Pathology of CNS Tumors, part II

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Introduction

By end of this lecture; you have to:

- Identify different types of gliomas and describe pathological features.
- Describe main characters of medulloblastoma.
- Identify origin, types and morphology of meningioma.
- Describe causes and effects of high intracranial tension

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

C.Meningeal cells

D.Endothelial cells

Introduction

WHO classification of CNS tumors

Diffuse astrocytic and oligodendroglial tumours
Diffuse astrocytoma, IDH-mutant
  Gemistocytic astrocytoma, IDH-mutant
Diffuse astrocytoma, IDH-wildtype
Diffuse astrocytoma, NOS

Anaplastic astrocytoma, IDH-mutant
Anaplastic astrocytoma, IDH-wildtype
Anaplastic astrocytoma, NOS

Glioblastoma, IDH-wildtype
  Giant cell glioblastoma
  Gliosarcoma
  Epithelioid glioblastoma
Glioblastoma, IDH-mutant
Glioblastoma, NOS

Diffuse midline glioma, H3 K27M–mutant

Oligodendroglioma, IDH-mutant and 1p/19q-codeleted
Oligodendroglioma, NOS

Anaplastic oligodendroglioma, IDH-mutant and 1p/19q-codeleted
Anaplastic oligodendroglioma, NOS

Oligoastrocytoma, NOS
Anaplastic oligoastrocytoma, NOS

Other astrocytic tumours
Pilocytic astrocytoma
  Pilomyxoid astrocytoma
Subependymal giant cell astrocytoma
Pleomorphic xanthoastrocytoma
Anaplastic pleomorphic xanthoastrocytoma

Ependymal tumours
Subependymoma
Myxopapillary ependymoma
Ependymoma
  Papillary ependymoma
  Clear cell ependymoma
  Tanycytic ependymoma
Ependymoma, *RELA* fusion–positive
Anaplastic ependymoma

Other gliomas
Chordoid glioma of the third ventricle
Angiocentric glioma
Astroblastoma

Choroid plexus tumours
Choroid plexus papilloma
Atypical choroid plexus papilloma
Choroid plexus carcinoma

Introduction

WHO classification of CNS tumors

Neuronal and mixed neuronal-glial tumours
Dysembryoplastic neuroepithelial tumour
Gangliocytoma
Ganglioglioma
Anaplastic ganglioglioma
Dysplastic cerebellar gangliocytoma
(Lhermitte–Duclos disease)
Desmoplastic infantile astrocytoma and ganglioglioma
Papillary glioneuronal tumour
Rosette-forming glioneuronal tumour
Diffuse leptomeningeal glioneuronal tumour
Central neurocytoma
Extraventricular neurocytoma
Cerebellar liponeurocytoma
Paraganglioma

Tumours of the pineal region
Pineocytoma
Pineal parenchymal tumour of intermediate differentiation
Pineoblastoma
Papillary tumour of the pineal region

Embryonal tumours
Medulloblastomas, genetically defined
Medulloblastoma, WNT-activated
Medulloblastoma, SHH-activated and *TP53*-mutant
Medulloblastoma, SHH-activated and *TP53*-wildtype
Medulloblastoma, non-WNT/non-SHH
Medulloblastoma, group 3
Medulloblastoma, group 4
Medulloblastomas, histologically defined
Medulloblastoma, classic
Medulloblastoma, desmoplastic/nodular
Medulloblastoma with extensive nodularity
Medulloblastoma, large cell / anaplastic
Medulloblastoma, NOS

Embryonal tumour with multilayered rosettes, C19MC-altered
Embryonal tumour with multilayered rosettes, NOS
Medulloepithelioma
CNS neuroblastoma
CNS ganglioneuroblastoma
CNS embryonal tumour, NOS
Atypical teratoid/rhabdoid tumour
CNS embryonal tumour with rhabdoid features

Tumours of the cranial and paraspinal nerves
Schwannoma
Cellular schwannoma
Plexiform schwannoma

Introduction

Common 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**

Tumors of meninges

- **Meningioma**.

Metastatic tumours

GLIOMAS

Astrocytoma

- Tumors arising from astrocytes
- **Types**
  1. Pilocytic astrocytoma (WHO Grade I)
  2. Fibrillary astrocytoma (WHO Grade II)
  3. Anaplastic astrocytoma (WHO Grade III)

- **Clinically**:
  - Seizures (convulsions)
  - Symptoms of ICT (Headache, vomiting, blurred vision)
  - Focal neurological deficit

GLIOMAS

Astrocytoma

A. Pilocytic astrocytoma (WHO Grade I):

- Typically occurs in children and young adults.
- The common site is the cerebellum.
- Slowly growing and rarely infiltrative.

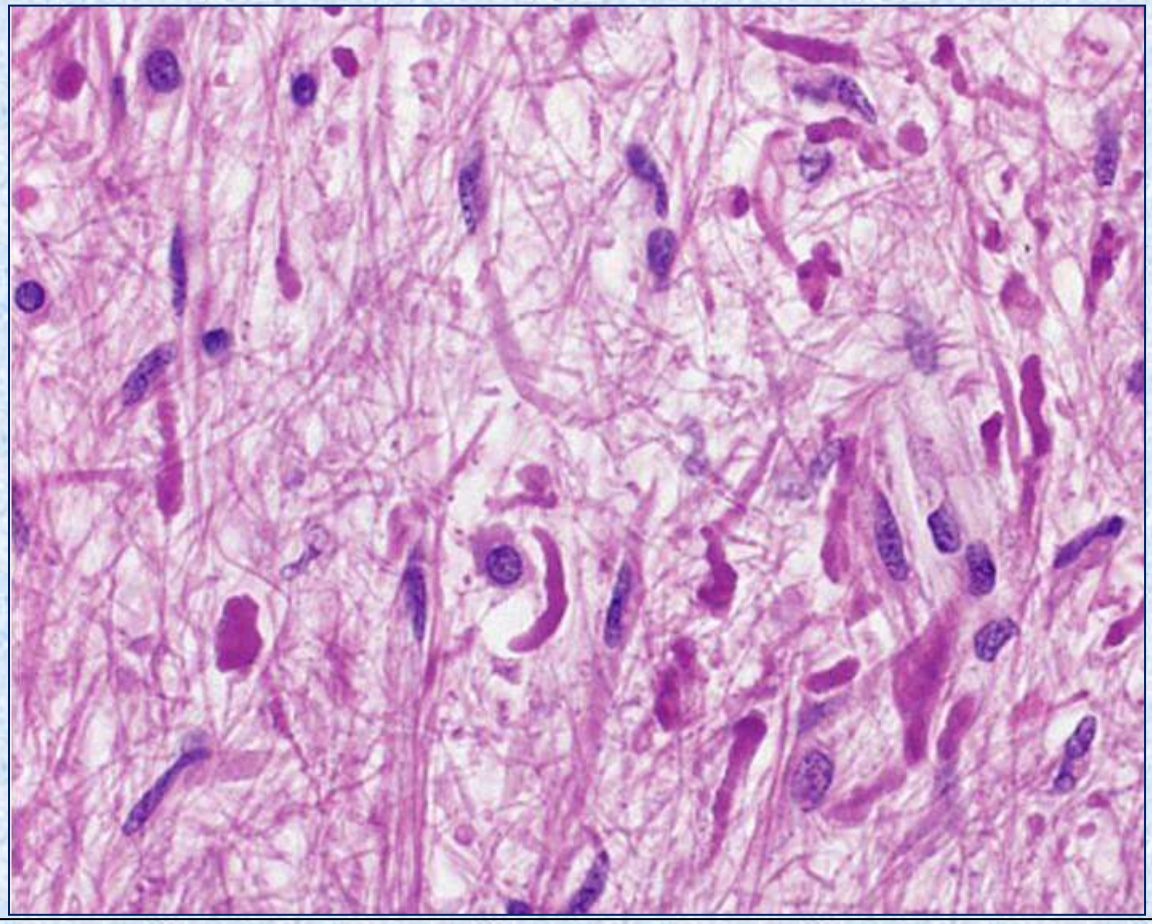
- ***Grossly***: well-circumscribed, often cystic tumor with a mural nodule attach to cyst wall.

- ***Microscopically***: tumor consists of bipolar cells with long, thin processes (hair cell) with no features of malignancy

GLIOMAS

Astrocytoma

A. Pilocytic astrocytoma (WHO Grade I):



Bipolar cells with long, thin processes (hair cell)

GLIOMAS

Astrocytoma

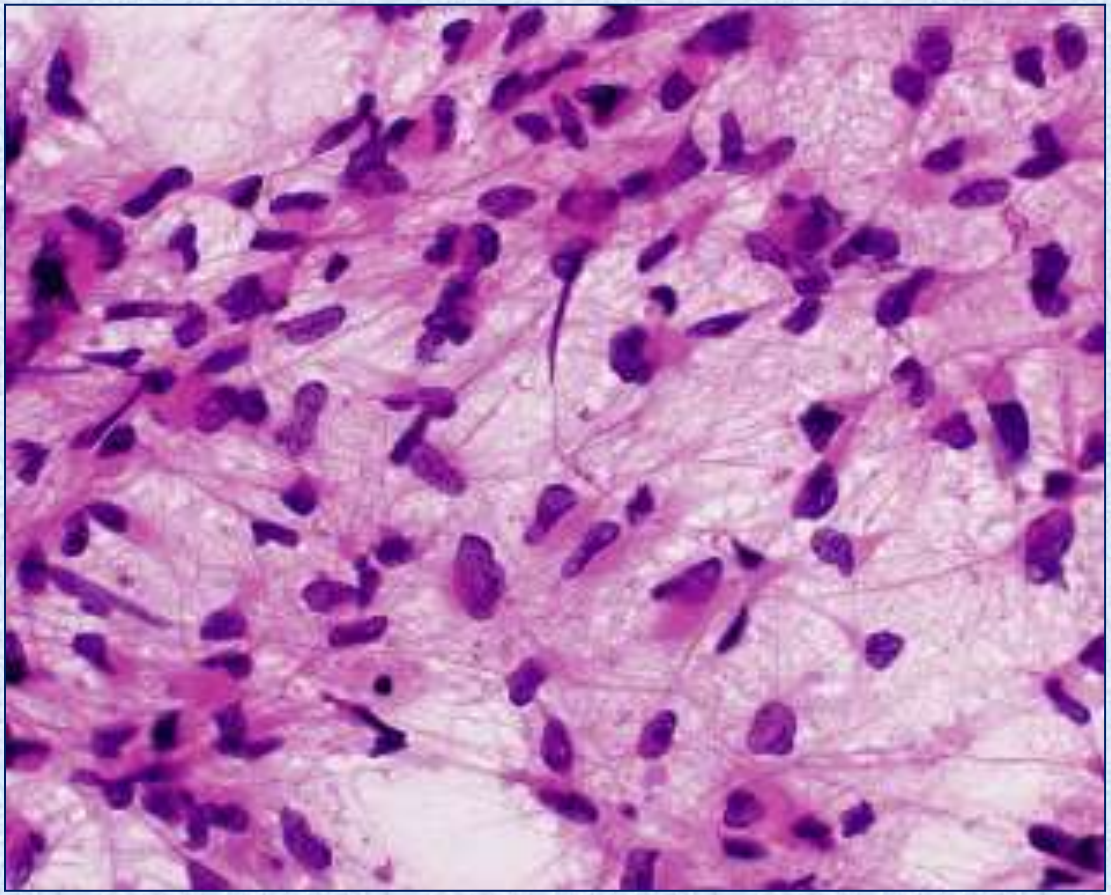
A. Diffuse or fibrillary astrocytoma (WHO Grade II):

- Represent about 2/3 of young adult brain tumors.
- Common site is cerebral hemispheres in adults and brain stem in children.
- ***Grossly***: poorly defined, diffuse growth, gray white, infiltrative, expand and distort the brain tissue
- ***Microscopically***:
  - Histologically benign
  - Astrocytic cell proliferation in a fibrillary stroma.
  - The nuclei are pleomorphic and hyperchromatic.
  - No mitotic figures.

GLIOMAS

Astrocytoma

A. Diffuse or fibrillary astrocytoma (WHO Grade II):



Fibrillary stroma with slightly pleomorphic and hyperchromatic cells

GLIOMAS

Astrocytoma

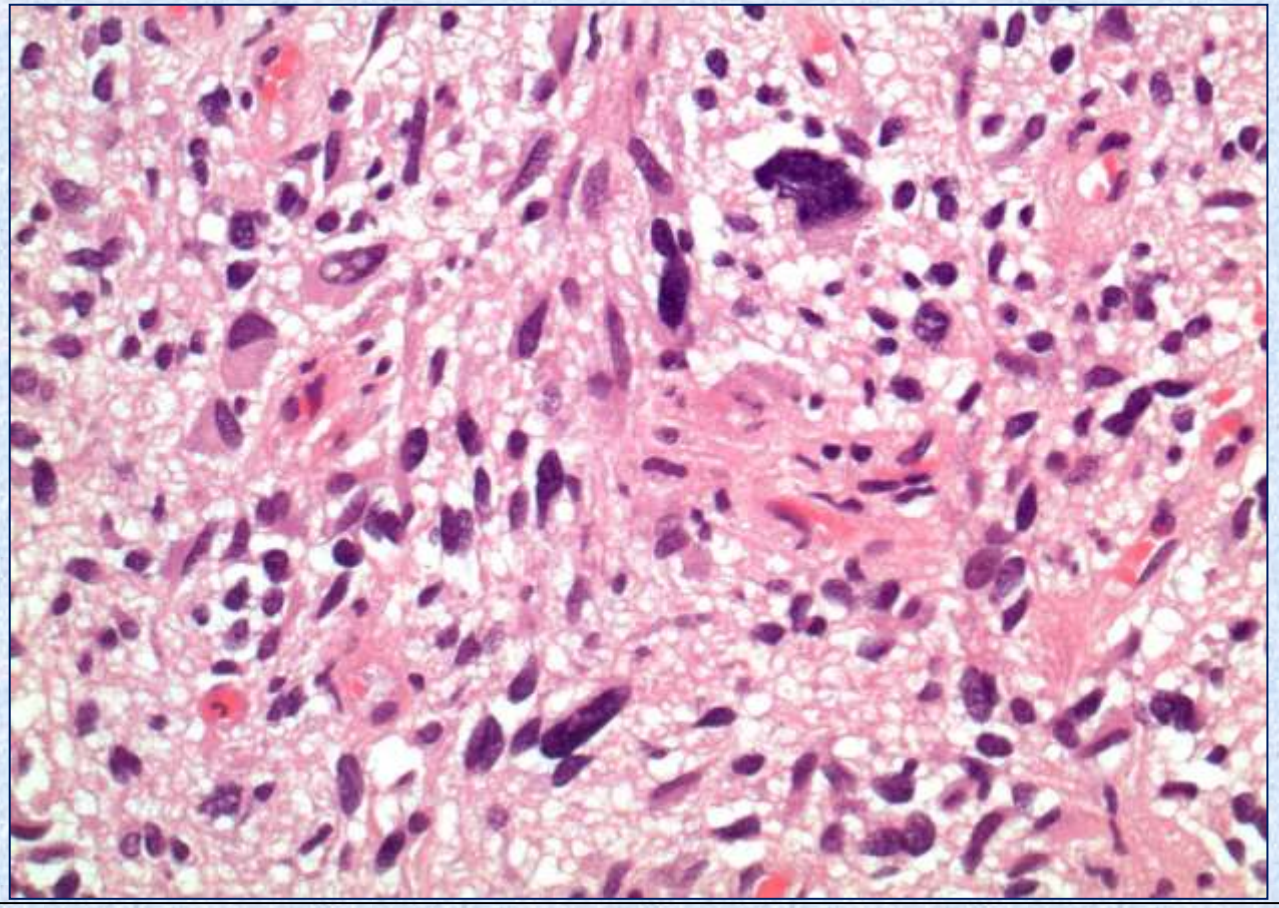
A. Anaplastic astrocytoma (WHO Grade III):

- An Aggressive tumor
- Commonly affects adults
- Site: cerebral hemispheres.
- ***Microscopically***:
  - high cellularity
  - nuclear anaplasia (tumor giant cells)
  - mitotic activity.
  - NO necrosis

GLIOMAS

Astrocytoma

A. Anaplastic astrocytoma (WHO Grade III):



Cellular tumor with nuclear anaplasia and tumor giant cells

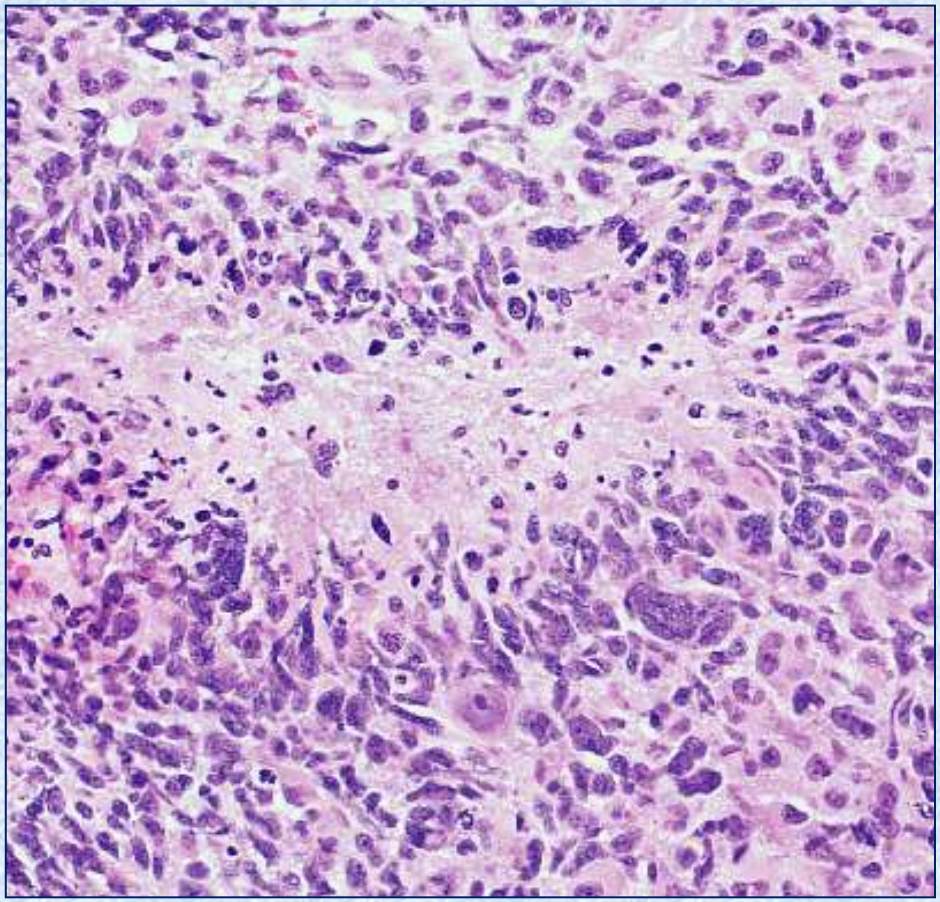
GLIOMAS

Glioblastoma multiform (GBM); WHO grade IV

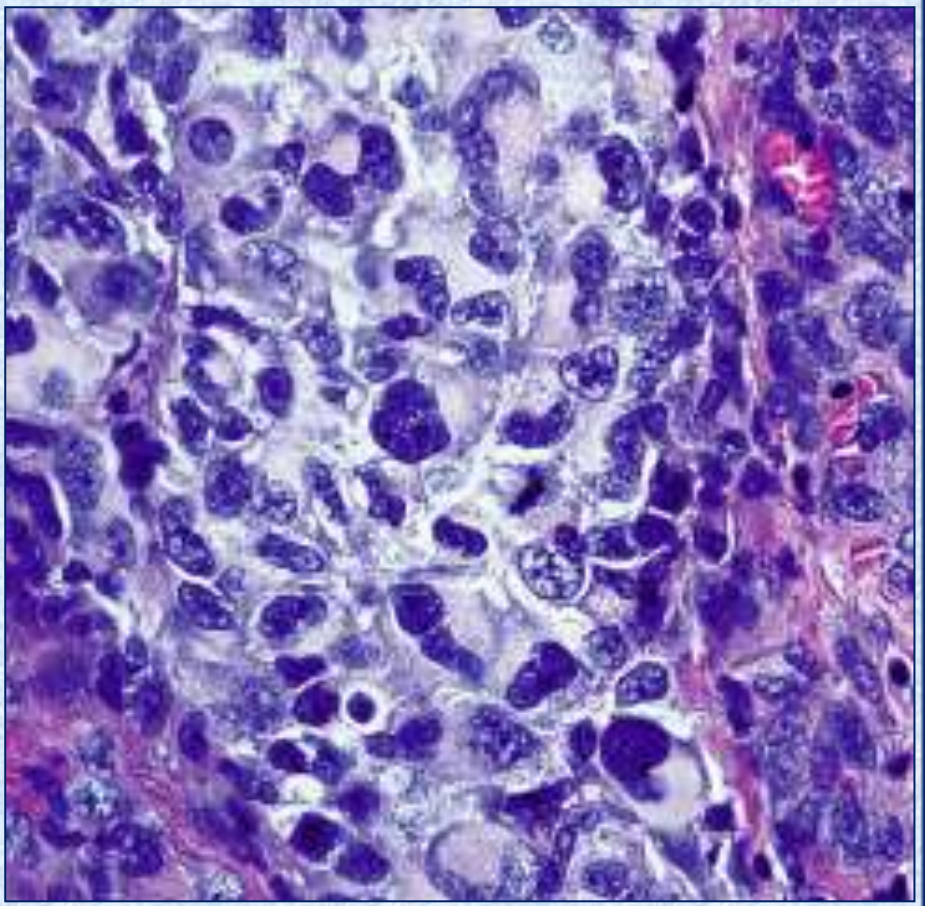
- The most aggressive malignant glial tumors.
- Commonly affects adult
- Site: cerebral hemispheres.
- ***Grossly***: solitary, butterfly with foci of hemorrhage, necrosis and cyst formation.
- ***Microscopically***:
  - Highly cellular tumor, with prominent nuclear anaplasia, high mitotic activity and tumor giant cells.
  - Proliferation of small vascular spaces (micro-vascular proliferation)
  - Palisade necrosis: tumor cells arrange around areas of necrosis

GLIOMAS

Glioblastoma multiform (GBM); WHO grade IV



Palisade necrosis



Highly cellularity with nuclear anaplasia and tumor giant cells

GLIOMAS

	Pilocytic astrocytoma	Fibrillary astrocytoma	Anaplastic astrocytoma	Glioblastoma multiformis
Site	Cerebellum	Cerebral hemisphere sand brain stem	Cerebral hemisphere	Cerebral hemisphere and basal ganglia
Age	Children and young adults	Children and young adults	Adults	Adults
GP	-Well defined nodule -May be cystic	-Poorly-defined -Infiltrative -Gray white	-Poorly-defined -Infiltrative -Gray white	-Poorly-defined -Infiltrative -Soft and firm areas -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 proliferation

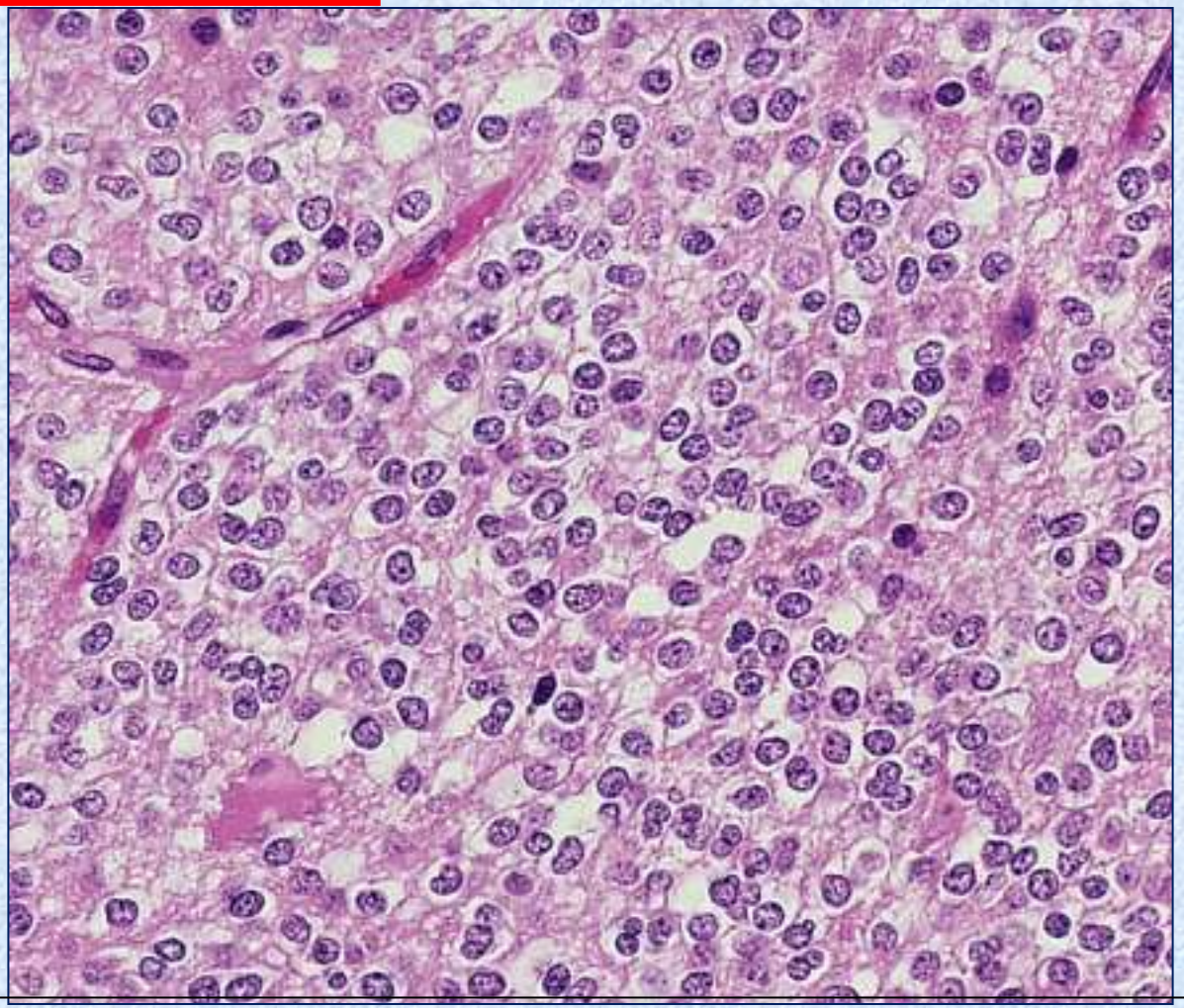
GLIOMAS

Oligodendroglioma

- A rare slowly growing tumor (WHO II)
- Frequent cystic changes and calcification.
- ***Microscopically***:
  - Sheets of rounded or polygonal cells with small rounded nuclei
  - Characterized by peri-nuclear pale halo.
  - Scanty stroma with numerous thin walled blood vessels.
  - Anaplastic oligodendroglioma (WHO III) shows increased cellularity, mitosis and nuclear atypia

GLIOMAS

Oligodendroglioma



Polygonal cells with rounded nuclei surrounded by peri-nuclear halo

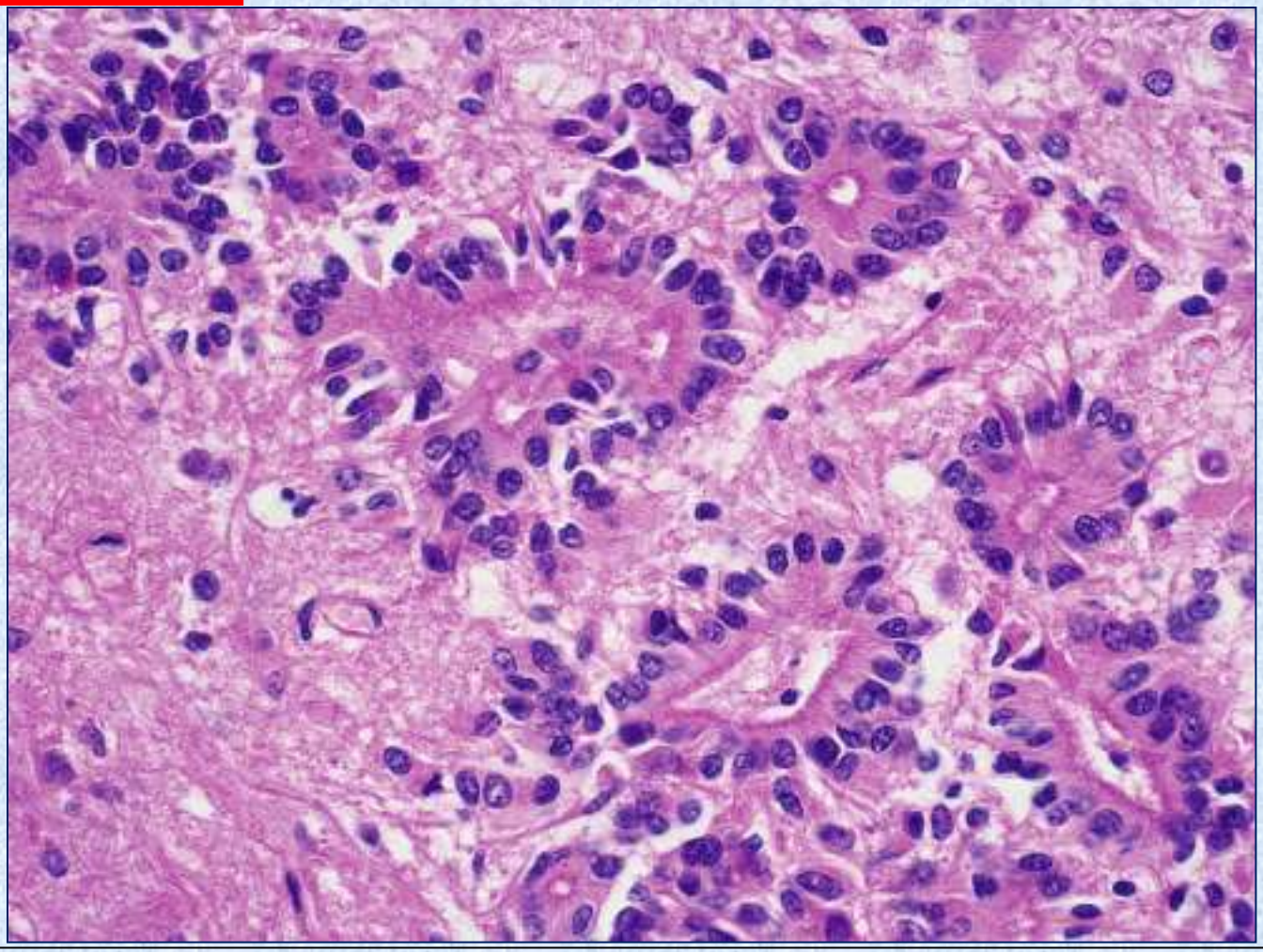
GLIOMAS

Ependymoma

- Arises from ependymal cell lining of ventricles and spinal canal.
- Affects children and young adult
- Associated with hydrocephalus
- ***Grossly***: 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)

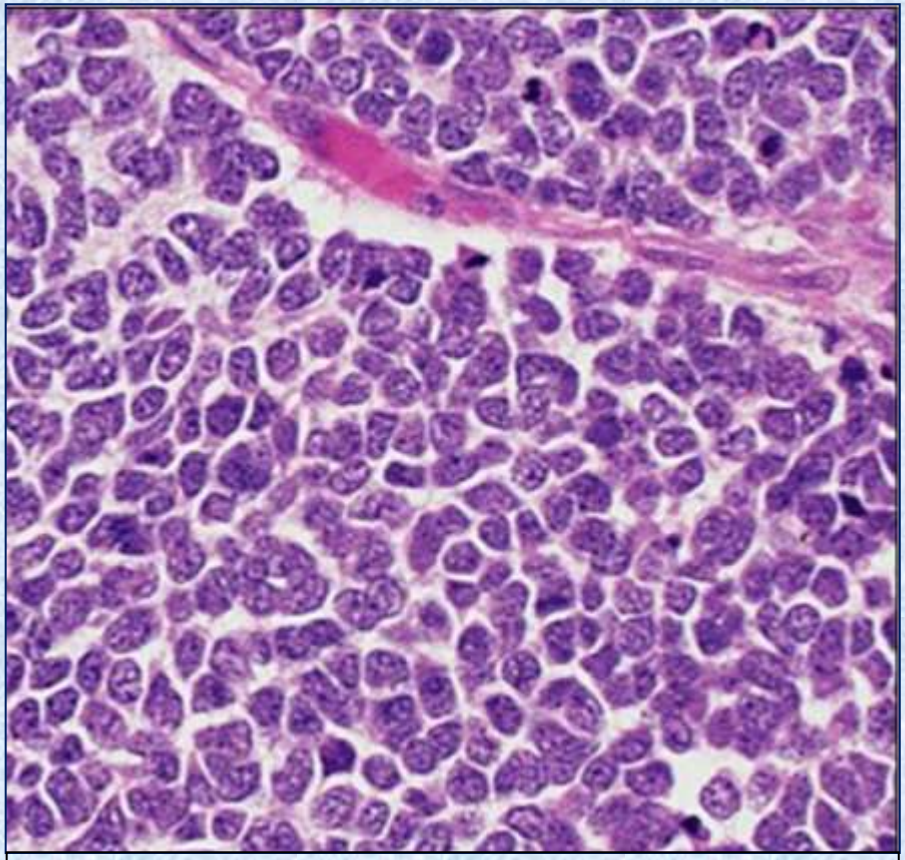
Tumors of primitive cells

Medulloblastoma

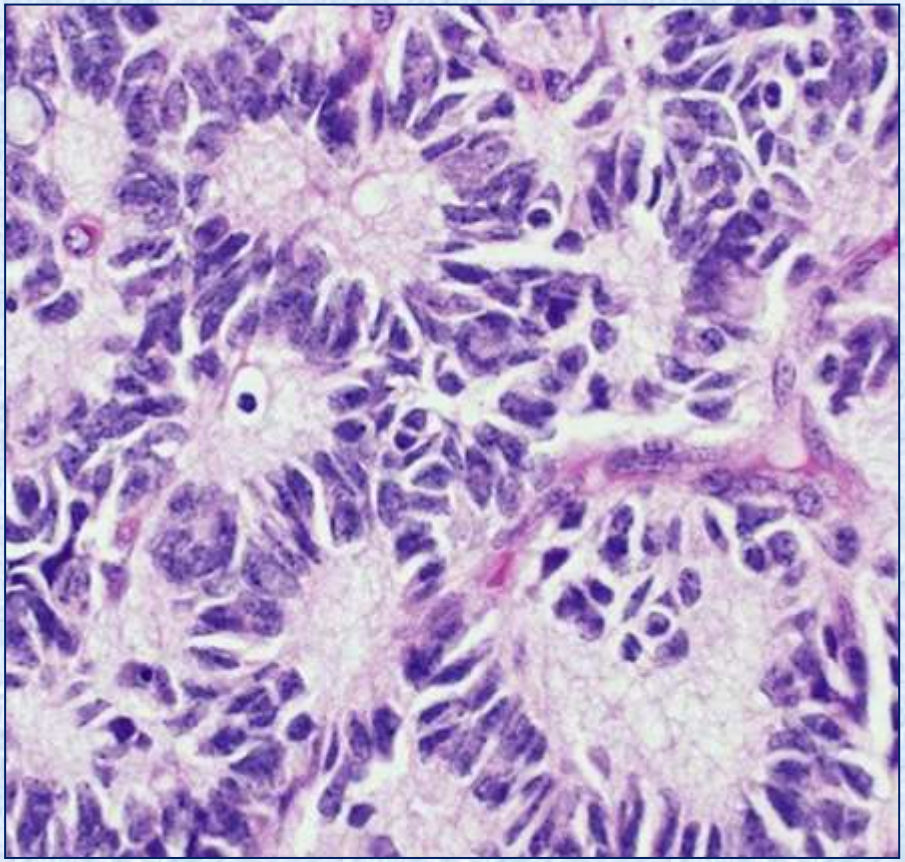
- One of primitive tumors (blastomas) that affect children
- Rapidly growing malignant neoplasm (WHO grade IV)
- Site: commonly cerebellum
- ***Grossly***:
  - Fleshy grayish pink mass projecting into lumen of fourth ventricle and obstructing the pathway of the C.S.F.
  - The tumor disseminates through the CSF.
  - Cut surface shows hemorrhage and necrosis.
- ***Microscopically:***
  - Cellular tumor
  - Formed of small round cells with scanty cytoplasm and small central hyperchromatic nuclei.
  - Frequent mitosis
  - Tumors cells show focal pseudo-rosettes around blood vessels

Tumors of primitive cells

Medulloblastoma



Small round cells with scanty cytoplasm, hyperchromatic nculei and frequent mitosis



Focal pseudo-rosette formation

Tumors of meninges

Meningioma

- **Origin**: arise from the meningothelial cell lining of meninges
- **Behaviour**: commonly benign tumor, but can be malignant
- **Age**: The tumor occurs in adults.
- **Site**: Any site but commonly in relation to superior sagittal sinus or at base of brain.
- **Gross Picture**:
  - Variable sized round or oval capsulated tumor attached to under surface of the dura.
  - Firm in consistency.
  - Cut surface is grey white and shows whorly appearance.

Tumors of meninges

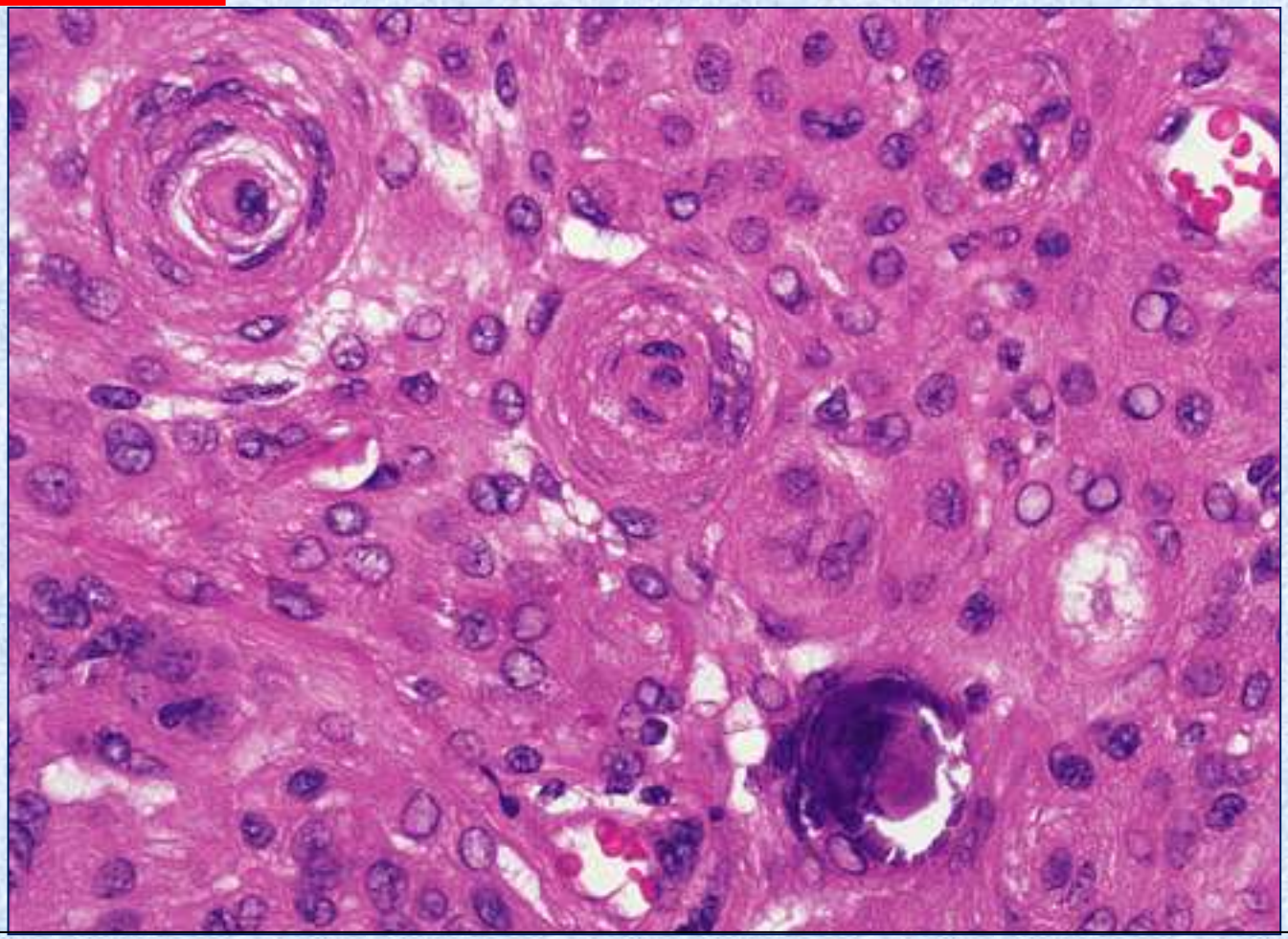
Meningioma

- **Microscopic Picture**:
  - Groups of large round or oval cells separated by variable amount of stroma
  - The cells arrange concentrically in whorls (diagnostic)
  - The cells have indistinct cell borders, eosinophilic cytoplasm and small round or oval nuclei.....syncytial appearance
  - The central cells undergo hyalinosis and calcification, psammoma bodies (diagnostic).
  - Microscopic variants:

1-Psammomatous	2-Fibroblastic
3-Meningiothelial	4-Papillary
5-Angiomatous	6-Metaplastic

Tumors of meninges

Meningioma



Syncytial tumor cells arrange in whorls with central hyalinized cells and calcification (psammoma bodies)

Increased intracranial tension

- **Aetiology**
  - Intracranial tumours (**mention**)
  - Intracranial inflammation:
    - Acute: -Suppurative: as septic meningitis
      -Non-suppurative: as viral meningitis
    - Chronic: -Non-specific: as chronic abscess
      -Specific: as TB, Gumma of syphilis
  - Intracranial vascular disorders (**mention**)
  - Hydrocephalus
- **Symptoms:**
  - Persistent headache
  - Vomiting
  - Blurred vision

Increased intracranial tension

- **Effects**
  1. Flattened brain convolution on same side of the lesion
  2. Shift of midline structures to opposite side
  3. Papillaedema: due to compression of retinal veins at sub-arachenoid space
  4. Skull changes: as thining of skull bone over the lesion (in chronic cases)
  5. Intracranial herniation:

Tentorial herniation

Tonsillar herniation:

Increased intracranial tension

- **Effects**

Tentorial herniation:

- Herniation of the cerebrum through tentorium cerebelli
- Occurs when the space occupying lesion is located above tentorium cerebelli
- Effects:

a. Compression of midbrain and aquiduct of Sylvious → Hydrocephalus
b. Compression of 4^{th} and 6^{th} cranial nerves → distorted eye movement
c. Compression of posterior cerebral artery → posterior cerebral infarction

Tonsillar herniation:

- Herniation of the brain stem through foramen magnum
  - Compression of medulla oblongata → compress respiratory center

Good luck

Dr Ahmed Roshdi