CNS Tumors

Part II

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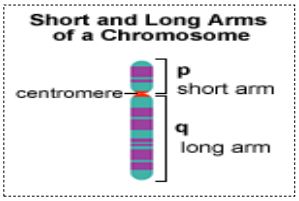
Oligodendroglioma (2nd type of gliomas):

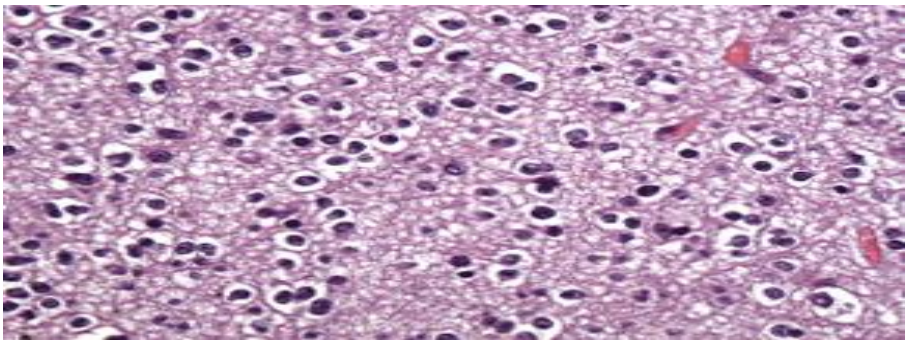


- **The most common genetic findings** are loss of heterozygosity for **chromosomes 1p and 19q**
- **Age group:** adults in their fourth and fifth decades (old)
- **Location:** cerebral hemispheres (supratentorial) , with a predilection (preference) for white matter.
- **Grading :** regular oligodendroglioma is grade 2

  anaplastic oligodenroglioma is grade 3
- **Prognosis:** better than do patients with astrocytomas (5 to 10 years with treatment). However, its anaplastic form is worse.
- **Histological findings:**

1. "fried egg" appearance : cells show round nuclei, often with a cytoplasmic halo
2. "chicken wire" pattern : blood vessels show thin and can form an interlacing –mingling- where capillaries anastomose .

Loss of heterozygosity (LOH) : is the loss of normal function of one allele of a gene in which the other allele was already inactivate



→*What additional features are needed for anaplastic oligodendroglioma?*

1. *Cyst formation*
2. *Focal hemorrhage*
3. *Calcification (in about 90% of oligodendroglioma cases)*

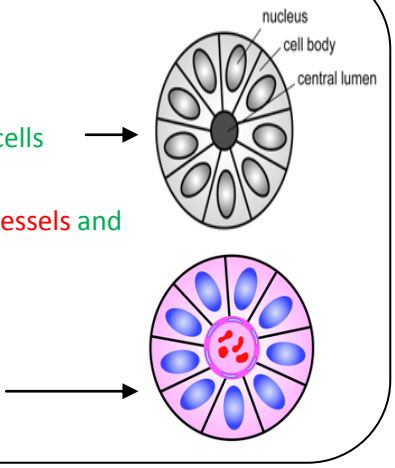
Ependymoma (3rd type of gliomas):

- **Origin**: most often arise next to the ependyma-lined ventricular system, including the central canal of the spinal cord
- **Age group:** Occurs in the first two decades of life (young)
- **Location:** they typically occur near the fourth ventricle, infratentorial (in children) and the spinal cord (in adults) are their most common location
- **Grade:** II
- **Histological findings:**
    1. **Rosettes** which are tumor cells may form round or elongated structures.
    2. **perivascular pseudo-rosettes**

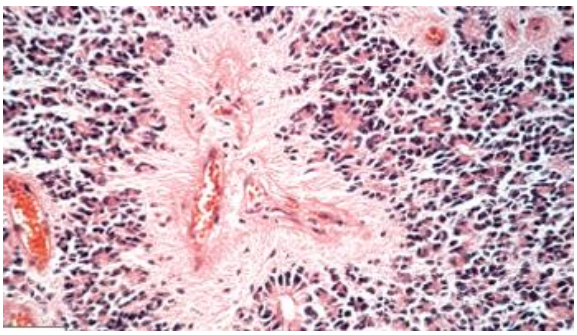
If a lumen is found then it is a true rosette. However if not, then it's a pseudo-rosette

what is a rosette?

A: **Rosette** consists of a halo or spoke-wheel arrangement of cells surrounding a central core or hub. It can be arranged around vessels and their ependymal processes extending to the wall of that vessel **(perivascular pseudo-rosette)**



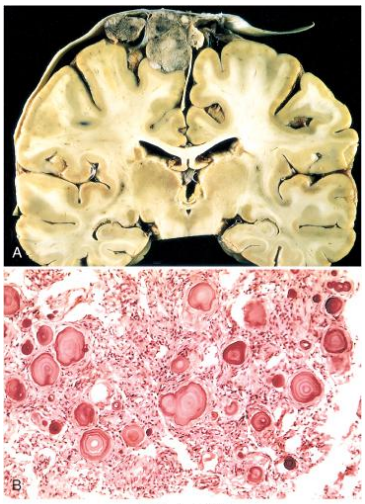
Anaplastic ependymomas: show increased cell density, high mitotic rates, necrosis and less evident ependymal differentiation



Meningioma (tumor of the covering cells)

- **Type:** mostly benign tumors.
- **Age group:** adults (not common in children).
- **Origin:** meningothelial cell of the arachnoid.
- **But it** is attached to the dura with compression of underlying brain
- Well demarcated (it has boundary) so most of it is easily separated from underlying brain, **however** if it invades the brain, it leads to increasing the risk of recurrence.

- **Histological findings:**
    1. It has **"Whorled"** (Spiral) pattern of cell growth and **"Psammoma Bodies"**, which are a round collection of calcium, (this can be also seen in papillary carcinoma of the thyroid).



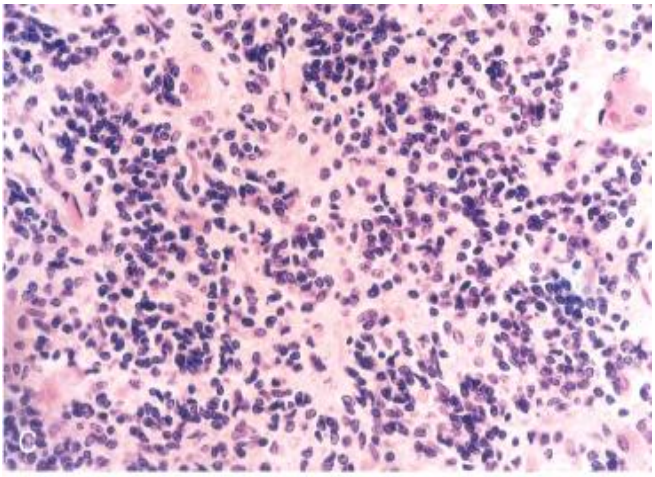
- **Main subtypes:**

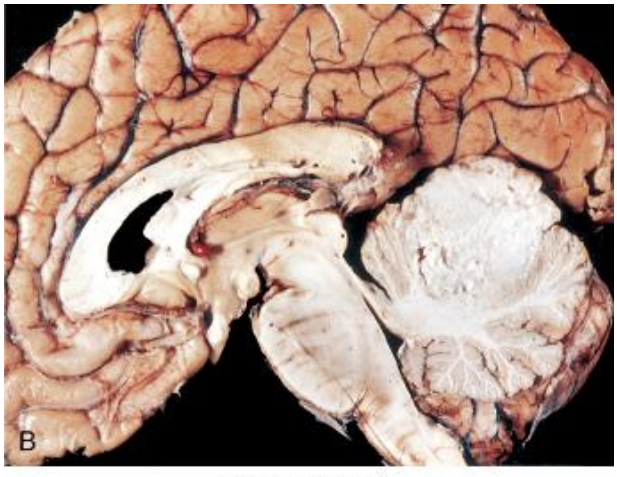
1. **Non-Malignant (benign, grade I):**
    A. Syncytial: clusters of cells without visible cell membrane between them.
    B. Fibroblastic: elongated cells with collagen fibers between them.
    C. Transitional: a case that shares characteristics of both syncytial and fibroblastic.
2. **Atypical meningiomas grade II (they can be non-malignant or malignant)**
3. **Anaplastic (malignant) meningiomas grade III**

Medulloblastoma

- **Type:** the tumor is highly malignant (Grade IV).
- **Age group:** children.
- **Location:** exclusively in the cerebellum.
- Often largely undifferentiated (primitive). However, neuronal and glial markers may be expressed.
- **Prognosis:** for untreated patients are very dismal (bad); however, it is greatly radiosensitive.
- With total excision and radiation, the 5-year survival rate may be as high as 75%.
- **Histological findings:**
    1. Extremely cellular, with sheets of anaplastic ("small round blue") cells.
    2. Little cytoplasm and hyperchromatic "carrot shaped" nuclei, mitoses are abundant.

Radiosensitive means that tumor is potentially treatable with radiation therapy.





Schwannoma (peripheral nervous system tumor)

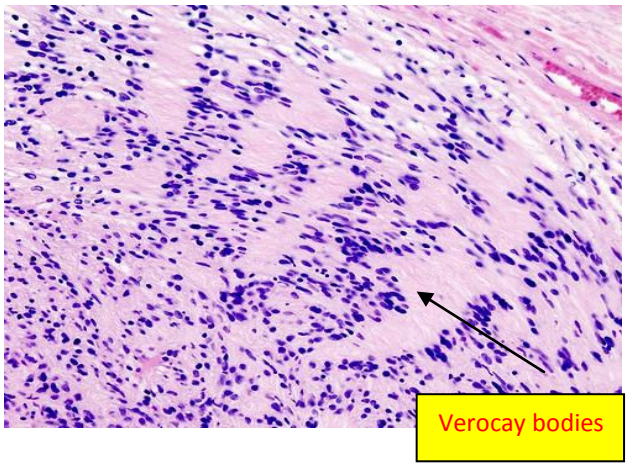
- **Type:** benign
- **Location:** In the CNS, they are often encountered within the cranial vault in the cerebellopontine angle, where they are attached to the vestibular branch of the (8^{th} cranial nerve) → (acoustic schwannoma) causing tinnitus and hearing loss but it can be separated from it.

The cerebellopontine angle: is a structure at the margin of the cerebellum and pons.

- **Classes :**
    1. Sporadic schwannomas: are associated with mutations in the neurofibromatosis *(NF2)* gene
    2. Bilateral acoustic schwannoma: is associated with neurofibromatosis type 2 syndrome

Sporadic means: at irregular intervals. Moreover, occurring singly; not grouped.

- **Histological findings:**
    1. Cellular (Antoni A) pattern and less cellular (hypocellular component) (Antoni B).
    2. Nuclear-free zones of processes that lie between the regions of nuclear palisading are termed Verocay bodies.



Neurofibroma (peripheral nervous system tumor):

- It is familial tumor – runs in family- and has 2 types: neurofibroma **type I** and neurofibroma **type II**.
- **Examples(patterns):**
    1. *cutaneous neurofibroma* in skin
    2. *solitary neurofibroma* in peripheral nerve

The previous two can be arising sporadically or in association with type 1 neurofibromatosis, (rarely malignant)

3. *plexiform neurofibroma*, mostly arising in individuals with NF1, potential malignancy

- Neurofibromas cannot be separated from nerve trunk (in comparison to schwannoma)

Metastatic tumors:

- About 25-50% of brain tumors are metastatic in origin.
- The commonest types are those from : lung, breast, skin (melanoma), kidney, and gastrointestinal tract

Familial tumor syndroms :

- Describe the inheritance pattern and the main features of:

  Type 1 Neurofibromatosis: autosomal dominant

  Type 2 Neurofibromatosis: autosomal dominant

- Which one of these two syndromes has a tendency for the neurofibromas to undergo malignant transformation at a higher rate than that observed for comparable tumors in the general population?

  Individuals with NF1, this is especially true for plexiform neurofibromas.