

CNS pathology  
Third year medical students

Dr Heyam Awad

Lecture 12: CNS tumours 3/3

# meningioma

- Arise from arachnoid meningotheial 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

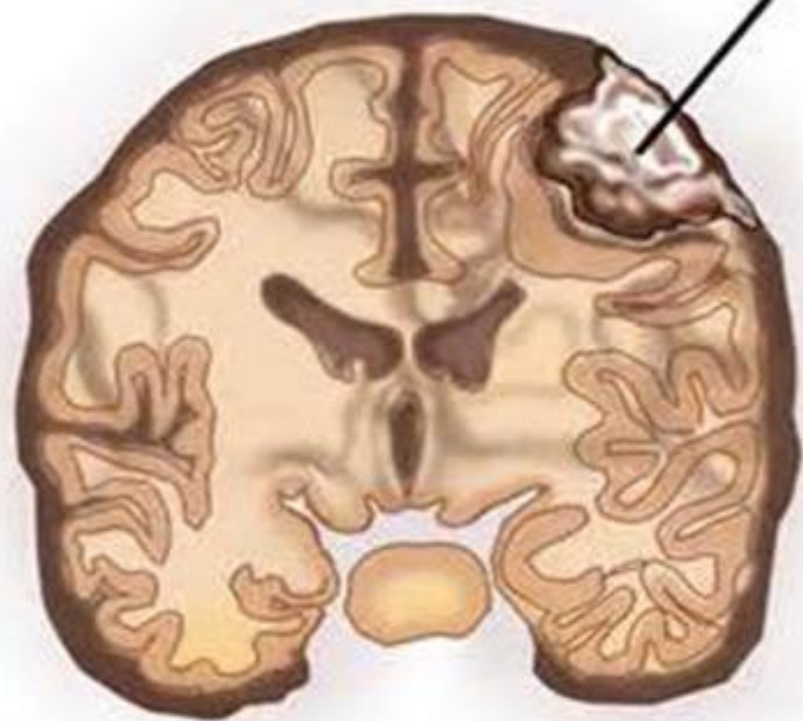
# Histological grades

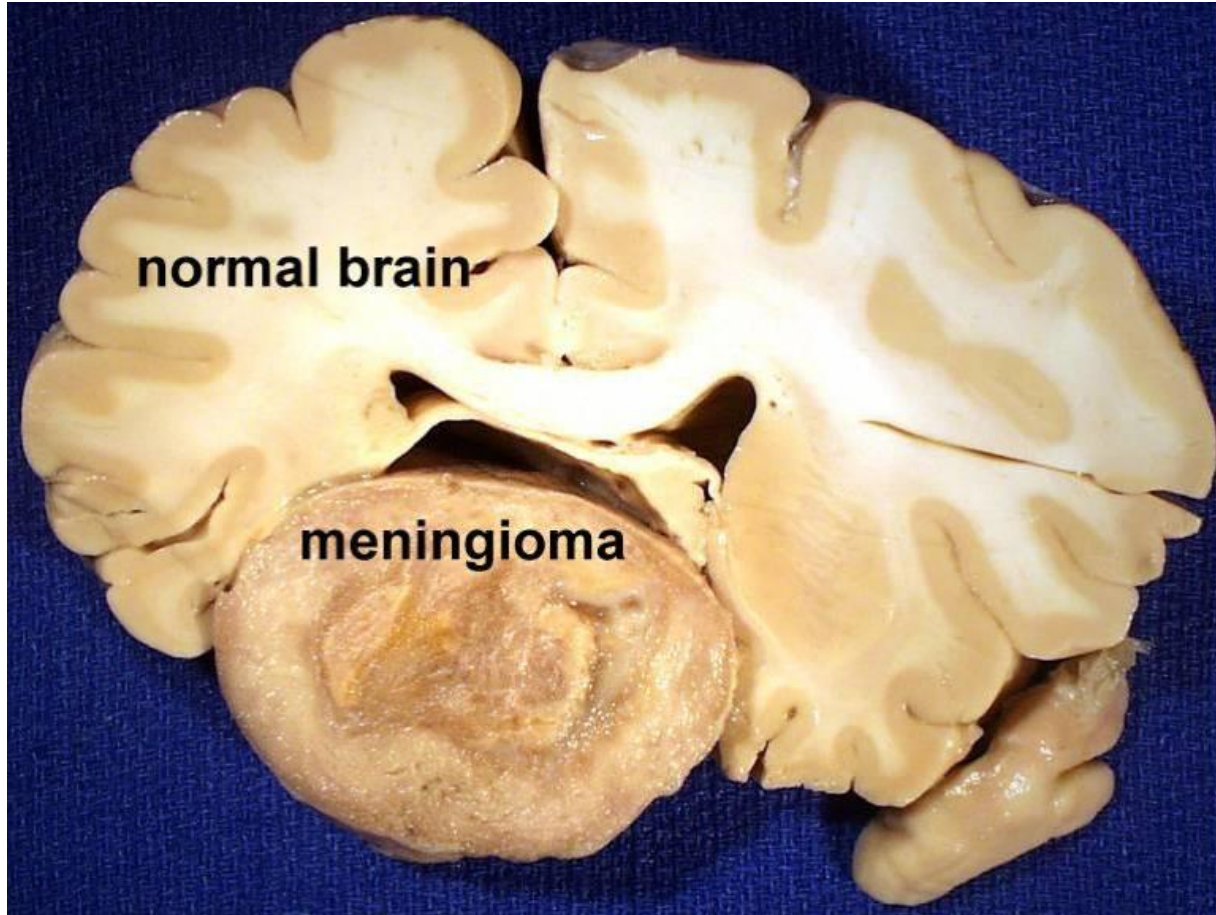
- WHO I: (well diff) meningioma
- WHO II: atypical meningioma
- WHO III: anaplastic ( malignant ) meningioma

# Grade 1 meningiomas

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

**Meningioma**





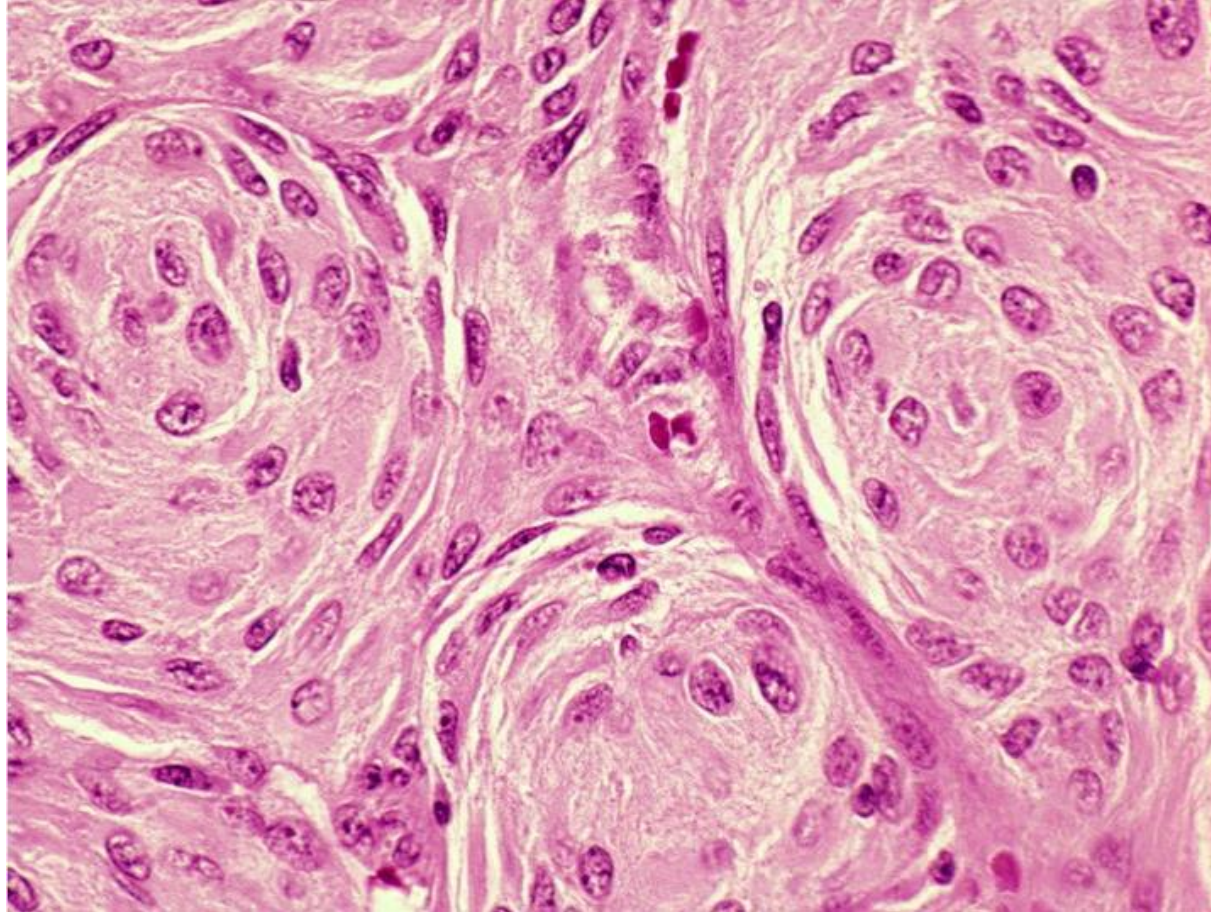
**normal brain**

**meningioma**

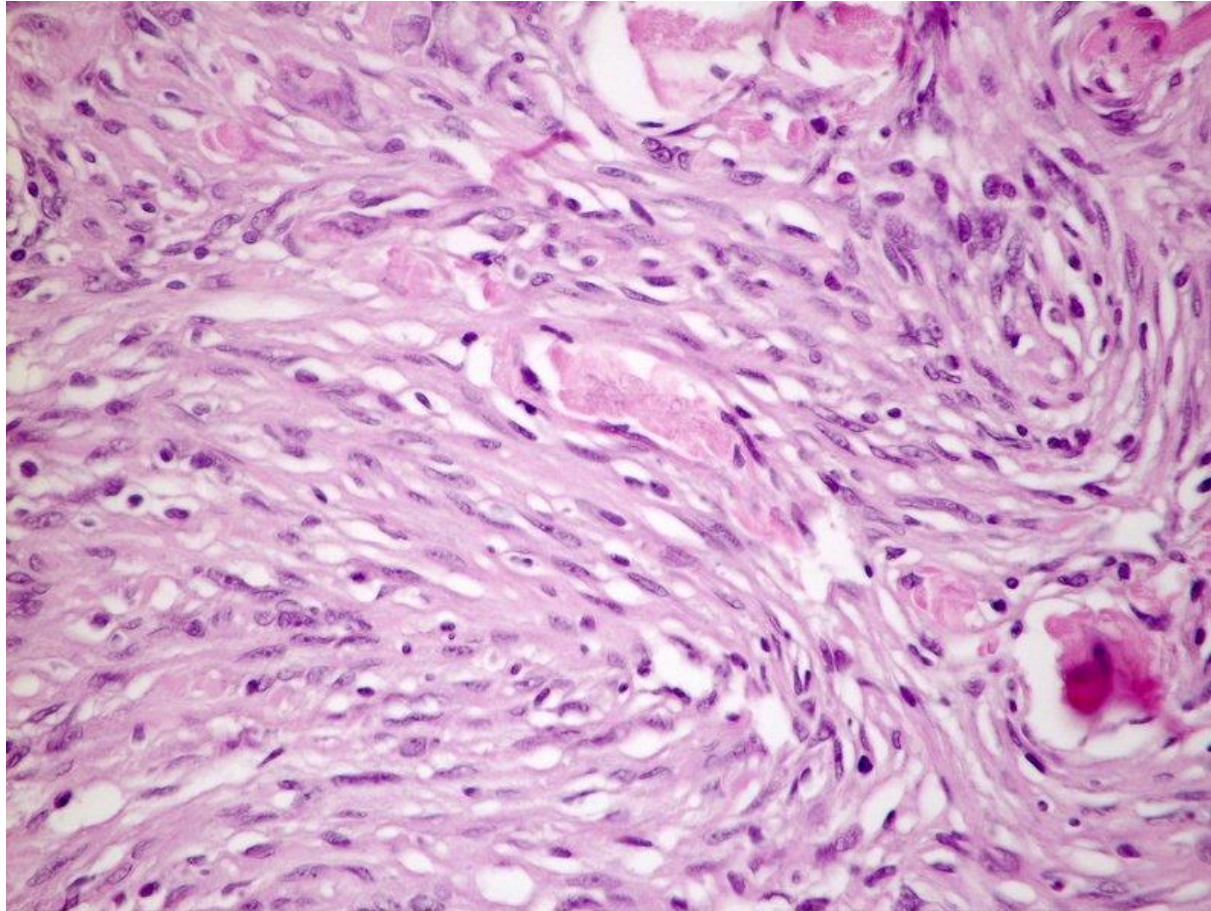
- Grade 1 meningiomas/ histological types
- •Syncytial: whorled clusters without visible cell membranes.
- •Fibroblastic: elongated cells and abundant collagen
- •Transitional: features of both, syncytial and fibroblastic
- •Psammomatous: numerous psammoma bodies



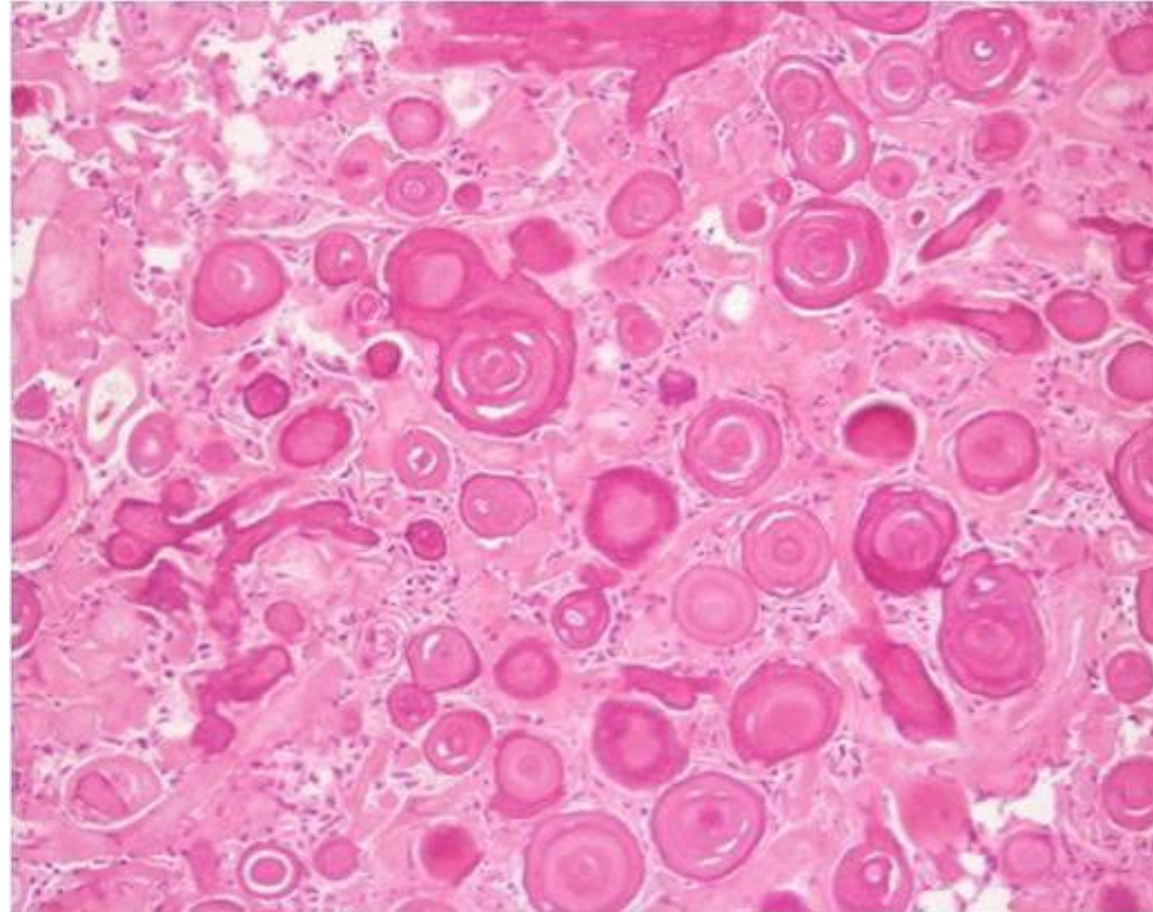
syncytial



fibroblastic



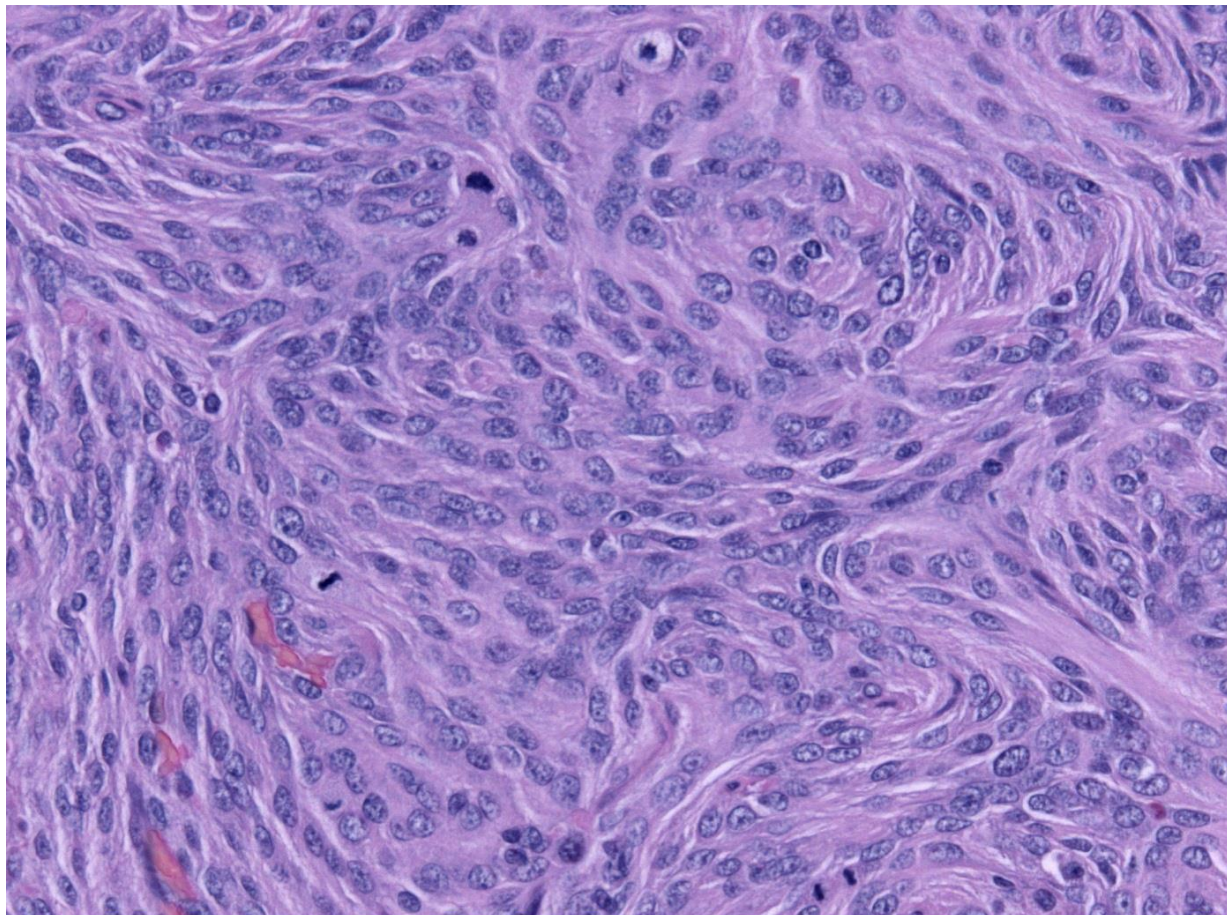
pasammomatous



# Atypical meningioma WHO grade 2

- High cellularity
- Prominent nuclei
- High mitotic rate
- More aggressive than grade 1
- Recur

# Atypical meningioma



# Anaplastic meningioma

- Anaplastic meningioma WHO grade 3
- Highly aggressive
- Resemble sarcomas

# 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)

# 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.



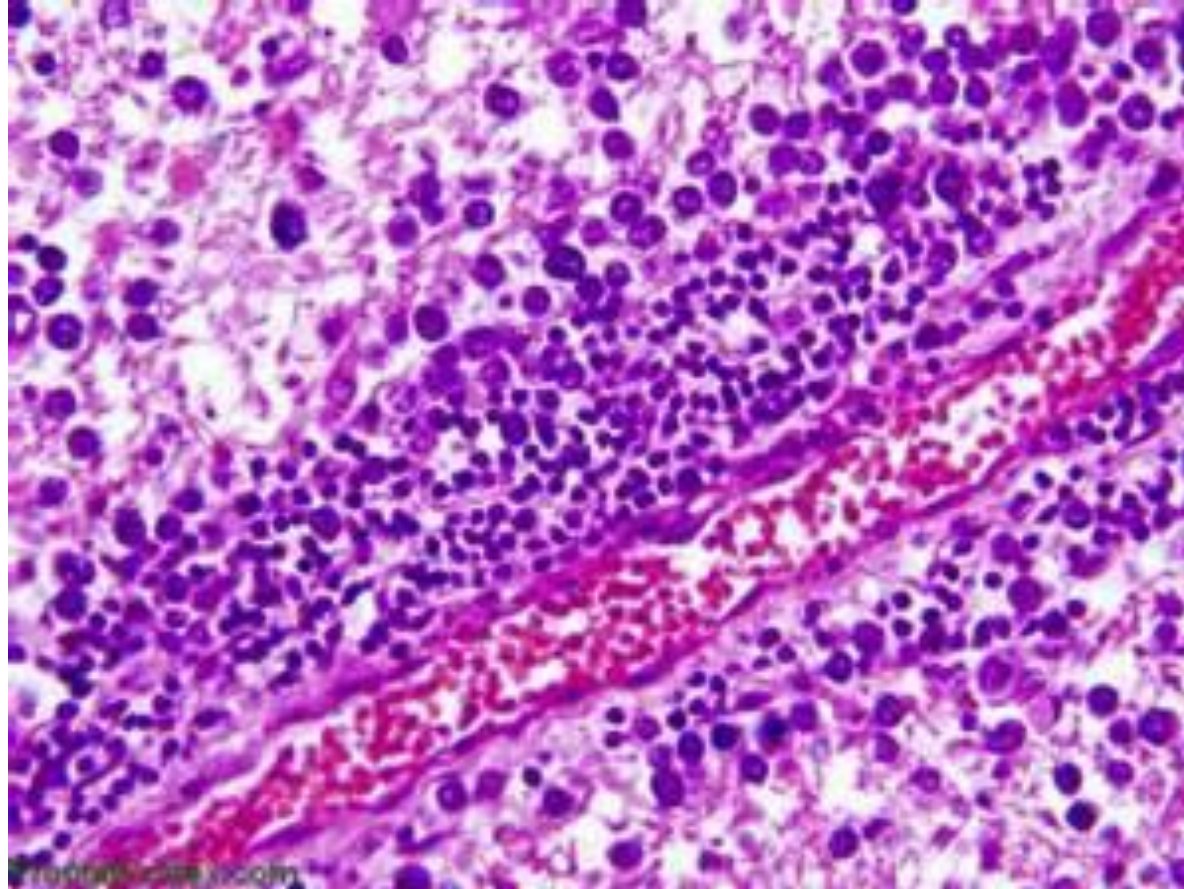
# lymphoma

- 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

lymphoma: note the multiplicity of the lesions



lymphoma



# Metastatic tumors

-  $\frac{1}{4}$  to  $\frac{1}{2}$  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 suppressor 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
- Hamartomas and benign neoplasms in brain and other sites
- CNS tumors: cortical tubers and subependymal hamartomas

- Tuberosclerosis
- 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

## TUBEROUS SCLEROSIS COMPLEX. Tubers

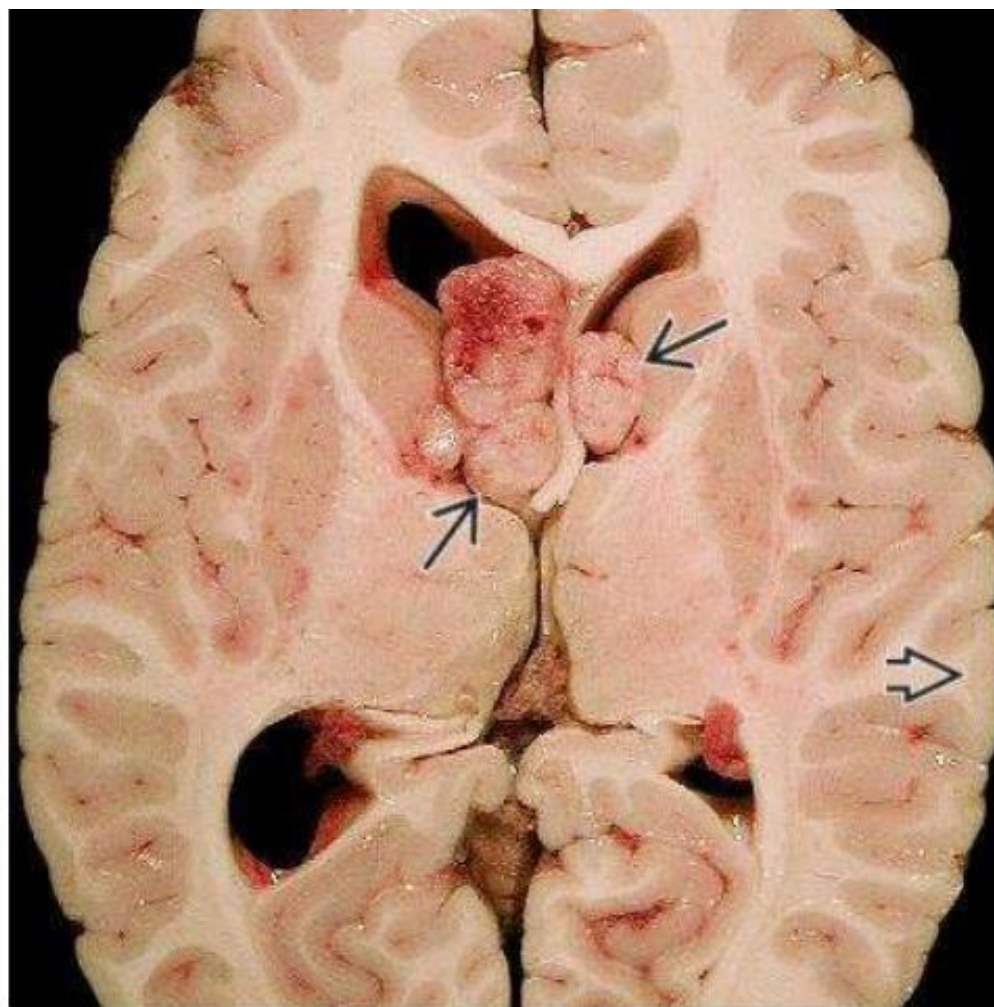


▲ Subcortical tuber (*arrow*) in left parietal lobe demonstrated by sagittal T1-weighted image as a hypointense and well-defined area.

Although MRI is the best imaging modality for TSC study, subtle bilateral cortical and subcortical tubers (*arrows*) can also be seen in axial computed tomography scan. ▼



Tuberous sclerosis.. potatoes in the brain!



- Subependymal tubers :Similar to cortical tubers
- Can cause hydrocephalus

- Tuberosous 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 syndrome

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

	Most common age	most common location	main histologic criteria	Genetic mutations	Prognosis	Notes
<b>Pilocytic astrocytoma</b>	Children	Cerebellum	Micro cysts Rosenthal fibers low cellularity	BRAF	Excellent, grade I tumour	
<b>diffuse astrocytoma</b>	adults 40-60	Cerebral hemispheres	Astrocytic cells in fibrillary background GFAP positive	IDH 1 or 2 Mutation	Mean survival more than 5 years	to be called grade III: increased cellularity, mitosis and polymorphism.
<b>Glioblastoma</b>	Adults Primary: over 55 years Secondary : younger: 50	cerebral hemispheres	Necrosis, usually palisading OR vascular proliferation	IDH mutated have better prognosis than IDH wild type	mean survival 15 months	Enhancing lesions on radiology. Can be primary (90%) or secondary (10%)
<b>oligodendroglioma</b>	Younger than astro 40-50	white matter	Rounded nuclei surrounded by a clear halo (fried egg appearance)	IDH PLUS 1p 19 q cpdeletion.	grade II: 10-20 years survival grade III : 5-10 years	grade III: same histological criteria of grade III astro
<b>Ependymoma</b>	adults and children	Adults: spinal cord <20 years: near fourth ventricle	True rosettes around empty spaces ( canals) Pesudoreosettes around blood vessels		better if resectable Spinal tumors easily resectable, so better prognosis than posterior fossa ones	
<b>Medulloblastoma</b>	Children	Exclusively in the cerebellum	Primitive cells( small round blue cells) Homer Wright rosettes around neuropil	Myc : poor prognosis WNT: beter prognosis	grade IV tumors highly aggressive Can metastasise to bone	WNT mutation can be tested by B catenin stain.
<b>meningioma</b>	Middle age	Dura based lesions	Meningeal cells Psammoma bodies		generally good Depends on grade	

Thank  
you