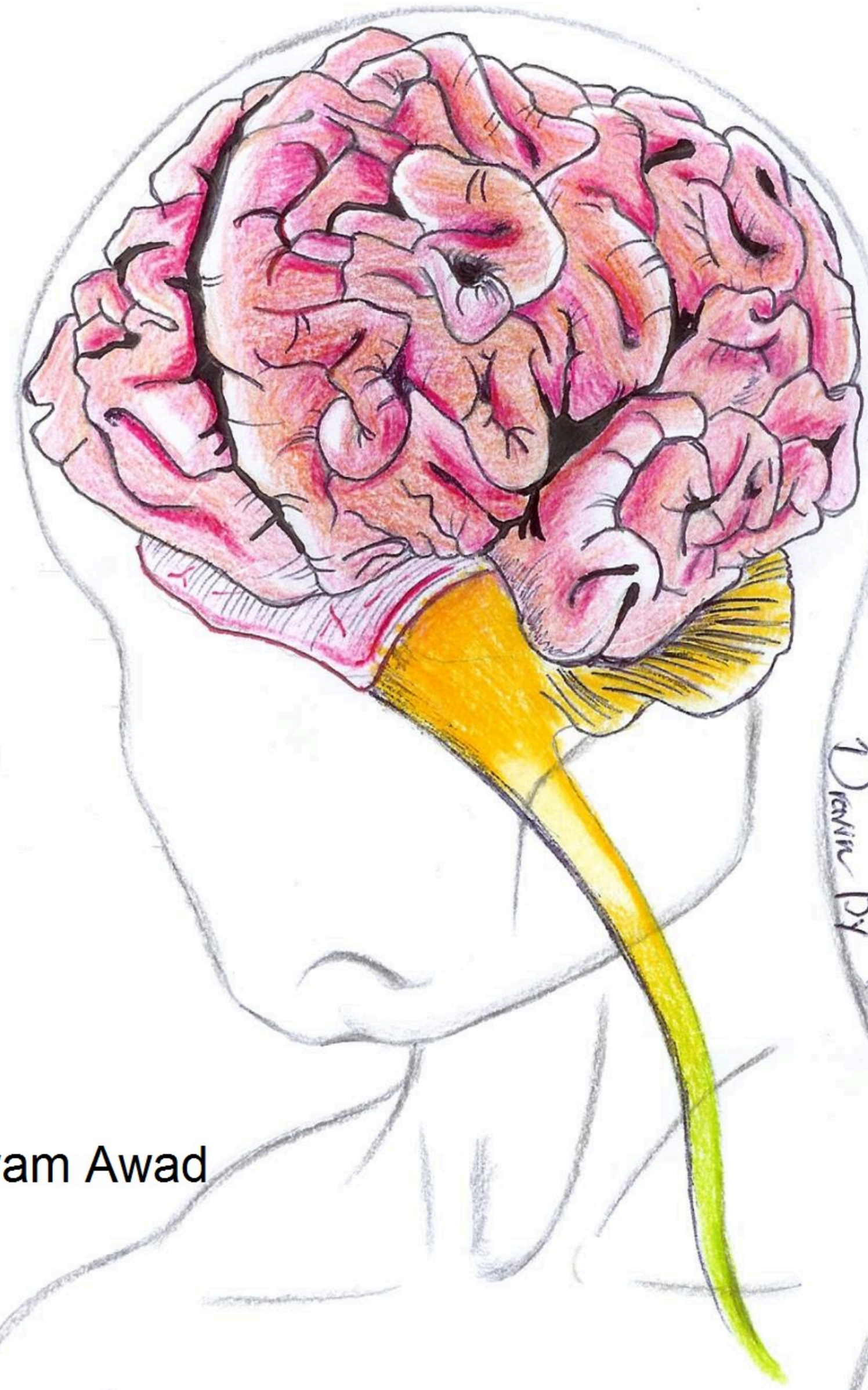
CENTRAL NERVOUS SYSTEM

- ○ Handout
- ○ Sheet
- ○ Slide

- ○ Anatomy
- ○ Physiology
- ■ Pathology
- ○ Biochemistry
- ○ Microbiology
- ○ Pharmacology
- ○ PBL



Done By:

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Lec #: 9

Drawn By Tarig Bushnaq ...

CNS 9
CNS tumors/2

DR Heyam Awad
FRCPath

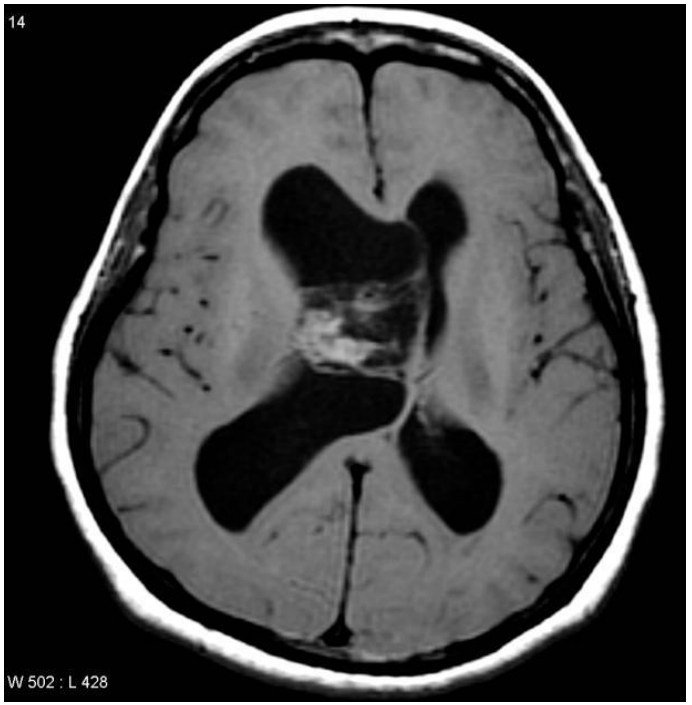
Brain tumors

- Gliomas
- Neuronal tumors
- Embryonal neoplasms
- Lymphoma
- Germ cell tumors
- Meningioma
- metastatic

Neuronal tumors

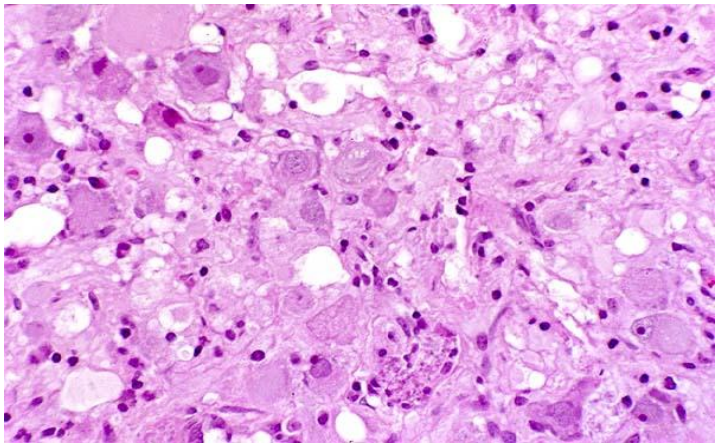
- 1. **Central neurocytoma**: low grade neoplasm within and adjacent to ventricular system.

  round uniform nuclei and islands of neuropil.



- 2. **ganglioglioma :** glial elements and mature appearing neurones.

usually slow rgowing but the glial element can progress



- **3.dysembryoplastic neuroepithelial tumor**: low grade childhood tumor .

  -good prognosis after resection

- Manifests with : seizures
- Mostly: temporal lobe.

Embryonal neoplasms (primitive neoplasms)

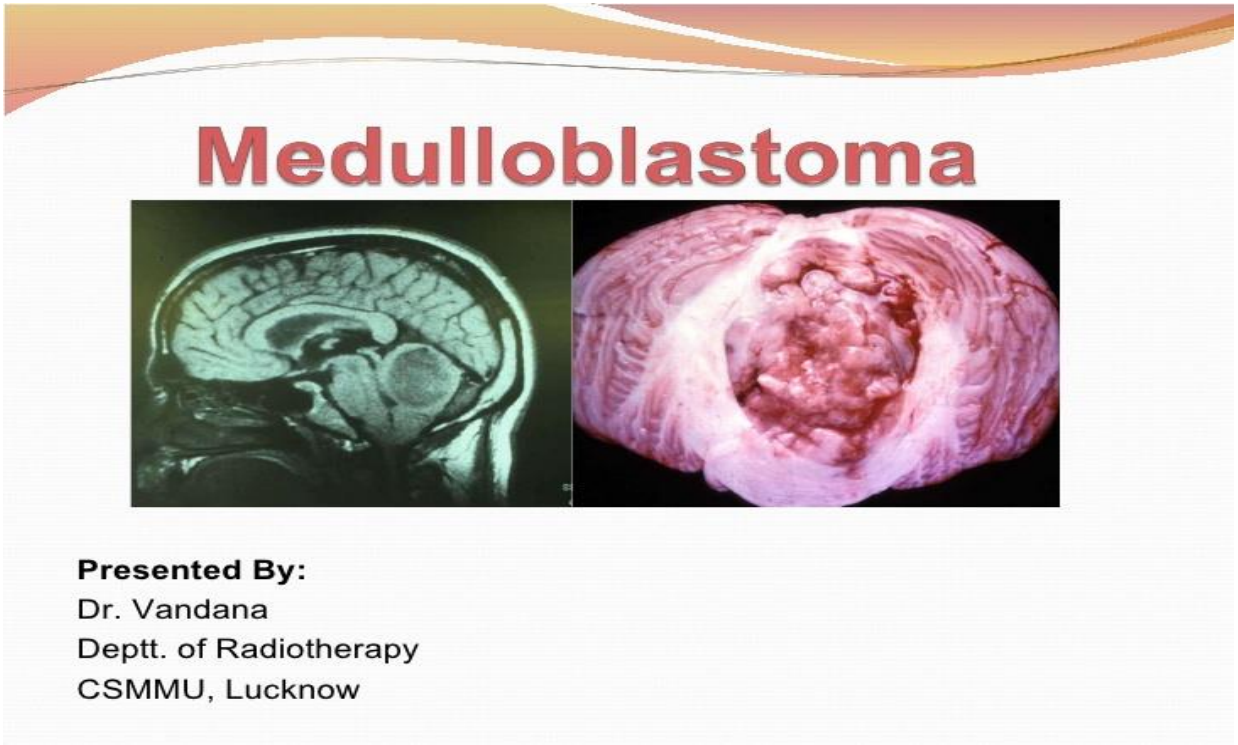
- Primitive appearance: small round cells, little cytoplasm
- Most common : medulloblastoma
- Medulloblastoma: 20% of paediatric brain tumors

medulloblastoma

- Occurs predominently in children
- Exclusively in the cerebellum
- Highly malignant if untreated
- Radiosensitive
- Surgery + chemo + radio.. 5 year survival reaches 75%
- Similar tumors outside the brain: PNETs= primitive neurosctodermal tumors

Macroscopic appearance

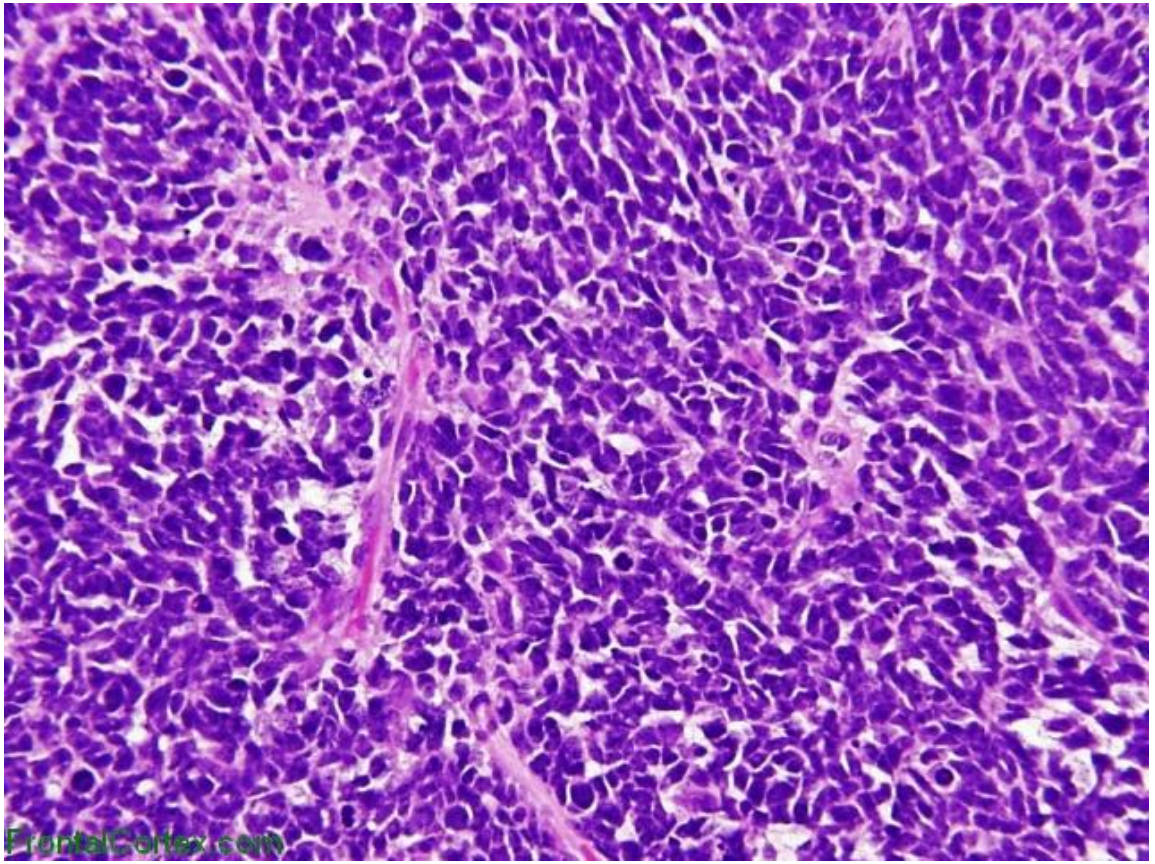
- Well circumscribed, grey, friable



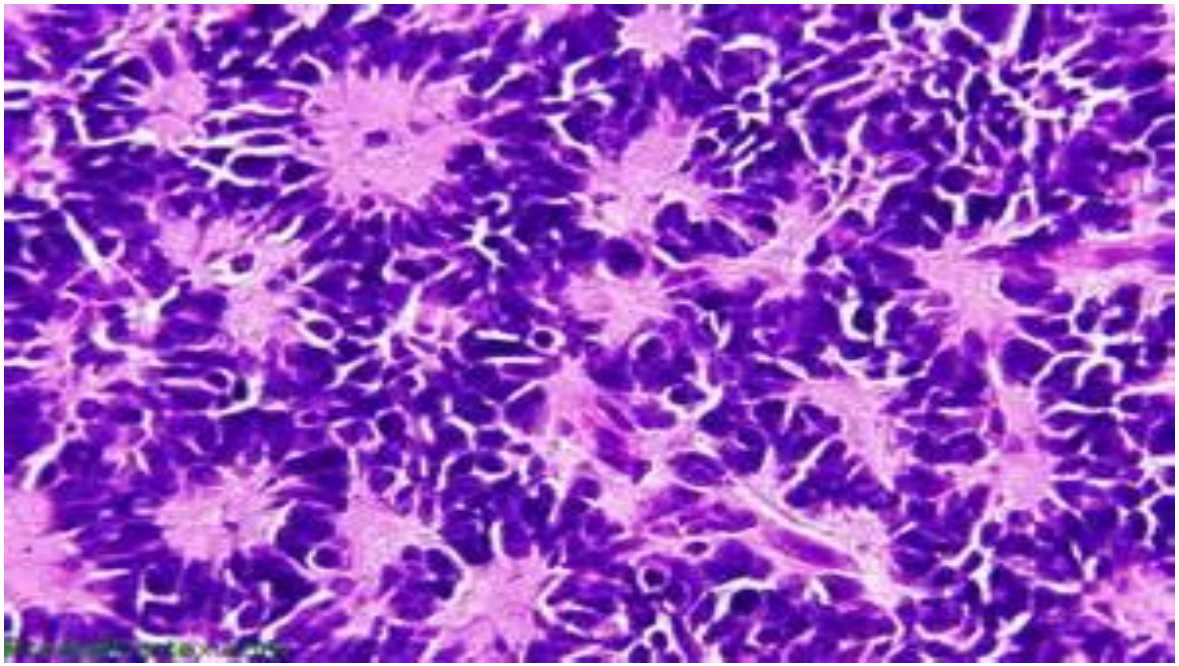
Microscopic appearance

- Highly cellular
- Sheets of small blue cells (small, rounded hyperchromatic nuclei, scanty cytoplasm)
- Many mitoses
- Homer Wright Rosettes= primitive tumor cells surrounding central neuropil (pink material formed by neuronal processes)

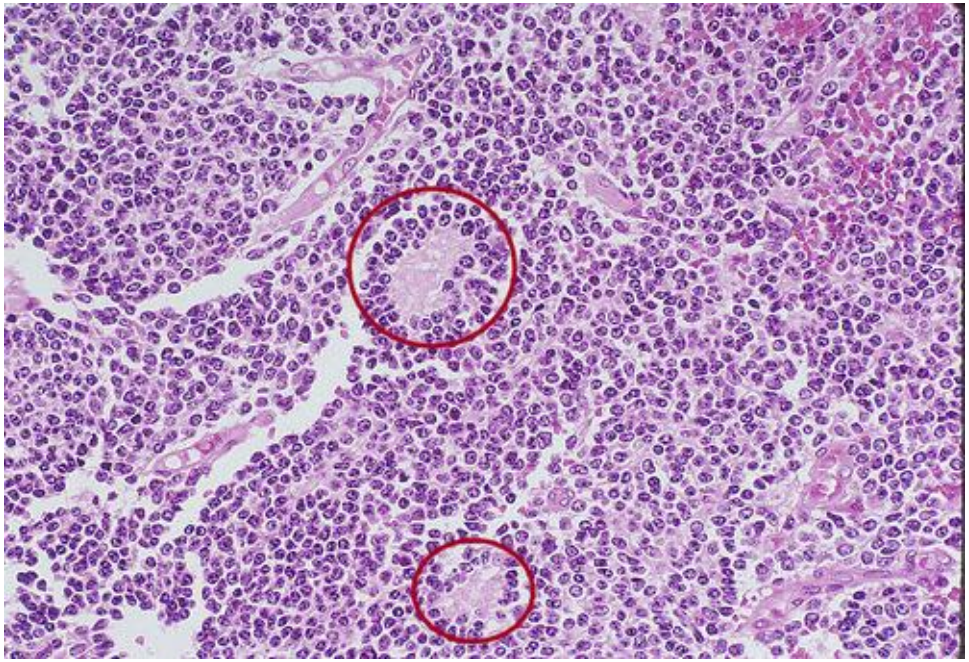
medulloblastoma



Homer Wright rosettes



Homer Wright Rosettes



genetics

- MYC amplification: poor prognosis
- WNT signalling pathway mutations: better prognosis
- These can help in developing new therapies… because it is better to avoid radiotherapy in young patients

Primary CNS lymphoma

- Mostly: diffuse large B cell lymphoma.
- 1% of intracranial tumors.
- Primary CNS lymphoma is the most common CNS neoplasm in the immunocompromised… in this situation they are almost always positive for EBV ((Epstein – Barr virus)

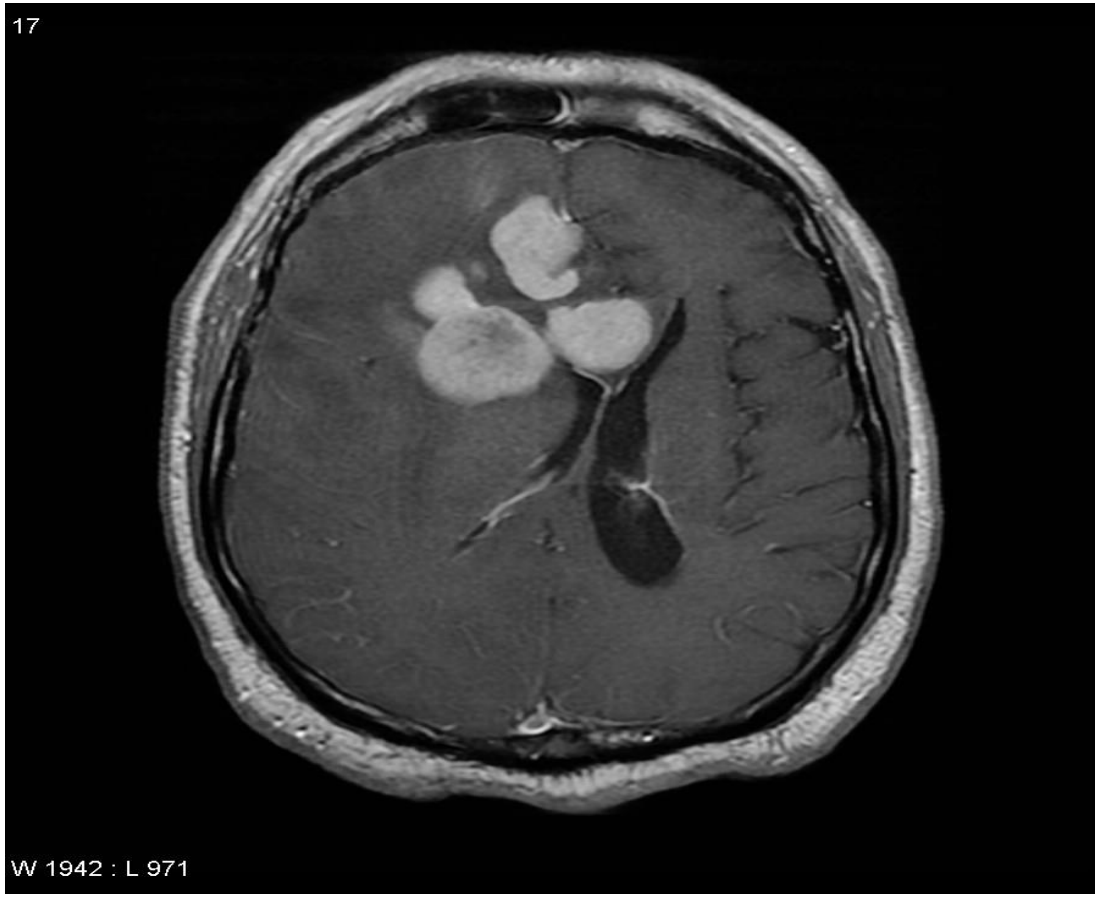
Primary CNS lymphoma

- Aggressive disease with poor prognosis
- **Poor response to chemotherapy as compared to peripheral lymphomas**
- Usually multiple nodules within the brain parenchyma
- Spreading outside the brain happens rarely and at late stages
- Peripheral lymphoma rarely spreads to the brain, if it does there is usually associated meningeal and CNS involvement.

morphology

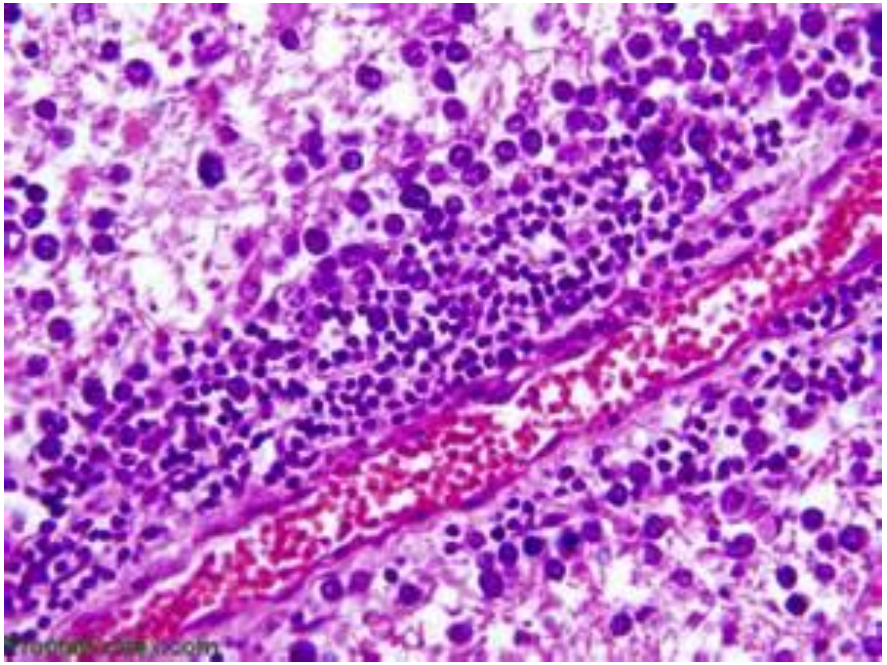
- Involves deep grey matter, white matter, cortex
- Periventricular spread is common
- Tumor nodules more defined than gliomas but less than metastases
- EBV positive tumors usually have extensive areas of necrosis
- Majority: diffuse large B cell lymphomas

Primary CNS lymphoma



Primary CNS lymphoma

- Tumor cells accumulate around blood vesseles



Germ cell tumors

- 0.2-1% of brain tumrs
- Higher incidence in Japan.. 10%
- Occur along the midline, mainly: pineal and suprasellar regions
- 90% occur in the first two decades of life
- Most common type: germinoma, resembles testicular seminoma
- **Secondary CNS involvement by metastatic germ cell tumors also occurs**

meningioma

- Arise from arachnoid meningothelial cells.
- Arise in adults
- Attached to the dura
- Can be seen at external surfaces of the brain or within the ventricular system

meningioma

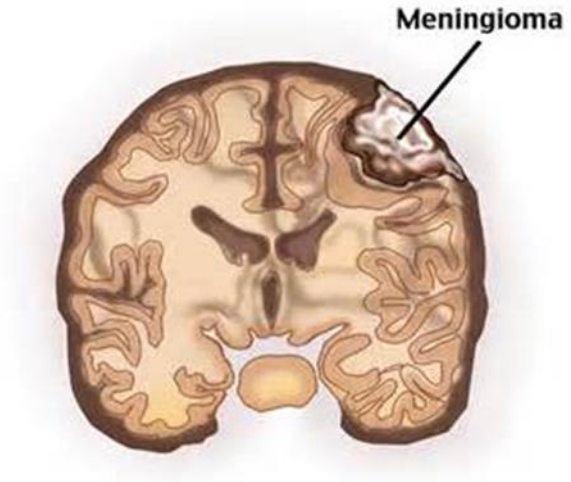
- Majority: can be easily separated from brain, but some are infiltrative
- Behaviour: benign but infiltrative lesions recur
- Outcome depends on: size, location, histological grade

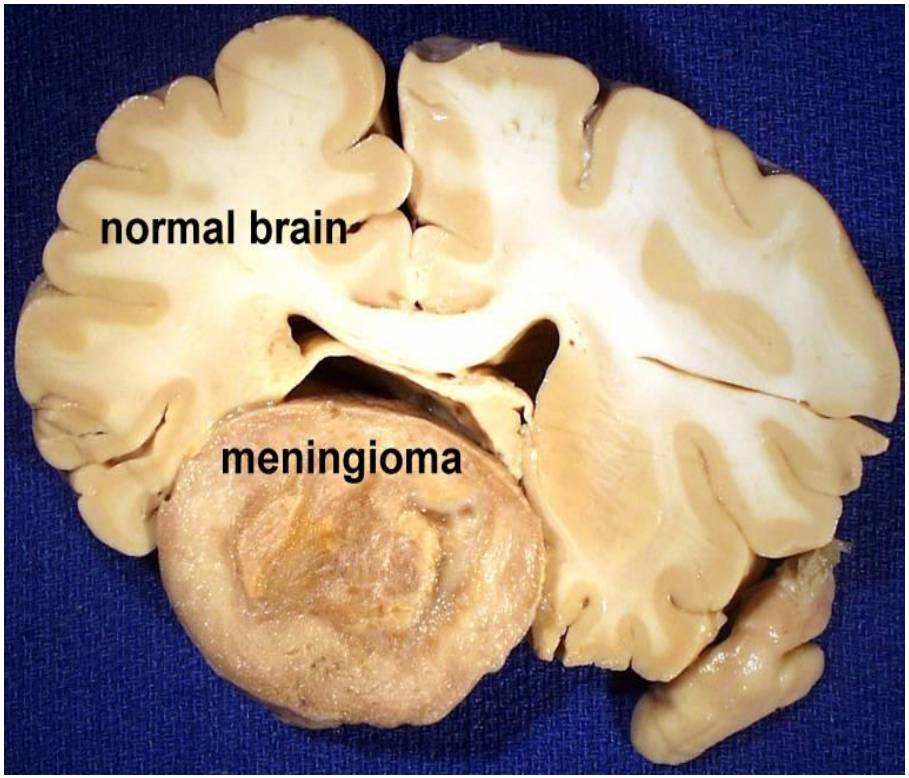
Histological grades

- WHO I: (well diff) meningioma
- WHO II: atypical meningioma
- WHO III: anaplastic (malignant) meningioma

Grade 1 meningiomas

- Well defined, dura based masses
- May compress but do not invade brain
- Can extend to overlying bone

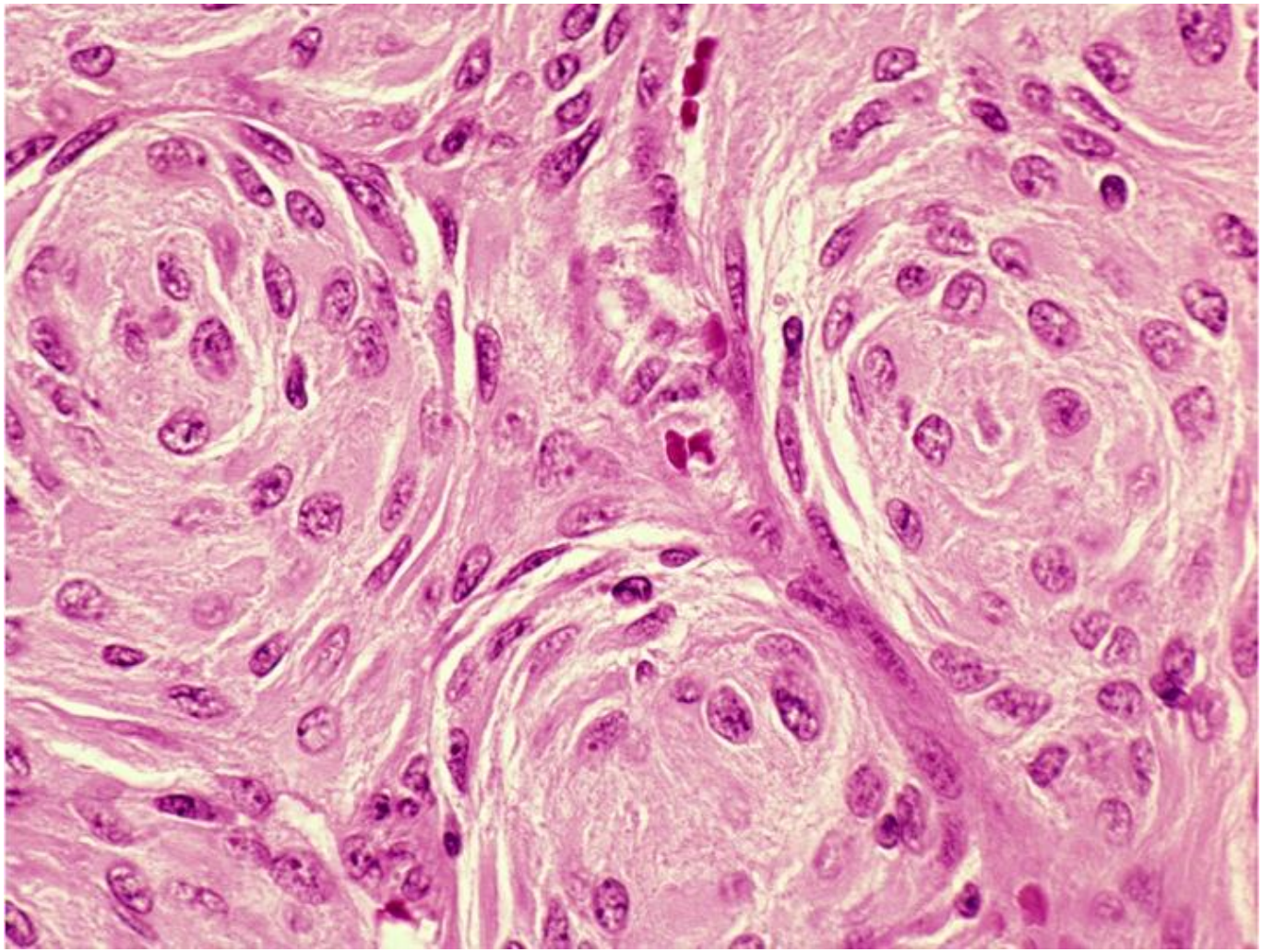



normal brain
meningioma

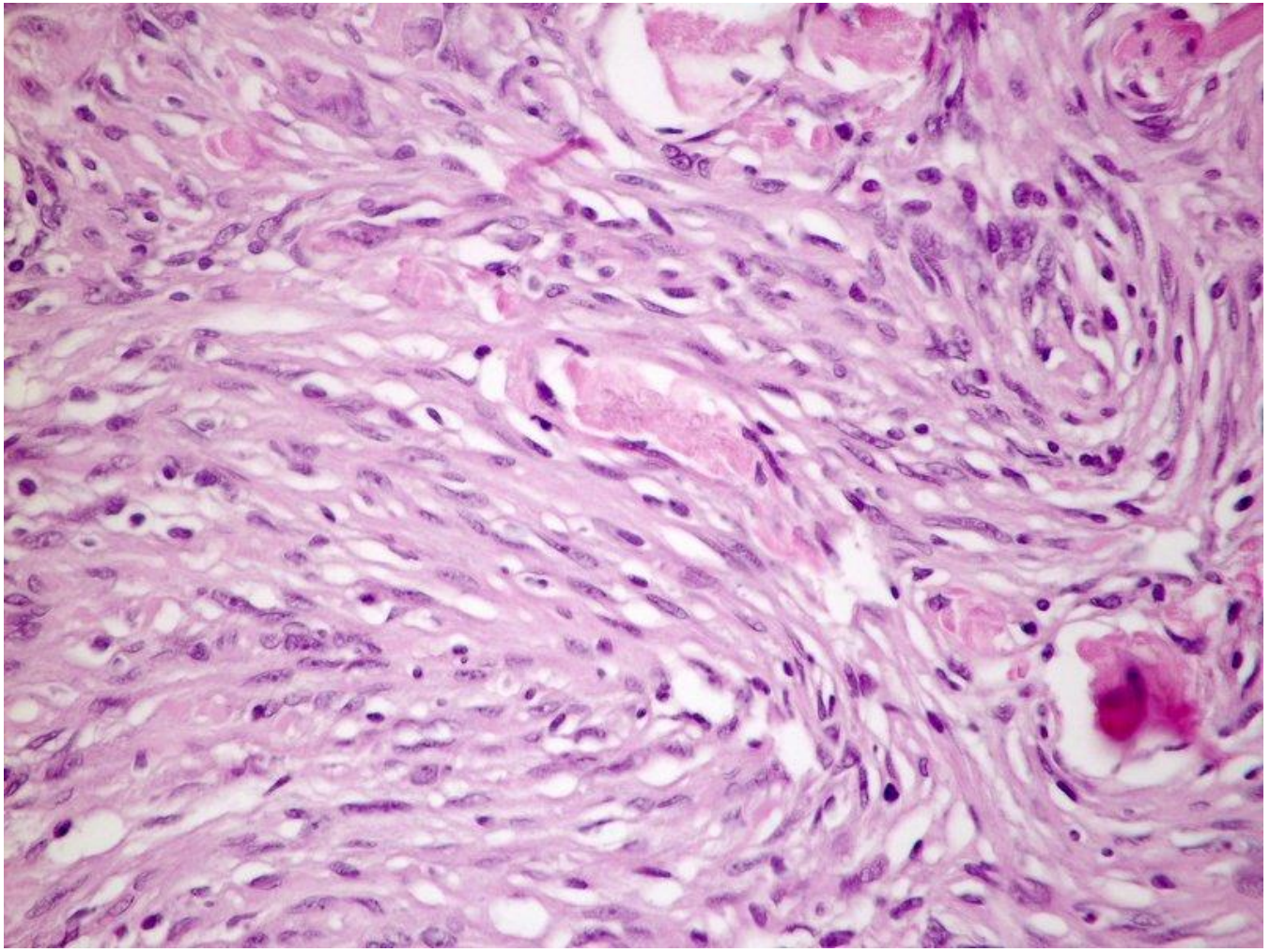
Grade 1 meningiomas/ histological types

- Syncytial: whorled clusters without visible cell membranes.
- Fibroblastic: elongated cells and abundant collagen
- Transitional: features of both, syncytial and fibroblastic
- Psammomatous: numerous psammoma bodies

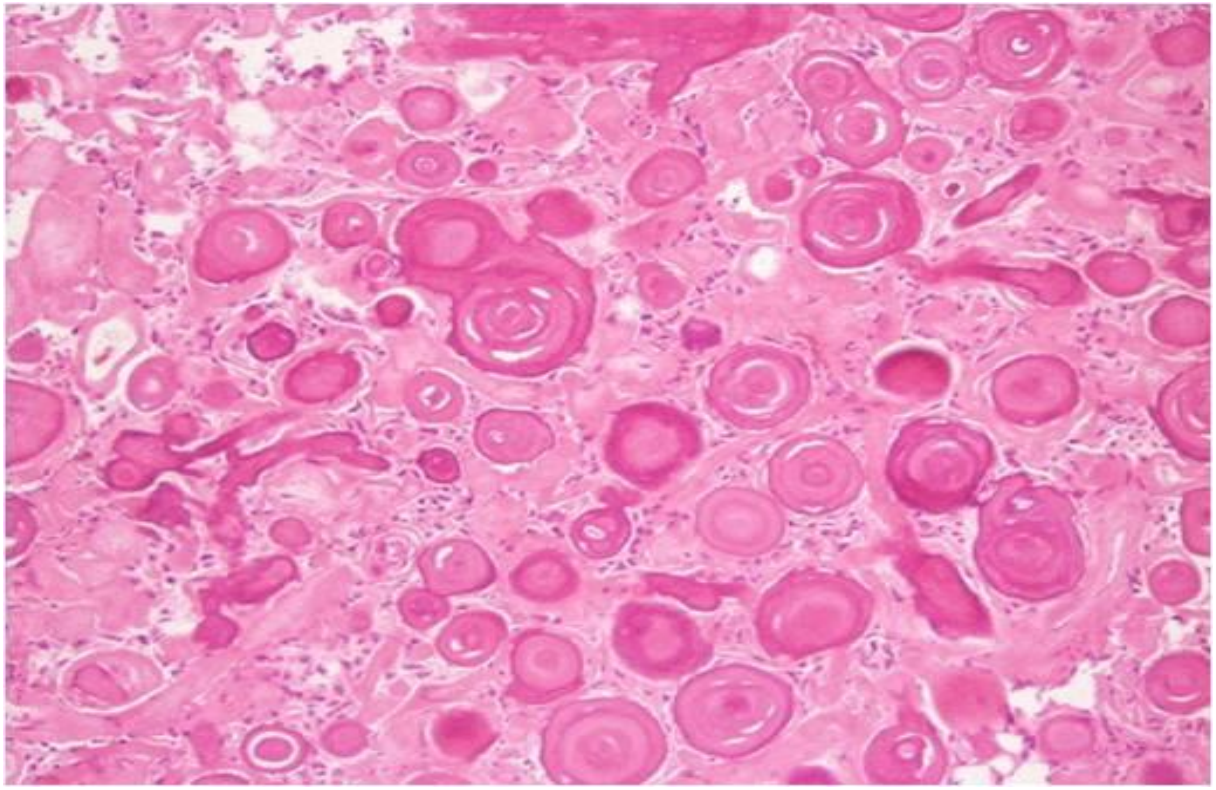
syncytial



fibroblastic



psammomatous

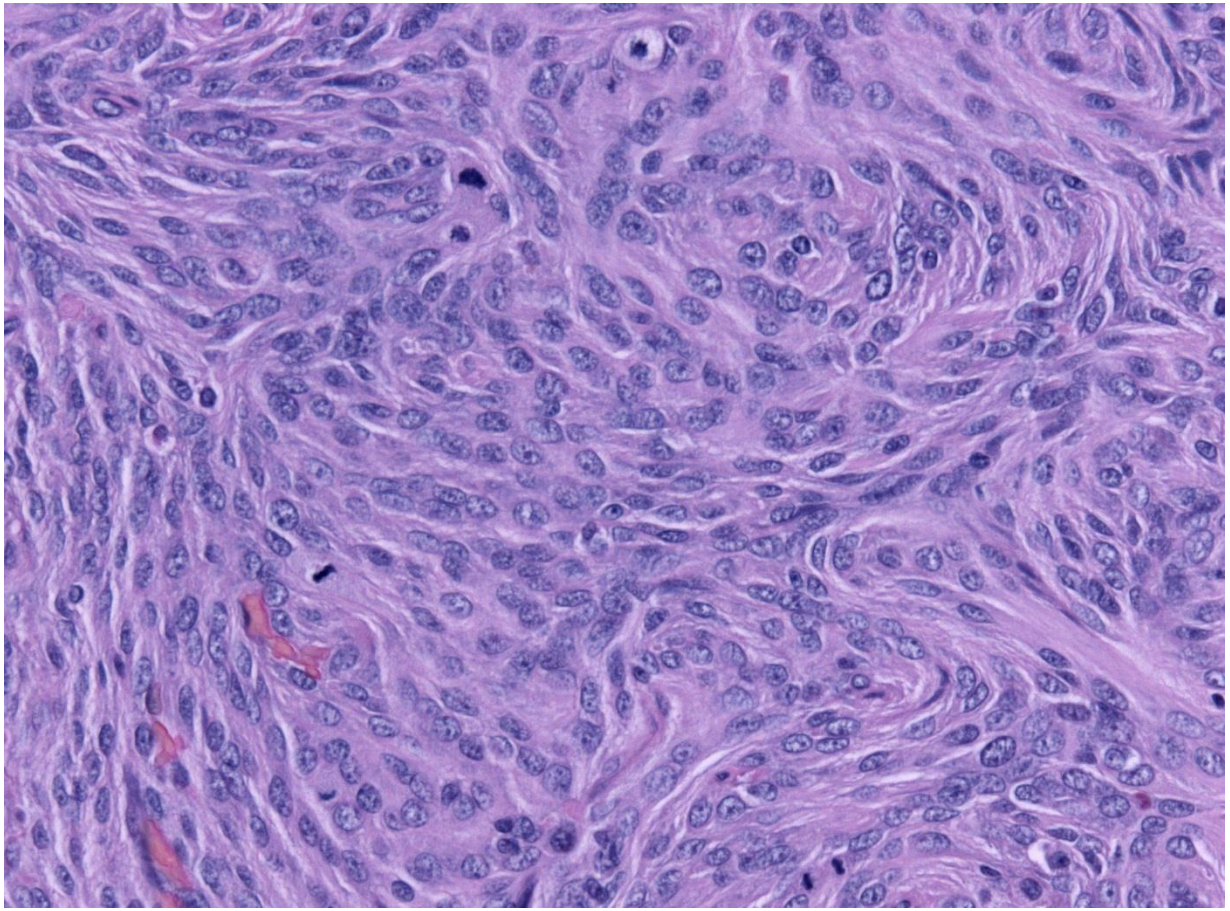


Atypical meningioma WHO grade 2

- High cellularity
- Prominent nuclei
- High mitotic rate

- More aggressive than grade 1
- Recur

Atypical meningioma
note the cellularity and mitoses



Anaplastic meningioma
WHO grade 3

- Highly aggressive
- Resemble sarcomas

Metastatic tumors

- ¼ to ½ of intracranial tumors
- Most common primary sites: lung, breast, melanoma, kidney and GIT.
- Form discrete well defined masses, can be multiple

Paraneoplastic syndromes

- CNS and peripheral nerves can be affected in disseminated cancer as part of the paraneoplastic syndromes
- These include several manifestations including dementia, ataxia, sensory neuropathy and psychosis

Familial tumor syndromes

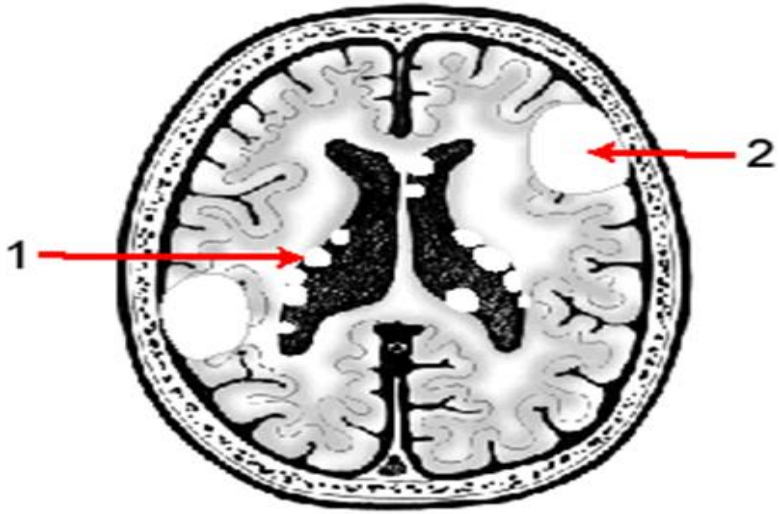
- Inherited syndromes
- Mutations in several tumor supressor genes
- Associated with increased risk of certain types of cancer
- 2 syndromes with CNS involvement: Tuberous sclerosis and von Hippel - Lindou

Tuberous sclerosis

- Autosomal dominant
- Hamartmas and benign neoplasms in brain and other sites
- CNS tumors: cortical tubers and subependymal hamartomas

Tuberous sclerosis

- Cortical tubers: look like potatoes!!
- Tuber: thickened underground part of a stem



Cortical tubers

- Hamartomas composed of haphazardly arranged large neurones.
- Mixture of glial and neuronal cells
- Cause seizures

Subependymal tubers

- Similar to cortical tubers
- Can cause hydrocephalus

Tuberous sclerosis/Extracerebral lesions:

- renal angiomyolipoma,
- retinal glial hamartomas,
- pulmonary lymphangiomatosis
- cardiac rhabdomyoma
- cysts in liver, kidney , pancreas.
- skin lesions: angiofibroma, hypopigmented areas, thichened patches.

Von Hippel Lindau

- Autosomal dominant
- Mutation in VHL tumor supressor gene.
- Hemangioblastomas mainly in cerebellar hemispheres, retina.
- Cysts in pancreas, liver kidney
- Increase risk of renal cell carcinoma.