Pathology of CNS tumors

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Introduction

By end of this lecture; you should:

- Identify classification of CNS tumors
- Describe grading of CNS tumors
- Identify different types of GLIOMAS
- Mention main clinical and pathological features of different types of gliomas

Introduction

Normal cells of CNS

A. Neuroglial cells

1. **Astrocytes**
   - Star-shaped
   - Responsible for nutritional supply and insulation of neurons
2. **Oligodendrocytes** — Form myelin sheath
3. **Ependymal cells** — Line ventricular chambers, aqueduct, central canal of spinal cord
4. **Microglia** — Native macrophages of the CNS

B. Neuronal cells: Nerve cells

C. Meningeal cells: Cells lining meinges

D. Endothelial cells: Cells lining blood vessels

Introduction

WHO classification of CNS tumors

Tumors of neuroglia (GLIOMAS)
- Astrocytoma.
- Glioblastoma multiforme.
- Oligodendroglioma.
- Ependymoma.

Choroid plexus tumors
- Choroid plexus papilloma.
- Choroid plexus carcinoma.

Tumors of primitive undifferentiated cells
- Medulloblastoma
- Neuroblastoma

Tumors of meninges
- Meningioma.

Metastatic tumours
- Most common tumors of CNS

Tumors of peripheral nerves
- Schannoma
- Neurofiboma

Introduction

WHO Grading of CNS tumors

Grading depends on

- Cellularity and anaplasia (features of malignancy)
- Microvascular proliferation
- Mitosis
- Necrosis

Four grades

- WHO Grade 1
- WHO Grade 2
- WHO Grade 3
- WHO Grade 4

Introduction

WHO Grading of CNS tumors

WHO grade 1

- Usually benign behaviour
- Low cellularity
- Minimal atypia
- NO vessel proliferation
- Very rare mitosis
- Absent necrosis

WHO grade 2

- Intermediate behaviour
- Moderate cellularity
- Mild atypia
- NO vessel proliferation
- Rare mitosis
- Absent necrosis

WHO grade 3

- Agressive behaviour
- High cellularity
- Moderate to high atypia
- Mild vessel proliferation
- Frequent mitosis
- Absent necrosis

WHO grade 4

- Malignant behaviour
- High cellularity
- High atypia
- Microvessel proliferation
- Frequent mitosis
- Frequent necrosis

Gliomas

GLIOMAS

- **Defintion:**

  Glioma is a term used to describe **ALL** tumors arising from neuroglia

- **Incidence:**
  - Gliomas are the most common **primary** brain tumors
  - They represent more than half of all brain tumors

- **Types**
  1. From astrocytes: astrocytoma
  2. From oligodendrocytes: oligodendroglioma
  3. From ependymal cells: ependymoma

GLIOMAS

- **Clinically:** gliomas can present with
  - ***Seizures*** (convulsions)
  - ***Symptoms of ICT*** (Headache, vomiting, blurred vision)
  - ***Focal neurological deficit*** as hemiplegia, hemipareseis, parathesia, arthralgia, affection of cranial nerves …. etc

- **Behaviour:**
  - Gliomas could spread to other parts of CNS by CSF
  - In general; gliomas rarely metastasize beyond the CNS

GLIOMAS

Astrocytoma

- **Defnition:**
  - Tumors arising from astrocytes
- **General features:**
  - The most common type of gliomas
  - Can affect any age
  - Mainly involve cerebral hemispheres and spinal cord.
  - Have tendency to progress from low grade to higher grades.
- **Types:**
  - Pilocytic (WHO grade 1)
  - Fibrillary (WHO grade 2)
  - Anaplastic (WHO grade 3)
  - Glioblastoma multiform (WHO grade 4)

GLIOMAS

Astrocytoma

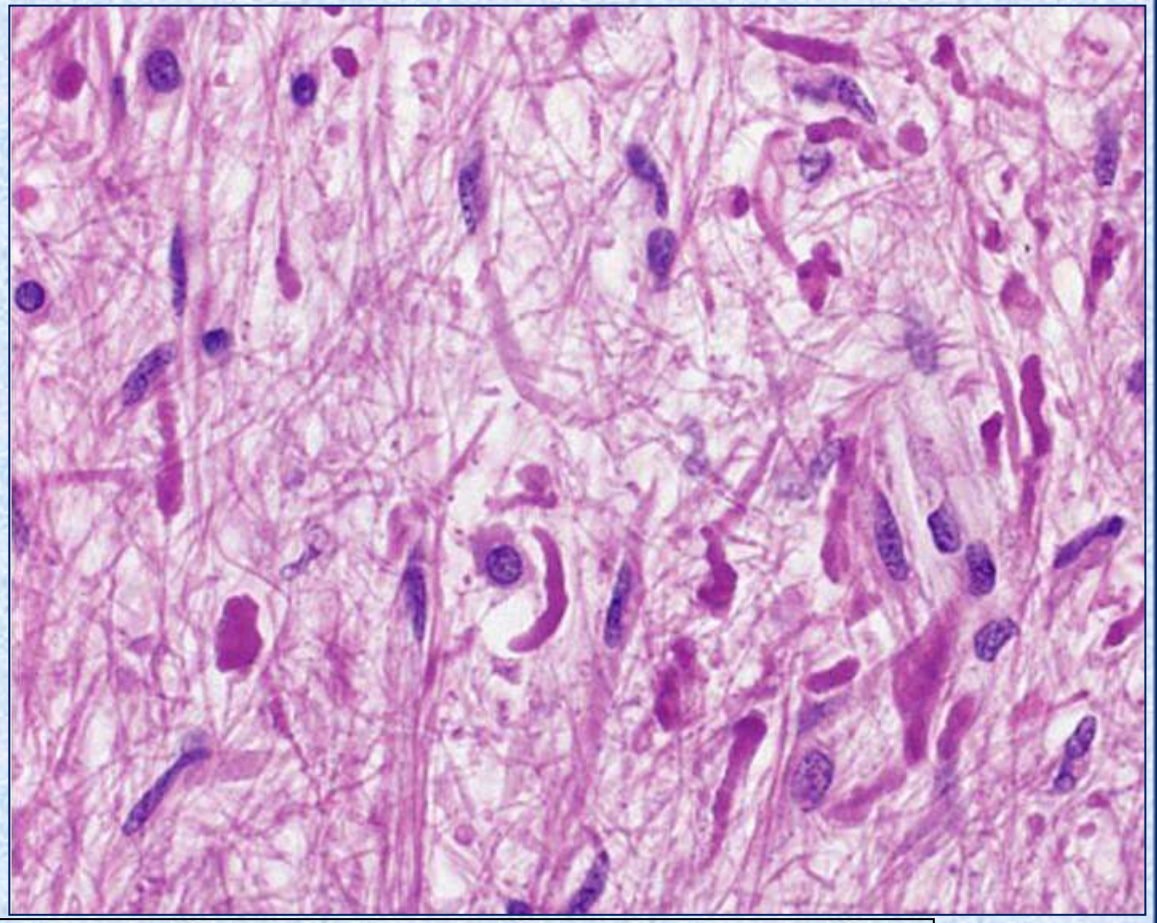
A. Pilocytic astrocytoma (WHO Grade I):

- Typically occurs in children and young adults.
- The common site is the cerebellum followed by third ventricle and optic nerve.
- Behaviour: Usually benign course (a slowly growing and rarely infiltrative tumor).
- **Grossly** (radiologically): well-circumscribed, often cystic tumor with a mural nodule attach to cyst wall.
- **Microscopically**:
  - Low cellularity
  - Proliferated bipolar cells with long, thin fibillary processes (hair cells)
  - No features of malignancy

GLIOMAS

Astrocytoma

Pilocytic astrocytoma (WHO Grade I):



Bipolar cells with long, thin processes (hair cell)

GLIOMAS

Astrocytoma

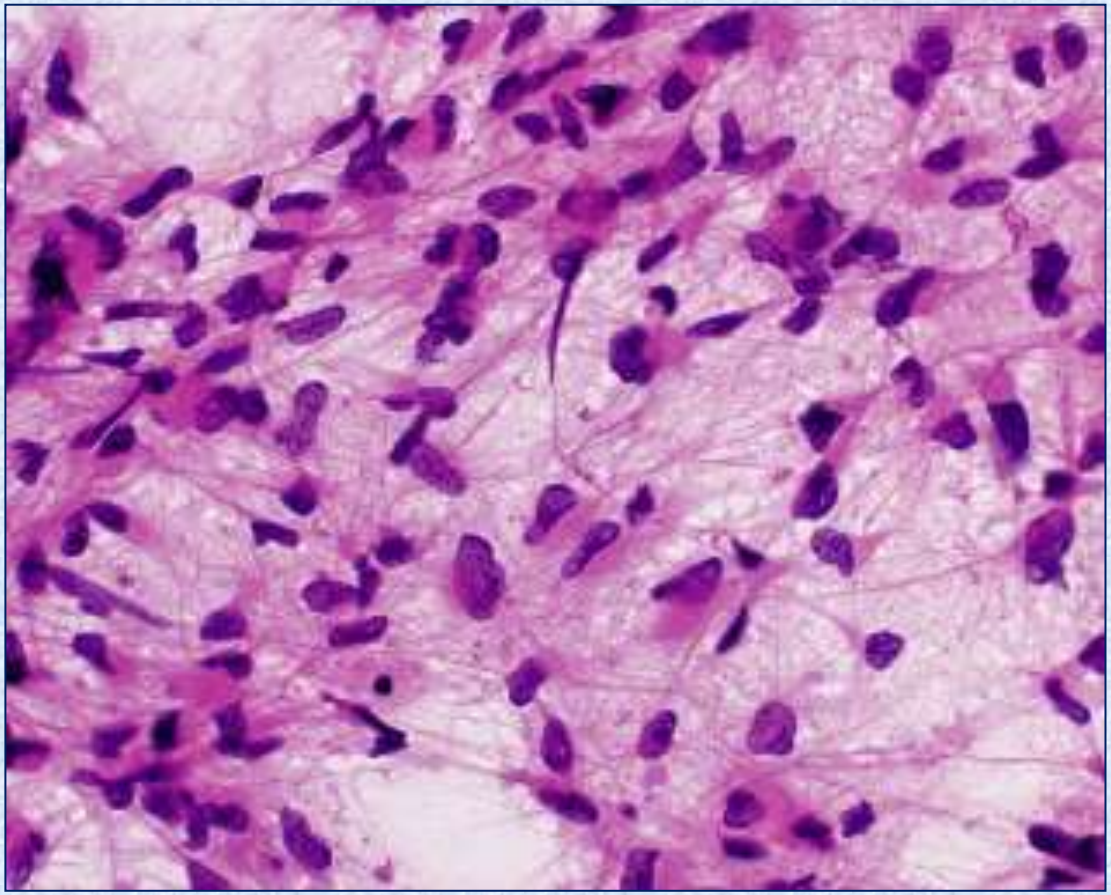
B. Well-differentiated or fibrillary astrocytoma (WHO Grade II):

- Commonly affect young adult brain.
- Common site is cerebral hemispheres in adults and brain stem in children.
- **Grossly**: poorly defined, gray white, infiltrative, expand and distort the brain tissue
- **Microscopically**:
  - Low to moderate cellularity
  - Proliferation of astrocytes with variable amount of fibrillary stroma.
  - The cells show slightly pleomorphic and hyperchromatic nuclei.
  - No mitotic figures.

GLIOMAS

Astrocytoma

Fibrillary astrocytoma (WHO Grade II):



Fibrillary stroma with slightly pleomorphic and hyperchromatic cells

GLIOMAS

Astrocytoma

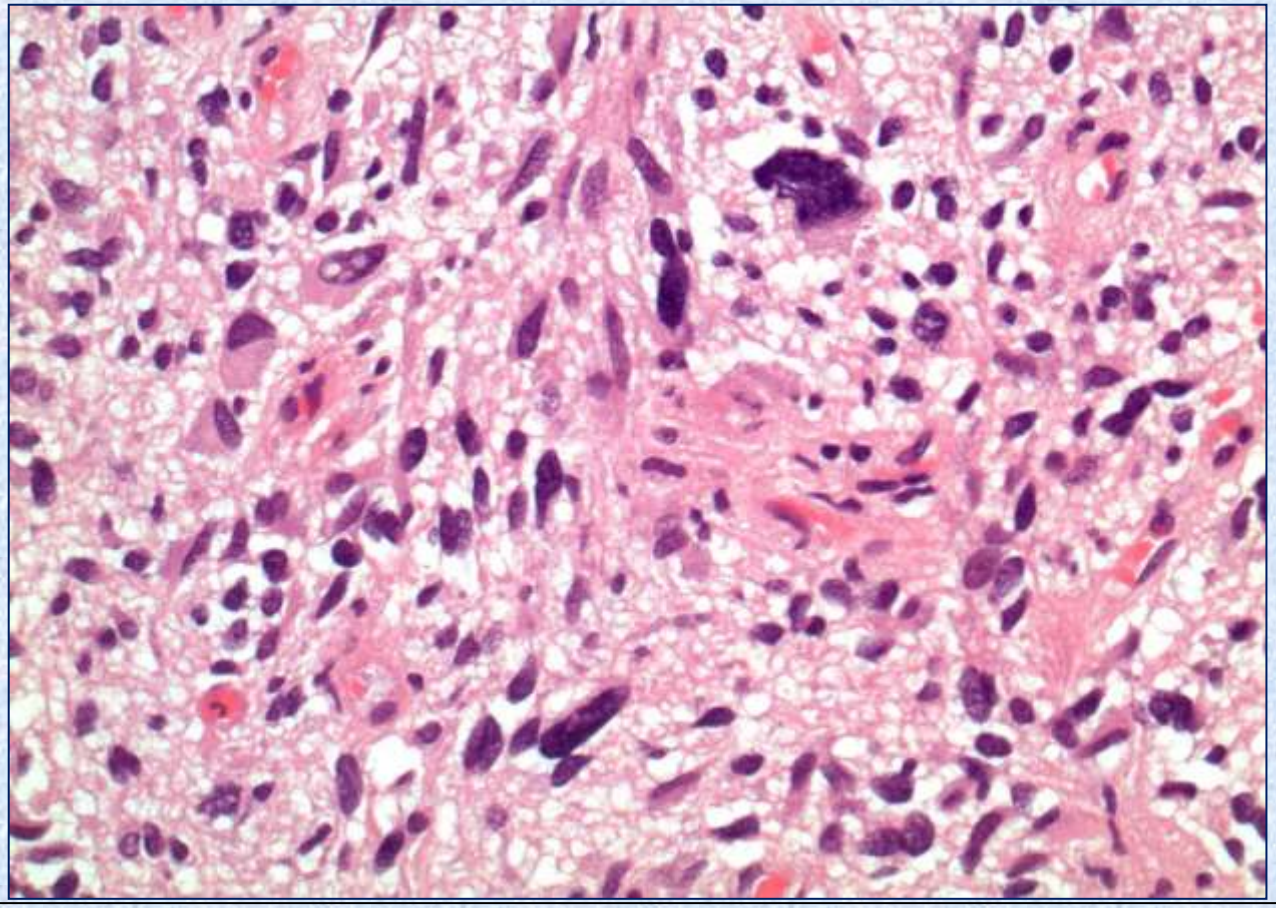
C. Anaplastic astrocytoma (WHO Grade III):

- An Aggressive tumor
- Commonly affects adults
- Site: cerebral hemispheres.
- ***Grossly***: an ill-defined, gray white tumor with infiltrative boredr
- ***Microscopically***:
  - high cellularity
  - nuclear anaplasia (tumor giant cells)
  - high mitotic activity.
  - NO necrosis

GLIOMAS

Astrocytoma

Anaplastic astrocytoma (WHO Grade III):



Cellular tumor with nuclear anaplasia and tumor giant cells

GLIOMAS

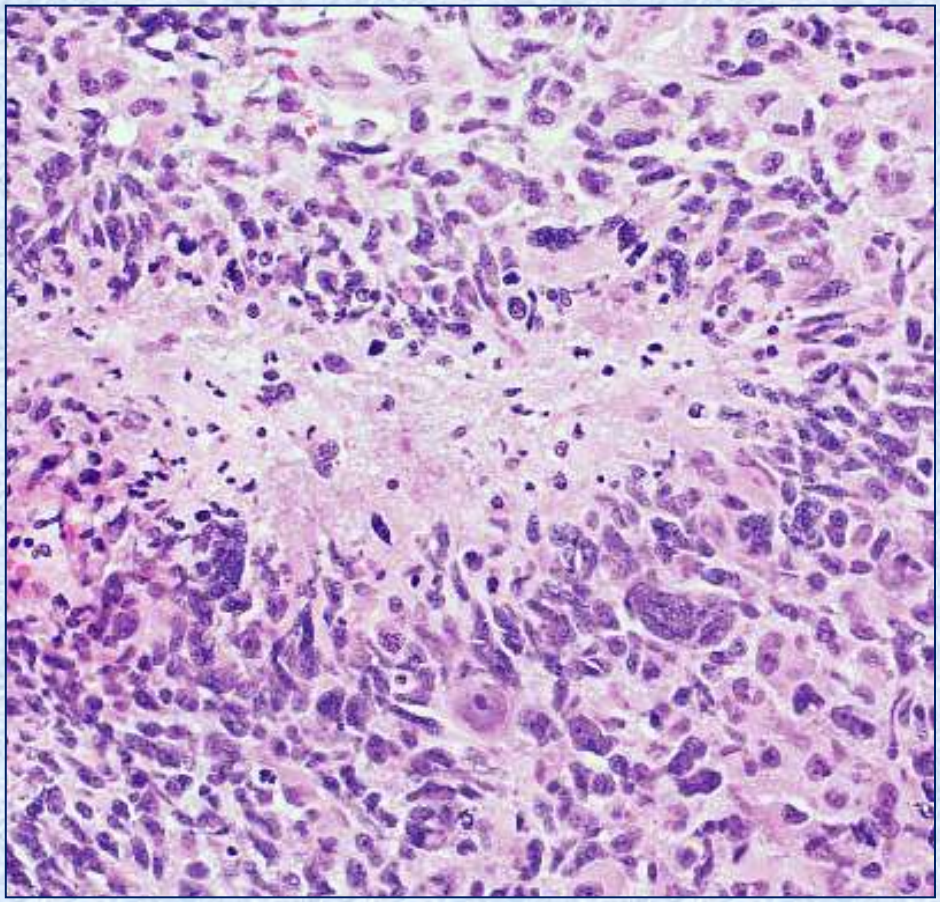
Astrocytoma

D. Glioblastoma Multiform; GBM (WHO Grade IV):

- The most aggressive malignant glial tumors.
- Commonly affects adults and elderly people
- **Site**: usually involves cerebral hemispheres.
- **Grossly**: an irregular tumor, may reach large size, with foci of hemorrhage, necrosis and cyst formation.
- **Microscopically**:
  - Highly cellular tumor
  - Prominent nuclear plromorphism, high mitotic activity and tumor giant cells.
  - Proliferation of small vascular spaces (micro-vascular proliferation)
  - Palisade necrosis: tumor cells arrange around areas of necrosis

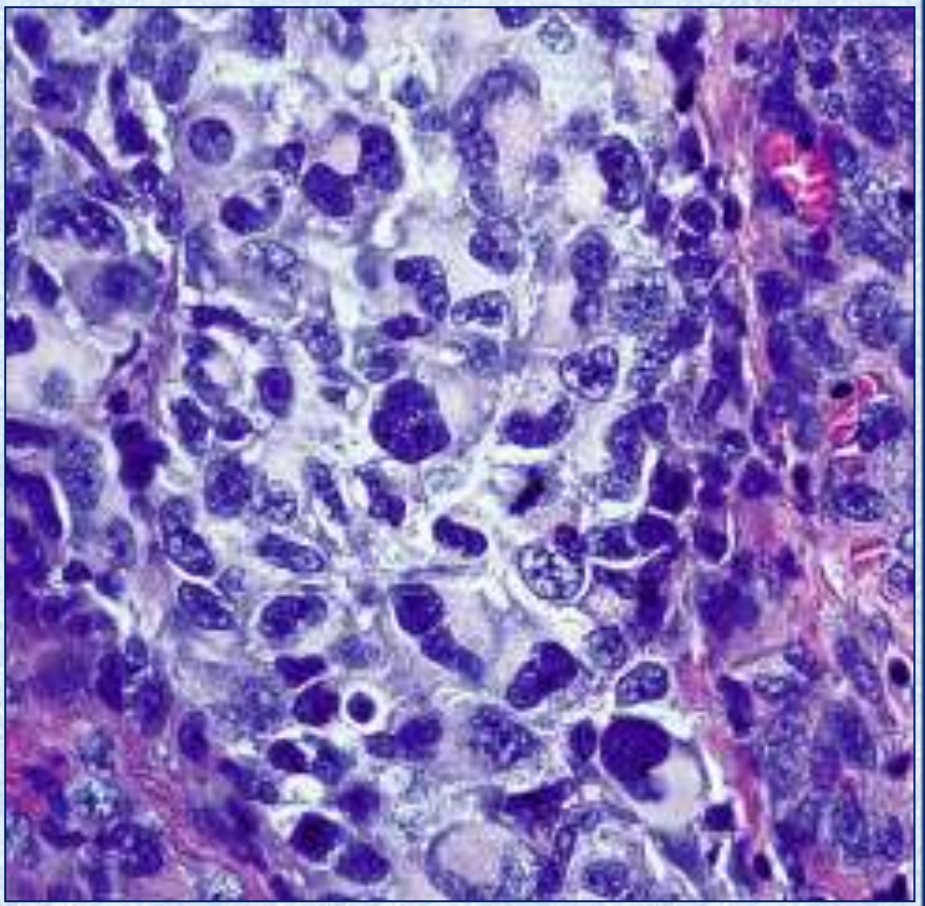
GLIOMAS

Astrocytoma



Palisade necrosis

Glioblastoma multiform



Highly cellularity with nuclear anaplasia and tumor giant cells

GLIOMAS

	Pilocytic astrocytoma	Fibrillary astrocytoma	Anaplastic astrocytoma	Glioblastoma multiformis
Site	Cerebellum	Cerebral hemisphere and brain stem	Cerebral hemisphere	Cerebral hemisphere and basal ganglia
Age	Children and young adults	Young adults	Adults	Adults and elderly
GP	-Well defined nodule -Commonly cyst	-Poorly-defined -Infiltrative -Gray white	-Poorly-defined -Infiltrative -Gray white	-Poorly-defined -Infiltrative -Hge and necrosis
MP	-Bipolar cells -With thin long processes -No mitosis	-Bipolar cells -Fibrillary stroma -Pleomorphic cells -No mitosis	-High cellularity -Pleomorphism -Mitosis	-High cellularity -Pleomorphism -Frequent mitosis -Giant cells -Palisade necrosis -Micro-vessel prolif

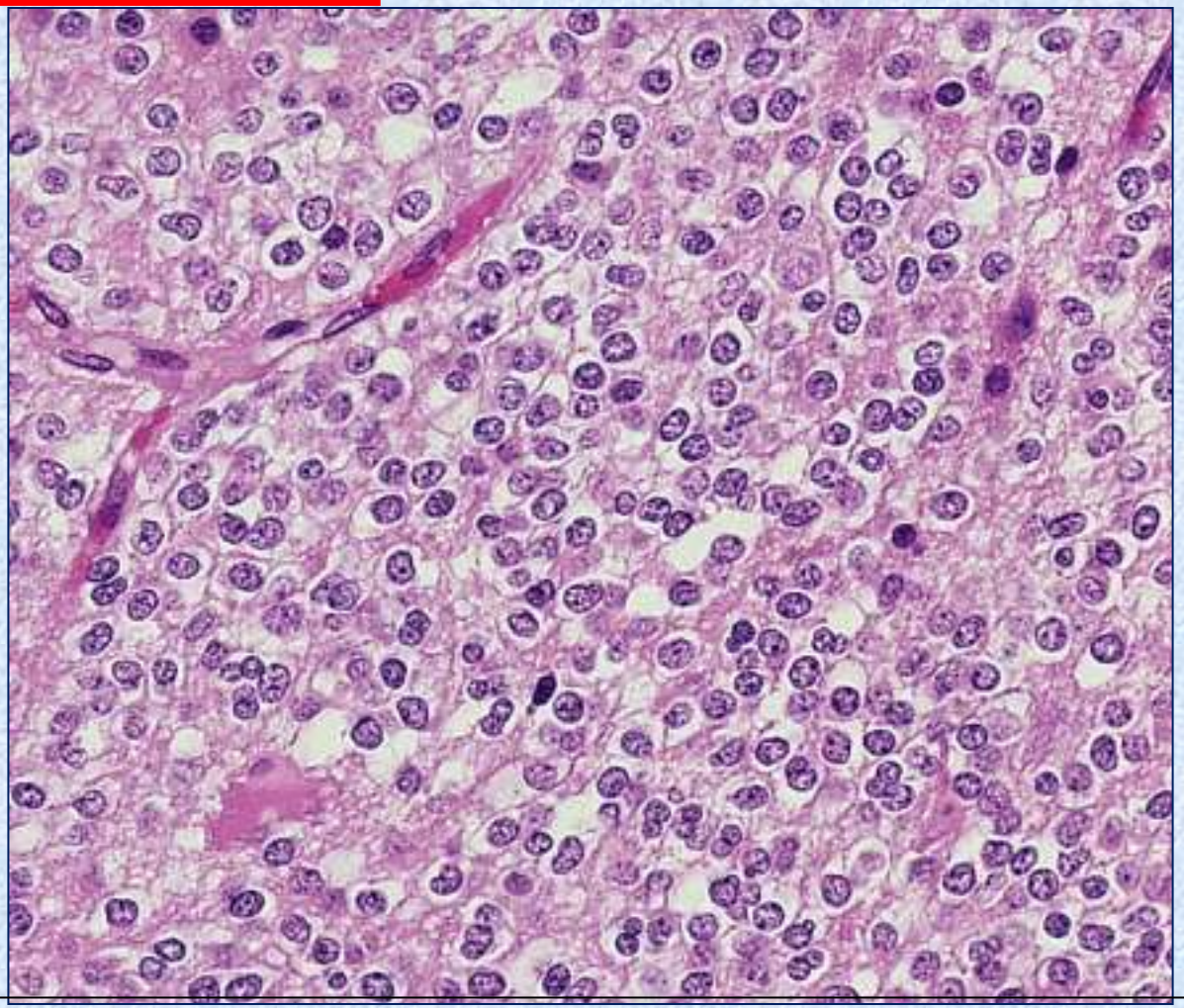
GLIOMAS

Oligodendroglioma

- Less common and usually slowly growing tumor
- Classified as WHO II tumor
- Occurs in cerebral hemispheres or within the ventricles
- ***Grossly***: Usually well-circumscribed, grey-white mass with frequent cystic areas and calcification
- ***Microscopically***:
  - Sheets of rounded or polygonal cells with small rounded nuclei
  - Characterized by peri-nuclear pale halo.
  - Scanty stroma with proliferation of small vessels.
  - Anaplastic change may occur (WHO III): increased cellularity, mitosis and nuclear atypia

GLIOMAS

Oligodendroglioma



Polygonal cells with rounded nuclei surrounded by peri-nuclear halo

GLIOMAS

Ependymoma

- Arises from ependymal cell lining of ventricles and spinal canal
- Commonly affects children and young adults
- Common sites:
  - In brain: 4th ventricle, lateral ventricles, 3rd ventricle.
  - In spinal cord, commonly involes lumbar spine
- In children; they can be associated with hydrocephalus
- A specific variant: myxopapillary ependymoma characteristically occurs in the region of cauda equina and originates from filum terminale.

GLIOMAS

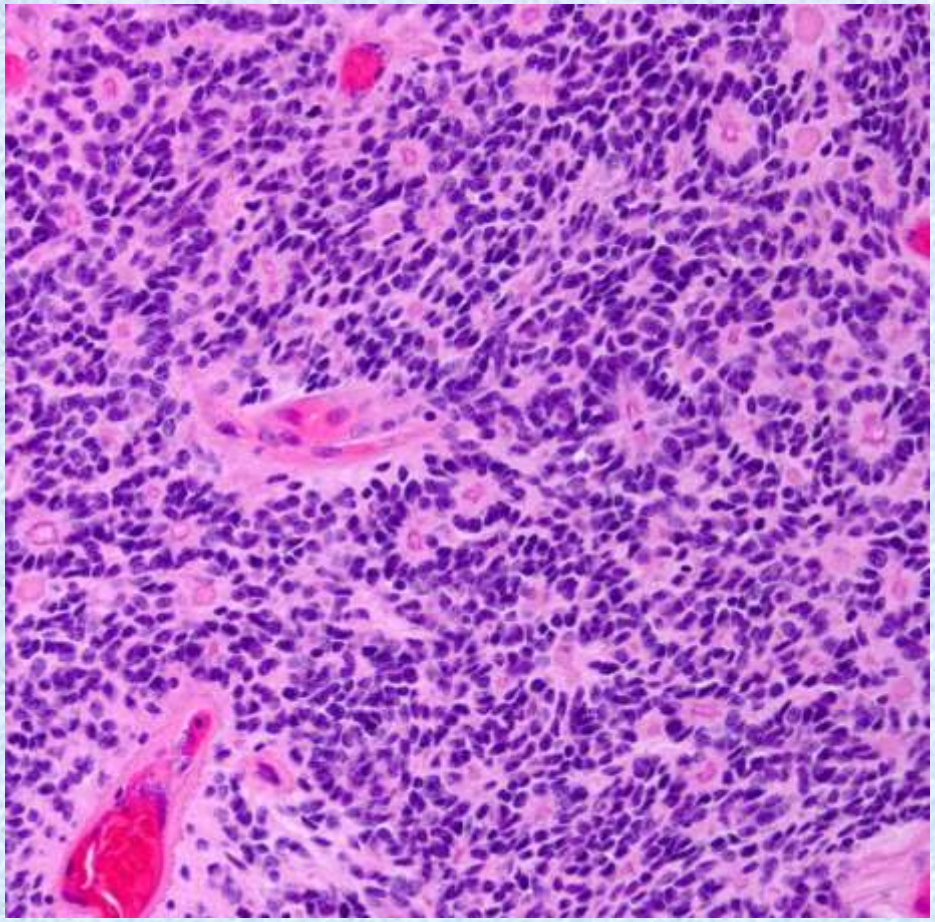
Ependymoma

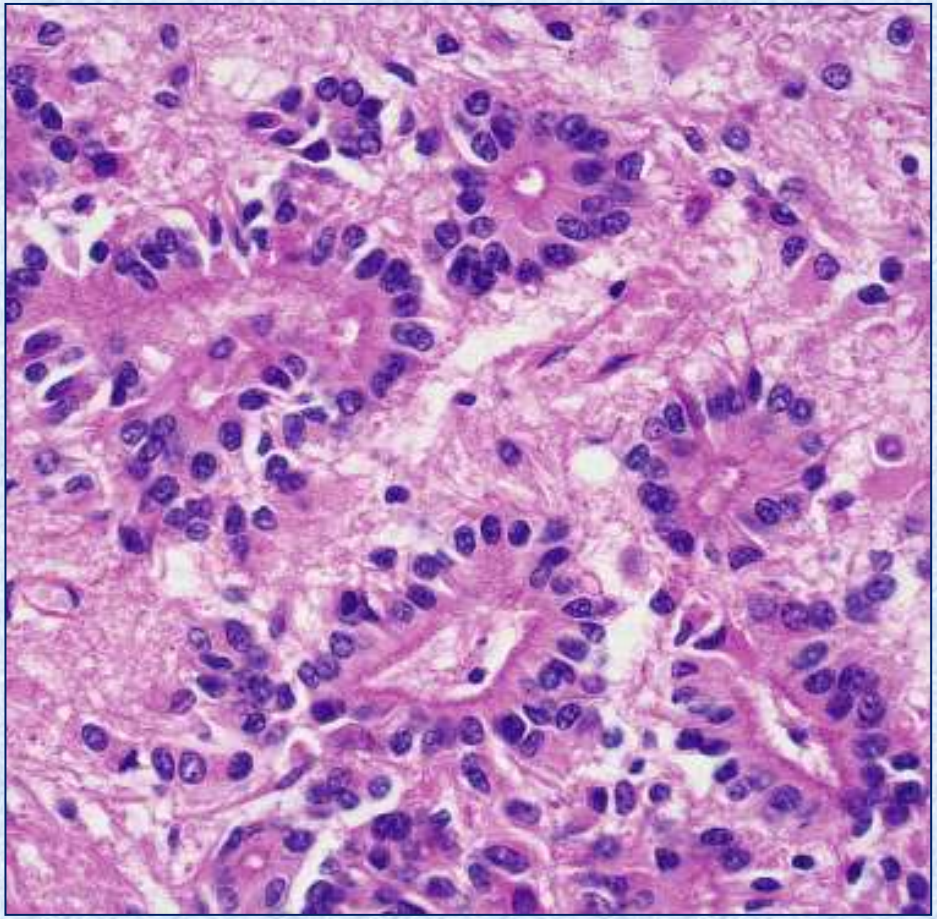
- ***Grossly***: well-defined; gray fleshy mass.

- ***Microscopically***
  - Cellular tumor.
  - Tumor cells have regular round to oval nuclei with granular cytoplasm.
  - Cytoplasmic processes of tumor cells condense around blood vessels to form pseudo-rosettes, or around central lumen to form rosettes (diagnostic).

GLIOMAS

Ependymoma





Tumor cells condense around blood vessels (pseudo-rosettes)

Test yourself

Which is a the most common tumors of CNS?

A. Gliomas
B. Meningioma
C. Medulloblastoma
D. Metastasis

Which of the following is a criterion of high grade gliomas?

E. Low tumor cellularity
F. Excess fibrillary background
G. Low mitotic activity
H. Necrosis

Test yourself

..... is a type with glioma with mostly benign behavior

A. Astrocytoma
B. Ependymoma
C. Oligodendroglioma
D. Glioblastoma multiform

..... commonly affects children

A. Pilocytic astrocytoma
B. Fibrillary astrocytoma
C. Anaplastic astrocytoma
D. Glioblastoma multiform

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Good luck

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