CNS pathology
Third year medical students

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Lecture 13: CNS tumours 3/3

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

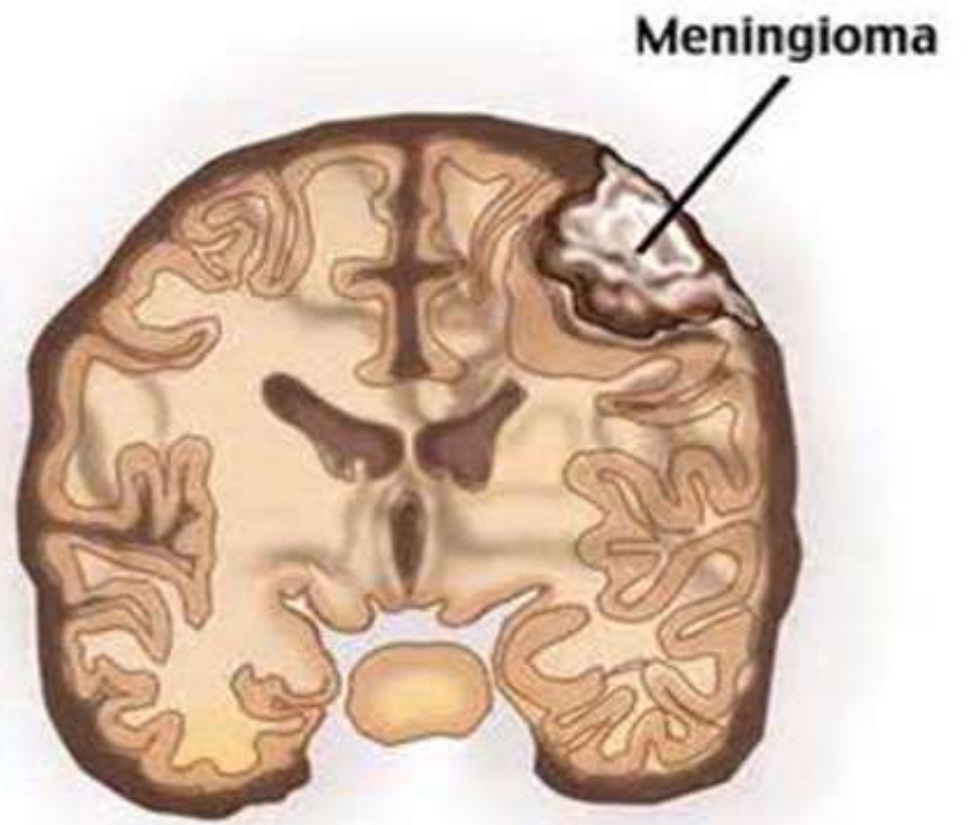
- Majority: can be easily separated from brain, but some are infiltrative
- Behaviour: benign but infiltrative lesions recur
- Outcome depends on: size, location, histological grade

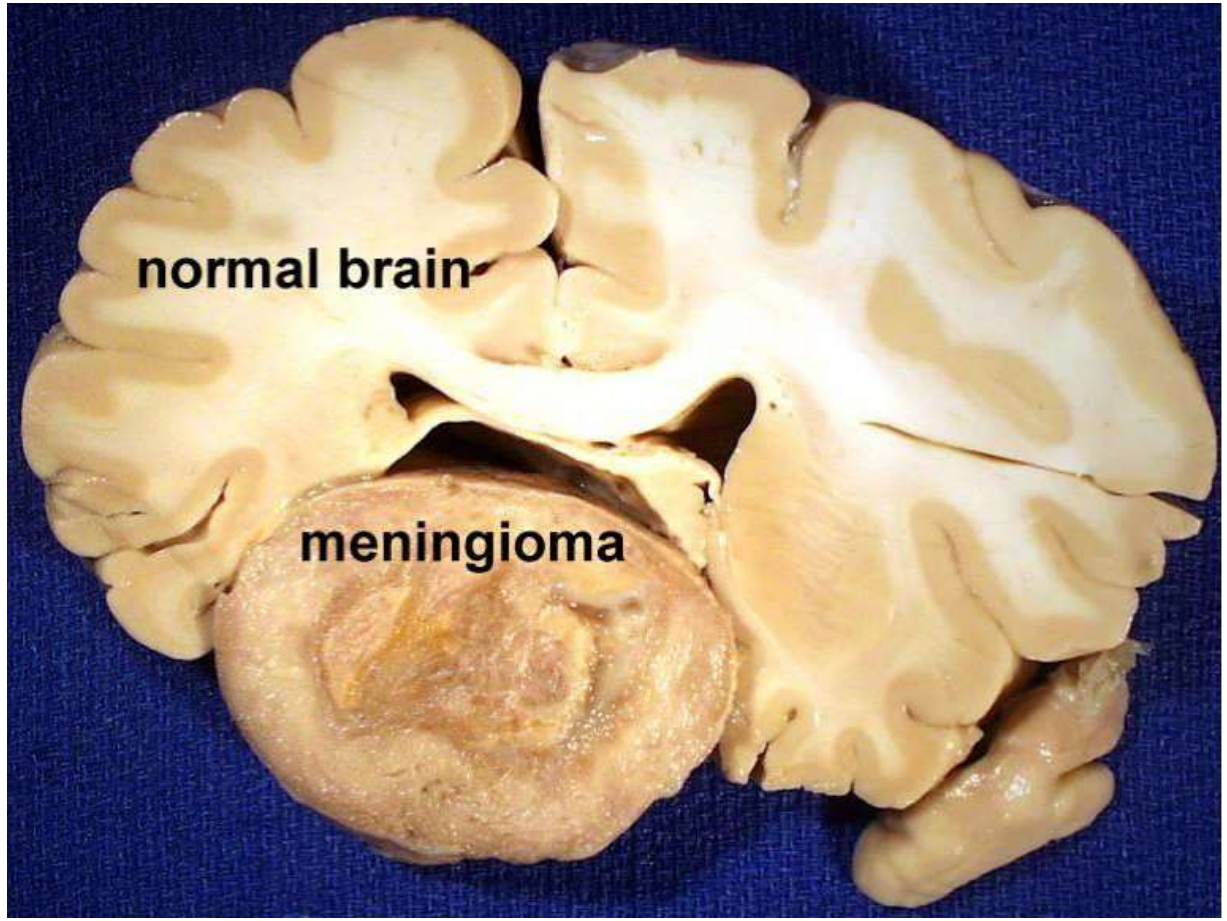
Histological grades

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

Grade 1 meningiomas

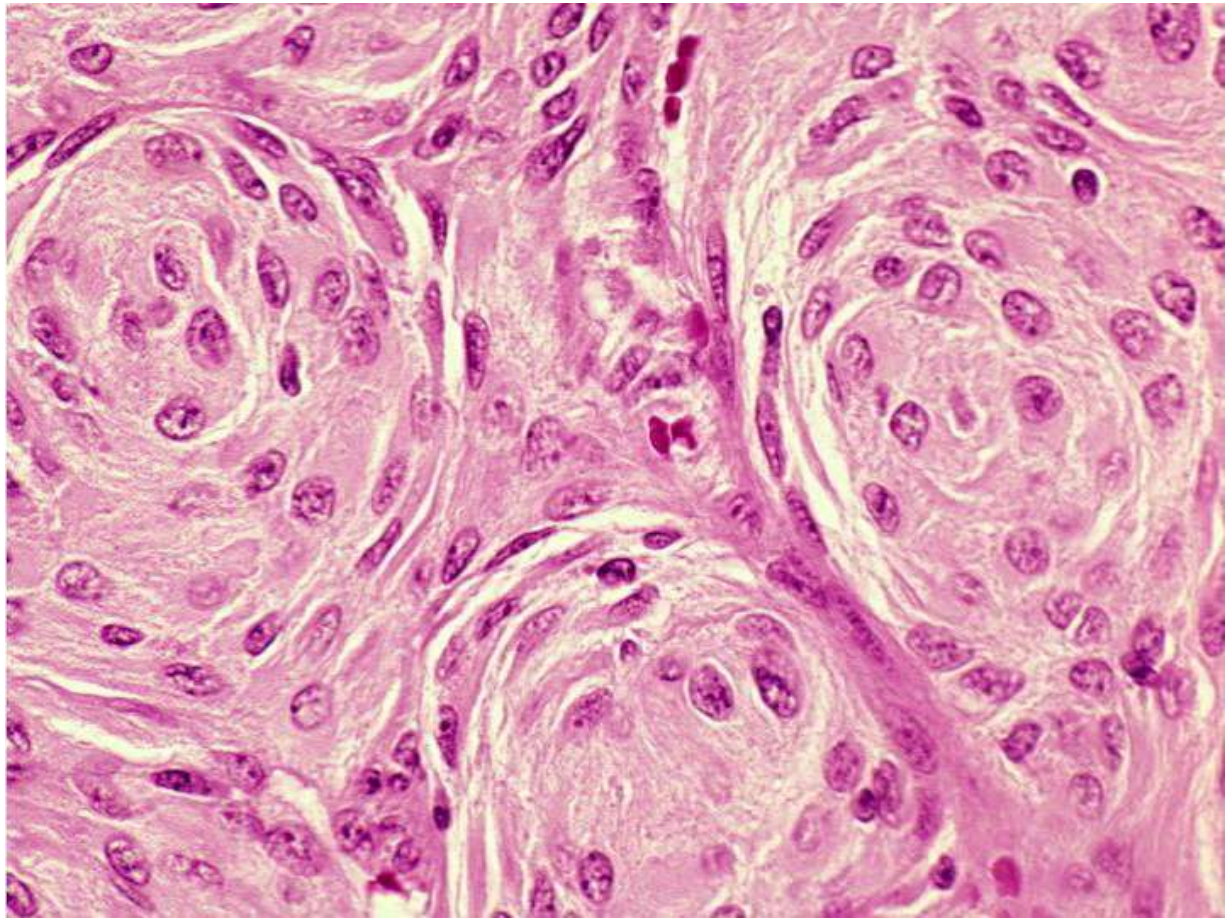
- Well defined, dura based masses
- May compress but do not invade brain
- Can extend to overlying bone


Meningioma

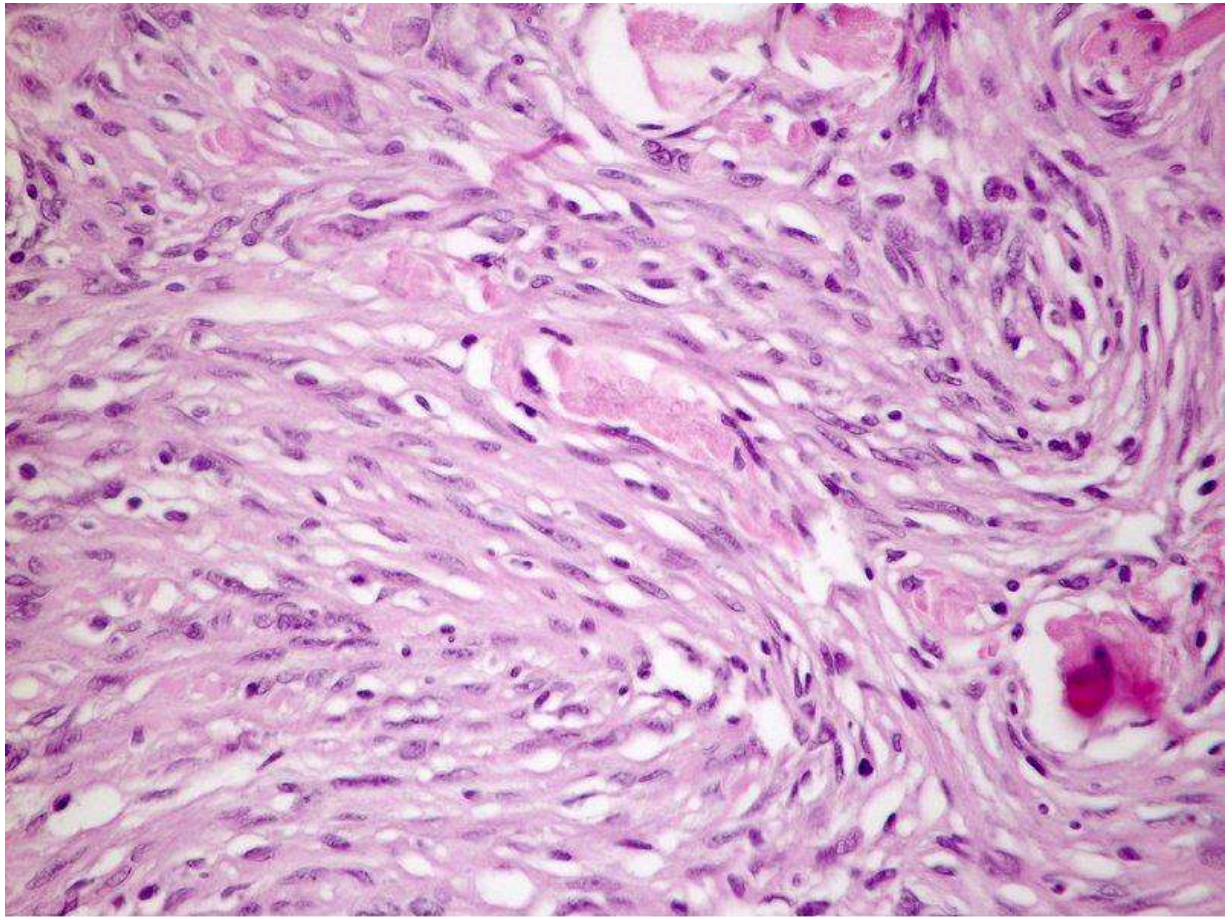

normal brain
meningioma

- 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

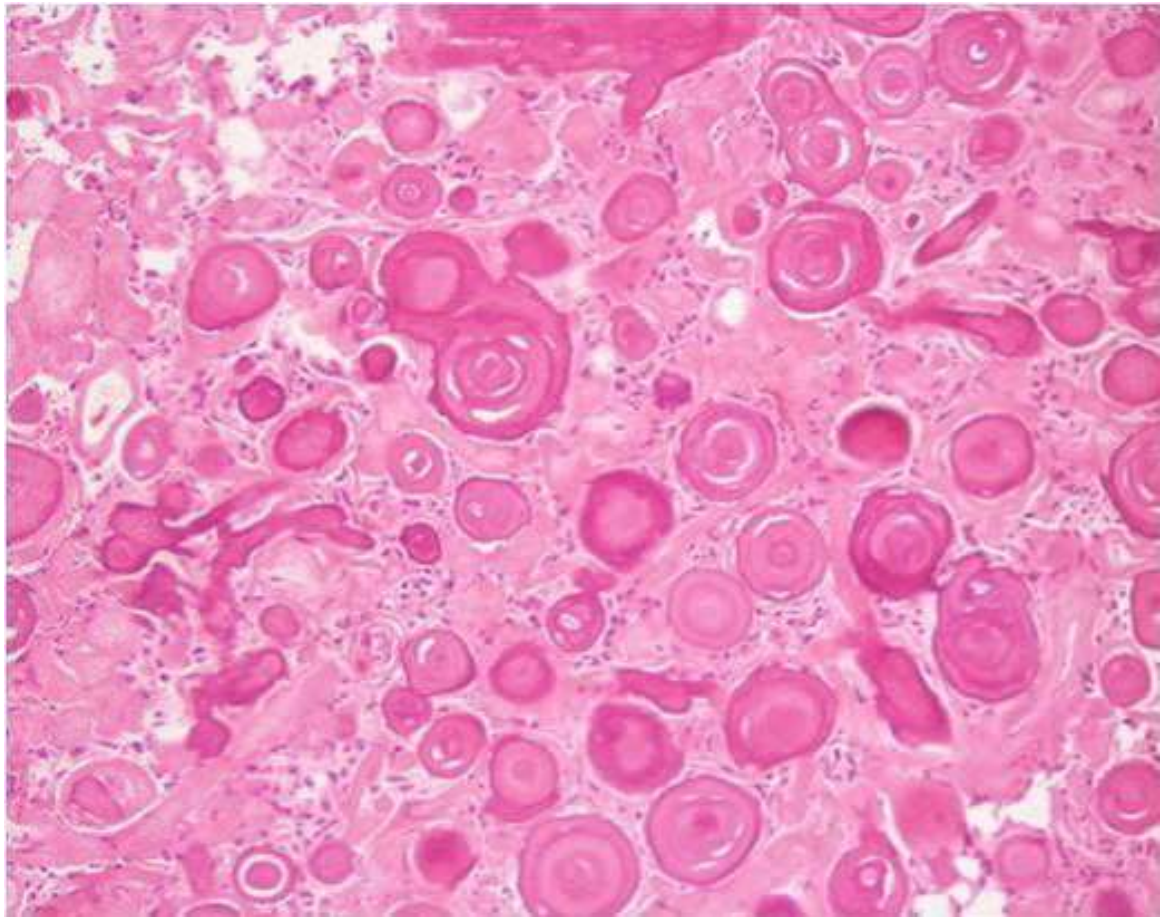
syncitial



fibroblastic



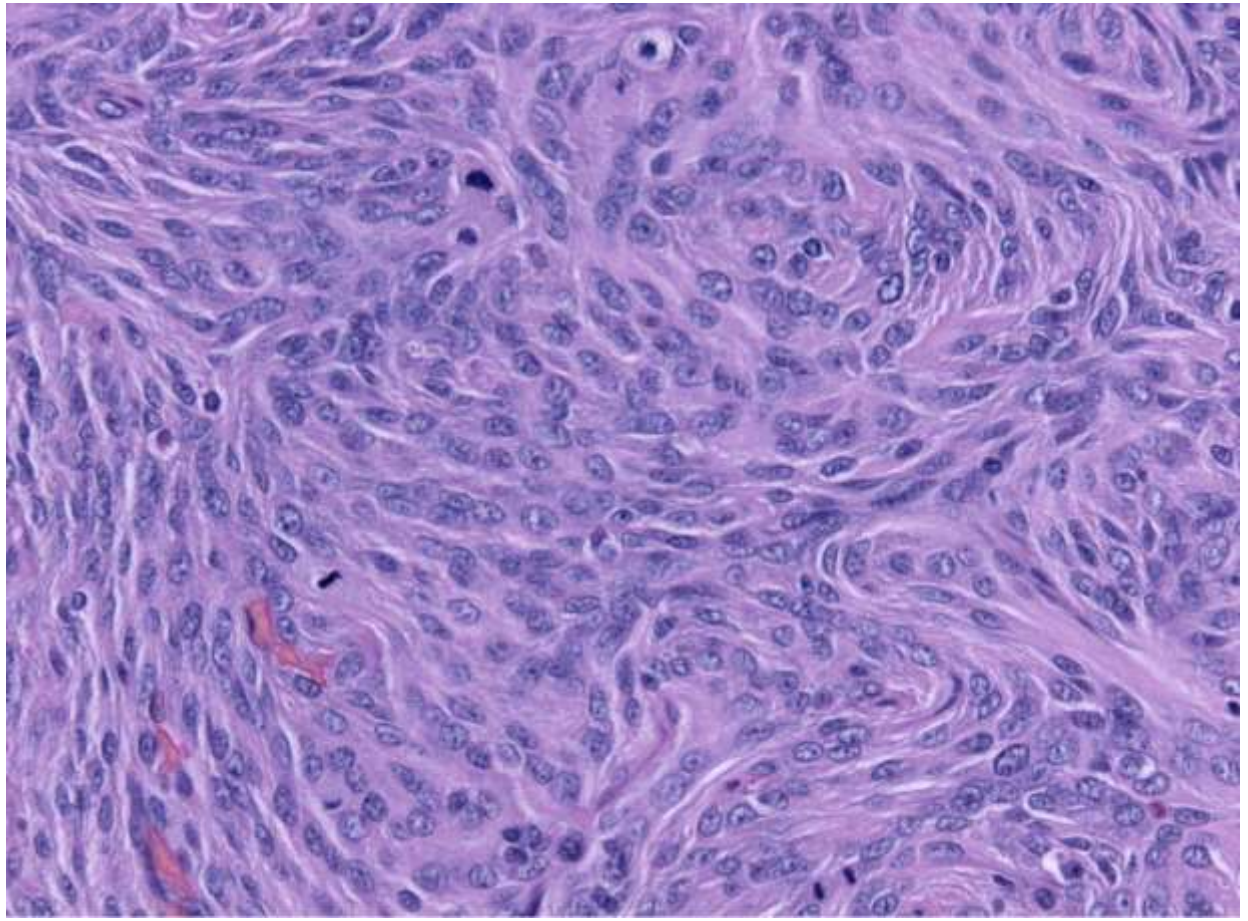
pasammomatous



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)

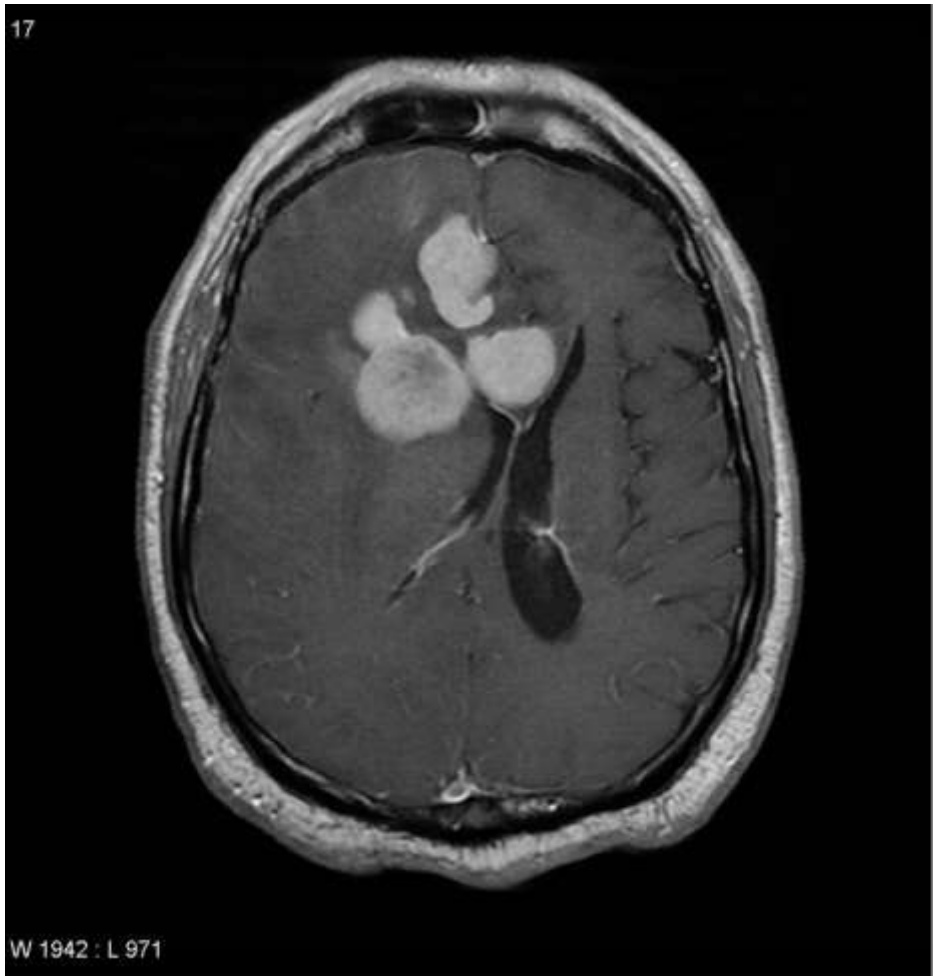
CNS lymphoma

- 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.

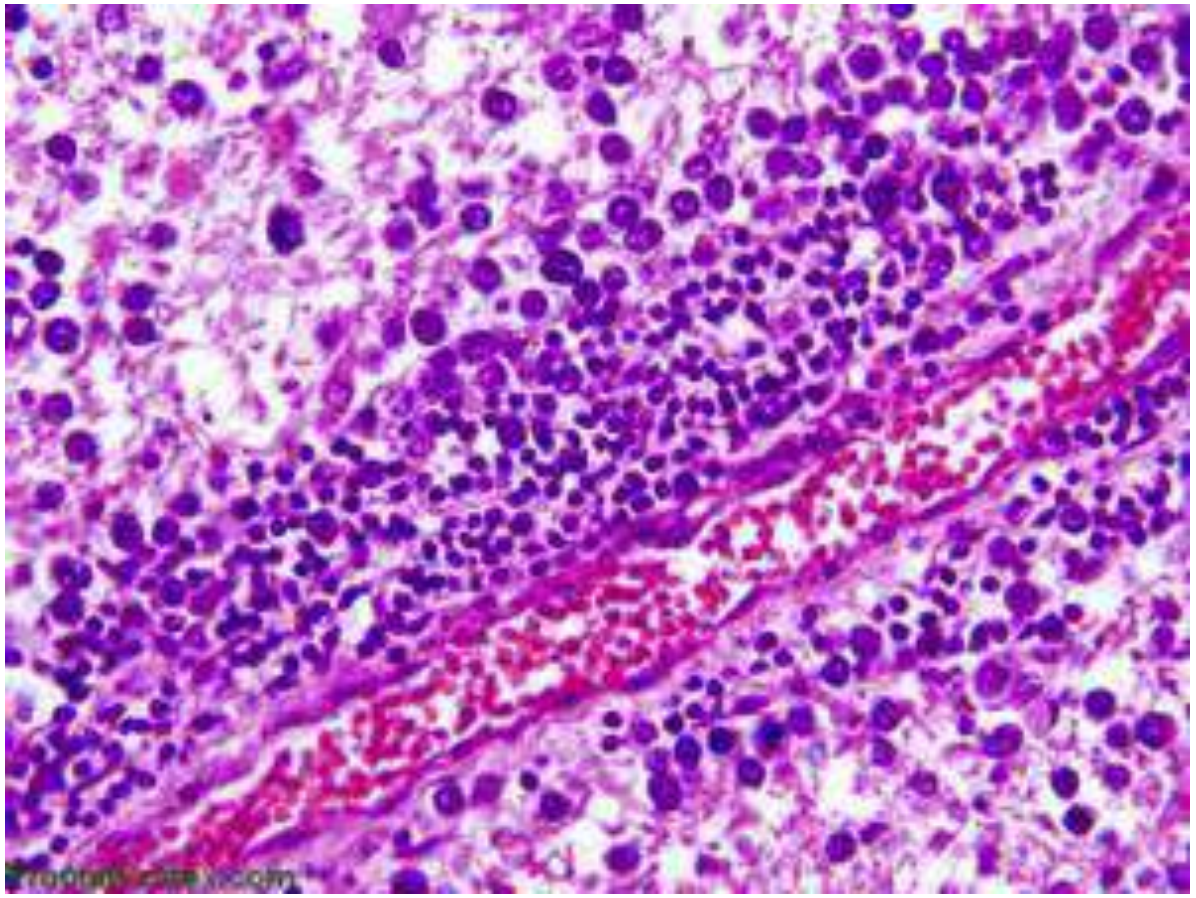
lymphoma

- 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

- 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 suppressor genes
- Associated with increased risk of certain types of cancer
- 2 syndromes with CNS involvement: Tuberous sclerosis and von Hippel - Lindou

•Tuberous sclerosis

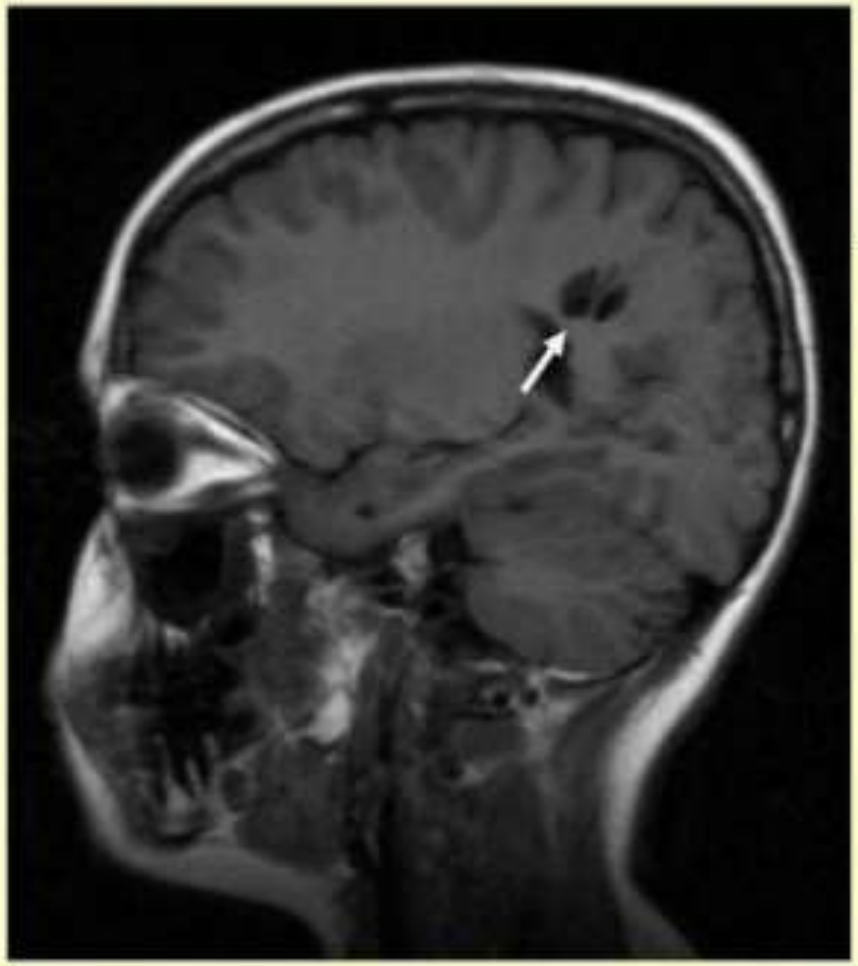
- Autosomal dominant
- Hamartmas and benign neoplasms in brain and other sites
- CNS tumors: cortical tubers and subependymal hamartomas

- Tuberous sclerosis

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

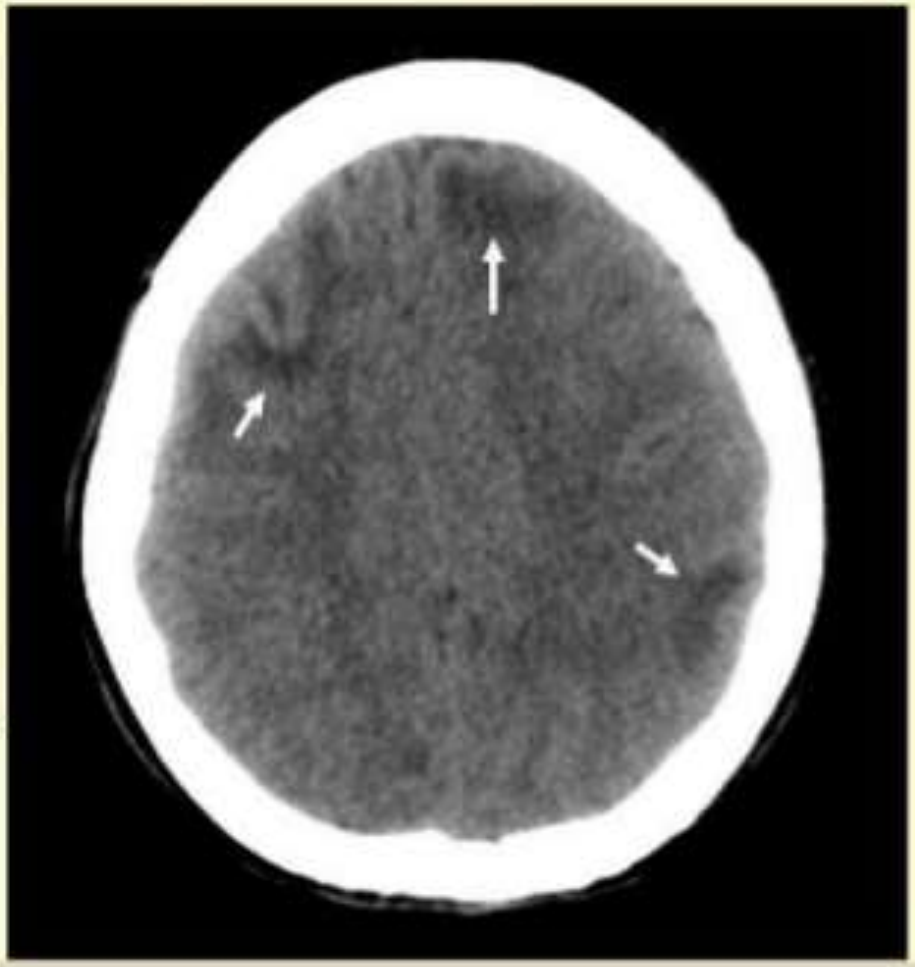
- Cortical tubers : Hamartomas composed of haphazardly arranged large neurones.
- Mixture of glial and neuronal cells
- Cause seizures

TUBEROUS SCLEROSIS COMPLEX. Tubers

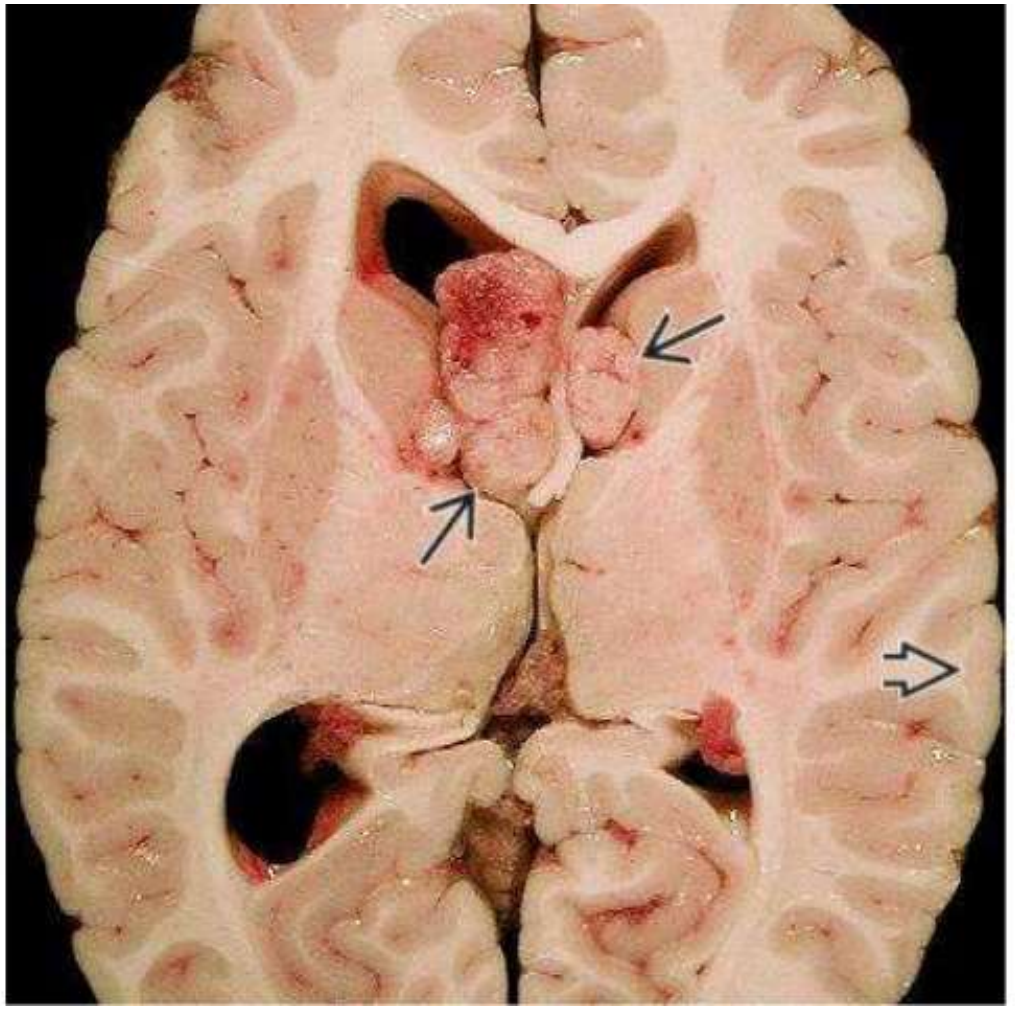


▲ Subcortical tuber *(arrow)* in left parietal lobe demonstrated by sagittal T1-weigthed image as a hypointense and well-defined area.

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

- 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 syndrome

- 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

	Most common age	most common location	main histologic criteria	Genetic mutations	Prognosis	Notes
Pilocytic astrocytoma	Children	Cerebellum	Micro cysts Rosenthal fibers low cellularity	BRAF	Excellent, grade I tumour	
diffuse astrocytoma	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.
Glioblastoma	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%)
oligodendroglioma	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
Ependymoma	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	
Medulloblastoma	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.
meningioma	Middle age	Dura based lesions	Meningeal cells Psammoma bodies		generally good Depends on grade	


thank
you