen at imaging in all three tumor types (Figures 12, 13, 14, 15 and 16).

**Meningioma:**

Intraventricular meningiomas account for 0.5%-3.7% of intracranial meningiomas. They are believed to arise from arachnoidal cap cells trapped in the choroid plexus or from the tela choroidea during embryologic formation of the choroid fissure and plexus. The most common location for intraventricular meningiomas is in the atrium of the lateral ventricles. Less commonly, they may arise in the third ventricle and rarely in the fourth ventricle. Like meningiomas elsewhere, they are most common in females (female-to-male ratio, 2:1) with a peak age range of 30-60 years. Meningiomas can also rarely affect the pediatric age group, accounting for less than 3% of intracranial neoplasms in this population. However, the intraventricular form accounts for 17% of
pediatric meningiomas.

These neoplasms usually reach a large size before patients become symptomatic; patients typically present with signs of increased intracranial pressure but may also present with contralateral sensory or motor deficits.

**Pathologic Findings:**

Gross inspection reveals a well-demarcated rubbery or firm mass. There is no difference in terms of histologic features between an intraventricular meningioma and one arising from a dural attachment. In adults, most intraventricular meningiomas are benign and of the meningothelial type. This form resembles the arachnoidal cap cell, consisting of uniform cells with oval nuclei containing delicate chromatin that may occasionally demonstrate central clearing. Psammoma bodies are not common. Another common histopathologic subtype is fibroblastic, but other forms have also been described.

**Imaging Features:**

CT reveals a well-defined, iso- to hyperattenuating globular mass. MR imaging demonstrates a mass that is iso- to hypointense on T1-weighted images and iso- tohyperintense on T2-weighted images. Owing to the highly vascular nature of these lesions, avid enhancement is seen on contrast-enhanced images (Figures 17, 18 and 19).

**Subependymal Giant Cell Tumor:**

SGCT is the most common cerebral neoplasm in patients with tuberous sclerosis (TS), developing in up to 16% of cases. It has previously been referred to as subependymal giant cell astrocytoma, but recent pathologic studies have revealed that it is a glioneuronal tumor; thus, the trend is away
from use of the term subependymal giant cell astrocytoma. SGCT is considered pathognomonic for TS.

There is a wide age range for presentation, from birth to the 5th decade (mean age, 11 years). These lesions arise near the foramen of Monro. They are slow-growing lesions, and due to their location they commonly manifest with symptoms of increased intracranial pressure from obstructive hydrocephalus. Indications for resection include increasing tumor size, hydrocephalus, a new focal neurologic deficit, or symptoms of increased intracranial pressure.

Pathologic Findings:

SGCT is a WHO grade I lesion that macroscopically appears as well-circumscribed solid intraventricular neoplasms occurring near the foramen of Monro. Histologic evaluation reveals large cells resembling astrocytes or ganglion cells with abundant cytoplasm. The cells may be polygonal, epithelioid, or spindle shaped. Nuclear pleomorphism, increased mitotic activity, occasional endothelial proliferation, and necrosis may occur but are not indicative of malignant transformation and have no implication for prognosis. The histogenesis of SGCT is unclear, but there is evidence to support both neuronal and astrocytic lineage. Immunohistochemical staining reveals markers for both glial and neuronal proteins.

Imaging Features:

Imaging reveals a well-circumscribed lesion that is typically larger than 1 cm and most commonly located near the foramen of Monro. Other subependymal nodules may be noted. Owing to its location, hydrocephalus may be seen. Variable degrees of calcification may be present, and hemorrhage may occasionally be seen. At CT, SGCTs are hypo- to isodense. MR imaging reveals a lesion that is hypo- to isointense to gray matter.
on T1-weighted images and iso- to hyperintense on T2-weighted images. At contrast-enhanced imaging, the lesions avidly enhance (Figures 20 and 21).

Metastases and Other Intraventricular Neoplasms:

Intraventricular metastases account for 0.9%-4.6% of cerebral metastases. In adults, renal, colon, and lung carcinoma are the most common causes; in children, neuroblastoma, Wilms tumor, and retinoblastoma are most common. Imaging findings of a solitary metastasis to the choroid plexus may be indistinguishable from those of a meningioma or choroid plexus neoplasm, and a history of a primary neoplasm should raise suspicion for metastasis. Avid enhancement is usually seen on contrast-enhanced images, and vasogenic edema may be seen in the adjacent brain parenchyma. Many other neoplasms involve the ventricular system, including lymphoma, low- and highgrade gliomas, intraventricular craniopharyngioma, and primitive neuroectodermal tumor.

Glioblastoma Multiforme:

GBM is the most common primary brain tumor, but intraventricular GBM is rare and only few cases have been reported in the literature. The authors report a case of 64-year-old man who had a remote history of previous periventricular intracerebral hemorrhage. Brain computed tomography (CT) and magnetic resonance (MR) imaging showed an intraventricular lesion with inhomogeneous enhancement, infiltrative borders and necrotic cyst, and obstructive hydrocephalus. The patient underwent surgical removal through transcortical route via the bottom of previous hemorrhage site and the final pathologic diagnosis was GBM (Figures 22, 23 and 24).

Colloid cyst of third ventricle

Colloid cysts are congenital lesions, benign and circumscribed, filled with mucinous material. Congenital neuroepithelial cysts almost always arise in the anterior aspect of
third ventricle, however, rare reports describe cysts in other locations. Correspond to 0.5-1% of primary CNS tumors and derive from primitive neuroepithelium of the tela choroidea or from endoderm. Typically assymptomatic, they are an incidentally observed in CNS image studies. Obstruction of Monro's foramen can cause symptoms like chronic headaches due to hydrocephalus.

**Phatologic features:**

Colloid cysts have their origin in embryonic endoderm and migration of this compound to vellum interpositum. The microscopic studies show a thin fibrous capsule, with a single layer of simple cuboidal or pseudoestafificated epithelium, resting on a thin collagenous membrane. Cystic formations, filled with amorphous material, faintly eosinophilic, that was strongly periodic acid-Schiff (PAS)-positive due to the presence of glycogen and mucosubstances. Perilesional edema can be found.

**Imaging features:**

CT findings include round masses, hyperdense for its high cholesterol levels and proteinaceous content. Isodense or hypodense patterns may be encountered. A nonenhancing lesion, hyperdense, in the anterior aspect of third ventricle at Monro's foramen is almost diagnostic but MRI is more conclusive.

A cystic lesion, in typical location, which demonstrates in MRI T1- weighted images, high signal intensity and, on post contrast T1, usually doesn't enhance. T2- weighted sequences show isointensity commonly or hypointensity and is a result of the proteinaceous content fluid, as well as the paramagnetic effects of the metal ions in the fluid and hemorrhage. This low intensity signal is related with high viscosity and difficulty in aspiration. No restricted diffusion on DWI (Figures 25, 26, 27, 28 and 29).

**Pilomyxoid astrocytoma**

Described recently, it’s an uncommon type of primary brain tumor, more aggressive than pilocytic astrocytoma, found in young children (mean age 18 months), with dissemination through CSF and high recurrence rates. Have predilection to hypothalamic-chiasmatic region, but can be found everywhere in neuroaxis. Clinically presents with increase in head size and focal signs as visual disturbances.

**Pathologic features:**
Histopathologically characterized by compact myxoid matrix, monomorphic cells and bipolar tumor cells, with an arrangement around vessels - pattern similar to rosettes. However, pilocytic astrocytomas differ from pilomyxoids by not having Rosenthal fibers and eosinophilic granular bodies. In some cases there may be simultaneous presentation of histological features of pilocytic astrocytoma and pilomyxoid and this group is called intermediate pilomyxoid astrocytoma.

**Imaging features:**

In MRI studies, tumors are in midline, in supra-sellar areas, commonly in hypothalamic-chiasmatic region, but may be present in atypical locations in older patients. It's a solid lesion with rare cystic compound. Lesions are isointense in T1-weighted images and have high signal in T2-weighted images and a variable enhancement. The predominantly solid portion does not enhance even in primary lesion or in disseminated lesions and this pattern is much more common in pilomixoid astrocytoma, due to myxoid compound, than in the pilocytic astrocytoma. Leptomeningeal dissemination can be found. Necrotic or haemorrhagic areas are responsible for T2 signal abnormalities. Spectroscopy evidences choline and lipid peaks and reduced creatine and N-acetyl-aspartate (Figures 30, 31, 32 and 33).

**Craniopharyngioma:**

Craniopharyngioma is a histologically benign, extra-axial, slow-growing tumor that predominantly involves the sella and suprasellar space.

Despite its histologic appearance, craniopharyngiomas occasionally behave like malignant tumors. They can metastasize, and patients can have severe symptoms that usually require surgery and/or radiation therapy (with intracystic chemotherapy in some pediatric patients). Recurrences, both local and along surgical tracts, have been reported, as has meningeal seeding. Characteristic radiographic findings help in differentiating craniopharyngiomas from other tumors that can occur in the same anatomic region. Zenker first described craniopharyngioma in 1857.

Craniopharyngiomas are believed to derive from Rathke's cleft rather than squamous cell rests along the craniopharyngeal duct as was previously thought. This histological appearances of the two subtypes are different, accounting for the different imaging features.

**Adamantinomatous**

This type is seen predominantly in children. It consists of reticular epithelial cells which have appearances reminiscent of the enamel pulp of developing teeth.
There may be single or multiple cysts filled with thick oily fluid high in protein, blood products, and/or cholesterol, creating the so called "machinery oil". "Wet keratin nodules" are a characteristic histological feature. Calcification is usually present: ~ 90%.

Papillary

The papillary sub type is seen almost exclusively in adults and is formed of masses of metaplastic squamous cells. "Wet keratin" is absent. Cysts do form, but these are less of a feature, and the tumour is more solid. Calcification is uncommon or even rare.

Imaging features:

Although similar in terms of location, radiographic features depend on the type, although due to a significant minority of tumour having both adamantinomatous and papillary components many show overlapping features (Figures 34, 35 and 36).

Location

In the vast majority of cases, craniopharyngiomas have a significant suprasellar component (95%), with most involving both the suprasellar and intrasellar spaces (75%). A minority are purely suprasellar (20%), whereas purely intrasellar location is quite uncommon (<5%), and may be associated with expansion of the pituitary fossa. Larger tumours can extend in all directions, frequently distorting the optic chiasm, or compressing the midbrain with resulting obstructive hydrocephalus.

Occasionally, craniopharyngiomas appear as intraventricular, homogeneous, soft-tissue masses without calcification (papillary sub type). The third ventricle is a particularly common location.

Rare / ectopic locations reported include: nasopharynx, posterior fossa, extension down the cervical spine.

Adamantinomatous

Typically adamantinomatous craniopharyngiomas have a lobulated contour as a result of usually multiple cystic lesions. Solid components are present, but usually form a relatively minor component of the mass, and enhance vividly on both CT and MRI. Overall, calcification is very common, but this is only true of the adamantinomatous subtype (90% are calcified).
These tumours have a predilection to be large, extending superiorly into the third ventricle, and encasing vessels, and even being adherent to adjacent structures.

CT

- cysts
  - typically large and a dominant feature
  - near CSF density
- solid component
  - soft tissue density
  - vivid enhancement
- calcification
  - seen in 90%
  - typically stippled and often peripheral in location

MRI

- cysts: variable but ~80% are mostly or partly T2 hyperintense
- solid component
  - T1: iso to lightly hypointense to brain
  - T1 C+: vivid enhancement
  - T2: variable / mixed
- calcification
  - difficult to appreciate on conventional imaging
  - susceptible sequences may better demonstrate calcification
- MR angiography: may demonstrate displacement of the A1 segment of the anterior cerebral artery
- MR spectroscopy: cyst contents may show a broad lipid spectrum, with an otherwise flat baseline

Papillary

Papillary craniopharyngiomas tend to be more spherical in outline and usually lack the prominent cystic component; most are either solid or contain a few smaller cysts. Calcification is uncommon or even rare in the papillary subtype, a fact often forgotten.

These tumours tend to displace adjacent structures.

CT

- cysts
  - small and not a major feature
  - near CSF density
- solid component
  - soft tissue density
  - vivid enhancement
- calcification
• uncommon - rare

MRI

• cysts
  • when present they are variable in signal
  • 85% T1 hypointense 4
• solid component
  • T1: iso to lightly hypointense to brain
  • T1 C+: vivid enhancement
  • T2: variable / mixed
• MR spectroscopy: cyst contents does not show a broad lipid spectrum as they are filled with water fluid

Imaging Differential Diagnosis of non-neoplastic Intraventricular Lesions:

Intraventricular neurocysticercosis.

Parasitic neurological disease, caused by *Taenia Solium* larva, which encystis in CNS. This is the most commom acquired disease of CNS and cause of secondary epilepsy. Humans are definitive and transient host, depending on the stage of disease. In CNS, can be found in subarachnoid space, ventricles and spinal cord.

Pathologic findings:

Cysticercosis is related to eggs ingestion of *T.solium*, from the feces of a tapeworm carrier and affects males and females equally, in early adult life (25-35 years). The contamination by fecal-oral route usually occurs drinking contaminated water or eating contaminated food and takes the eggs to the intestinal mucosa. The cysts, contained in the eggs, can pass through the intestinal mucosa and reach the systemic circulation and the capillaries of the central nervous system. Within the central nervous system, cysts differentiate into encysted larvae, but by the action of the blood-brain barrier is not observed any surrounding inflammatory reaction (noncystic phase). Increase in size or a cluster of cysts (racemose form) can be seen in white and gray matter junction (*vesicular phase*). In untreated patients or early treated, the inflammatory response settles, the cystic fluid becomes turbid, with surrounding edema and proeminent symptoms (*colloidal vesicular phase*). Cystic retraction occurs, with decrease of inflammatory response (*granular nodular phase*) and with drug treatment and death of the larva, calcification can be observed without edema (*nodular calcified phase*) and this is the last stage of this infection. Except for calcified phase, all other phases have active infections. The diagnosis of
neurocysticercosis is confirmed by clinical history, serology, CSF analysis, CT and RNM imaging evaluation.

The commonest location is the subarachnoid space over the cerebral hemispheres and adjacent to meninges, causing arachnoiditis and hydrocephalus. In parenchymal form, the parasites can be found in gray-white matter junction. In ventricles, usually appear as solitary foci, without a visible scolex, commonly in fourth ventricle (50% cases) and lateral ventricles (35% cases), causing ependymitis, ventriculitis and hydrocephalus.

**Imaging findings:**

Tomographic studies (CT) the majority of lesions located in ventricular system, as well as brainstem and cisterns can not be identified. In nonenhanced CT cystic lesions are isodense and may not be detected. CT helps to find structural alterations like ventricular distortion, signs of hydrocephalus and end staged calcified lesions. Intraventricular contrast injection may delineate cyst and the obstructed ventricular portion.

MRI studies are useful to detect ventricular cysts in neurocysticercosis. Cisternal and intraventricular cysts have different signal intensity from CSF, as a result of the relative T1 shortening but may present with the same signal intensity, requiring additional MRI sequences like three dimensional steady-state free precession or MR contrast enhanced MRI. The intraventricular cysts typically have 1-2 cm in diameter and can evidence pericystic ependymal inflammatory reaction. In T2-weighted images show periventricular high signal and hyperintense rim (pericystic ependymal inflammatory reaction), even as mass effect and CSF flow void next to the cysts (Figures 37, 38 and 39).
Fig. 1: Fetal Period - posterior horn of the lateral ventricle, choroid plexus of the third ventricle, suprapineal recess, interthalamic adhesion, aqueduct, and apertures in the roof of the fourth ventricle.

**Fig. 2:** Demographic and Imaging Features of Common Intraventricular Neoplasms


<table>
<thead>
<tr>
<th>Tumor</th>
<th>Patient Age</th>
<th>CT Appearance</th>
<th>MR Imaging Appearance</th>
<th>Hallmarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ependymoma</td>
<td>Mean age 6 y for tumors in fourth ventricle and 18–24 y for supratentorial lesions</td>
<td>Usually isoattenuated; calcification in 40–80% of lesions; enhancement usually intense but variable</td>
<td>Usually heterogeneous (calcification, hemorrhage, cysts); isointense at T1WI, hyperintense at T2WI</td>
<td>Constitutes 6%–12% of all pediatric brain tumors; 58% of lesions in fourth ventricle; supratentorial lesions usually extraventricular and cystic; recurrence and extraventricular extension common; WHO grade II</td>
</tr>
<tr>
<td>Subependymoma</td>
<td>82% of patients &gt;15 y, with middle-age and older males most common</td>
<td>Iso- to hypoattenuated; hydrocephalus in 85% of cases, calcification in 31%, cysts in 18%, hemorrhage rare; focal enhancement</td>
<td>Hyperintense at T1WI, hypointense at T2WI, variable enhancement</td>
<td>&gt;50% of lesions in fourth ventricle, lateral ventricular lesions common; extraventricular extension rare; recurrence very rare; WHO grade I</td>
</tr>
<tr>
<td>Central neurocytoma</td>
<td>20–40 y most common</td>
<td>Hyperattenuated; many small cyst-like areas, calcification in 50% of cases</td>
<td>Hyperintense at T1WI, solid portions hypointense and cysts hyperintense at T2WI</td>
<td>50% of lesions in lateral ventricle, 15% in lateral and third ventricles; WHO grade II</td>
</tr>
<tr>
<td>Subependymal giant cell astrocytoma</td>
<td>Children</td>
<td>Calcified nodule near fornix of Monro; intense enhancement</td>
<td>Hyperintense at T1WI, heterogeneously hyperintense at T2WI</td>
<td>All lesions in lateral ventricle near fornix of Monro; occasionally extends into third ventricle; associated with tuberous sclerosis; WHO grade I</td>
</tr>
<tr>
<td>Choroid plexus papilloma</td>
<td>50% of patients &lt;10 y for lateral ventricle tumors; fourth ventricular lesions seen in patients 0–50 y</td>
<td>Iso- to hyperattenuated, lobulated mass typically centered in atrium of lateral ventricle; calcification in 24% of cases; intense enhancement</td>
<td>Iso- to hypointense at T1WI, variably hyperintense at T2WI; flow voids common</td>
<td>50% of lesions in lateral ventricle, 40% in fourth ventricle, 5% in third ventricle; extraventricular extension common; prominent lobulation and hydrocephalus; WHO grade I</td>
</tr>
<tr>
<td>Choroid plexus carcinoma</td>
<td>Infants and young children</td>
<td>Heterogeneous attenuation; vasogenic edema</td>
<td>Heterogeneous intensity; vasogenic edema</td>
<td>Brain invasion, less hydrocephalus than in choroid plexus papilloma; WHO grade III</td>
</tr>
<tr>
<td>Intraventricular meningioma</td>
<td>Adults 30–60 y most common</td>
<td>Hyperattenuated arachnoid mass; calcification in 50% of cases; intense enhancement</td>
<td>Iso- to hypointense at T1WI, iso- to hyperintense at T2WI; intense enhancement</td>
<td>Axial mass in choroid plexus; usually WHO grade I</td>
</tr>
<tr>
<td>Metastasis</td>
<td>Adults more common</td>
<td>Iso- to hyperattenuated</td>
<td>H yp ointense at T1WI, hyperintense at T2WI; intense enhancement</td>
<td>Axial mass in choroid plexus; renal and lung carcinoma most common primary lesions</td>
</tr>
</tbody>
</table>

Note.—T1WI = T1-weighted imaging, T2WI = T2-weighted imaging.
**Fig. 3:** Ependymoma. Axial Flair and contrast-enhanced T1-Weighted images: a predominantly solid lesion that arises from the fourth ventricle and extends through the foramen of Luschka into the right cerebellopontine angle. The lesion is slightly hyperintense relative to the adjacent cerebellum. The mass results in hydrocephalus, with dilatation of the third and lateral ventricles.

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Fig. 4: Recurrent ependymoma. Axial T2-Weighted and contrast-enhanced T1-Weighted images: a solid cystic lesion that arises from the fourth ventricle and extends through the foramen of Luschka into the left cerebellopontine angle. The lesion is slightly hyperintense relative to the adjacent cerebellum.

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**Fig. 5:** Ependymoma: The image shows a neoplasm with moderate cellularity and numerous typical vascular pseudorosetas of ependymomas.

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**Fig. 6:** Subependymoma: Axial T2-Weighted and T1-Weighted axial and sagittal images show a fourth ventricle heterogeneous lesion filling and conforming to the fourth ventricle with extension through the foramen magnum with minimal enhancement. On the T2-weighted images, the neoplasm is hyperintense. Resultant hydrocephalus is present.

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Fig. 7: Subependymoma. Sagittal non enhanced T1-weighted image shows a hypointense lesion in the floor of the fourth ventricle with extension through the foramen magnum and syringomyelia of upper cervical spine.

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Fig. 8: Subependymoma. Contrast-enhanced sagittal T1-weighted image shows a hypointense lesion in the floor of the fourth ventricle with extension through the foramen magnum and syringomyelia of upper cervical spine.

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Fig. 9: Subependymoma: HE (200x) - image shows very paucellular tumor, arranged in small nodules inside a fibrillar background.

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**Fig. 10:** Atypical Central Neurocytoma. Axial Flair and T1-weighted images show a mass that is slightly hyperintense to gray matter located centrally around the septum pellucidum. Foci of hyperintensity consistent with cystic regions are noted. No increased signal intensity is appreciated in the adjacent brain parenchyma. Axial contrast-enhanced T1-weighted image shows moderate heterogeneous enhancement.

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Fig. 11: Neurocytoma: HE (200x)- lobular arrangements of cells with a small blood vessel at the centre, and polygonal small cells with a clear perinuclear halo.

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**Fig. 12:** Choroid Plexus Papilloma. Axial Flair, T2-Weighted and axial, sagittal and coronal, Enhanced and nonenhanced T1-Weighted images: Large heterogeneous and hypervascularized intraventricular lobulated mass in the atrium of the right lateral ventricle. Hydrocephalus is present.

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Fig. 13: Choroid Plexus papilloma. The image shows a well-differentiated neoplasm with well-formed papillae without atypia.

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Fig. 14: Choroid plexus carcinoma of the posterior horn of the left ventricle. Axial Flair and T2-Weighted images, sagittal, coronal and axial enhanced T1-Weighted images show multiple hypervascular lobulated masses and leptomeningeal spread lesions, a finding consistent with CSF dissemination.

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**Fig. 15:** Choroid plexus carcinoma. Axial contrast-enhanced CT image shows a hypervascular mass in the atrium of the left lateral ventricle.

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Fig. 16: Choroid plexus carcinoma: HE (400x) - image shows a papillary tumor growth pattern with severe atypia.

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Fig. 17: Intraventricular meningioma: Axial gradient-echo (1000/54) pMRI image with an rCBV color overlay demonstrates high rCBV throughout the lesion. Coronal T2-weighted image shows a slightly heterogeneous, predominantly isointense mass. No surrounding edema is present. Axial contrast-enhanced T1-weighted image shows avid homogeneous enhancement.

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Fig. 18: Meningioma within the atrium of the left lateral ventricle. Axial T2-weighted image shows a slightly heterogeneous, predominantly isointense mass. No surrounding edema is present. Non-contrast enhanced Axial and sagittal T1-weighted images and axial contrast-enhanced T1-weighted image shows avid homogeneous enhancement.

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Fig. 19: Meningioma HE, (200x): image shows a moderately cellular tumor with a growth pattern forming whorls and a calcification.

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Fig. 20: Subependymal giant cell tumor. Axial and Coronal contrast-enhanced T1-weighted images show avid enhancement of the neoplasm near the foramen of Monro. There were also multiple subcortical focal areas of hyperintensity on T2 WI, which were consistent with subcortical tubers.

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Fig. 21: Giant cell astrocytoma. Axial contrast-enhanced CT images show a well-circumscribed calcified lesion located near the foramen of Monro.

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Fig. 22: Glioblastoma multiforme. Coronal T2-Weighted image and T1-weighed Gd-enhanced MR images reveals an enhancing huge intraventricular mass in the temporal horn of the right lateral ventricle. Axial, sagittal and coronal images show that the heterogenously rim-enhancing mass is intraaxial, located from the periventricular region.

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Fig. 23: Glioblastoma multiforme. MRS of the right intraventricular mass, which revealed large accumulation of lipid and lactate (peaked at 0.8-1.33 ppm), marked decrease in peak NAA at 2 ppm, marked increase in the peak of Cho at 3.2 ppm, and decrease in the peak of Cr. MRS results were from the multi voxel square area.

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Fig. 24: Glioblastoma: HE (200x) - tumor shows severe atypic cells arranged around a necrotic area, typical of glioblastoma (pseudopalisade necrosis).

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**Fig. 25:** Colloid cyst. Axial and sagittal T1-Weighted and axial Flair images show a heterogeneous non-enhancing mass obstructing the third ventricle, located at the foramen of Monro, characteristically occur at the anterosuperior aspect of the third ventricle.

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Fig. 26: The Pterional Approach


Fig. 27: Colloid Cyst. Microsurgical removal of intraventricular lesion using endoscopic visualization and stereotactic guidance.

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**Fig. 28:** Colloid cyst: surgical specimen.

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Fig. 29: Colloid Cyst: HE (400x)- the image shows a cyst lined by a single layer of cuboidal, partially flattened, non-ciliated and ciliated epithelial cells.

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**Fig. 30:** Pilomyxoid astrocytoma. Axial ADC, DWI and T2-weighted images hyperintense multilobulated lesion on T2, demarcated poorly from the cerebral tissue and invaded into the left anterior cingulate gyrus and the corpus callosum, the mas fills the anterior horn of the left lateral ventricle. Enhanced T1-weighted sagittal image showing a midline mass that is solid in nature and hypervascular.

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Fig. 31: Pilomyxoid astrocytoma. Axial Flair image shows a hyperintense multilobulated lesion, demarcated poorly from the cerebral tissue and invaded into the left anterior cingulate gyrus and the corpus callosum, the mass fills the anterior horn of the left lateral ventricle. Nonenhanced sagittal and Enhanced T1-weighted sagittal and axial images demonstrates a multilobulated enhancing tumor.

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Fig. 32: Pilomyxoid astrocytoma: HE (200x): the image shows a moderately cellularity neoplasm, consisting of rounded and oval cells without significant atypia, embedded in a myxoid stroma.

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**Fig. 33:** Pilomyxoid astrocytoma. Pilomyxoid astrocytoma: HE (200x): the image shows a moderately cellularity neoplasm, consisting of rounded and oval cells without significant atypia, embedded in a myxoid stroma.

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Fig. 34: Adamantinomatous craniopharyngioma. The solid enhancing mass is centered in the third ventricle, expanding the supraoptic recess and outwardly bowing the floor of the third ventricle. Superiorly the mass extends to the foramina of Monro. There is a stippled and peripheral calcification.

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Fig. 35: Craniopharyngioma: T2-Weighted and contrast-enhanced T1-Weighted axial and sagittal images demonstrate a complex cystic mass in the suprasellar space, extending superiorly into the third ventricle, and encasing vessels, and being adherent to adjacent structures.

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Fig. 36: Craniopharyngeoma: HE (400x)- a tumor resembling epithelial tissue with queratinization, surrounding by brain parenchyma.

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Fig. 37: Fourth ventricle neurocysticercosis. Axial T2-weighted and Flair images show a cystic intraventricular lesion. Surrounding edema is present. CSF samples were positive by real-time PCR.

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**Fig. 38:** Fourth ventricle neurocysticercosis. Axial and coronal contrast-enhanced T1-weighted images show ring enhancement intraventricular lesion. Surrounding edema is present.

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**Fig. 39:** Anterior horn of lateral ventricle neurocysticercosis. Axial T2-weighted and Flair images show a cystic intraventricular lesion. The cysts typically do not have a visible scolex. CSF samples were positive by real-time PCR.

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Findings and procedure details

Typing intraventricular tumors is often an issue, even for an experienced Neuropathologist.

In this retrospective observational study, we reviewed the records of consecutively patients who underwent neuroendoscopic tumor excision at University Hospital of Clinicas of Porto Alegre from November 2009 to November 2013. We encountered specific entities, such as astrocytoma, ependymoma, subependymoma, central neurocytoma, subependymal giant cell tumor (SGCT), choroid plexus neoplasms, meningioma, metastases and other intraventricular neoplasms.
Conclusion

Although these neoplasms are easily detected with computed tomography (CT) and magnetic resonance imaging (MRI), both techniques are relatively unspecific in identifying the type of tumor. However, few imaging patterns are specific for a particular pathological process and useful conclusions can be made from the morphological appearance of the lesion, its location and enhancement pattern.
Personal information

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