Non-glial intracranial tumors: A radiologic - pathologic correlation

Poster No.: C-1286  
Congress: ECR 2011  
Type: Educational Exhibit  
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Keywords: Neuroradiology brain  
DOI: 10.1594/ecr2011/C-1286

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Learning objectives

- To revise the clinical, neuroimaging, and pathological features of the most common nonglial tumors in the intracranial CNS and surrounding tissues.

- To provide some useful clues, based on the direct correlation of imaging features and typical histologic appearance, for a comprehensive diagnostic approach.

- To enhance the usefulness of modern neuroimaging techniques for the demonstration of particular pathologic characteristics (high cellularity, increased perfusion, metabolic changes), crucial in the diagnostic workout.
Background

The brain is made up of about 100 billion neurons and trillions of support cells called **glia**, conforming what is called the neuropil. Glia is a varied group of cells, each with a dedicated and specific function (Figure 1):

- **Oligodendrocytes**: they surround tightly the axons to form the myelin sheath, which helps for a fast and optimal transmission of the electric signal (action potential).

- **Microglia**: they are a special type of immune cells. Their function is to save the brain from harmful microorganisms and damaged neurons.

- **Astrocytes**: this "funny looking" and ubiquitous cell has been classically considered to be the one to hold neurons (several of them at a time) in place and provide them with nutrients by connecting with blood vessels and acting as a bridge between them and neurons. Recently a new function as processors of information at the synapse, by the release by exocytosis of some gliotransmitters as glutamate, has been recognized.

- **Ependymal cells**: a type of glial cell conforming a thin membrane (ependyma) lining the ventricular system and the spinal cord. It is also involved in CSF production. Within the brain's ventricles, a population of modified ependymal cells and capillaries together form a system called the **choroid plexus**, which produces the CSF.

They all, along with neurons, conform the main solid structure of the CNS.

Intracranially, we might also consider other types of supporting, vascular, and functional tissues, each with its specific functional and typical tumoral behaviour:

- **Scalp tissues**: basically constituted by cutaneous and subcutaneous layers, in which epithelial, glandular, adipose and vascular cells can be found.

- **Skull**: both compact cortical (inner and outer tables) and trabecular spongy (diploe) bone tissues.

- **Meninges**: Duramater (the innermost layer of the external protective and supporting structures, in close contact with the periostium on the inner table), Arachnoid and Piamater (Leptomeninges as a whole, separated by the arachnoid space).
- **Schwann cells**: Schwann cells are the supporting cells of the peripheral nervous system (PNS). Like oligodendrocytes, Schwann cells wrap themselves around nerve axons, but the difference is that a single Schwann cell makes up a single segment of an axon's myelin sheath (Figure 1).

- **Vascular cells**: Endothelial and smooth muscle cells.

- **Paraganglia**: Collection of cells that came from embryonic nervous tissue, and are found near the adrenal glands and some blood vessels and nerves. Most paraganglia secrete epinephrine and norepinephrine.

- We should also consider all those tissues that, in an intracranial location and surrounded and somewhat connected with the CNS, derive from extraneuronal cellular lineages: Pineal, pituitary, haematopoietic or embryonal tissues.

Primary brain tumors constitute approximately 2% of all malignancies and 20% of malignancies in children. They seem to have increased in incidence over the past 30 years, but the rise probably results mostly from use of new neuroimaging techniques.

Tumors of the CNS are classified according to the consensus of an international Working Group of the World Health Organization (WHO) with the objective of establishing a classification and grading system that is accepted and used worldwide. Its most recently actualization (fourth edition) was published in 2007 and included concise sections on epidemiology, clinical signs and symptoms, imaging, prognosis and predictive factors (Figure 2 on page 7, Figure 3 on page 8).

Classification is made according to the cellular origin of each group of tumors, distinguishing between clinico-pathological entities (characterized by distinctive morphology, location, age distribution and biologic behaviour, and not simply by an unusual histopathological pattern), variants of entities (reliably identified histologically and having some relevance for clinical outcome, but as still being part of a previously defined, overarching entity) and histological patterns (identifiable histological appearances, but that do not have a distinct clinical or pathological significance).

Grading is the way of predicting the biological behaviour of a neoplasm, influencing the choice of therapies, particularly determining the use of adjuvant radiation and specific chemotherapy protocols. So we can consider this grading scheme as a malignancy scale rather than a strict histological grading system. **Grade I** tumors are low proliferative lesions prone to cure following surgical resection alone. Grossly, it is circumscribed or encapsulated, the tumor cells are well differentiated resembling the cell of origin and mitosis is absent or very rare; blood vessels are scanty and normal.
**Grade II** are generally infiltrative and with a risk of recurrence despite low-level proliferative activity. Some type II tumours tend to progress to higher grades of malignancy (low-grade diffuse astrocytomas that transform to anaplastic astrocytoma and glioblastoma; oligodendroglioma and oligoastrocytomas). **Grade III** is applied to lesions with histological evidence of malignancy, including nuclear atypia and brisk mitotic activity. In most settings, patients with grade III tumours receive adjuvant radiation and/or chemotherapy. WHO **grade IV** is assigned to cytologically malignant, mitotically active and necrosis-prone neoplasms, some of them accompanied by a widespread infiltration of surrounding tissue and a propensity for craniospinal dissemination. Grossly, they are ill-defined and surrounded by edema. They are typically associated with rapid pre- and postoperative disease evolution and a fatal outcome (glioblastoma, most embryonal neoplasms and many sarcomas) (Figure 4 on page 9).

Primary neoplasms are divided into 6 major categories (Figure 2 on page 7, Figure 3 on page 8). The largest is Tumors of Neuroepithelial tissue. As we mentioned before, the neuropil mostly consists of neuron and glial cells. Each glial cell type gives rise to a specific type of Glioma (Astrocytomas, Oligodendroglial tumors, Ependymal tumors, Choroid plexus tumors) of which Astrocytomas are by far the most common and subjected to thorough investigation.

In this presentation we have a look at those other tumors originating from non-glial cells or in which glial cells don’t have such a histological predominance. Apparently rare tumors, some of them are quite frequent and highly malignant, as Medulloblastoma, or are associated with prevalent diseases, as Ganglyonic tumors in epilepsy. They used to be classified based on clinical and radiological criteria into subgroups where those better defined and prone to a less traumatic curative treatment (surgical resection, radiosurgery) are separated from those of a diffuse and infiltrating nature, in which surgery may not be as beneficial. Neuronal and mixed neuronal-glial tumors, pineal region tumors, embryonal tumors, meningeal tumors, tumors of cranial nerves, lymphoma, hemangioblastoma, germ cell tumors, sellar region tumors and pseudotumoral vascular lesions (cavernoma) as long as lesions affecting the CNS by external compression (arising from the skull, cerebrospinal fluid (CSF) spaces or extraaxial nervous and vascular elements) are included. We also give a quick look at metastases, although not originating from nervous tissue strictly speaking, they account for near 50% of adult brain neoplasms.

All of them are pictured in detail, facing them from a histological point of view and with a thorough clinical and neuroimaging correlation that will try to provide us with a comprehensive guide for a correct diagnostic approach. Histologic diagnosis of CNS tumors in surgical or autopsy specimens usually requires immunohistologic techniques using antibodies to tumor-specific antigens as a supplementation of standard histologic stains (Figure 5 on page 10).

For educational purposes, we classify non-glial tumors in 11 main groups:
1. Neuronal and mixed neuronal-glial tumors.

2. Tumors of the pineal region.

3. Embryonal tumors.

(These three first groups belong to the group Tumors of Neuroepithelial tissue).

4. Tumors of cranial nerves.

5. Tumors of the meninges.


7. Lymphoma.

8. Germ cell tumors.

9. Tumors of the sellar region.

10. Metastatic tumors.

11. Other tumors (includes tumors not classified into de WHO groups, as Epidermoid cyst, Chordoma or Cavernous angioma, or those belonging to an isolated group, as Haemangioblastoma, classified as the only neoplasm into the "other neoplasms related to the meninges"). All of them display very particular neuroimaging features and clinical interest that make them deserve to be included in this presentation.
Fig. 0

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### Tumours of Neuroepithelial Tissue

<table>
<thead>
<tr>
<th>Tumour Type</th>
<th>Subtypes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Astrocytic tumours</td>
<td>Pilocytic astrocytoma, pilomyxoid astrocytoma, subependymal giant-cell astrocytoma, diffuse astrocytoma, fibrillary astrocytoma, gemistocytic astrocytoma, pleomorphic xanthoastrocytoma, anaplastic astrocytoma, giant cell glioblastoma, glioblastoma, gliomatosis cerebri</td>
</tr>
<tr>
<td>Choroid plexus tumours</td>
<td>Choroid plexus papilloma, atypical choroid plexus papilloma, choroid plexus carcinoma</td>
</tr>
<tr>
<td>Other neuroepithelial tumours</td>
<td>astroblastoma, choroid glioma of the third ventricle, angiocentric glioma</td>
</tr>
<tr>
<td>Neuronal and mixed neuronal-glial tumours</td>
<td>dysplastic gangliocytoma of cerebellum (Lhermitte-Duclos), desmoplastic infantile astrocytoma, ganglioglioma, dysembryoplastic neuroepithelial tumour, gangliocytoma, ganglioglioma, anaplastic ganglioglioma, central neurocytoma, extraventricular neurocytoma, cerebellar liponeurocytoma, papillary glioneuronal tumour, Rosette-forming glioneuronal tumour of the fourth ventricle, paraganglioma</td>
</tr>
<tr>
<td>Tumours of the pineal region</td>
<td>pineocytoma, pineal parenchymal tumour of intermediate differentiation, pineoblastoma, papillary tumour of the pineal region</td>
</tr>
<tr>
<td>Embryonal tumours</td>
<td>medulloblastoma, desmoplastic/nodular medulloblastoma, medulloblastoma with extensive nodularity, anaplastic medulloblastoma, large cell medulloblastoma, CNS primitive neuroectodermal tumour, CNS neuroblasticoma, CNS ganglioneuroblastoma, medullopithelioma, ependymoblastoma, atypical teratoid/rhabdoid tumour</td>
</tr>
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</table>

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**Fig. 0**

Fig. 0

### WHO Grading of Tumours of the Central Nervous System.

<table>
<thead>
<tr>
<th>Neuronal and mixed neuronal-glial tumours</th>
<th>I</th>
<th>II</th>
<th>III</th>
<th>IV</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gangliocytoma</td>
<td>X</td>
<td></td>
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<tr>
<td>Dysembryoplastic neuroepithelial tumour</td>
<td></td>
<td>X</td>
<td></td>
<td></td>
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<tr>
<td>Central neurocytoma</td>
<td></td>
<td></td>
<td>X</td>
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</tbody>
</table>

#### Pineal tumours

| Pineocytoma                             | X |     |     |    |
| Pineoblastoma                           |   |     | X   |    |

#### Embryonal tumours

| Medulloblastoma                          |   |     |     | X  |
| CNS primitive neuroectodermal tumour (PNET) |   |     | X   |    |
| Atypical teratoid / rhabdoid tumour      |   |     |     | X  |

<table>
<thead>
<tr>
<th>Tumours of the cranial and paraspinal nerves</th>
<th>I</th>
<th>II</th>
<th>III</th>
<th>IV</th>
</tr>
</thead>
<tbody>
<tr>
<td>Schwannoma</td>
<td>X</td>
<td></td>
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<td></td>
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<tr>
<td>Malignant peripheral nerve sheath tumour (MPNST)</td>
<td></td>
<td></td>
<td>X</td>
<td>X</td>
</tr>
</tbody>
</table>

#### Meningeal tumours

| Meningioma                                 | X |     |     |    |
| Atypical meningioma                        |   |     |     | X  |

| Anaplastic / malignant meningioma           |   |     |     | X  |

| Haemangioblastoma                          | X |     |     |    |

#### Tumours of the sellar region

| Craniopharyngioma                          | X |     |     |    |

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**Fig. 0**

### Major Tumor-Related Immunohistologic Stains

<table>
<thead>
<tr>
<th>Protein</th>
<th>Astrocytic tumors</th>
<th>Ependymoma</th>
<th>Oligodendroglioma</th>
<th>Neuronal/Glial tumors</th>
<th>Neuronal/Glial tumors</th>
<th>Medulloblastoma</th>
<th>Pineal gland tumor</th>
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</thead>
<tbody>
<tr>
<td><strong>GFAP</strong></td>
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<tr>
<td><strong>Vimentin</strong></td>
<td>Meningioma</td>
<td>Hemangiendoblastoma</td>
<td>Hemangiopericytoma</td>
<td>Choroid plexus papilloma</td>
<td></td>
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<tr>
<td><strong>Chromogranine</strong></td>
<td>Neuroendocrine tumors</td>
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<tr>
<td><strong>S-100 protein</strong></td>
<td>Astrocytic tumors</td>
<td>Ependymoma</td>
<td>Oligodendroglioma</td>
<td>Neuronal/Glial tumors</td>
<td>Choroid plexus papilloma</td>
<td>Neurinoma</td>
<td>Neurofibroma</td>
</tr>
<tr>
<td><strong>Synaptophysin</strong></td>
<td>Neuronal/Glial tumors</td>
<td>Medulloblastoma</td>
<td>Pineal gland tumor</td>
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<tr>
<td><strong>Cytokeratin</strong></td>
<td>Craniopharyngioma</td>
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<tr>
<td><strong>EMA</strong></td>
<td>Meningioma</td>
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<tr>
<td><strong>PLAP</strong></td>
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<tr>
<td><strong>AFP</strong></td>
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<td></td>
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<td></td>
<td>Embryonal carcinoma</td>
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</tr>
</tbody>
</table>

GFAP, glial fibrillary acidic protein; EMA, epithelial membrane antigen; PLAP, placental alkaline phosphatase; AFP, α-fetoprotein.


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**Fig. 0**

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Imaging findings OR Procedure details

Imaging findings:

1. Neuronal and mixed neuronal-glial tumors:

This group comprises relatively benign lesions (WHO grades I and II), more frequently encountered in children and young adults. Symptoms of long-standing increased intracranial pressure (ICP) are common in the clinical setting.

1.1 Dysembryoplastic neuroepithelial tumor (DNET) (WHO grade I).

Initially described by Daumas-Duport et al in 1988, it is considered an hamartomatous, low-grade lesion that constitutes 1.5% of all pediatric intracranial tumors. It is commonly located in the cerebral cortex (more often in the mesial structures of the temporal lobe), where it usually manifests as medically refractory epileptic syndrome. At diagnosis, the patients are often in the 2nd or 3rd decade.

**Histology:** it is characterized by a glioneuronal element composed of bundles of axons oriented perpendicularly to the cortex and surrounded by a specific oligodendroglia-like cell element distributed within a mucinous matrix in which normal and dysplastic ganglion-like neurons appear to be floating (“floating neurons”). Foci of cortical dysplasia are associated with the tumor (Figure 1 on page 35).

**Neuroimaging:** On CT they appear as hypo- or isodense lesions with calcifications in up to 36% and on MR as well-defined nodular lesions, hypointense on T1 and hyperintense on T2 relative to gray matter. Due to its long-standing nature, calvarial scalloping is reported in around 50% of cortical based tumors. They typically cause gyrus enlargement and gray-white matter interface blurring, with cysts seen in around 30% and focal enhancement in up to 66% (so the neuroimaging spectrum varies from a macrogyric to a pseudo- or multicystic appearance). The absence of mass effect and no peritumoural edema are important criteria for differentiating between DNETs and gliomas (Figure 2 on page 35, Figure 3 on page 36).

It has an excellent prognosis without progression over many years even after subtotal resection. (FIGURE 4 on page 37)

1.2 Gangliocytoma (WHO grade I).

Gangliocytoma (GC) is a rare neuronal tumor, in several aspects closely related to Ganglioglioma (GG). Both commonly considered together, GC and GG represent 0.4%
of all CNS tumors and 1.3% of all brain tumors. It usually appears in children and young adults (mean age at diagnosis: 8.5-25 years) with a slight female predominance. Its common site is the cerebral hemispheres (temporal and frontal lobes). Symptoms vary according to tumor size and site. They are the most common tumors associated to chronic temporal lobe epilepsy.

**Histology:** Although far less common than GG, we focus on GC due to its composition, based on a large neoplastic, mature ganglion cell population within a fibrillary matrix, wether GG contains a mixture of neoplastic neurons and also glial cells, mostly astrocytes. We can consider them opposite ends into the differentiated ganglion cell tumors group, in which GC corresponds to the non-aggressive WHO grade I lineage (Figure 5 on page 38).

**Neuroimaging:** On imaging it is indistinguishable from GG, appearing as a well-defined solid or mixed solid-cystic mass (cyst with a mural nodule) with minimal or no mass effect, of low-intensity on T1 and high-intensity on T2, and common enhancement after Gadolinium administration that varies in intensity and pattern (solid, rim or nodular) (Figure 6 on page 39, Figure 7 on page 40). Calcifications are common. Some author refers the presence of a dural tail mimicking menigioma in those more cortically located.

(FIGURE 8 on page)

### 1.3 Central neurocytoma (CN) (former terms: Intraventricular oligodendroglioma, Ependymoma of the foramen of Monro) (WHO grade II).

CN comprises approximately 0.25-0.5% of all intracranial tumors. It is predominant in the 2nd - 4th decades, with no gender predilection. It is typically located in the anterior portion of one of the lateral ventricles (with a preference for the left) near the foramen of Monro, attached to the septum pellucidum an with an intraventricular growth. Increased ICP symptoms due to obstructive hydrocephalus conform the prominent clinical spectrum. The term central neurocytoma should be restricted to neoplasms located within the cerebral ventricles. The term extraventricular neurocytoma is now given to neoplasms that arise within the CNS parenchyma and share histological features with the more common CN.

**Histology:** This rare tumor consists of a small, round and uniform neuronal cell population, embedded in clusters in a variably developed fibrillary matrix, in which the formation of an artificial perinuclear halo is the histological hallmark. Typical neuronal ultrastructural characteristics along with the expression of neuronal marker proteins in immunohistochemical studies allow the differentiation with other tumors with which share some histologic features, as oligodendroglioma or ependymoma (Figure 9 on page 42).
**Neuroimaging**: On CT, it appears as a lobulated mass iso- or slightly hyperdense relative to gray matter, with calcifications in up to 50% and a mild to moderate irregular enhancement. On MR, CN is an heterogeneous lesion, iso-to hypointense relative to cortex in T1 and iso- to hyperintense on T2. Small tumor cysts and hemorrhage may be present. Some CN may disclose anaplastic features (Figure 10 on page 43).

Surgery is the treatment of choice, though complete tumor resection is accomplished in around 35% of cases due to the difficulty that implies the intraventricular location and often large tumor size. Radiotherapy should be the choice in those cases. Early #-knife surgery seems to be a treatment of choice early in small tumor remnants after incomplete resections, minimizing the risks of conventional radiotherapy. (FIGURE 11 on page 44).

1.4 Paraganglioma (Jugulotympanic chemodectoma, glomus jugulare tumour, jugular glomus tumour, glomus tympanicum tumour, tympanic glomus tumour).

This tumor arises from glomus bodies composed of chemoreceptor cells derived from primitive neural crest (paraganglion) of an adrenal or extra-adrenal origin. The extra-adrenal paraganglia can be divided into sympathetic and parasympathetic types. Although they are indistinguishable at the cellular level, they differ in their anatomic distribution and secretory products. Parasympathetic paraganglia are localized almost exclusively in the head and neck along the branches of the glossopharyngeal and vagus nerves (main sites are at the bifurcation of the common carotid artery, in the middle ear - temporal bone, along the course of the vagus nerve, and exceptionally in the orbit, nasal cavity, paranasal sinuses, nasopharynx, larynx, trachea and thyroid).

Of our interest in this presentation are those located in the jugular foramen area (glomus jugulare paraganglioma) or in the promontorium of the middle ear cavity (glomus tympanicum paraganglioma). The distinction between jugular and tympanic paragangliomas can easily be made by modern imaging methods with which the jugular neoplasm is identified as arising from the jugular bulb region and may show evidence of invasion of the petrous bone, while the tympanic neoplasm is confined to the middle ear. Jugular paraganglioma is the most common jugular foramen tumor and the second most common temporal bone tumor (tympanicum paraganglioma is the first). It may be multicentric (5-10% of sporadic cases) or coexist with tumours of other types. They may be bilateral in the same patient and coexist with carotid body paragangliomas which may also be bilateral.

Most patients present with conductive hearing loss. Pain in the ear, facial palsy, haemorrhage, and tinnitus are also described. On examination, a red vascular mass is seen either behind the intact tympanic membrane or sprouting through the latter into the external canal. Surgical approach to the mass at biopsy often results in severe bleeding.
Histology: In the jugular variety, the petrous temporal bone and the middle ear space are largely replaced by red, firm material as far as the tympanic membrane. The otic capsule is rarely invaded by paraganglioma. The histological appearances of the jugular and tympanic paragangliomas are similar, resembling that of the carotid body paraganglioma. Epithelioid, small, uniform cells, with finely granular cytoplasm are separated by numerous blood vessels. The tumour cells often form clusters or "Zellballen" with peripheral flattened cells. Nuclei are usually uniform and small, but diagnosis is sometimes made difficult by the presence of bizarre or multinucleate cells which, however, do not indicate malignancy. A prominent fibrous stroma is sometimes present (Figure 12 on page).

Neuroimaging: The glomus jugulare paraganglioma appears as a jugular foramen mass with permeative bone changes along superolateral margin of jugular foramen with common erosion of the jugular spine on CT. Jugulotympanicum paragangliomas depict clearly the portion of tumor curving up from jugular foramen through middle ear floor, terminating on cochlear promontory (coronal images) (Figure 13 on page 46). On MR, it shows a classic "salt and pepper" appearance on T1 due to the high-velocity flow voids and the focal hemorrhagic areas shown inside the lesion on the different sequences ("pepper" refers to multiple black dots representing high flow voids and "salt" refers to hyperintense foci representing hemorrhage or slow flow), and a heterogeneous hyperintensity on T2 with multiple hypointense foci ("pepper"). They are hypervascular lesions that show an avid enhancement after contrast administration (Figure 14 on page 47).

(FIGURE 15 on page 48)

2. Tumors of the pineal region.

The term "pineal region tumors" includes both the neoplasms originating from the pineal body and those originating from the adjacent structures. We focus on the former, specifically in the neoplasms of the pineal parenchyma (1% of all primary intracranial tumors, and 15-30% of all tumors in the pineal region), as a second main group of neoplasms, the germ-cell tumors, will be discussed later.

2.1 Pineocytoma (WHO grade I).

It is a slowly growing, benign tumor originating from mature parenchymal cells with neuroendocrine and photosensory functions. Representing around 45% of all pineal parenchymal tumors, it is more common in adolescence and adulthood. It accounts for less than 1% of all intracranial neoplasms. It is located in the pineal region and rarely extends into the IIIrd ventricle (it compresses but does not invade adjacent structures). It generally causes increased ICP symptoms due to obstructive hydrocephalus or impairment of ocular movements, as Parinaud’s syndrome (upward gaze palsy, related
to compression of the tectum of the midbrain, with three main neuronal structures near
to the pineal gland: the posterior comissure, the rostral interstitial nucleus of the medial
longitudinal fasciculus, and the interstitial nucleus of Cajal). Also endocrine dysfunction
can be present, due to compression of the wall of the IIIrd ventricle. Pineocytoma is less
prone than Pineoblastoma to seed the CSF, having a 86% 5-year survival rate.

**Histology:** Pineocytoma is composed by uniform, mature cells resembling pineocytes
with a small nucleus and a moderate amount of cytoplasm containing secretory granules,
forming lobules or rosettes around large acellular zones of fibrillarity, that resemble
Homer-Wright rosettes (pineocytomatous rosettes) (Figure 16).

**Neuroimaging:** Imaging of pineocytoma may be nonspecific, sharing most features with
other pineal parenchymal tumors and even with germ-cell tumors of the pineal region.
On CT, it appears as a iso- or hypodense well-defined mass, less than 3 cm in diameter,
that tipically "explodes" the pineal calcium to its periphery. Cysts can be present and
prevail, mimicking a pineal cyst. Enhancement is variable, typically heterogeneous. On
MR, it appears as iso- to hypointense on T1 and hyperintense on T2 and FLAIR. T2* GRE
depicts the “blooming” artifact due to the presence of peripheral calcium in many cases.
Enhancement is common, solid or peripheral. Hydrocephalus is a common associated
finding (Figure 17 on page 51, Figure 18 on page 50).

2.2 Pineoblastoma (WHO grade IV).

It is the highly malignant version of the tumors of the pineal parenchyma, composed by
immature neoplastic cells. It behaves as a fast growing, invasive mass, with tendency to
seed within the subarachnoid space and that may contain hemorrhage and necrosis. It
is more common in children an adolescents. It shares with pineocytoma the main clinical
symptoms.

**Histology:** It is a highly cellular lesion, composed by small cells with round to somewhat
irregular nuclei and scant cytoplasm (high nuclear:cytoplasmic ratio), arranged in
sheets or forming rosettes. Pineoblastomas resemble other small cell, or primitive
neuroectodermal tumors of the CNS ("pineal PNET").

**Neuroimaging:** It overlaps considerably with pineocytoma and germ cell tumors. They
tend to be larger (# 3 cm), lobulated, heterogeneous, and with poorly delineated margins
than pineocytoma. Calcifications are less frequent. On CT, it depicts as a mixed tumor
with a frequently hyperdense solid portion. On MR it appears as a heterogeneous mass,
iso- or hypointense on T1 and iso- to minimally hyperintense to gray matter on T2. Other
MRI-associated techniques, as Diffusion weighted imaging (DWI) can be useful in the
differential diagnosis with pineocytoma and other less agressive lesions. DWI usually
demonstrates water restriction (hyperintensity on DWI image and hypointensity on ADC
maps), due to its high cellularity and high nuclear:cytoplasmic ratio.
Pineoblastoma has a poor prognosis. Surgery plus cranial/spinal radiation and chemotherapy is the optional treatment.

(FIGURE 19 on page 52).

3. Embryonal tumors.

3.1 Medulloblastoma / Primitive Neuroectodermal Tumor (PNET) (WHO grade IV).

Medulloblastoma was named by Baily and Cushing in 1925. It is a malignant, invasive embryonal tumor with neuronal or glial differentiation, and an inherent tendency to metastasize via CSF pathways. Medulloblastoma and PNETs are the main entities into the group of embryonal tumors according to the new WHO classification, all of them characterized by their high aggressiveness (grade IV). In fact, medulloblastoma has been commonly referred as a PNET of the posterior fossa. Recent investigations support the theory that medulloblastoma has more than one cell of origin, one associated with the ventricular system, and the other derived from the ventricular matrix and the external granular layer of the cerebellum. They tipically occur in children (peak age at presentation, 7 years), being considered the most common malignant CNS tumor (25% of all pediatric brain tumors) and the most common posterior fossa tumor (up to 38% of all posterior fossa tumors) at this age range.

Cerebellar vermis is the usual site of origin (85% of cases), with a higher tendency to grow from the lateral hemispheres in older children and adults. Hydrocephalus and signs and symptoms related to a rapid increase in the ICP and cerebellar destruction (headache, morning vomiting with or without nausea, papilledema, ataxia, cranial nerve palsy) are the common clinical onset. Signs seen in babies include macrocephaly, rapid head growth, bulging anterior fontanelle, downward deviation of the eyes (sundowning), and irritability.

**Histology:** This highly cellular tumor is characterized by densely packed round cells with high nuclear-to-cytoplasmic ratio (*blue-cell tumor*), conforming neuroblastic (Homer-Wright) pseudorosettes in < 40% of cases. As for other highly malignant tumors, mitoses and apoptosis are frequent, and necrosis commonly encountered. It is highly vascular, with its principle vascular supply arising from the posterior inferior cerebellar artery (PICA). Four different histological subtypes are recognized, with increasing grade of malignancy: *Medulloblastoma with extensive nodularity* (also known as cerebellar neuroblastoma, can be seen as an intracranial manifestation of Gorlin syndrome) has an expanded lobular architecture and is characterized by a higher degree of neuronal differentiation and a better prognosis; *Desmoplastic* (10-20% of cases) consists of paucicellular islands of well-differentiated cells surrounded by large amounts of reticulin and collagen, as well as more indifferntiated cells; *Classic* (75% of cases), consists of
sheets of small blue cells; and **Large cell/Anaplastic medulloblastoma** (5%), showing areas of large cells with anaplasia (Figure 20 on page 53).

**Neuroimaging:** Medulloblastoma shows signs of hypercellularity and high aggressiveness. On CT, classic medulloblastoma in children appears as a well-defined and hyperdense mass of the cerebellar vermis (85% of cases) with surrounding edema and occasional cyst formations and calcifications (5-10%). Homogeneous enhancement is commonly found. On MR, an iso- or hypointense mass relative to white matter in T1 is the rule, with a variable degree of heterogeneity in T2. After Gadolinium administration, irregular enhancement in the solid portions of the mass is the most common finding. MRI is an excellent way to determine if the tumor has invaded the floor of the fourth ventricle or extended out through the outlets of the fourth ventricle. DWI clearly depicts its high cellularity (high nucleus/cytoplasma ratio) as increased diffusion restriction (increased signal on DWI images and low signal ADC maps) in the solid portions of the tumor (Figure 21 on page 54, Figure 22 on page 55, Figure 23 on page 56).

Magnetic Resonance Spectroscopy (MRS) may show a Choline (Cho) increase, N-acetylaspartate (NAA) decrease, and occasional increase of lipids and lactate.

CT and MR appearances can vary according to the histological type. Medulloblastoma in adults can more frequently manifest as a poorer-defined hemispheric mass in which cyst formations are much more commonly seen.

Leptomeningeal seeding is a common risk that occurs in up to 33%-40% of medulloblastomas at the time of diagnosis. It carries a poorer prognosis, and a thoroughful seek by an extensive contrast-enhanced screening of the skull and spine is mandatory before treatment protocol. Sulcal and cisternal effacement, and diffuse and nodular enhancement of the nerve roots, spinal and tentorial surface and thecal sac are the common findings on CT and MR after contrast administration (Figure 24 on page 57).

Ependymoma is the main differential diagnosis in the fourth ventricular region. Ependymoma is a glial tumor prone to calcification, often more heterogeneous and malleable, anchored to the floor of the fourth ventricle. Other tumors to include in the differential diagnosis are the other subtypes of PNET (Ependymoblastoma, Medulloepithelioma), Atypical teratoid/rhabdoid tumor, Glioblastoma multiforme, and those expressing high cellularity, as metastatic small-cell carcinoma of pulmonary origin, or primary CNS lymphoma. Patients younger than 3 years at diagnosis, CSF dissemination, and bulky residual disease after surgery have poorer prognosis. Combination of surgery and radiation is the most common therapy. However, the side effects derived from radiation use (development of telangiectasia, cavernous malformation, mineralizing microangiopathy, intellectual deterioration, growth retardation) have led to the avoidance of craniospinal radiation in children younger than 2 years unless there is documented evidence of CSF dissemination or recurrence. The advent of chemotherapy in the treatment of medulloblastoma in the 1980s has been
associated with an increase in survival rates in high risk children and in patients with recurrent or advanced disease. Before that time, the overall 5-year survival rate was 2-30%. Since then, individual reports have described improved 5-year survival rates between 50-70% and over 80% in some reports.

**Other PNETs.**

Hart and Earle described PNETs in 1973 as tumors similar to Medulloblastomas in morphology but occurring outside the cerebellum. In 1993 the WHO recommended to classify the PNETs and Medulloblastomas under a single category, namely PNET. Nowadays it has become clear that the prognosis and molecular features of supratentorial PNETs (sPNET) are different from the infratentorial PNET or Medulloblastoma (Figure 25 on page 58). For these reasons they are classified separately although into the same group of Embryonal tumors. PNETs are composed of undifferentiated neuroepithelial cells with a capacity of differentiation to astrocytic, ependymal, neuronal, muscular or melanotic tissue. They are also more common in childhood (1% of all pediatric brain tumors), with a median age at diagnosis of 35 months. They usually appear as large, heterogeneous hemispheric masses with minimal peritumoral edema and prone to disseminate by subarachnoid space. Calcification, hemorrhage and necrosis are common. Histologically they appear quite similar to medulloblastoma, with which also share its high aggressiveness (WHO grade IV), harboring a poorer prognosis (30-35% 5-year survival rate).

**Ependymoblastoma:** is a highly malignant embryonal tumor of infancy and young childhood (WHO grade IV). Histologically, the tumor cells are arranged in multilayered rosettes consisting of an outer rim of tumor cells merging with the surrounding undifferentiated neuroectodermal cells. It usually presents in a paraventricular supratentorial location, although it may arise in the posterior fossa, or as a primary leptomeningeal or extradural sacrococcygeal tumor. Infratentorial ependymoblastomas present with signs and symptoms of increased ICP and cerebellar signs (coordination symptoms). Its imaging appearance is similar to those of other PNETs, with a large, heterogeneous, well-circumscribed mass with occasional calcification or hemorrhage and no significant surrounding edema.

**Medulloepitelioma:** It is a rare tumor of neonates and young children, in which primitive epithelial cells are arranged in a tubular or papillary fashion that resembles the epithelial lining of the embryonic neural tube.

*(FIGURE 26 on page 59)*

**3.2 Atypical Teratoid-Rhabdoid tumor (AT/RhT) (WHO grade IV).**

Defined in 1987, AT/RhT is a highly malignant, embryonal CNS tumor of the early childhood. It appears as a bulky, heterogeneous tumor in children < 3 years, 50%
infratentorial and off-midline (cerebello-pontine angle, cerebellar hemisphere) and 40% supratentorial. In 15-20% of cases it appears as a disseminated disease.

**Histology:** It contains rhabdoid (eosinophilic, homogeneously stained cytoplasm, and an eccentric nucleus with vesicular chromatin structure and prominent nucleolus) cells with or without primitive neuroectodermal tumor-like areas (may resemble medulloblastoma/PNET) and/or tumor cells showing epithelial, mesenchymal, glial, or neuronal differentiation (which can mimic the appearance of carcinoma, sarcoma, or glioma, respectively) (Figure 27 on page , Figure 28 on page ).

**Neuroimaging:** On CT it usually appears as a hyperdense mass, commonly containing cysts, hemorrhage or calcification, and with an heterogeneous enhancement (Figure 29 on page 62). On MR it shows as an heterogeneous lesion, depending on the amount of cysts, hemorrhage and calcium (the latter two with the typical hypointense blooming artifact on GRE-T2*), and with the typical appearance of an hypercellular tumor in the solid portion (water restriction on DWI) (Figure 29 on page 62, Figure 30 on page 63). Leptomeningeal seeding depicts as diffuse lineal or multiple nodular enhancement of the CSF spaces.

AT/RhT has a very poor prognosis, with an overall death rate of 85% and a median survival of 6 months.

(FIGURE 31). on page 64

4. Tumors of cranial nerves.

Several tumors arise from the cranial or peripheral nerve sheaths: schwannomas, neurofibromas, perineurinomas and malignant peripheral nerve sheath tumors (MPNST). We focus on the by far most common tumor of the intracranial nerves, the schwannomas.

4.1 Schwannoma (Neurinoma, Neurilemoma) (WHO grade I).

Schwannoma is an encapsulated benign tumor that account for 8% of intracranial tumors, developing in most cranial nerves except I and II (which do not have Schwann cells). They appear most often in adults (70% before age 30), with no gender predilection. It is the second most common intracranial extraaxial tumor. Vestibular schwannoma accounts for 95% of intracranial schwannomas although neural sheath tumor can develop in all the other nerves of the posterior fossa, especially the trigeminal, facial, and nerves IX to XII. Cranial nerves schwannomas have a higher incidence in Neurofibromatosis (NF) type 2. Symptoms depend principally on the nerve associated with the tumor and the size of the tumor (facial neuropathy in trigeminal schwannomas; vertigo, tinnitus and hearing loss in vestibular schwannomas; swallowing difficulties in caudal nerves schwannomas). Large schwannomas may compress the brainstem as in cases of vestibular schwannomas.
with a large extension out of the internal auditory canal (IAC), when most of the tumor is located in the cerebellopontine angle (CPA). Most nerve tumors are benign (WHO grade I). Although schwannomas associated with NF 2 may have a higher proliferative activity, this does not indicate malignant behaviour. In contrast, MPNST or malignant schwannomas represent WHO grades III and IV tumors.

**Histology:** Schwannomas involve sensory nerves more frequently than motor roots. The vestibular schwannoma arises from the vestibular division of the VIII nerve root. It is a well-encapsulated tumor composed by differentiated neoplastic Schwann cells in which two histological patterns are distinguished: The Antoni A schwannoma, with spindle-shaped cells with bipolar processes arranged in interwoven fascicles, with unusual mitosis and nuclei aligned in rows giving a palisading pattern. Palisades around eosinophilic fibrillar areas form the typical Verocay bodies; and Antoni B schwannoma with a loose structure and stellate and fusiform cells in a mucoid matrix. Cysts, hemorrhage and necrosis are common findings (Figure 32 on page ).

**Neuroimaging:** On CT appears as a iso- to hyperdense extraaxial mass, with occasional intratumoral cysts or hemorrhage, variable calcification, and strong homogeneous postcontrast enhancement. MR depicts the solid portion as iso- or mixed iso/hypointense on T1 and hyperintense on T2 with no water restriction on DWI and common peritumoral edema. After contrast an strong solid enhancement is seen in 70% of cases, and ring-enhancement in 30%. Vestibular schwannomas usually show the typical enhancing "icecream cone" lesion (Figure 33 on page 66). The anatomic location of the tumor, their extension following the course of the nerves, and the enlargement of their foramina are key features for diagnosis.

Schwannoma is a slowly growing tumor with exceptionally rare malignant degeneration. Microsurgical resection or radiosurgery are the elective treatment. Postsurgical recurrence rate < 10% has been reported.

(Figure 34 on page 67)

5. **Tumors of the meninges.**

Although we find them in a same group into the 2007 WHO classification, we separate the Meningothelial tumors of the meninges (meningioma), from the mesenchymal tumors (lipoma, condroma, etc..), for a practical radiological purpose.

5.1 **Meningioma** (WHO grades I-III).

The term *meningioma* was coined in 1938 by Harvey Cushing to describe a benign tumor arising from the meninges of the CNS. Meningioma is the most common non-glial
primary tumor of the CNS and the most common extraaxial neoplasm. They represent approximately 15% of all intracranial tumors. They are usually benign (grade I) and have been found to arise from the arachnoid cap or cluster cells in close proximity to the dura mater (that is, the meningothelial cells of the arachnoid) known to be most prevalent near collections of arachnoid villi at the dural venous sinuses and large tributary veins (parasagittal/convexities), over the cribriform plate (olfactory plate) and medial regions of middle cranial fossae (sphenoid ridge). Its appearance as a bulky extraaxial lesion attached to the dura mater allows an easy identification and diagnosis in most cases. In 9-15% of cases it is located in the posterior fossa, including the CPA (2-4%), the clivus (<1%), the cerebellar convexity (5%), or the tentorium cerebelli (2-4%). In the CPA, meningioma is the second most common mass lesion after vestibular schwannoma. Intraventricular meningiomas are rare tumors thought to arise from meningothelial cells located in the choroid plexus or tela choroidea. Meningiomas occur two to three times more commonly in female patients, especially those in the 4th-6th decades. Patients with NF-2 have an increased risk of meningioma. Less commonly, meningiomas show anaplastic or atypical histological features that are associated with an increased likelihood for recurrence and/or aggressive behaviour.

**Histology:** The histology of meningioma is variable. The WHO classification includes 9 different histological variants with a benign grade I behaviour, including the meningothelial or syncytial, fibrous/fibroblastic and transitional variants and 6 more aggressive and with a higher tendency to recurrence, as the anaplastic, papillary and rhabdoid meningiomas (grades II-III). 85-90% of all meningiomas belong to the benign group. The meningothelial meningioma consists of cells with round to oval nucleus with scanty chromatin, homogenous cytoplasm, and poorly defined cellular membrane. Some nuclei appear vacuolated from cytoplasmic invagination (pseudoinclusion). The cells form whorls separated by a fine reticulin network. Psammoma bodies may be also seen. (Figure 35 on page  ).

**Neuroimaging:** On CT, they appear as bulky and well-circumscribed homogeneously hyperattenuating masses with a broad-based dural attachment. After contrast, it enhances homogeneously. It may be associated to hyperostosis of the adjacent skull. Necrotic and cystic changes are rare. Calcification can be seen in 20-25%. On MR, meningioma shows the typical characteristics of extraaxial masses, including the sharply defined margins, the inward displacement of the cortical gray matter, or the entrapment of cortical vessels and CSF clefts. They are iso- to hypointense on T1, iso- to hyperintense on T2, and enhances homogeneously after Gadolinium administration. Meningiomas often compress and displace but do not usually invade the adjacent brain tissue (Figure 36 on page 69, Figure 37 on page 70). In the CPA, it usually doesn’t involve the ICA, which is a very common characteristic of schwannomas. Meningioma en plaque or intraosseous meningioma describe a "flat" meningioma with a tendency to invade through the skull, usually located in the sphenoid wing and affecting the orbit (Figure 38 on page 71).

6.1 Lipoma (WHO grade I).

Intracranial lipoma is a rare tumor, comprising 0.1-0.5% of all primary brain neoplasms. Interhemispheric lipomas are the most frequent (40-50% of all intracranial lipomas), commonly located in the subarachnoid space and associated with dysgenetic corpus callosum. Other sites to be found are the CPA, chiasmatic or perimesencephalic cisterns, pineal region, sylvian fissure or in close apposition to the brainstem, especially the cuadrigeminal plate.

Although rarely symptomatic, they can manifest with seizures and headache.

**Histology**: Lipoma represents a developmental malformation of the CNS, resulting from a lipomatous differentiation of the persistent meninx primitiva, the mesenchymal derivative of the neural crest which envelops the developing embryo, and which forms the subarachnoid space after its reabsorption. This is why they are usually found in the close vicinity of the subjacent brain tissue, commonly traversed by the vessels and cranial nerves.

**Neuroimaging**: Lipomas appear as fat, with a negative attenuation value on CT and homogeneous high signal on T1-images on MR (isointense to orbital or subcutaneous fat), which decreases on fat-suppressed images, and hypointense and frequently more heterogeneous on T2. They do not enhance after contrast administration (Figure 40 on page ). Lipoma has a very low proliferative activity and a very good prognostic outcome even without surgery.

(FIGURE 41 on page 74).

6.2 Chondroma (WHO grade I) and Chondrosarcoma (WHO grades I-IV).

Benign and circumscribed Chondroma and its malignant variant Chondrosarcoma are rare tumors of the skull. They develop from mesenchymal cells or embryonic cartilaginous remnants enclosed in the bones of the skull base (clivus, skull base). They arise as lytic bone lesions from the paramedian synchondrosis (petrous apex or sphenoid ridge) and cause symptoms by involvement of cranial nerves and mass effect on the brain stem and cerebellum (a similar presentation to chordoma with which share a similar biological behaviour). They predominate in men and present within the 2nd - 3rd decades. Chondrosarcoma is the third most common malignant tumor of bone, following multiple myeloma and osteogenic sarcoma.
**Histology:** Chondroma consists of benign chondrocytes in scattered lacunae with bone being formed by enchondral ossification. Chondrosarcomas can be divided into three histologic types: Classic, mesenchymal and dedifferentiated. The classic form is the most common and characterized by large, atypical chondrocytes within a hyaline cartilaginous matrix background.

**Neuroimaging:** They appear as expansive, lobulated, soft tissue masses with curvilinear matrix calcifications. On CT, they usually appear as low density lesions with a slight to moderate enhancement. On MR, a hypointense signal on T1 and hyperintense on T2 with internal low intensity foci due to calcification is the most common finding. Lytic destruction of adjacent bones (clivus, sella, skull base) is clearly depicted on CT, and MR is preferable to evaluate invasion of posterior fossa, sellar or retropharyngeal spaces, sinonasal structures and compression of vascular elements (Figure 42).

Treatment of choice consists of total surgical resection followed by local radiation.

(FIGURE 43 on page  )

7. **Lymphoma.**

Primary CNS lymphoma (PCNSL) refers to the occurrence of craniospinal lymphoma in the absence of primary tumor elsewhere in the body. PCNSL is encountered frequently nowadays, in both immunocompromised and immunocompetent patients. Recent epidemiologic data show an increased incidence of PCNSL in low-risk immunocompetent patients. Histology usually reveals a diffuse, large-cell, intermediate- to high-grade extranodal non-Hodgkin’s lymphoma of B-cell type. It affects all ages groups with a peak incidence in the 5th-7th decades in the non-AIDS patients. It commonly arises as a deep periventricular white matter mass, although other periventricular locations as corpus callosum, cerebellum, brainstem, and cranial nerves can harbour it. Symptoms vary according to the location of the lesions and the immune status of the patient.

**Histology:** PCNSL usually appears as masses with porly defined margins, reflecting their tendency to infiltrate white matter tracts including the corpus callosum. The infiltrating B-cells follow the walls of the ventricular system from which they likely invade the CSF spaces. Microscopically, there is a typical angiocentric growth pattern of cuffs of tumor cells within and around the cerebral blood vessels (Figure 44 on page , Figure 45 on page 78).

**Neuroimaging:** In immunocompetent, it usually presents as an homogeneous hyperdense enhancing mass on CT. Hemorrhage and calcification are rarely seen (more frequent in immunocompromised patients and after chemotherapy or radiation treatment).
MR imaging shows hypointense mass signal on T1, and iso- to hypointense signal relative to gray matter on T2. DWI typically depicts a restricted diffusion, with homogeneous hyperintensity in DWI and hypointense signal relative to surrounding parenchyma in ADC map. This characteristic, along with its CT and MR features and a typical homogeneous enhancement after Gadolinium (in contrast to the ring-enhancement more typical of immunocompromised patients), translate its typical histology of a high density mass with a large nucleus/cytoplasm ratio (Figure 46 on page __). Hemorrhage and necrosis may appear. Calcifications can be found after therapy. MRS usually shows an increase of Choline and decrease of NAA, with lipid and lactate peaks sometimes reported. MR-Perfusion depicts elevation of CBVr, not as much as in Glioblastoma Multiforme (in lymphoma there is a perivascular infiltration by B-cells leading to an augmented vascular permeability rather than a real neoangiogenesis) (Figure 47 on page 80).

(FIGURE 48 on page 81)

8. Germ cell tumors.

Germ cells tumors are those arising in the CNS from germ cell nests displaced during embryogenesis that are morphological and immunophenotypical homologous of those originating in the gonads or in other extragonadal sites outside the CNS. They are classified according to gonadal germ-cell neoplasms (germinoma, embryonal carcinoma, endodermal sinus tumor (yolk sac tumor), choriocarcinoma, immature teratoma, mature teratoma, teratoma with malignant transformation, and mixed germ-cell tumor), and predominate in the midline (more than 80% in the pineal and IIIrd ventricular region; suprasellar area). Multifocal germ-cell tumors usually involve the pineal region and suprasellar compartment simultaneously or sequentially. Germinoma constitutes more than half of all germ-cell tumors in the pineal region (with a male predominance) and thus is the most common type of intracranial germ-cell tumor. In general, germ-cell tumors account for less than 0,5% of all primary brain tumors, although in far-east Asia the incidence rises to 3% (Japan). The incidence peak is established between 10-12 years of age. They tend to disseminate along the CSF pathways. The typical clinical manifestations vary according to histological type and location. Those in the pineal region often compress and obstruct the aqueduct, resulting in progressive hydrocephalus with intracranial hypertension. Parinaud’s syndrome (due to invasion of the tectal plate), visual field defects, diabetes insipidus and pituitary failure and precocious puberty (due to production of human chorionic gonadotrophin - HCG- by the tumor) are other possible symptoms.

8.1 Germinoma (Dysgerminoma, extragonadal seminoma, atypical teratoma).
**Histology**: It is the homologue to testicular seminoma and ovarian dysgerminoma, respectively. It is composed by two kind of cell population: large polygonal neoplastic cells and small reactive lymphocytes. The former are relatively uniform, round cells with large vesicular nuclei, prominent nucleoli and pale, often vacuolated, glycogen rich (PAS-positive) cytoplasm. Mitoses are usually identified but necrosis is uncommon. The tumor is traversed by fibrous septa that are densely infiltrated by T-cells lymphocytes that are thought to represent host reaction to the tumor (Figure 49 on page 82). Immunohistochemistry for β-fetoprotein and HCG confirms the germ-cells origin of the tumor.

**Neuroimaging**: Germinoma usually appears as a circumscribed, dense pineal region mass on CT, that engulfs the pineal gland. It shows a prominent contrast enhancement that may extend to the walls of the IIIrd ventricle when CSF seeding occurs. Cystic, necrotic and hemorrhagic changes are not uncommon in larger tumors. On MR, Germinoma shows features of a highly-cellular tumor, with iso- to hypointensity relative to gray matter on T1 and T2, and water restriction on DWI (Figure 50 on page 83). MR examination of the entire neuroaxis should be mandatory before treatment to exclude subarachnoid dissemination.

Radiotherapy +/- adjuvant chemotherapy is the treatment of choice, with a 5-year survival rate of 91%.

**8.2 Teratoma.**

**Histology**: Teratomas are composed of an admixture of tissues derived from all three germinal layers: endodermal, mesodermal, and ectodermal differentiation. Some teratomas are composed of well-differentiated, mature tissue without mitotic activity resembling benign adult organs (mature variant). The ectodermal components commonly encountered include skin, brain and choroid plexus. The mesodermal representatives include cartilage, bone, fat and muscle. Glands lined by epithelia of respiratory or enteric type are the usual endodermal elements. Other teratomas are composed of immature tissue resembling fetal tissue (immature variant) with mitotically active stroma and primitive neuroectodermal elements. Histologically they contain a mixture of epithelial, mesenchymal and glandular elements (Figure 51 on page 84).

**Neuroimaging**: Teratomas are commonly situated in the pineal region or suprasellar area and rarely within the parenchyma. Their typical appearance on CT is that of a midline lobulated mass containing calcification, soft tissue, cysts and fat. Holocranial teratomas may be huge. Soft tissue components enhance heterogeneously. On MR teratomas depict the marked internal heterogeneity due to their varied histological composition. GRE-T2* sequence is helpful in determining calcified areas. Fat-suppressed sequences are recommended for confirmation of fatty globules (Figure 52 on page 85).

(FIGURE 53 on page 86).
8.3 Embryonal carcinoma.

**Histology**: Embryonal carcinoma consists of large, cuboidal to columnar epithelial cells that grow in sheets or chords and form abortive papillae or line irregular, gland-like spaces. Tumor cells may exceptionally replicate the structure of the early embryo forming "embryoid bodies" replete with germ discs and miniature amniotic cavities. The tumor cell nuclei are enlarged and contain prominent nucleoli. Mitotic activity is high and areas of coagulation necrosis are common.

**Neuroimaging**: This tumor has similar radiologic characteristics and location to those previously described for other germ-cell tumors. It appears as a heterogeneous mass with enhancing solid portion that may contain calcification and hemorrhagic foci. CSF dissemination may also occur.

9. Tumors of the sellar region.

The two most common tumors affecting the sellar region are pituitary adenomas and craniopharyngiomas. They appear as lobulated masses centered on the pituitary fossa that displace supra- and parasellar structures and commonly invade the cavernous sinus. When large, they can destroy the dorsum sellae and compress the anterior surface of the brainstem. Adenomas were not included in the 2000 and 2007 WHO classifications, but since they are among the most common brain tumors encountered, their most important features will be described.

9.1 Craniopharyngioma (WHO grade I).

Craniopharyngioma is a benign epithelial tumor of the sellar region related to Rahtke’s pouch (craneopharyngeal duct). Two histological forms (adamantinomatous and papillary) can be found. They represent 3% of all intracranial tumors and 5-10% of pediatric intracranial tumors (most common pediatric tumor of non-glial origin). Adamantinomatous form occurs in all age groups, whereas papillary craniopharyngiomas is virtually restricted to adults. Its usual symptoms are morning headache, visual disturbances (bitemporal hemianopsia), endocrine disturbances and those other derived from hydrocephalus.

**Histology**: Adamantinomatous form resembles odontogenic tumors (calcifying odontogenic cysts and ameloblastomas) with multistratified squamous epithelium with nuclear palisading, nodules of “wet” keratin, and dystrophic calcifications. The papillary craniopharyngioma is constituted by sheets of squamous epithelium forming pseudopapilla that cover fibrovascular cores. Keratinization and calcification are absent (Figure 54 on page ).
**Neuroimaging:** Adamantinomatous craneopharyngiomas locate in the suprasellar area in 75% of cases, in the suprasellar + intrasellar area in 21%, and exclusively intrasellar in 4% of cases. It often extends into multiple cranial areas (anterior, middle, posterior fossae, retroclival). Its appearance is that of a multilobulated and multicystic lesion, usually large (> 5 cm), in 90% of cases with a mixed structure (solid-cystic) and with calcifications. Solid portion use to enhance heterogeneously and cysts wall strongly. On MR, signal varies according to cysts content (hyperintense on T1 and FLAIR and DWI variable when rich in protein content) and presence of calcified foci (T2*) (Figure 55 on page 88). MRS may show broad peaks in the lipid spectrum (0,9-1,5 ppm).

(Figure 56 on page 89)

**9.2 Pituitary Adenoma** (WHO grade I).

They are benign epithelial tumors derived from secretory cells of the adenohypophysis, which are classified according to their immunohistochemical expression pattern of hormones. By definition, those < 10 mm in diameter are called microadenomas and those > 10 mm macroadenomas. Due to the importance of a correct differential diagnosis with other sellar-suprasellar lesions already mentioned, we focus on the histologic and neuroimaging characteristic of macroadenomas. Macroadenomas are represented as sellar masses > 10 mm in diameter without separate identifiable pituitary gland ("the mass is the gland"). Its most common presentation is as a sellar-suprasellar mass in an adult (3rd-6th decades). It represents 10-15% of primary intracranial tumors. 75% of adenomas are endocrinologically active and determine the clinical symptomatology along with those symptoms derived from sellar and extrasellar mass effect (Figure 57 on page 90). The remaining one-third are hormonally inactive (null-cell adenomas). Prolactine (PRL)-secreting adenomas or prolactinomas are the most common of functioning adenomas and are predominant in women. Pituitary apoplexy due to a hemorrhagic infarction is a serious complication in about 1% to 2% of adenomas. It may be the first manifestation of the tumor, presenting acutely with headache, visual impairment, oculomotor palsy, and altered mentation.

**Histology:** The tumor cells have small round or oval nuclei with stippled chromatin (typical of hormone-secreting tumors). Mitoses and cellular pleomorphism may occur, but are not considered as evidence of aggressiveness. The cellular pattern may be uniform (sheets of cells with a delicate vascular network) or may show a papillary, glandular, perivascular, or trabecular arrangement. Hormone immunohistochemical studies identify the specific hormone produced by the adenoma (Figure 58 on page 91). Null-cell adenomas are chromophobic and are immunonegative.

**Neuroimaging:** On CT macroadenomas use to appear as sellar masses isodense to gray matter with moderate enhancement. When they extend to suprasellar area they adopt the typical "figure-of-eight" or "snowman" morphology due to the constriction
caused by the diaphragma sellae. When very large and aggressive they may erode adjacent bony structures (dorsum sellae, clivus, sphenoid sinus) and invade cavernous sinus, prepontine CSF spaces with brainstem compression, suprasellar cystern with compression of optic quiasm and IIIrd ventricle, and anterior, middle or posterior fossae. They may show cystic changes and necrosis and hemorrhage in 10% of cases. Calcification is rare. On MR macroadenomas appear as isointense relative to gray matter on T1 and T2 and with an early, intense, but heterogeneous enhancement after contrast. When hemorrhage is present, it is seen as hyperintense on T1 (subacute stage), and with the typical "blooming" artifact on T2* (Figure 59 on page 92). When rarely hemorrhage occurs into cysts or in pituitary apoplexy, it can adopt a fluid-fluid level appearance. Cysts and necrotic changes are seen as hypointense on T1 and hyperintense on T2.

FIGURE 60 on page

10. Metastasis.

Metastases are the most common brain tumors, with an incidence and prevalence 10 times larger than those of primary neoplasms. 20-40% of patients with cancer develop brain metastases. An increase in the incidence of brain metastases has been described, due to a longer survival of oncologic patients and the ability of the current neuroimaging techniques (especially MR) in detecting small lesions. The primary tumors that more frequently metastasize to the brain are lung (30-60% of all CNS metastases), breast (10-30%), melanoma (5-21%), and renal cancer in adults. In children, in descending order, leukaemia, lymphoma, osteogenic sarcoma, rhabdomyosarcoma and Ewing sarcoma. Unknown origin of the primary tumor is found in 10-15% of cases. Metastatic spread into the CNS occurs hematogenously (since lymphatic drainage is absent in the brain), and has a higher tendency to localize in the borderzone areas of the arterial vascular system, what accounts for the relative distribution of the lesions: 80% in cerebral hemispheres, 15% in the cerebellum, and 5% in the brainstem. Metastases are multiple in 60-80% of cases.

They use to be well-defined lesions with a heterogeneous internal structure where tumoral tissue merges with hemorrhagic and necrotic areas. The surrounding parenchyma is characterized by edema, reactive gliosis, microvascular proliferation and lymphocytic infiltrates. Intralesional hemorrhage is common in metastases of renal carcinoma, melanoma, and choriocarcinoma.

Histology: Metastasis in the brain often form circumscribed and rounded with variable central necrosis and surrounding edema. The histological and immunohistochemical features are as diverse as in the primary tumor from which they arise. These tumors often expand by growth of groups of tumor cells that lead to destruction of the neuroglial tissue, rather than infiltrate. Several reactive changes as gliosis, inflammation and microvascular proliferation are common. In leptomeningeal metastases the tumor cells are dispersed in
the subarachnoid space and may invade the adjacent parenchyma by expansion by the Virchow-Robin spaces (Figure 61 on page ).

**Neuroimaging**: MR turned to be superior to CT in the evaluation of CNS tumors. It is able to depict more lesions (mainly those of a small size, less than 10 mm), delineates better those in the posterior fossa, and allows the evaluation of leptomeningeal metastases. CT is a better option to evaluate bone metastases and the presence of intratumoral calcium or hemorrhage. On CT, metastases appear as solitary or multiple solid and well-defined lesions, iso- or hypodense to normal parenchyma, and a central hypodensity in case of necrosis. Hyperdense lesions can be found, due to the presence of calcium, hemorrhage, or a high cellularity (ie lymphoma, small-cell tumors, and adenocarcinoma). They use to enhance homogeneously or in a ring-like fashion after contrast administration. On MR metastases use to appear as iso- or hypointense relative to gray matter on T1, with surrounding edema that can be absent in small lesions. On T2, they appear iso- or hyperintense to gray matter with a higher hyperintensity of the edema and the central necrosis. Signal of hemorrhagic metastases depend on the stage of hemorrhage. Metastases from melanoma can appear hyperintense on T1. Hypercellular and calcified lesions can show hypointensity on T2. Most of them enhance after Gadolinium administration with a nodular or ring-like pattern, usually with a thick and irregular wall (Figure 62 on page ). Enhancement can decrease after corticosteroid treatment, chemotherapy and radiotherapy. Differential diagnosis include abcess, demyelinating diseases, and high-grade primary tumors (Glioblastoma Multiforme).

Progression in number and size along their natural history is typical. Younger age is associated with a longer survival. Median survival with whole brain radiotherapy is between 3 and 6 months.

(FIGURE 63 on page 96)

**11. Other tumors.**

**11.1 Hemangioblastoma** (WHO grade I).

Hemangioblastomas are well-defined tumors, tipically located in the posterior fossa (it is the most frequent intraaxial primary neoplasm of the posterior fossa in adults). They arise from the endothelial cells of the CNS, and although histologically benign, their development is often unfavorable due to high frequency of recurrence and multicentricity, specially in the 20-25% of cases in which it is associated with Von-Hippel-Lindau (VHL) disease, an autosomal dominant phakomatosis characterized by visceral cysts and neoplasms.
They are considered grade I WHO tumors and comprise 1-3% of CNS primary neoplasms. Patients with VHL used to have complications early in life from CNS hemangioblastomas or renal cell carcinoma.

Most lesions are infratentorial, being the cerebellum the common site of location, followed by the brainstem and the spinal cord. Brainstem hemangioblastomas typically cause lower cranial nerve dysfunction, long-tract signs, sensory impairment, and gait abnormalities.

**Histology:** They appear as circumscribed, cystic or solid tumors, highly vascular, in which hemorrhage may occur but necrosis is rare. Four different types can be distinguished macroscopically: Type 1 (simple cyst without mural nodule) and type 4 (solid form with microcysts), comprise 10% of all hemangioblastomas. The most frequent is type 2, a macrocyst with a mural nodule, 65% of all. Type 3, the hypervascularized solid form, comprises the 25%. Histologically, hemangioblastomas have two components: a capillary network lined with endothelial cells and large foamy stromal cells. These cells immunoreact for vimentin and may contain lipid droplets (Figure 64 on page 97).

**Neuroimaging:** In type 2, the solid portion usually has a typical peripheral and subpial location, hypointense signal on T1 and hyperintense on T2 on MR, and a marked and homogeneous enhancement after contrast administration. High-flow vessels can be observed as flow voids in the nodular portion (Figure 65 on page 98, Figure 66 on page 99).

Complete excision is the goal of surgery. The cyst wall needs not to be removed. Some authors advocate the use of preoperative embolisation of the mural nodule.

(FIGURE 67 on page 100)

### 11.2 Epidermoid cyst.

This tumor-like lesion is also known as Primary cholesteatoma or Ectodermal inclusion cyst. It is a congenital inclusion cyst which arises from the slow accumulation of cholesterol and keratin produced in the desquamation of normal squamous epithelium included during neural tube closure. Its typical sites of location are the cerebello-pontine angle, the parasellar region, or the diploe of the skull. It is a slow-growing lesion with symptoms depending on location and growth pattern (headache, neuropathy, hypopituitarism, seizures).

**Histology:** Grossly, the cyst wall is smooth or nodular and contains white, grossly material resembling mother of pearl ("pearly tumor"). The fibrous capsule is lined with stratified squamous epithelial cells and the tumor contains concentrically arranged keratin from the desquamation of squamous epithelial cells (Figure 68 on page 101).
**Neuroimaging**: It appears as a lobulated, hypo- or isoattenuating to CSF lesion on CT that encases nerves and arteries rather than displaces them. At MR, epidermoid cyst usually shows a very similar appearance to arachnoid cysts (its main differential diagnosis). FLAIR, and more recently DWI, have evolved as especially useful sequences in this differential diagnosis, and in confirming the presence of residual postoperative tumor. In FLAIR, due to its capacity of supressing the CSF signal, epidermoid usually has a high signal, whereas the signal of arachnoid cysts is suppressed. DWI typically shows a marked diffusion restriction in epidermoid cyst compared to arachnoid cyst, with a high signal intensity on DWI images and hypointensity on ADC maps (Figure 69 on page 102, Figure 70 on page 103).

(FIGURE 71 on page 104)

11.3 Chordoma

Chordoma is a midline tumor that arises from notochordal remnants being the skull base the second most site of origin following the sacrococcygeal area. They account for 0.1-0.7% of all intracranial tumors and 6% of primary tumors of the skull base and are thought to originate specifically from the sphenoid-occipital synchondrosis. They used to become symptomatic between the 3rd-5th decades. Symptoms depend on the location and extension of the lesion (headache, diplopia, endocrine dysfunction and low cranial nerves pareses being the most common).

**Histology**: Chordoma are described to contain fibrous strands which create lobulations and pseudoencapsulation. These lobules are found to contain either sheets of physaliferous cells or pools of mucin. Physaliferous cells contain varying amounts of cytoplasmic mucin, giving these cells their characteristic vacuolated appearance.

**Neuroimaging**: Chordoma appears as a slow-growing, lobulated, expansile mass arising from the midline (clivus) with lytic destruction of the adjacent bone and a tendency to invade the sellar and parasellar regions (cavernous sinus), sphenoid sinus, retropharingeal space, prepontine cystern and posterior fossa. They can have the appearance of chondroma/chondrosarcoma, with low density on CT and the presence of calcification in 47-74%, due to sequestered bony fragments within the tumor. Multiple hypodense zones can be seen throughout the mass, probably representing mixoid or gelatinous material. On MR, they usually appear as lobulated masses, hypointense on T1 and hyperintense on T2, with a slight to moderate enhancement after contrast administration. Some tumors show multiple small internal foci of very high signal on T1, which are thought to represent small areas of hemorrhage and mucinous collection (Figure 72 on page ). MR angiography may be a complement to CT and MR in cases of significant displacement or encasement of vertebrobasilar and intracranial internal carotid systems.
The main challenge in the treatment of chordomas is local recurrence, thus a combination of local resection and high-dose radiation therapy is required.

( on page 106 FIGURE 73 on page 106).

**11.4 Cavernous angioma.**

Cavernoma or cavernous angioma is a benign vascular hamartoma, consisting of sinusoidal immature vascular spaces with slow flow ("caverns"), areas of internal hemorrhage and no intervening neural tissue. It can be sporadic (75%) or appear as a familiar entity with autosomal dominant inheritance (10-30%). In a scarce percentage of patients it can be the consequence of an insult to the brain (ie, cerebral irradiation). Its frequency has been reported as 0.4-0.8% in some autopsy or MR-based series, though its real incidence is unknown, due to a commonly asymptomatic natural history.

It is commonly found in the supratentorial compartment of young adults, with a slight male predominance. Location in the posterior fossa is found in 10-23% of cases, mostly in the protuberance. The risk of hemorrhage seems to be larger in this location, with a rate of bleeding of 6% per year. The association with other vascular lesions (ie, developmental venous anomalies) is common and may play a role in their origin and recurrence. In 97% of cases a focal neurological deficit appears as the main symptom due to bleeding. Other symptoms may appear between second and fifth decades, and are related to the amount and frequency of bleeding inside the lesion (vertigo, headache, hemihypoestesia, ataxia and nerve palsy).

**Histology:** Macroscopically it appears as a well circumscribed lesion, dark blue or purple, frequently described as mulberry or grapelike in appearance. The vascular channels are covered by a single endothelial layer, and separated by thin walls of collagen. Calcification, hemosiderin deposition in their walls, and vascular thrombosis are commonly encountered features (Figure 74 on page 107).

**Neuroimaging:** CT has a reported sensitivity of 70-100% and a specificity of 50% in the diagnosis of cavernoma. It usually appears as a 0.5-4 cm lesion, moderately hyperdense, with calcifications in a third of cases and a hypodense peripheral halo due to gliosis. A patchy enhancement can be observed due to blood ectasia in the vascular channels.

MR is considered the most accurate technique for diagnosis, and T2*-GRE sequence as the most sensible for its detection. Zabramski defined four major types of cavernous malformation, representing the type 2 the most typical appearance: a well-defined lobulated mass on T2 with central heterogeneity (bleeding in different stages) and a hypointense peripheral ring (hemosiderin deposit) is considered patognomonic ("popcorn ball"). T2*-GRE is very useful in determining the typical hypointense "blooming" artifact due to the prominent susceptibility effect (Figure 75 on page 108, Figure 76 on page 109).
Cavernous angioma is considered the most common angiographically "occult" vascular malformation

(FIGURE 77 on page 110)

**QUIZ CASES**
Fig. 0: The "specific glioneuronal element" is the hallmark of this tumour. In a columnar or alveolar pattern we can see normal-appearing neurons floating in a pale matrix surrounded by small oligodendroglia-like cells.

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**Dysembryoplastic Neuroepithelial Tumor (DNET)**

A, B: Left frontal gyrus enlargement due to a markedly hypointense, irregular and cortically-based lesion with a cystic appearance. A slight degree of cortical bone remodeling may be suspected in the axial image.

C: Multicystic form of this DNET in an adult woman with refractory epilepsy. A well defined hyperintense lesion on T2 can be depicted in the left temporal amygdala and head of Hippocampus (C). Neither perilesional edema nor contrast enhancement are shown on T1 before (D) and after (E) Gadolinium.

**Fig. 0**

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Fig. 0

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Remember about DNET…

- Partial seizures with or without secondary generalization before the age of 20.
- No progressive neurological deficit.
- Predominantly supratentorial, cortically located.
- Macrogryric, pseudo- or multicystic appearance.
- No mass effect, peritumoral edema, or significant enhancement.

Fig. 0

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**Fig. 0:** Irregular groups and clusters of large dysplastic neurons with non-neoplastic glial component.

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Gangliocytoma

A: Frontal cortical GC. Very well-circumscribed, hyperintense and homogeneous lesion without peripheral edema.

FLAIR (B), T1 (C), and T1 CE (D) of a small GC of the posterior temporobasal cortex. Be aware of the intense and solid enhancement.

Fig. 0

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26 y.o woman with refractory partial complex seizures (refused contrast). MRI shows a fairly well-defined cortical right frontal lesion with a hyperintense on T2 cystic pole (F), and scattered small hypointense foci with a slight blooming artifact on T2* (E), probably representing calcifications. A Tc99m-HMPAO SPECT was performed before surgery. A clear accumulation of the radiotracer is seen by the lesion (G,H,I) (a subclinical ictal activity was suspected during SPECT)

Fig. 0

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Remember about Gangliocytoma…

- GG-GC most common tumors associated with chronic temporal epilepsy in young adults.
- GC indistinguishable from GG on neuroimaging.
- Location: temporal and frontal lobes.
- Classic pattern: cyst with mural nodule. But not infrequently shown as a solid mass, even with a dural tail.

Fig. 0

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Fig. 0: Sheets of uniform-appearing round cells with arborizing capillary vascular pattern and fibrillar nucleus-free neuropil-like areas.

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Fig. 0

A lobulated and slightly hyperdense mass is seen on CT filling the left frontal ventricular horn, near the foramen of Monro (A,B,C). Gross calcifications in the centre of the mass are depicted (A). MR better displays its heterogeneity, with cystic components, and its connection with the septum pellucidum (D-H). No significant enhancement is demonstrated after contrast on CT or MR (C,H).
Remember about Neurocytoma…

- **Location** is the neuroimaging hallmark: Tumor of the lateral ventricle, near the foramen of Monro.
- Increased intracranial pressure symptoms due to obstructive hydrocephalus.
- Variable pattern at neuroimaging, with calcification, small cysts and hemorrhage as fairly common findings.

**Fig. 0**

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**Fig. 0:** Typical Zellballen architecture with chiefs cells disposed in nets or lobules (red circle), surrounded by a single layer of sustentacular cells and a delicate capillary network.

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Jugulotympanicum paraganglioma: On CT, a permeative erosion of the left jugular foramen (A), with an avidly enhancing mass expanding to left middle ear (B, C, D).

Tympanic paraganglioma (E): Solid enhancing mass filling the right middle ear, with no ossicles or peripheral bone erosion associated.
Tympanic paraganglioma. MR: There is a mass filling the middle ear and the external auditive canal (red arrow), with a hypointense signal on T1 (A), and hyperintense on T2 (B). After contrast, a homogeneous avid enhancement is depicted (C). Note the accompanying left serous mastoiditis, without enhancement of the mastoid cells content after contrast (white arrow).

Fig. 0

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Remember about Paraganglioma…

- Jugulare paraganglioma is the most common jugular foramen tumor.
- On bone CT it appears as a jugular foramen mass with permeative destructive change of the adjacent bone (Check the jugular spine).”
- Hypervascular mass, with a typical “salt and pepper” appearance on MR due to numerous high velocity flow voids and hemorrhage.
- Think of it when a pulsatile tinnitus and a vascular retrotympanic mass is present.
- When sporadic, it may be multicentric in 5-10% of cases.
- Check the middle ear floor when a jugulotympanic paraganglioma is suspected.

Fig. 0

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Fig. 0: Patternless sheets of small uniform round cells with nucleus-free areas filled with a fine meshwork of cell processes (arrow) (pineocytomatous rosettes).

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**Pineocytoma (PC)**

Pineocytoma in a 77 y.o man. CT shows a pineal solid mass displacing calcifications posteriorly, with small scattered central calcifications. Blood in the IIIrd ventricle due to previous ventriculostomy (A). High cellularity is shown as a diffuse hypointensity of the lesion on T2 (E) and water restriction on DWI and ADC map (F,G). Diffuse, heterogeneous enhancement is shown after contrast (C). Active secondary hydrocephalus is present.

**Fig. 0**

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Fig. 0

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Pineocytoma: Well-defined mass in the pineal region, with a predominant cystic component mimicking a pineal cyst is seen on the sagittal plane (A, B). A thin solid rim which displaces the pineal calcifications to the periphery is well depicted on T2 and T2* (B, D). The mass, displaced to the right, slightly compresses the midbrain tectum. Secondary hydrocephalus is present. Note the higher-to-CSF signal of the cystic portion on FLAIR (E, F).
Remember about Pineocytoma-Pineoblastoma…

- Elevated ICP (hydrocephalus) + Parinaud’s syndrome + endocrine-hypotalamic symptoms = think of a tumor of the pineal parenchyma.
- PC and PB share neuroimaging features.
- PB depicts high cellularity and malignant neoplastic conditions in functional-MRI techniques (DWI, MRS)...but also germ-cell tumors.
- PC can resemble pineal cyst.
- PC “explodes” pineal calcifications.
- PB is prone to CSF dissemination, so evaluate spinal MR.

Fig. 0

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Fig. 0: Sheets of densely packed undifferentiated round cells with highly hyperchromatic atypical nuclei and scanty cytoplasm.

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Fig. 0

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Contrast enhancement in Medulloblastoma: it may vary from faint (A), to intense but somewhat irregular (B,C). It is not unusual to find cystic and non-enhancing structures in the periphery or center of the tumor (D through F).

Fig. 0

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An heterogeneous solid-cystic mass with intense enhancement is seen growing from the cerebellar vermis, with left hemisphere invasion and cranial expansion through the tentorium (A, B). Marked compression of the brainstem is depicted in sagital images (A). Fiber tractography shows marked displacement of the left corticospinal tract to the right (C, D).

Fig. 0

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Fig. 0

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### Comparison between sPNETs and Medulloblastomas

<table>
<thead>
<tr>
<th></th>
<th>sPNETs</th>
<th>Medulloblastomas</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Prevalence</strong></td>
<td>&lt;1% of pediatric CNS tumors</td>
<td>About 20% of pediatric CNS tumors</td>
</tr>
<tr>
<td><strong>Mean age of presentation</strong></td>
<td>3 years</td>
<td>5 years</td>
</tr>
<tr>
<td><strong>Sex</strong></td>
<td>M=F</td>
<td>M&gt;F</td>
</tr>
<tr>
<td><strong>Imaging</strong></td>
<td>Heterogeneous enhancement, distinct border</td>
<td>Homogeneous enhancement</td>
</tr>
<tr>
<td><strong>Histopathology</strong></td>
<td>Poorly differentiated neuroepithelial cells, small round nuclei</td>
<td>Same</td>
</tr>
<tr>
<td><strong>Molecular Biology</strong></td>
<td>No specific chromosomal abnormalities</td>
<td>Deletion 17p and gain 17q</td>
</tr>
<tr>
<td><strong>Metastases</strong></td>
<td>5-39%</td>
<td>40-60%</td>
</tr>
<tr>
<td><strong>Treatment</strong></td>
<td>Surgery + craniospinal radiotherapy + chemotherapy</td>
<td>Same</td>
</tr>
<tr>
<td><strong>3-year progression-free Survival</strong></td>
<td>45-47%</td>
<td>&gt;80%</td>
</tr>
</tbody>
</table>


**Fig. 0**

Remember about Medulloblastoma…

- MBs and PNETs are considered the most common CNS malignant tumor of childhood.
- MB: more common infratentorial location (vermis). PNETs: more common supratentorial.
- Almost indistinguishable in imaging and histologically.
- Beware of possibility of CSF dissemination: Evaluate total spinal MR.
- Highly cellular tumors: tend to be hyperdense on CT and show restriction on DWI.
- May be heterogeneous, with cysts, calcium and hemorrhage, but less than Ependymoma (DD with MB).

Fig. 0

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Fig. 0: Embryonal tumour with carcinoma-like areas and frequent typical rhabdoid cells with eccentric nuclei, prominent nucleoli and cytoplasmic pink body inclusion (arrow).

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**Atypical Teratoid Rhabdoid tumor**

![Images of Rhabdoid cells showing strong smooth-muscle actin (SMA) immunoreactivity and loss of INI-1 nuclear expression (IHC).](image_url)

**Fig. 0:** Rhabdoid cells show strong smooth-muscle actin (SMA) immunoreactivity and loss of INI-1 nuclear expression (IHC).

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Fig. 0

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Atypical Teratoid Rhabdoid tumor

Medulloblastoma-like AT/RhT tumor of the cerebellar vermis, with a highly cellular solid portion and central cystic component. Beware of the severe and active hydrocephalus and the marked water restriction on DWI.

Fig. 0

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Remember about Atypical Teratoid-Rhabdoid tumor…

- Everytime you see a Medulloblastoma/PNET in a child under 3 y, think of AT/RhT.
- Usually round, bulky tumor, with an heterogeneous structure (cysts+calcium+hemorrhage+highly cellular solid portion).
- 50% offline infratentorial lesion; 40% supratentorial.
- Need to stablish differential diagnosis with other highly malignant tumors (High –grade glioma, sarcoma, carcinoma), due to the combination of epithelial, mesenchymal and neuroectodermal elements in its composition.
- 15-20% present with CSF dissemination.

Fig. 0

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Fig. 0: Spindle-shaped Schwann cell neoplasm composed of cellular Antoni A areas with Verocay bodies. These bodies are formed by parallel and compact arrays of elongated nuclei forming palisades which are separated by dense nuclei-free fibrillary areas (arrows).

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Fig. 0

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Remember about Schwannoma…

- **Most of intracranial nerve tumors are schwannomas** (unless I and II nerves).
- Benign, encapsulated extraaxial tumor of adults.
- **95% vestibular or acoustic** schwannomas.
- On imaging, heterogeneous tumor with not highly cellular solid portion, and variable cysts, hemorrhage and calcium.
- Strong enhancement, mostly with a solid pattern (remember *ice-cream cone*).
- Symptoms depend on nerve affected and size (hearing loss, tinnitus, vertigo, brainstem compression, hydrocephalus).

Fig. 0

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**Fig. 0:** Lobules of uniform arachnoidal cells with oval nuclei and syncytial appearance surrounded by collagenous septae. We can see numerous concentric onion-bulb structures and whorl formations (black arrow), and psammoma bodies (red arrow).

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Meningioma. MR features: Inward displacement of the cortical gray matter and cortical vessels, iso-intensity of the solid portion of the tumor to the gray matter, and CSF clefts are shown in these meningiomas of the left convexity (A) and the planum Sphenoidale (B). (C) Bulky calcifications are depicted on T2*. (D) A dural tail is clearly seen on T1 after contrast administration (arrow).
Fig. 0

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Fig. 0

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Remember about Meningioma…

- Meningioma is the **most common extraaxial neoplasm**.
- **85-90% have a benign behaviour.**
- Most common sites of origin: parasagittal, convexities, olfactory plate, sphenoid ridge.
- Remember other less frequent locations: intraventricular, meningioma en plaque.
- Bulky, **hypervascularized** lesion attached to the dural surface...look for the **dural tail**.
- Schwannoma is the most common differential diagnosis when located in the CPA.

---

**Fig. 0**

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**Fig. 0**

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Remember about Lipoma…

- Represents a developmental malformation of the CNS.
- More often located in the midline (corpus callosum).
- Homogeneous fat without solid nodules or cysts: Differential diagnosis with dermoid tumor.
- Benign lesion commonly asymptomatic.

Fig. 0

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Chondroma / Chondrosarcoma

Chondrosarcoma of the cavernous sinus. Calcifications of the chondroid matrix are well demonstrated on CT (A) and T2* MR (B).
(C) Hyperintense signal with internal low intensity foci on T2.
(D,E) Lobulated margins and heterogeneous enhancement.

Fig. 0

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Remember about Chondroma-Chondrosarcoma...

- Rare slow-growing tumors of the clivus, skull base.
- Originate from embryonic cartilaginous remnants enclosed in the bones.
- Very similar to chordoma in biological and neuroimaging behaviour.
- They locate paramedially (synchondrosis) in contrast to chordoma that is a midline tumor.
- Resemble cartilaginous tissue on neuroimaging (iso-hypodense on CT; hypointense on T1; hyperintense on T2), with scattered foci of curvilinear calcified matrix.

Fig. 0

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**Fig. 0:** Brain parenchyma with diffuse infiltration of CD20-positive atypical lymphocytes proliferation with a prominent angiocentric pattern forming concentric perivascular cuffs of tumour cells (arrows).

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**Fig. 0:** Brain parenchyma with diffuse infiltration of CD20-positive atypical lymphocytes proliferation with a prominent angiocentric pattern forming concentric perivascular cuffs of tumour cells.

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This left frontal lesion shows typical features of PCNSL in an immunocompetent patient: 1. Homogeneous hyperdensity on TC and hypointense T1 and T2 signal, and marked water restriction on DWI (signs of high cellularity), and 2. Avid and diffuse enhancement after contrast.

Fig. 0

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Fig. 0

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Remember about Lymphoma…

- Increased incidence of PCNSL in low-risk immunocompetent patients (remember: the immunological status influences the biological behaviour and neuroimaging appearance of lymphoma).
- Most of PCNSL are of the diffuse and large B-cells type.
- Nodular, single or multiple, periventricular or in basal ganglia, solid masses with high cellularity criteria on CT and MR.
- CSF dissemination frequent.
- CBVr higher than normal, but lower than in high-grade glial tumors.

Fig. 0

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Fig. 0: Sheets of uniform large cells with vesicular round nuclei, prominent nucleoli, and abundant clear glycogen-rich cytoplasm, associated with a lymphoid infiltrate.

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Pineal Germinoma (A,B,C). A lobulated, heterogeneous pineal mass with cysts and calcifications is seen on T2 (A) and T2* (B). After contrast (C), irregular enhancement is shown along with another solid-cystic lesion in the lower medulla (CSF dissemination).

Hypothalamic Dysgerminoma (D): A solid, lobulated and enhancing mass is depicted, widening the pituitary stalk and protruding into the anterior recess of the III ventricle.

Fig. 0

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Fig. 0: Tumour composed of fully differentiated adult-type tissue. We can see fat tissue (*), nerves (black arrows) and different types of glands (red arrows).

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Mature intraventricular teratoma: Lobulated and well-defined mass conformed predominantly by cystic-like components of highly varying intensities. Fatty elements are clearly shown as hyperintensity similar to subcutaneous fat on T1 and T2 (A,B,E), which saturates on T1 fat-sat sequence or T2* (C,D).
Remember about Germ-cell tumors...

- Germ cells tumors derive from germ cell nests displaced during embryogenesis. Analogous in morphology and immunophenotype to gonadal and other extragonadal GCT.
- Midline tumors, more than 80% located in the pineal-IIIrd ventricle areas.
- Tendency to disseminate throughout CSF spaces. Remember evaluate neuroaxis before treatment.
- Tumors included vary from highly cellular, like germinoma (remember use of DWI for confirmation), to markedly heterogeneous as Teratoma or Embryonal tumor (remember use of T2* or fat-saturated sequences for depiction of hemorrhage-calcium or fat respectively).

Fig. 0

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Fig. 0: Proliferation of nets and cords of squamoid cells. The nets are rimmed by palisades of basaloid cells (arrows). The internal layer become looser and form a spongy reticulum. Presence of compact wet keratine and cyst formations.

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Fig. 0

A well circumscribed mass with cystic appearance, very hyperintense signal relative to CSF and a faint fluid-fluid level is shown in a suprasellar location (A,B). Punctate peripheral calcifications are seen on T2* (arrow) (E). A slight compression of the pituitary gland can be seen in the sagittal images (A,D). There is a marked postero-inferior expansion of the lesion, with severe mass effect on the pons and medulla, and a thin linear enhancement of the cyst wall after contrast (D).
Remember about Craniopharyngioma...

- **Slow growing**, benign neoplasm, usually large at diagnosis, with tendency to great extension to other intracranial areas.
- Think of craniopharyngioma when a **midline, suprasellar heterogeneous mass** (cystic-solid plus calcifications) in a child.
- Take advantage of neuroimaging to depict structural features (FLAIR-T1-DWI for determine complex cystic component; T2* for calcifications, contrast for solid portions).

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Fig. 0

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Tumors of the pituitary gland

Hormones secreted by anterior pituitary adenomas.

**Prolactin:** F: Amenorrhea, Galactorrhea.
M: Hypogonadism.

**Growth hormone:** Acromegaly.

**ACTH:** Cushing syndrome.

**TSH:** Pituitary Hyperthyroidism.

**Neurohypophysis**

Astrocytoma
Granular cell tumor
**Fig. 0:** Adenoma (prolactinoma) Sheet of monomorphic round cells with round-oval nuclei, abundant acidophilic cytoplasms and distinct cell borders. Marked cytoplasmic immunoreactivity with Prolactin (IHC).

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Fig. 0

64 y.o man with diplopia due to paralysis of right VI nerve. On CT (A,B), an heterogeneous solid mass expands the sella with focal bone erosion of the left sphenoid sinus wall. On MR, the lesion has a small suprasellar component, with a high heterogeneous signal on T1 (C) due to internal hemorrhage (pituitary apoplexy). Heterogeneity is also visible on T2, and a slight compression of the optic chiasm and the right cavernous ICA (D). After contrast (E), only a linear enhancement of the diaphragma sellae is present.
Remember about Pituitary Adenoma...

- Sellar mass without identifiable normal pituitary tissue ("the mass is the gland").
- Always histological benign lesions although highly aggressive when large.
- Remember your figure of eight or snowman when suprasellar growth.
- Check bony erosions and extension or compression of adjacent areas (sphenoid sinus, cavernous sinus, clivus, brainstem, suprasellar cystern, optic ways, II\textsubscript{I}rd ventricle).
- Mostly solid and homogeneous, isointense to gray matter. Possible cysts and hemorrhage (pituitary apoplexy): evaluate T1 without contrast and T2*.
- \textit{75\% endocrinologically active:} Check hormones levels (PRL).

Fig. 0

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Fig. 0: Brain metastases of carcinoma. A clear-defined boundary is shown between the highly cellular metastatic area (*) and the normal cortex (+). Compression prevails over infiltration.

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Fig. 0

(A-D) Intraaxial metastases. T1 CE images demonstrate small well-circumscribed nodules with solid or anular enhancement in a predominantly cortical distribution of both cerebral hemispheres and cerebellum (A,B). Note the high cellularity (restriction on DWI), and the significant peripheral edema in this left parietal metastases of bladder carcinoma (C,D).

(E,F) T1 fat-sat CE images of a dural metastases of breast carcinoma. A nodular enhancing mass originates from the right sphenoidal lesser wing.

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Remember about metastases…

- The most common brain tumors, with an incidence and prevalence 10 times larger than those of primary neoplasms.
- Well-defined, round lesions, in the gray-white interface of cerebral hemispheres. Necrosis and hemorrhage common, depending on the histological features of the primary tumor from which they arise.
- Metastases better displace rather than infiltrate tissue.
- Spontaneous intracranial hemorrhage or new onset seizures in elderly patients may be caused by metastases.
- MR superior to CT to define, count and characterize them.

Fig. 0

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**Fig. 0:** Tumour composed by large vacuolated stromal cells and a rich capillary meshwork.

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Hemangioblastoma

Hemangioblastomas of the cerebellar vermis. On DPw (A), a dense vascular net can be seen as irregular flow voids in the solid portion of the tumor. After contrast (B,C), the typical appearance of a type 2 hemangioblastoma is better demonstrated. The avidly enhancing solid nodule in close contact to the parenchyma surrounded by a descending cyst with no mural enhancement.

In a second patient (D,E), DWI shows the absence of water restriction in the cystic component of the mass (E).

Fig. 0

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Two cases of medullar hemangioblastomas. On sagital T2 (A) and CET1 (B), the first lesion is shown as an ovoid cyst surrounded by vasogenic edema with slight medullar expansion. After contrast a typical enhancing mural nodule is depicted. Solid-type enhancing HB are seen in the left cerebellar hemisphere of the same patient (C).

A second patient, who refused contrast, shows on T2 a marked medullar and cervical cord expansion due to an elongated cystic lesion surrounded by edema.
Remember about Hemangioblastoma...

- Benign tumor of an uncertain hystogenesis, classically classified as a meningeal tumor.
- **90-95% HB locate in the posterior fossa**...But in middle-aged and older adults the most common posterior fossa tumor is metastases and not HBs.
- Several forms of presentation, the most common as a cyst with an enhancing mural nodule (60%).
- Although benign, HB has an uncertain prognosis due to its high frequency of recurrence and multicentricity, specially when associated to Von-Hippel-Lindau disease (look for other signs of the disease and screen the entire neuroaxis for other HBs).

Fig. 0

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Fig. 0: Cyst lined by squamous epithelium and filled with keratinous lamellar keratin.

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Fig. 0

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Epidermoid cyst (A-C) versus Arachnoid cyst (D-F): Note the differences between these two extraaxial lesions of the left PCA cistern:
1. More lobulated margins of the EC.
2. Tendency of the EC to vascular/nerves encasement (left trigeminal nerve, arrow).
3. Slight hyperintense signal on FLAIR in EC.
4. Marked diffusion restriction of EC on DWI.

Fig. 0

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Remember about Epidermoid cysts…

- EC are **congenital inclusion cysts.**
- Appearance of a lobulated, cystic mass **isointense to CSF** with a tendency to surround/encase surrounding structures.
- Two clues: 1. **FLAIR** may show an incomplete CSF nulling, with slight diffuse hyperintensity and 2. **DWI** depicts a marked restriction (**GO FOR IT WHEN DD WITH Arachnoid cysts**).
Chordoma: Clival lobulated mass with suprasellar expansion, with heterogeneous signal on axial T1 (A), and marked mass effect on the pons and fourth ventricle. On T2 (B), chordoma shows a predominant hyperintense signal with internal septa of low intensity. The T2* (C) depicts the foci of calcification as susceptibility artifact focal areas.

Fig. 0

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Remember about Chordoma…

- Chordoma is a midline tumor of bony structures which originates from notochordal remnants.
- Clivus is the most common site of origin (remember: chordoma-midline/chondrosarcoma-paramedian).
- Slow-growing mass with lytic bone destruction and high tendency of expansion to adjacent spaces.
- Calcification is common (sequestered bone instead of matrix ossification).

Fig. 0

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**Fig. 0:** Compact mass of vessels closely packed with great diversity in caliber and degree of collagenization and any or little interstitial parenchyma.

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Cavernous angioma

Cavernoma: Sagital T2 image (A), shows a cavernous angioma of the pons as a mixed hypo- and hyperintense lobulated lesion. (B) Axial T2* shows a characteristic blooming artifact due to hemosiderin deposition. On contrast-enhanced T1 (C), a slight heterogenous enhancement can be seen.

Fig. 0

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CTs of a left frontal cavernoma before (A) and after (B) a bleeding episode. T1 (C) shows the typical “popcorn” appearance. T2* (D) demonstrate an expanded “blooming” artifact due to the bleeding.

T2* (B,C) helps to determine the great number of micro-cavernomas in this case of familial Cavernomatosis.
Remember about Cavernous angioma...

- Cavernoma can be considered an **hamartomatous vascular proliferation** more than a real neoplasm.
- In 10-30% of cases appears as a multiple, familial syndrome.
- A **risk of hemorrhage** appears in **special location** (posterior fossa), or when associated with other vascular anomalies (developmental venous anomalies).
- Type 2 has the typical MR appearance: **popcorn-ball** (remember: “**blooming**” peripheral artifact on T2*).
- The **most common angiographically occult vascular lesion**.

Fig. 0

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OK, now a few cases to check how much we have learned...

Fig. 0: QUIZ CASES

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Case 1

10 y.o. boy with visual defect & hypopituitarism.

Choose the correct diagnosis:

a) Pituitary non-functioning Adenoma.
b) It is an extraaxial lesion, so it should be a sellar meningioma.
c) Craniopharyngioma.
d) Sellar- hypothalamic Germinoma.
e) So heterogeneous in a child?. Let’s say an Atypical Teratoid - Rhabdoid tumor

Fig. 0: QUIZ CASES

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a) Pituitary non-functioning Adenoma.

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e) So heterogeneous in a child?. Let’s say an Atypical Teratoid - Rhabdoid tumor
Fig. 0: QUIZ CASES

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a) Such increase of Choline level indicates a III-IV grade astrocytoma.
b) DWI-ADC show a high cellularity in the solid portion of the tumor.
c) There is a secondary hydrocephalus.
d) Severe decrease of the NAA/Cho ratio appears on highly aggressive tumors.
e) Presence of lipids and lactate may indicate necrotic-cystic transformation.
Cerebellar Medulloblastoma

a) Such increase of Choline level indicates a III-IV grade astrocytoma.

b) DWI-ADC show a high cellularity in the solid portion of the tumor.

c) There is a secondary hydrocephalus.

d) Severe decrease of the NAA/Cho ratio appears on highly aggressive tumors.

e) Presence of lipids and lactate may indicate necrotic-cystic transformation.
Case 3

Young man with refractory Temporal Epilepsy.

Choose the best option:

a) Metastases.
b) Solid type of Hemangioblastoma.
c) Tumor of ganglyonic origin.
d) Cavernous angioma.
e) Differential diagnosis should be established between b and c.

Fig. 0: QUIZ CASES

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That's right ji.
Anyway... we forgot to tell you that the patient suffered from the Von-Hippel Lindau Disease..., so... Which option shoud you choose now the best?...

a) Metastases.
b) Solid type of Hemangioblastoma.
c) Tumor of ganglyonic origin.
d) Cavernous angioma.
e) Differential diagnosis should be established between b and c.

Fig. 0: QUIZ CASES

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OK |j| Much better...

a) Metastases.
b) Solid type of Hemangioblastoma.
c) Tumor of ganglyonic origin.
d) Cavernous angioma.
e) Differential diagnosis should be established between b and c.

Fig. 0: QUIZ CASES

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Case 4
62 y.o woman found comatose...
Choose the correct option

a) There is a triventricular hydrocephalus secondary to massive intraventricular hemorrhage.
b) This type of bleeding used to be secondary to subependymal AVMs.
c) Aren’t we talking about tumors? So let me think about an intraventricular tumor prone to bleeding...
d) There is an associated midline congenital lesion.
e) A and d are correct options.

Fig. 0: QUIZ CASES

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Fig. 0: QUIZ CASES

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Case 5. 69 y.o woman with a rapidly evolving cognitive decline

And your diagnosis is ...

a) Pineal and suprasellar Germinoma with CSF dissemination.
b) Multiple metastasis.
c) Familial cavernomatosis.
d) Primary Lymphoma of the CNS.
e) Gliomatosis cerebri

Fig. 0: QUIZ CASES

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Remember?

- Periventricularly located masses.
- High cellularity (hyperdense on CT, hypointense on T2, water restriction on DWI).
- Homogeneous & intense enhancement with no necrosis.

….. This woman was not immunocompromised. Cognitive decline with non other significant neurological symptoms is a commonly encountered clinical onset.

a) Pineal and suprasellar Germinoma with CSF dissemination.

b) Multiple metastasis.

c) Familial cavernomatosis.

d) Primary Lymphoma of the CNS.

e) Gliomatosis cerebri.

Fig. 0: QUIZ CASES

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BRAVO ii

Thank you for your attention

Fig. 0: QUIZ CASES
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Conclusion

- Non-glial tumors conform a wide group of lesions with varying grades of biological aggressiveness.

- They can be considered of special significance in several diseases and demographic groups (epilepsy, children). Neuroimaging has played a main role in the "discovery" of some of these lesions.

- We can limit the differential diagnosis for the most common nonglial tumors by using clinical and demographic data and imaging findings, what in most cases will be a translation of the histologic characteristics of these lesions.
Personal Information

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References


DNET:

Gangliocytoma:

Central Neurocytoma:

Parangangioma:

Fig. 0: REFERENCES

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Fig. 0: REFERENCES

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Lymphoma:

Metastases:

Hemangioblastoma:

Epidermoid Cyst:

Chordoma:

Cavernous Angioma:

Fig. 0: REFERENCES

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