CNS pathology
Third year medical students
lecture 11 CNS tumors/ 3

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FRCPath

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Contents:
-Meningiomas
-CNS lymphomas
-Metastasis
-Paraneoplastic syndromes affecting the CNS
-Familial tumor syndromes

Meningioma. *A benign tumor*

- 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
- Behavior: benign but infiltrative lesions recur
- Outcome depends on: size, location, histological grade

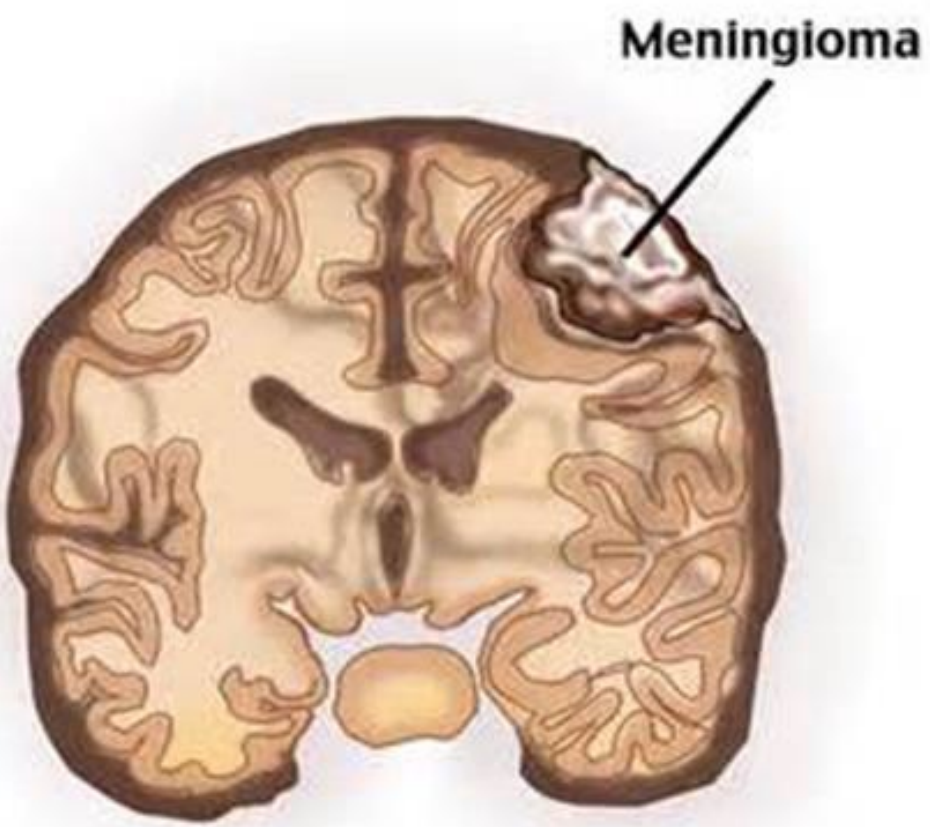
Histological grades

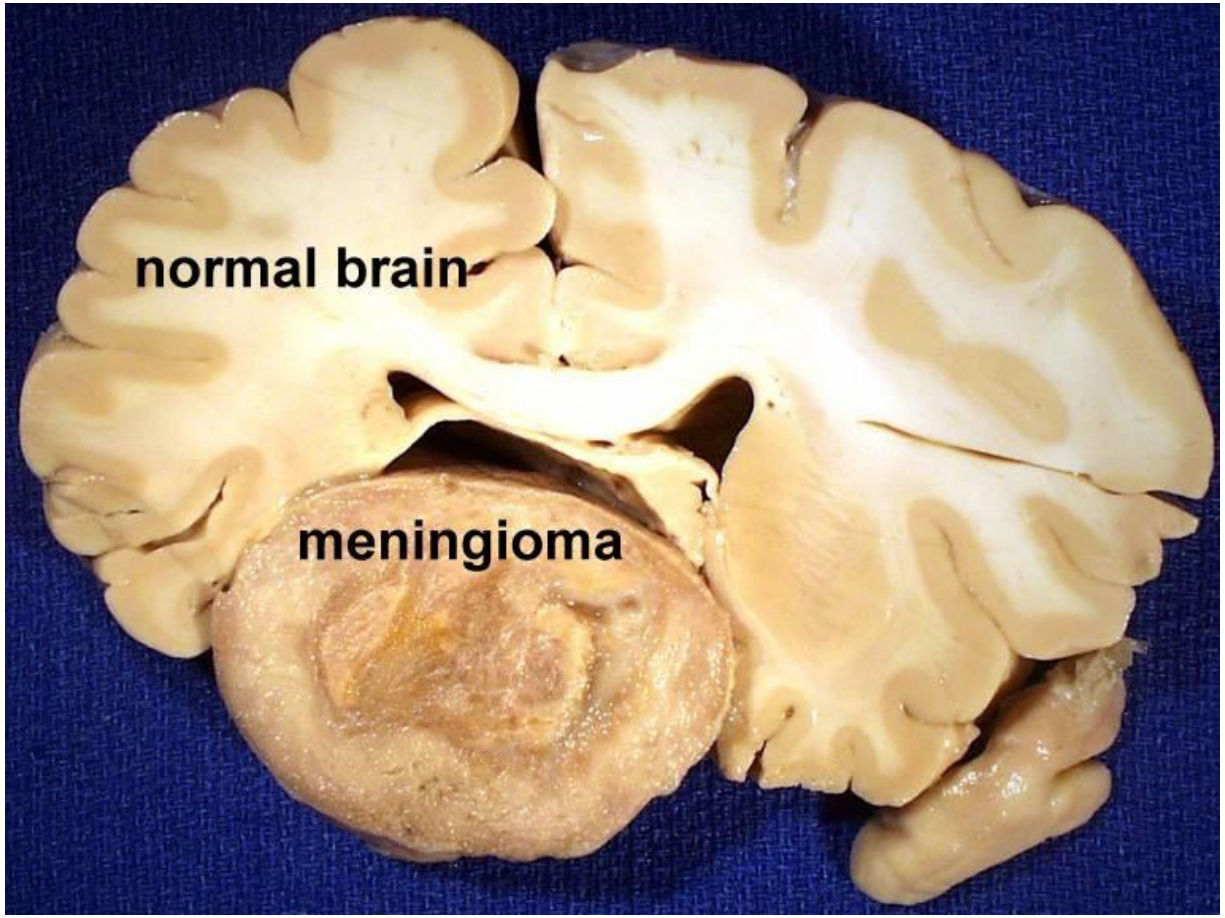
- WHO I: (well diff) meningioma .. *Further discussion in next slides*
- WHO II: atypical meningioma
- WHO III: anaplastic (malignant) meningioma (*looks like sarcoma*)

Grade 1 meningiomas

- Well defined, dura based masses
- May compress *causing seizures and blurred vision* but do not invade brain
- Can extend to overlying bone

Imaging shows a round mass attached to the dura



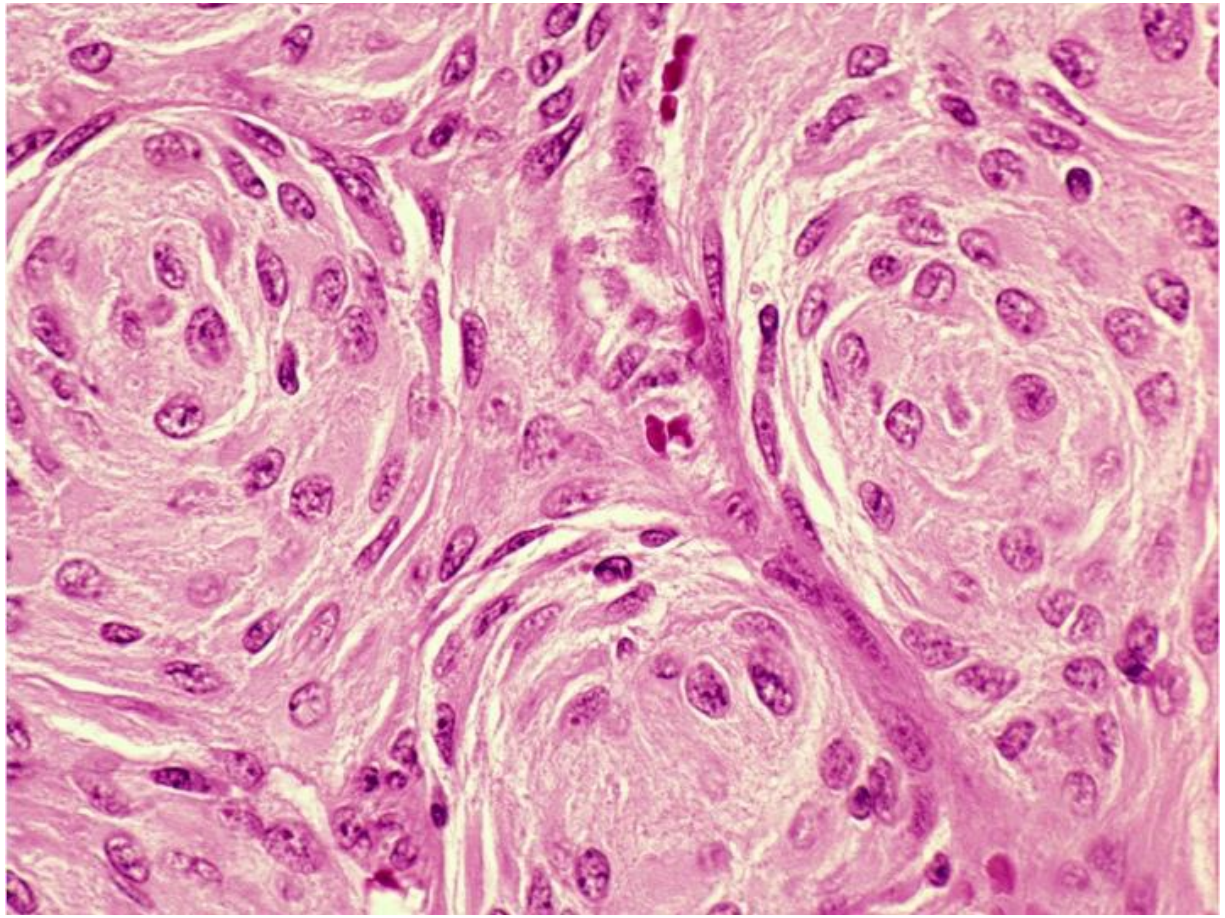

normal brain
meningioma

Grade 1 meningiomas/ histological types

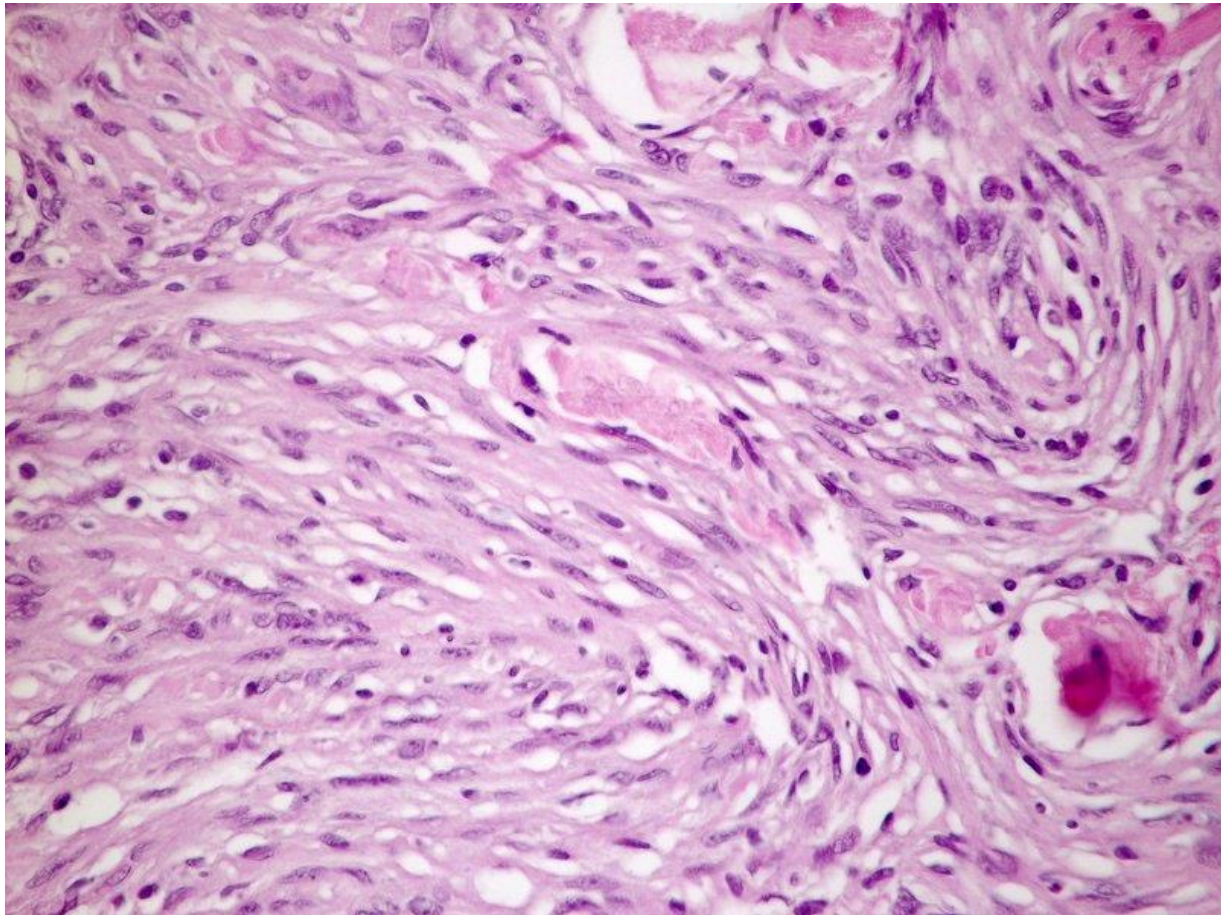
- Syncytial: whorled clusters without visible cell membranes.
- Fibroblastic: elongated cells and abundant collagen
- Transitional: features of both, syncytial and fibroblastic
- Psammomatous: numerous psammoma bodies

Tumors in which you see psammoma bodies:
1. Meningeoma
2.Papillary thyroid carcinoma
3.Serous ovarion adenocarcinoma

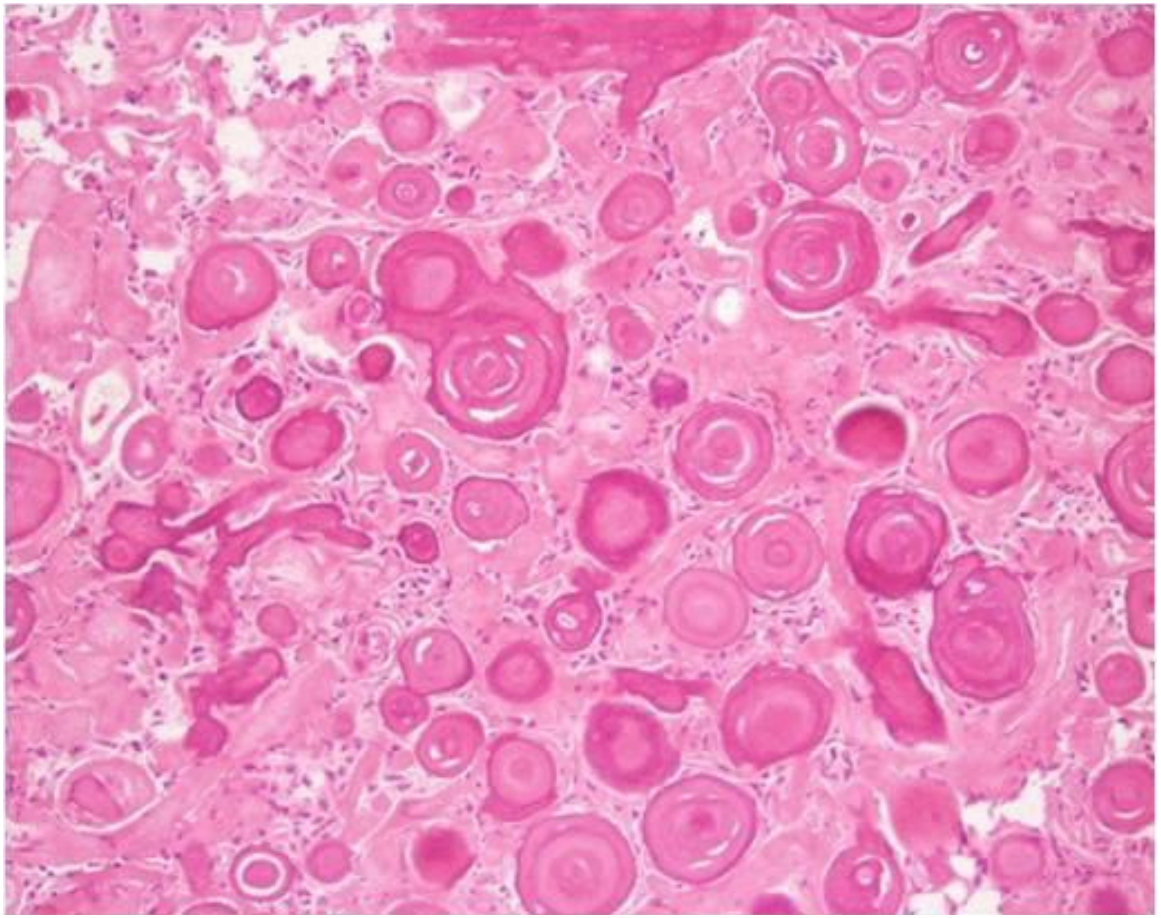
syncitial



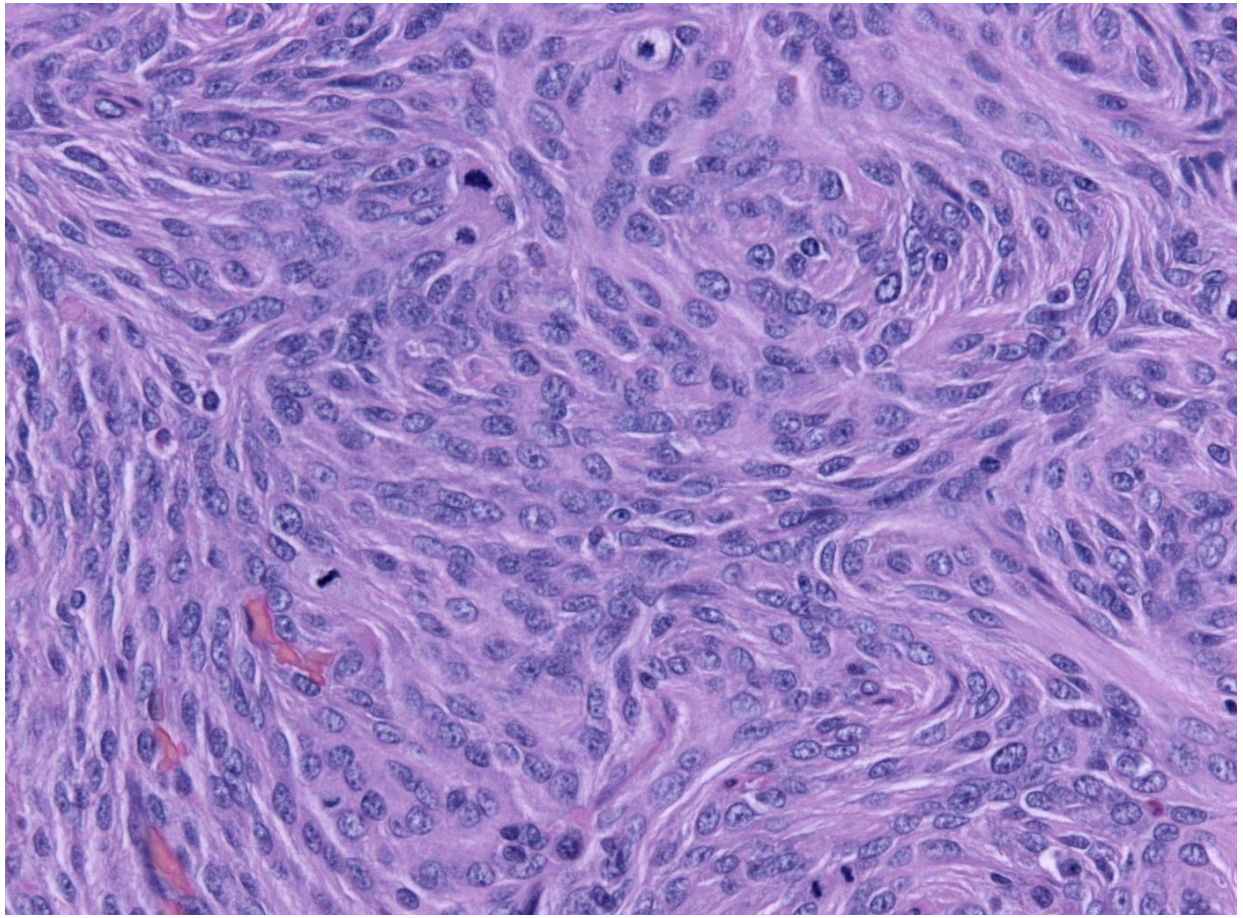
fibroblastic



pasammomatous



Atypical meningioma



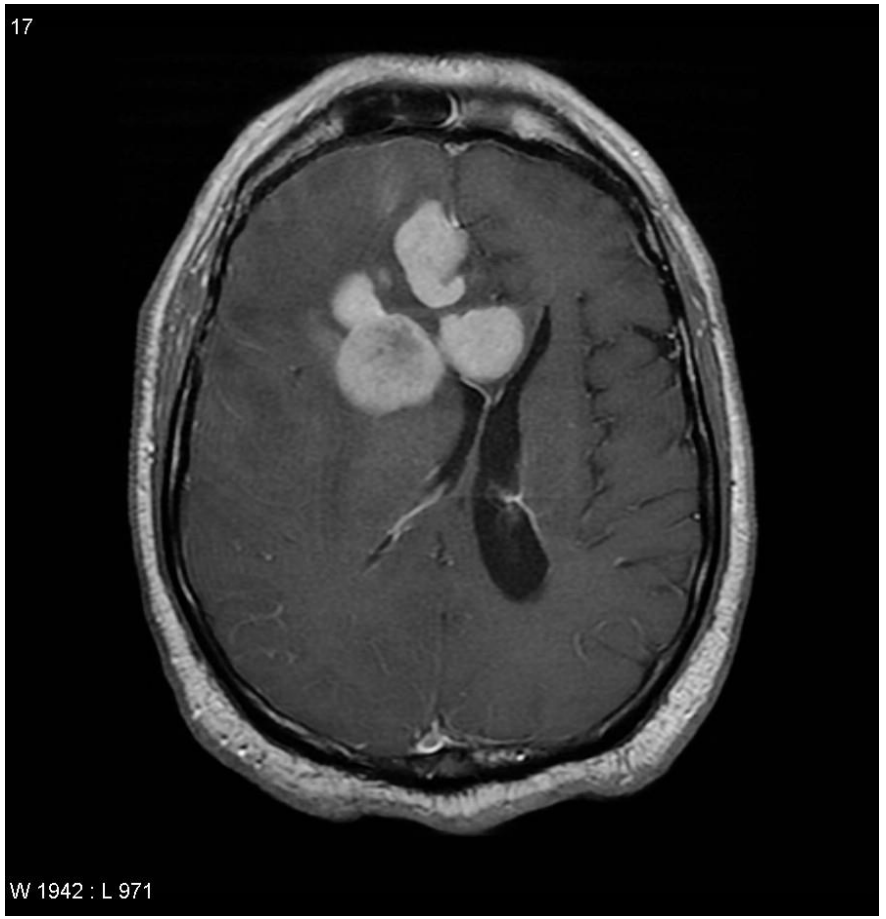
Anaplastic meningioma

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

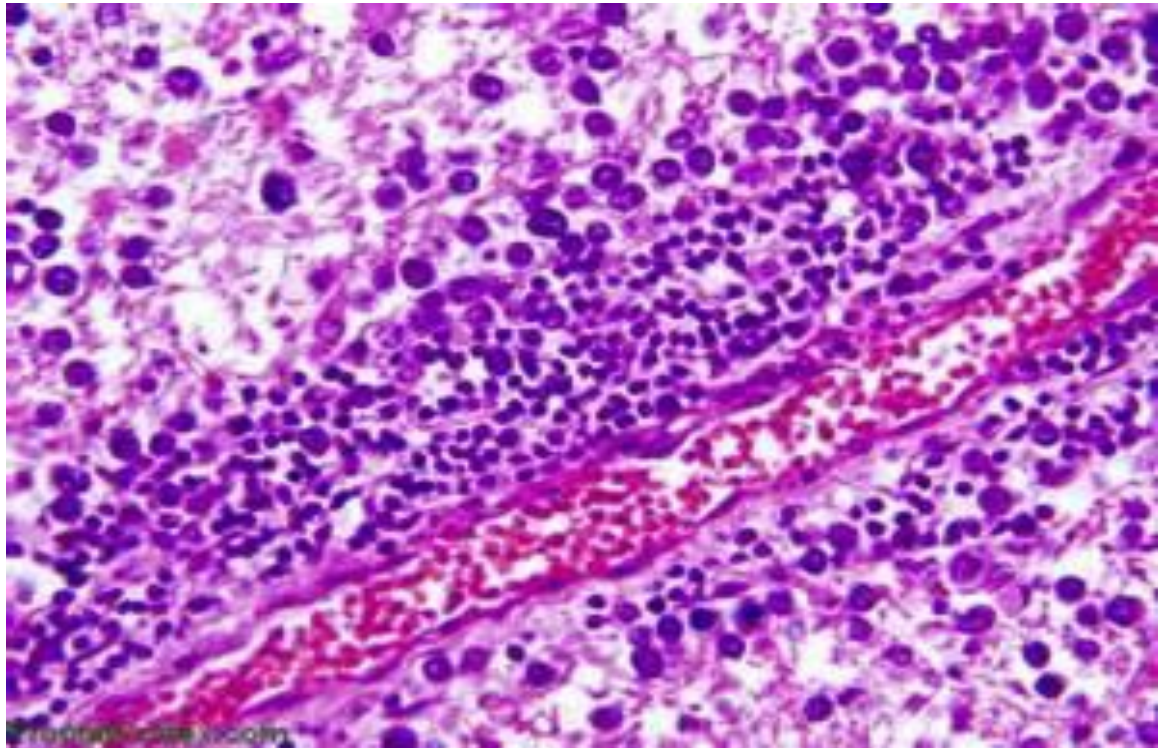
Primary CNS lymphoma

lymphoma

*Imaging shows **multiple masses**.*
And when you see multiple masses on imaging you should consider metastatic cancer as well.



Lymphoma/ note arrangement around blood vessels (*remember that CNS lymphoma arises from lymphocytes initially circulating in blood)*



- 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

Metastatic tumors

- 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

Paraneoplastic syndrome: Symptoms that occur in patients with cancer and that cannot be readily explained by 1.location of the primary tumor 2.distant spread of the tumor or 3.by the endogenous secretion of hormones

- 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

So if a patient that has cancer presented with CNS symptoms it is not necessiraly that his cancer metastasized to the CNS. It's probably a paraneoplastic syndrome

Familial tumor syndromes

The predisposition to these tumors shows an **autosomal dominant** pattern of inheritance because inheritance of a single mutant gene greatly increases the risk of developing a tumor.

- 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

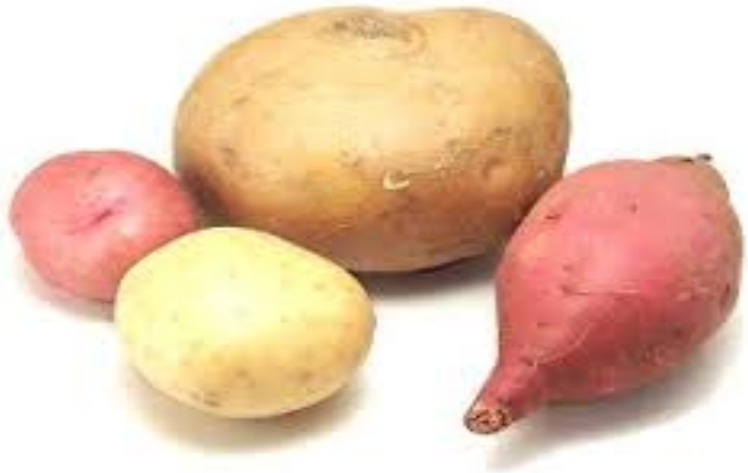
Tuberous sclerosis *tuberous=الدرنات*

- Autosomal dominant
- **Hamartomas** and benign neoplasms in brain and other sites
- CNS tumors: cortical tubers (*seizures*) and subependymal (*hydrocephalus*) hamartomas

Hamartoma is a mass of disorganized tissue indigenous to the particular site. i.e. normal tissue components but messy arrangement.

Cortical tubers

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



Cortical tubers

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

Sub ependymal tubers

- Similar to cortical tubers
- Can cause hydrocephalus

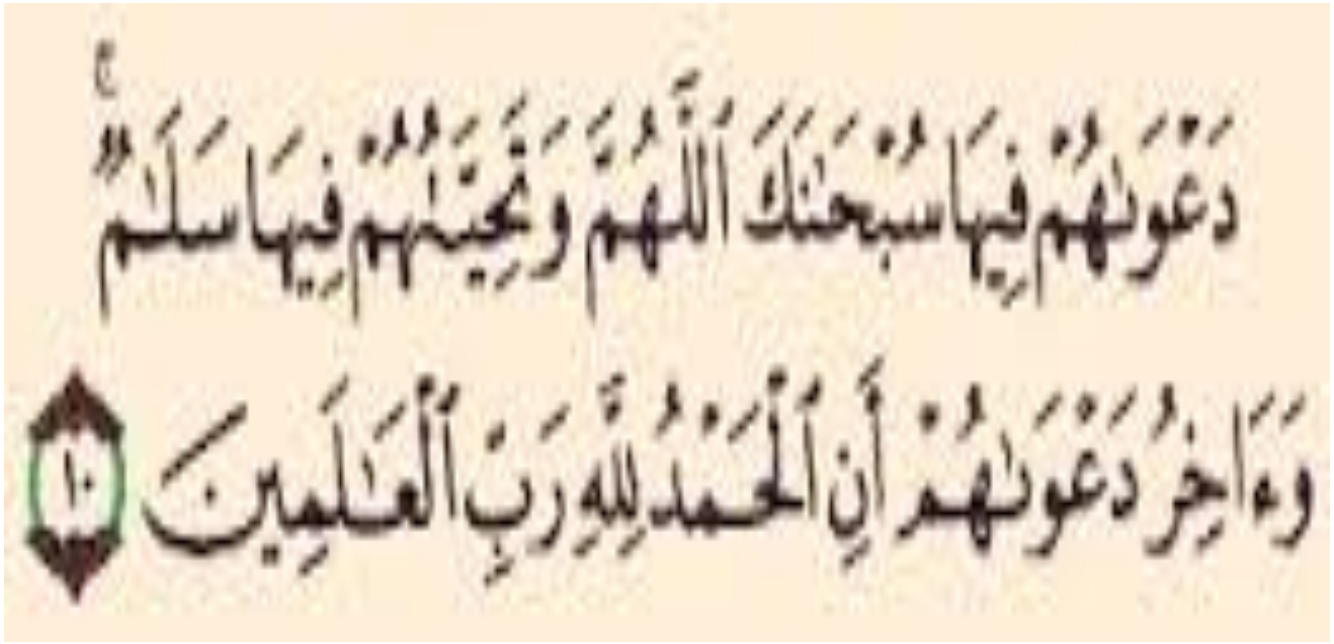
Tuberous sclerosis/Extra-cerebral lesions:

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

Von Hippel Lindau

von Hippel–Lindau protein (VHL) normally controls **angiogenesis**, If mutated: sustained angiogenesis. And that explains why it produces **Hemangioblastomas** (Hemangio=Blood vessels)

- 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 *(no increased risk in tuberous sclerosis)*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وَءَاخِرُ دَعْوَىٰهُمْ أَنِ ٱلْحَمْدُ لِلَّهِ رَبِّ ٱلْعَٰلَمِينَ ﴿١٠﴾