

# CNS Tumours and Raised intracranial pressure (ICP)

# CNS tumours

## Objectives

- Classification of CNS tumours
- Morphology of common tumours of the CNS

# Pathogenesis

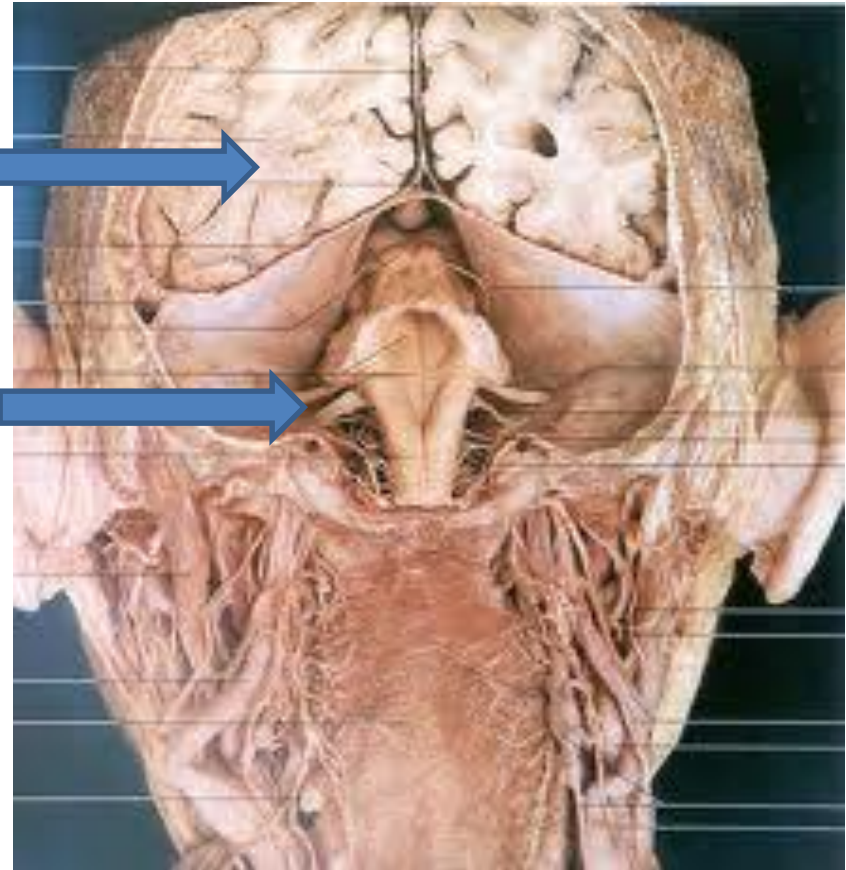
- Majority unknown
- Genetic alterations
  - Astrocytoma - inactivation of P53
- Associated with syndromes (rare)
  - Neurofibromatosis - Ependymoma
  - Tuberous sclerosis
  - Von-Hippel Lindau syndrome
- Immunosuppression - CNS lymphoma
- Chemicals
- Viruses
- Radiation

# CNS tumours - Unique features

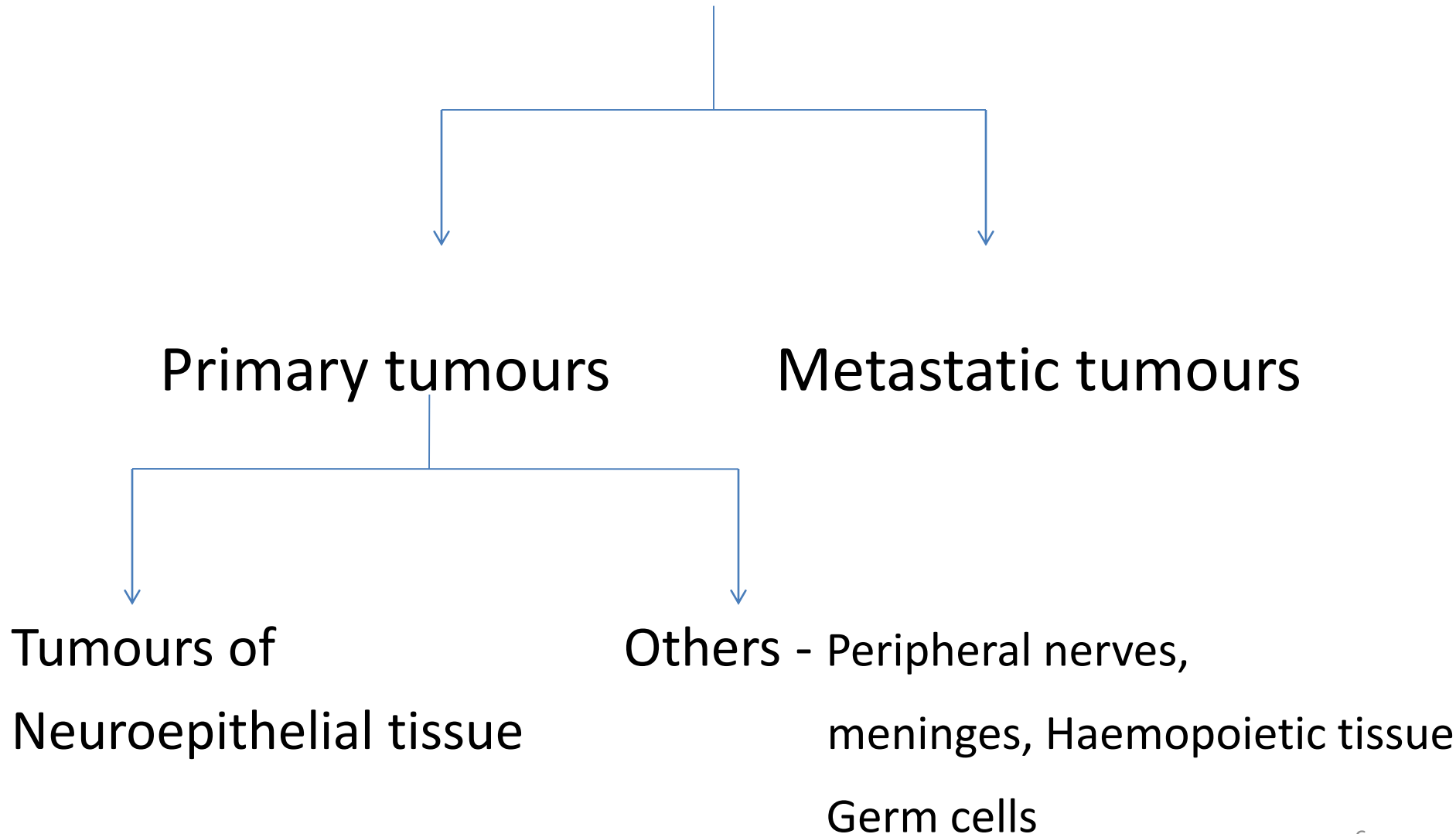
- Anatomic site
- Limited ability for resection
- Tumour spread through subarachnoid space
- Subtle histologic distinction between benign and malignant lesions

# Classification according to the location

- Supratentorial tumours  
Common in adults
- Infratentorial tumours  
Common in children



# Tumours of the CNS



# CNS tumour classification

- Glial tissue (65%)
  - Astrocytoma
  - Oligodendroglioma
  - Ependymoma
- Choroid plexus
  - Papilloma
  - Carcinoma
- Primitive neuroectodermal (Embryonal) - Medulloblastoma
- Meningoendothelial (arachnoid) cells - Meningioma
- Nerve sheath cells
  - Schwannoma, Neurofibroma
- Lymphoreticular cells
  - Lymphoma

# Tumours of Neuroepithelial origin

- Glial tissue (commonest)
  - Astrocytoma
  - Oligodendroglioma
  - Ependymoma
- Choroid plexus
  - Papilloma
  - Carcinoma
- Primitive neuroectodermal (embryonal)
  - Medulloblastoma



# Astrocytoma

- Commonest primary brain tumour in adults (80%)
- Usually involves cerebral hemispheres (supratentorial)
- Signs and symptoms - Seizures, headaches, and focal neurologic deficits
- Not defined as benign/malignant as all tumours are infiltrative
- Resection is difficult
- Recurrence - common
- Metastasis outside the brain - unlikely
- Occasionally spread through the sub arachnoid space

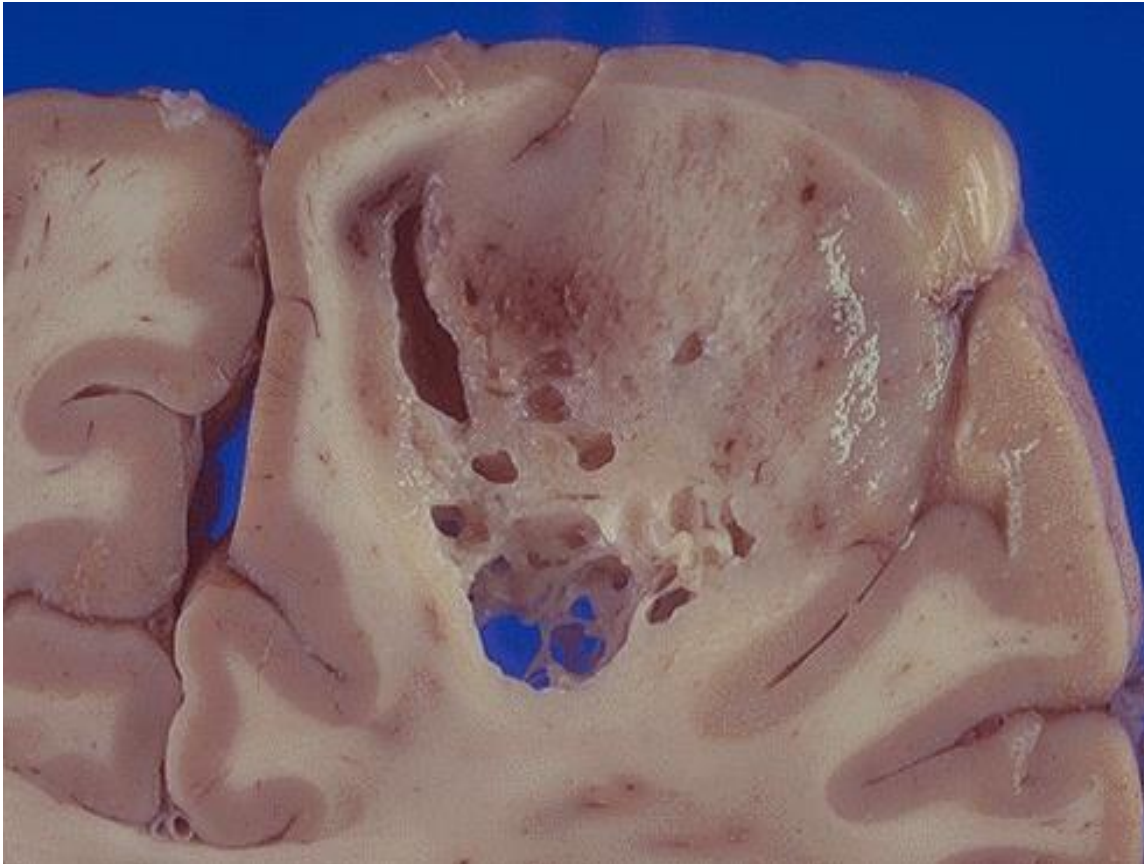
# Astrocytoma

- Subtyping is according to cellularity, cellular atypia, mitoses and necrosis
- Diffuse astrocytoma - Well differentiated tumour
- Anaplastic astrocytoma - Increased cellularity, cellular atypia, increased mitoses
- Glioblastoma multiforme - Poorly differentiated astrocytes, marked nuclear atypia and mitoses, microvascular proliferation , necrosis

# Astrocytoma

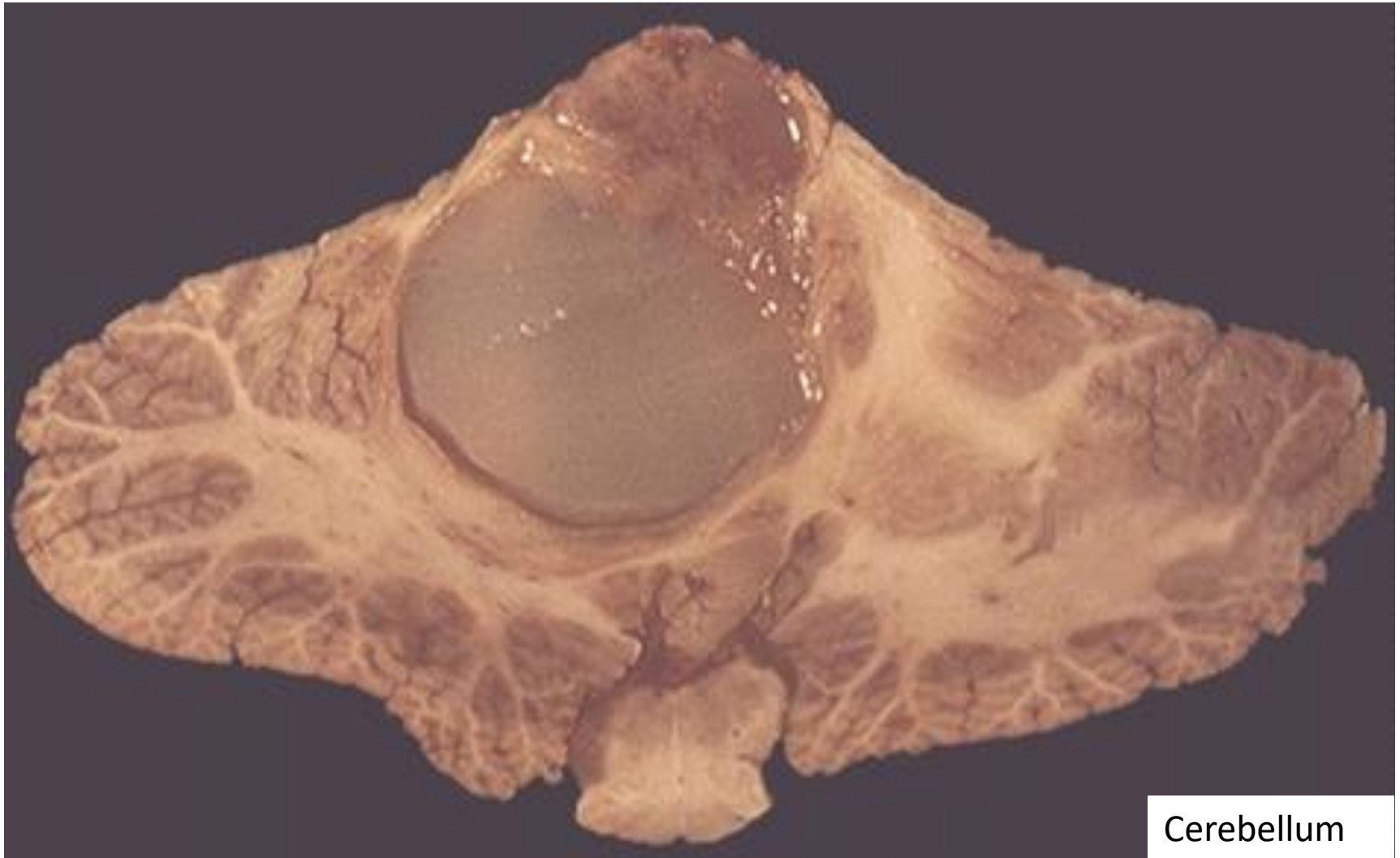
- Macroscopy
  - Poorly defined grey color tumour
  - Expands and infiltrates brain
  - No destruction of brain tissue
  - Cut surface - Firm / soft and gelatinous
    - Gritty if calcification is present
    - Cystic changes may be seen

# Astrocytoma - Macroscopy



- Note: - Poor demarcation
- Expands and infiltrates brain
  - Cystic changes

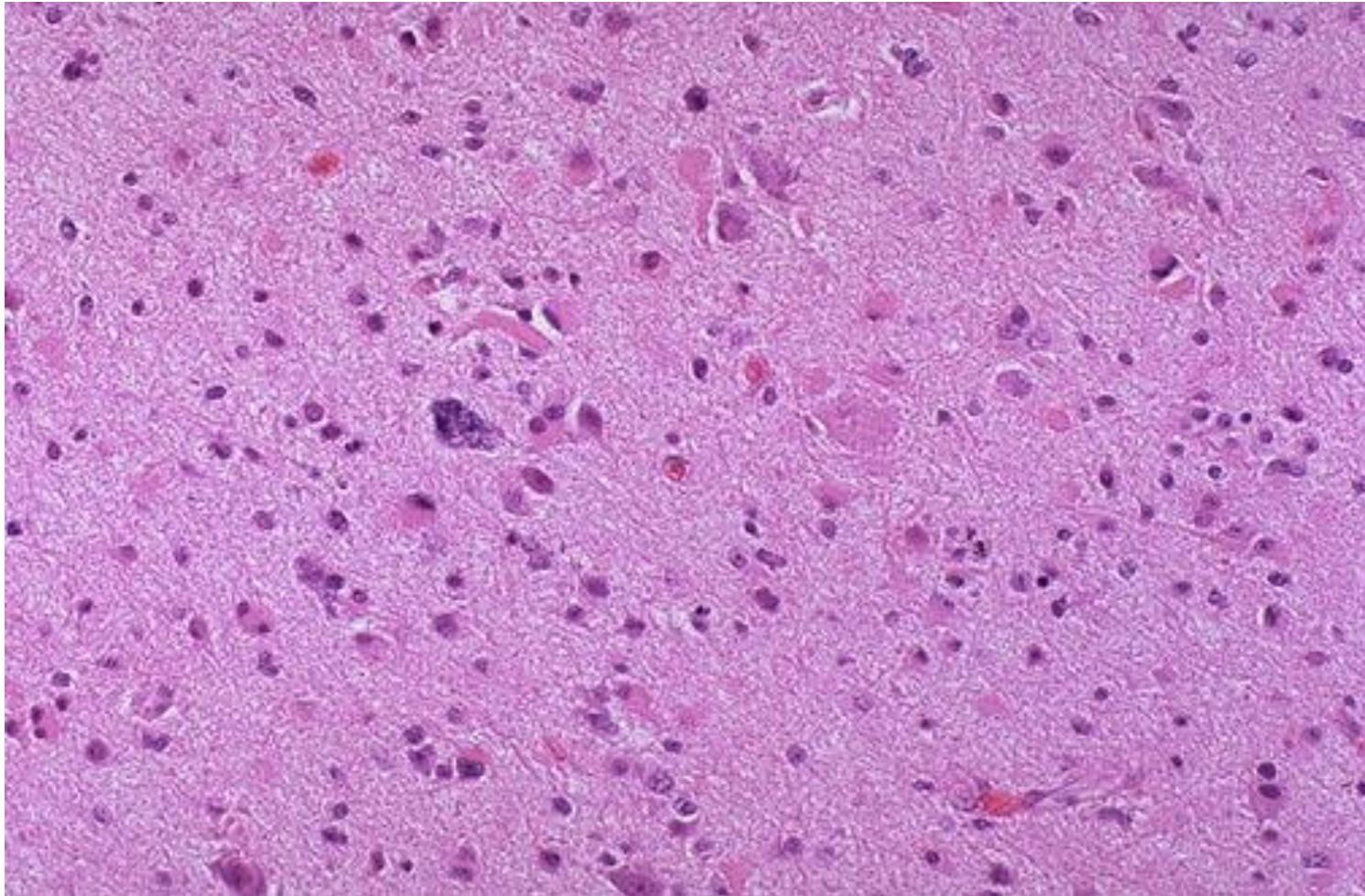
# Astrocytoma - Macroscopy



Note: Single large cyst filled with gelatinous material

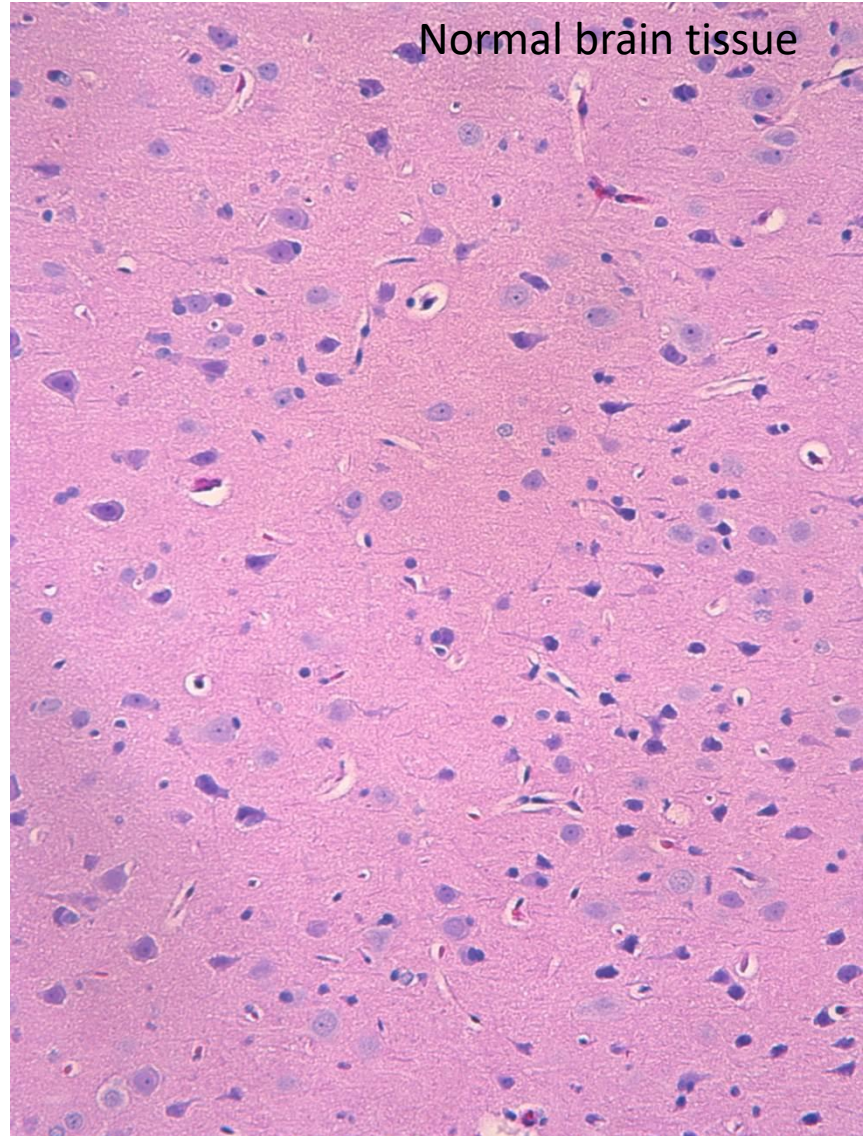
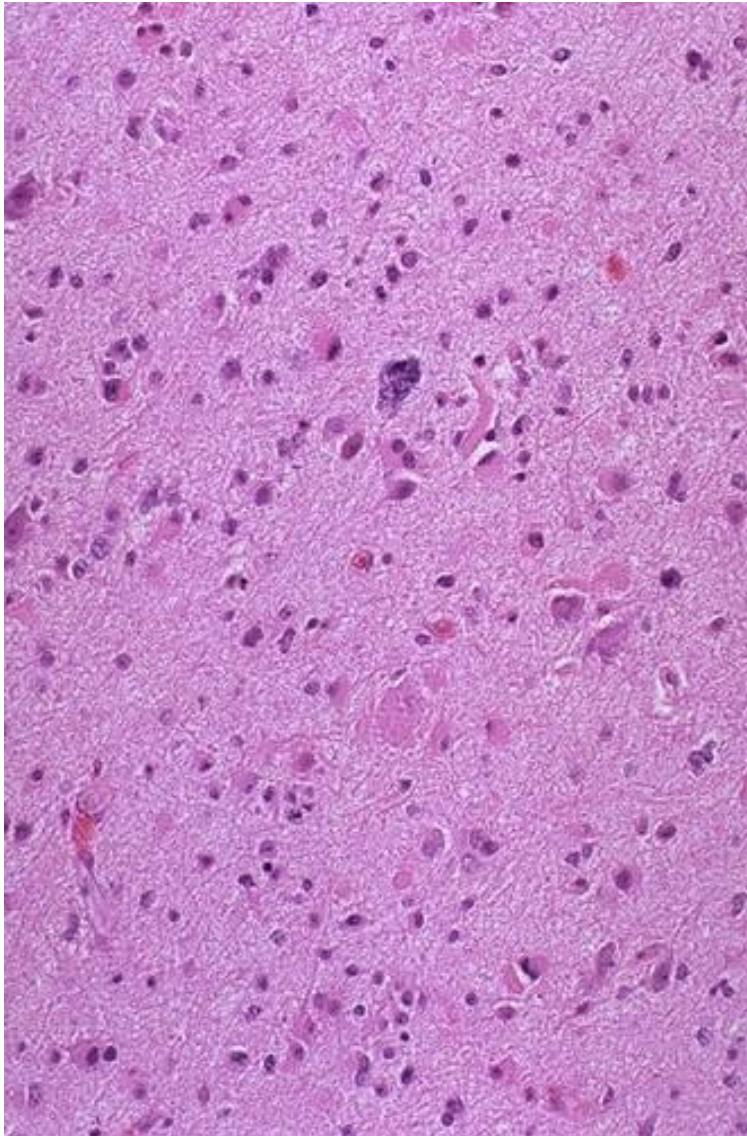


# Astrocytoma - Microscopy





# Astrocytoma - Microscopy



Normal brain tissue

May be difficult to differentiate from the normal brain tissue

# Glioblastoma

- Usually >50 yrs
- Primary / secondary (arise from an astrocytoma)

## Macroscopy

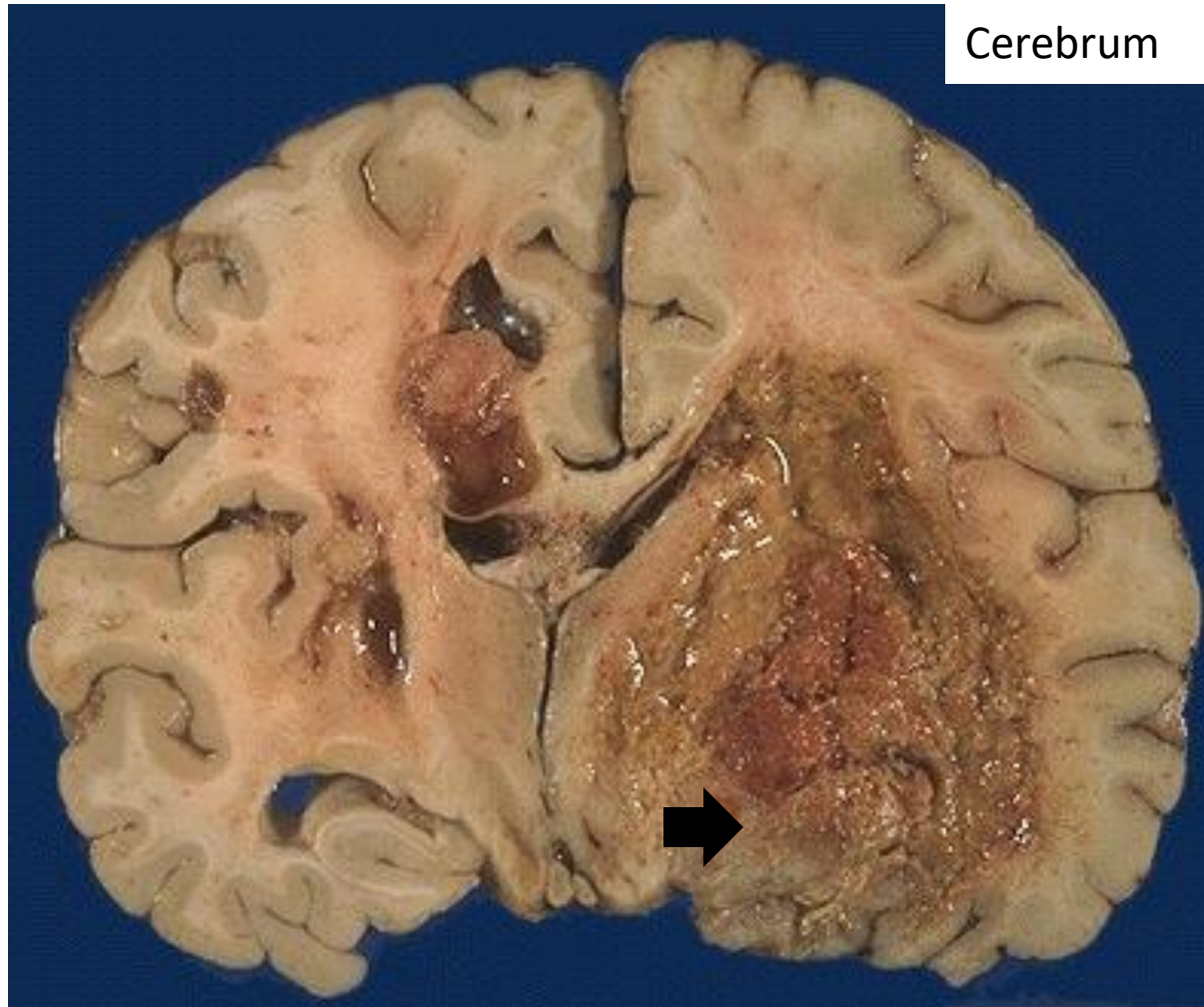
- Poorly demarcated tumours
- Grey-yellow colour
- Necrosis and haemorrhages

## Microscopy

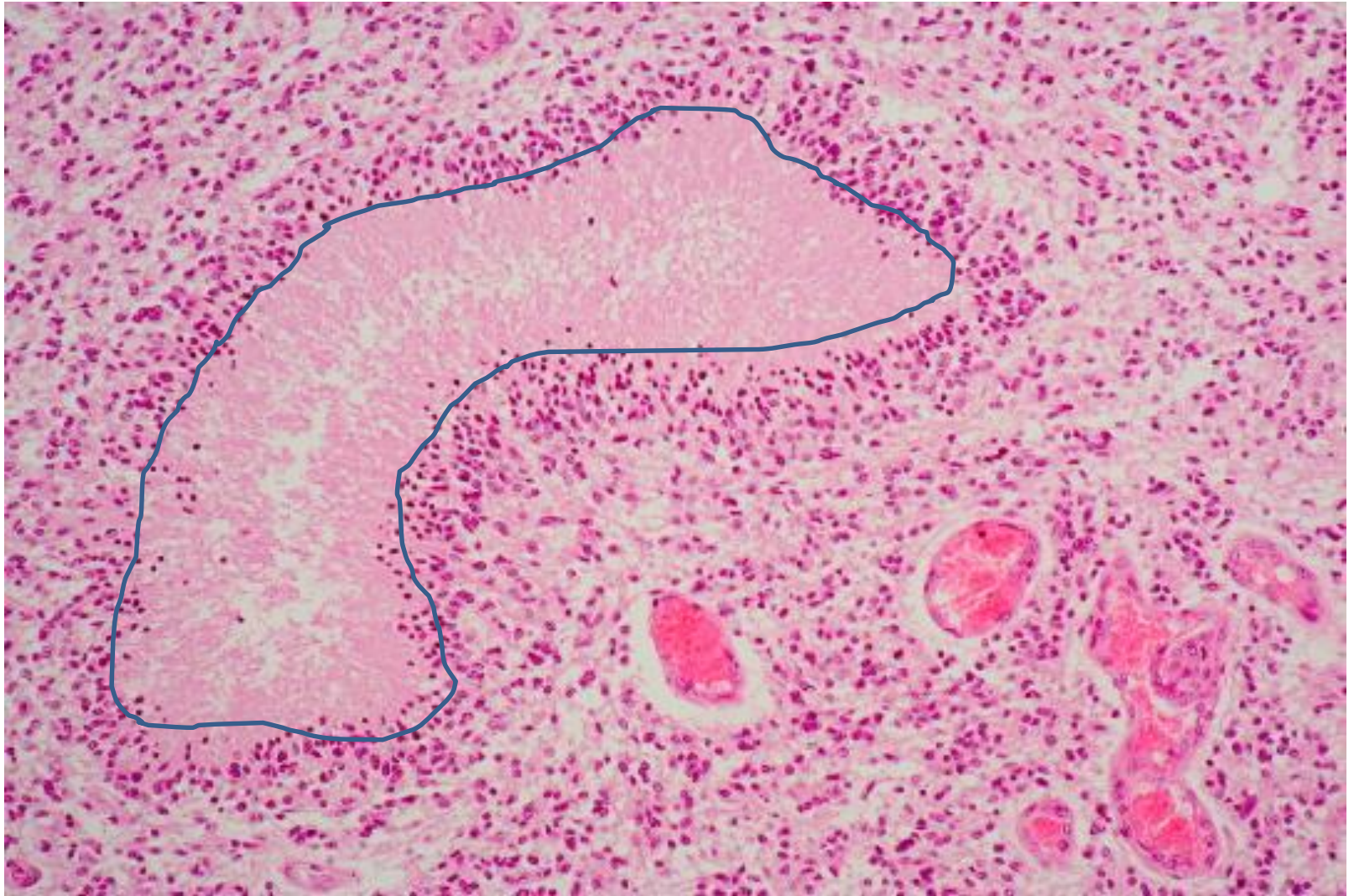
- Cellular atypia
- Necrosis
- Mitoses
- Endothelial cells proliferation



# Glioblastoma - Macroscopy



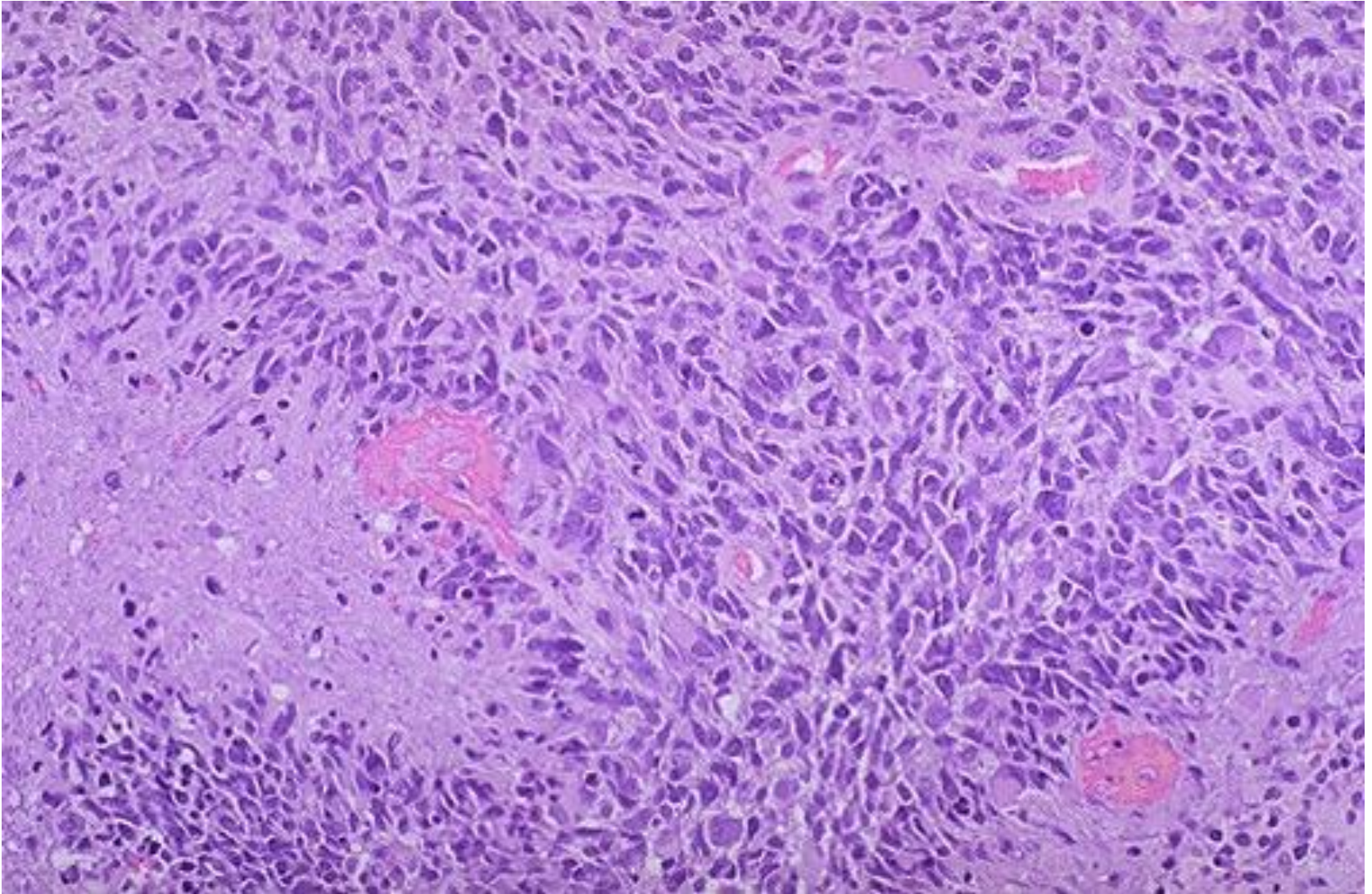
# Glioblastoma - Microscopy



Note: Tumour necrosis



# Glioblastoma - Microscopy



Note: Cellular atypia

# Oligodendroglioma

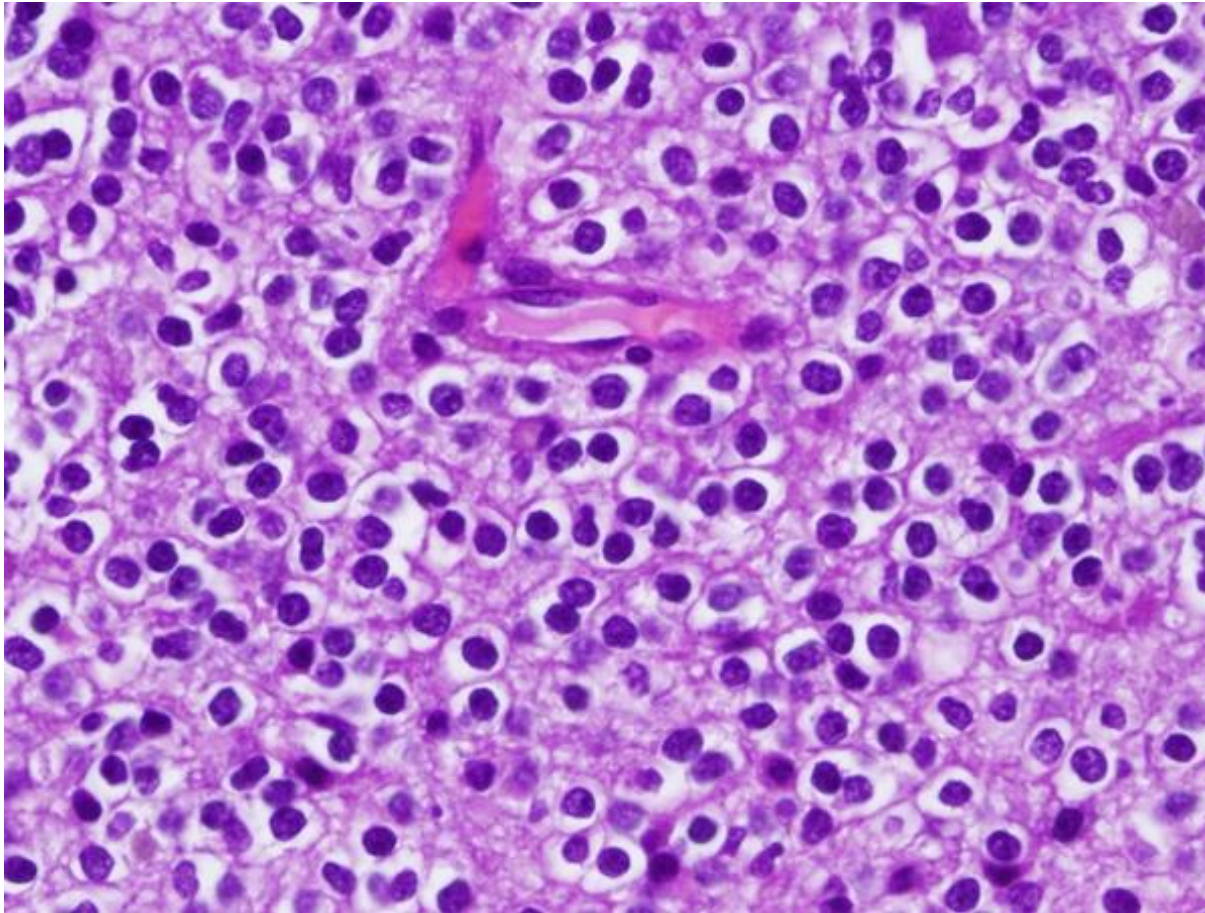
- Well differentiated, diffusely infiltrating tumours
- Usually in adults
- Slowly growing tumours
  - May present with long history of neurological symptoms
- Mostly in the cerebral hemispheres
- Predilection for white matter

# Oligodendroglioma

- Macroscopy
  - Fairly circumscribed, gelatinous tumours
  - Cystic changes
  - Focal hemorrhage
  - Calcification



# Oligodendroglioma - Microscopy



- Note:
- Uniform small cells
  - Round uniform nuclei and
  - Clear cytoplasm with distinct cell borders

# Ependymoma

- Arises from the ependymal lining of the cerebral ventricles / central canal of spinal cord

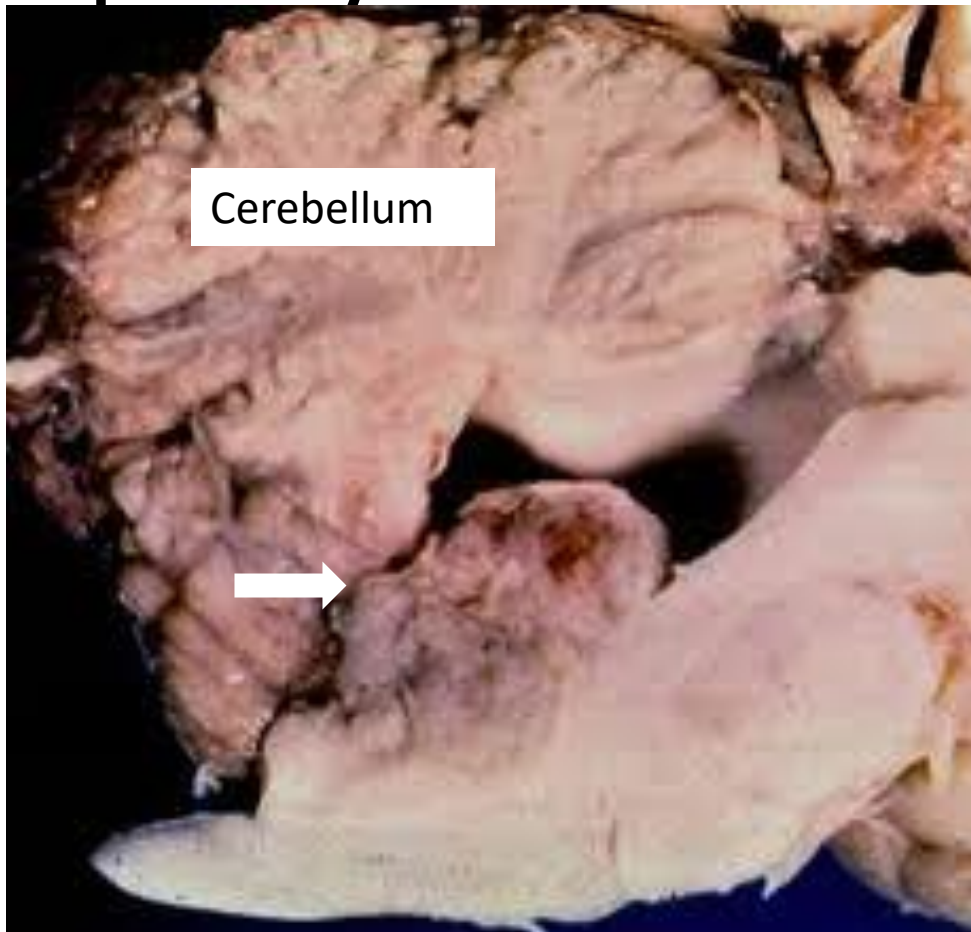
- In children - 4<sup>th</sup> ventricle is a common site

In adults - spinal cord is the most common location

- Associated with neurofibromatosis type 2

- High grade variant seeds through the CSF
- Posterior fossa tumours
  - Hydrocephalus secondary to progressive obstruction of the fourth ventricle

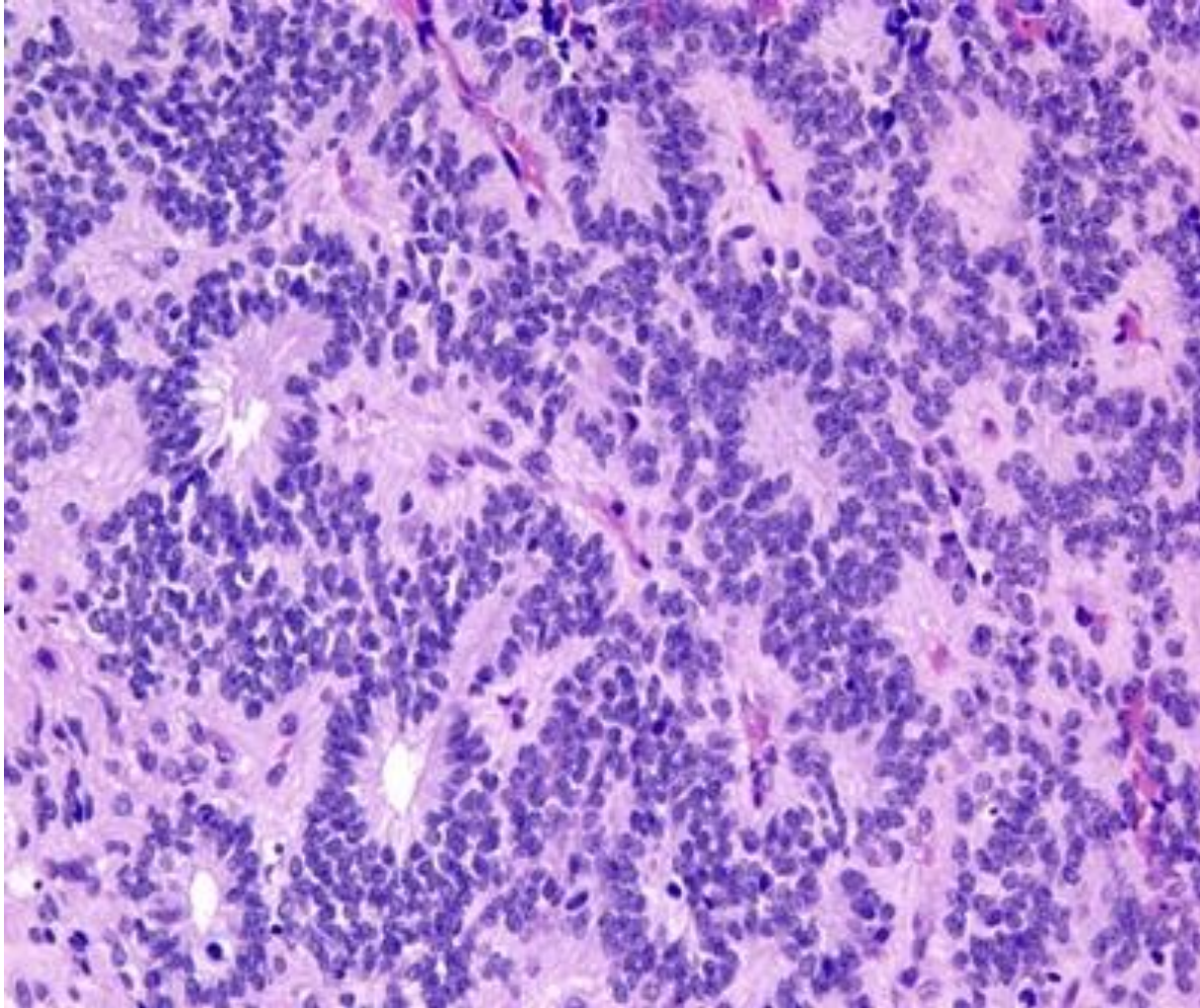
# Ependymoma - Macroscopy



- Note:
- Tumour rising from the floor of the fourth ventricle
  - Well demarcated , grey-red colour tumour
  - Solid/ papillary mass
  - Haemorrhagic and necrotic areas



# Ependymoma - Microscopy

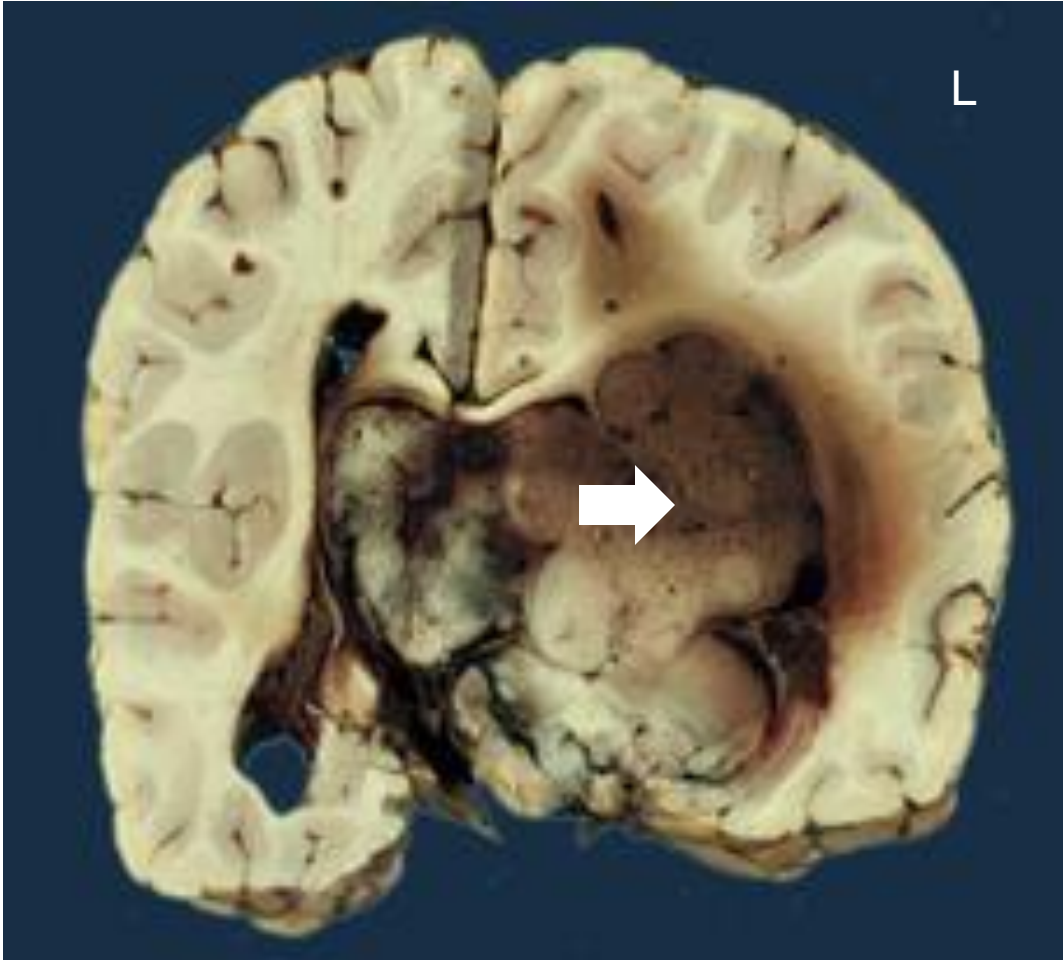


Uniform tumour cells arranged around blood vessels  
“Perivascular pseudorosettes”

# Choroid plexus papilloma

- Benign tumour
- Common in children
- Usually presents with hydrocephalus due to obstruction of the ventricular system by tumour or to overproduction of CSF
- Arise from the epithelium of the choroid plexus of the cerebral ventricles
- Macroscopy : Circumscribed cauliflower like mass attached to the ventricular wall

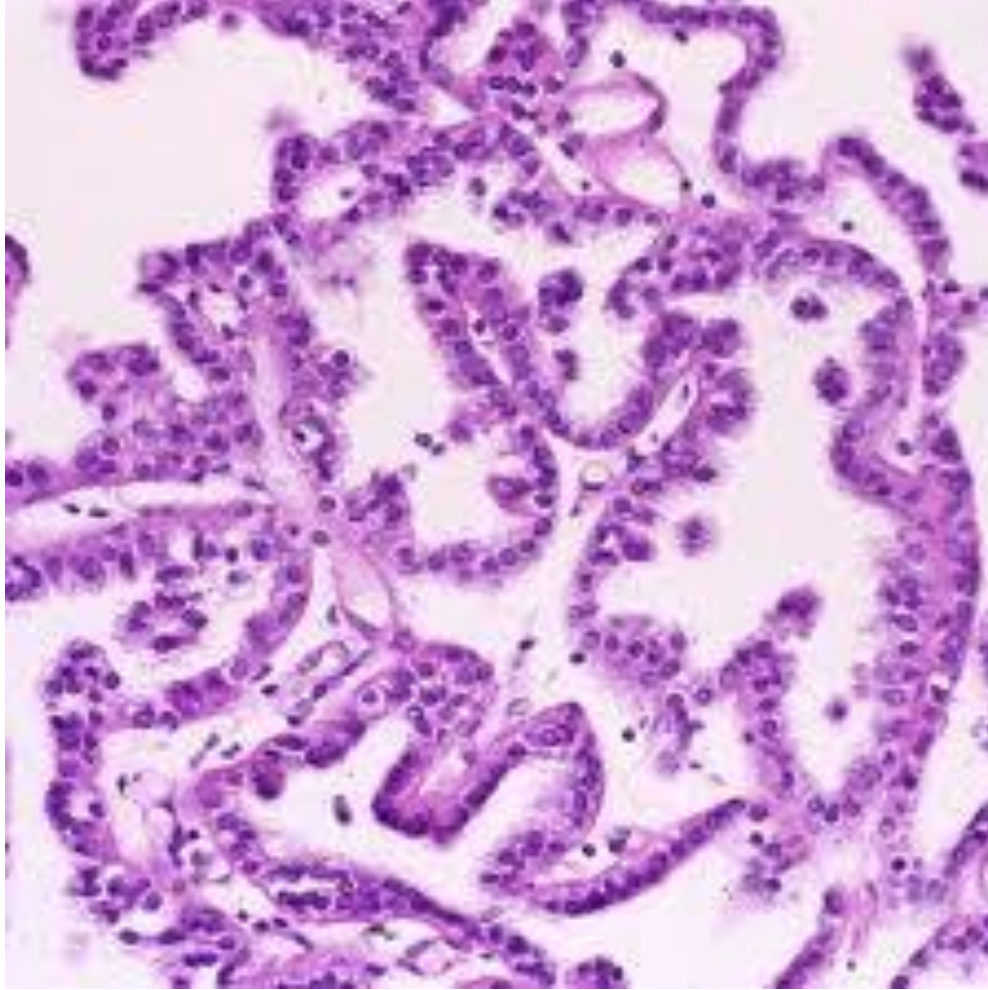
# Choroid plexus papilloma - Macroscopy



Note:

Involvement of the  
left lateral ventricle

# Choroid plexus papilloma - Microscopy



- Note: - Delicate papillary structures lined by a single layer of uniform cuboidal to columnar cells
- Basally located round to oval uniform nuclei

# Medulloblastoma

- Poorly differentiated tumour
- Predominantly in children
- Exclusively in the cerebellum (usually midline)
- Rapidly growing
- May occlude the flow of CSF → Hydrocephalus
- Extensive local infiltration with meningeal infiltration → CSF seeding



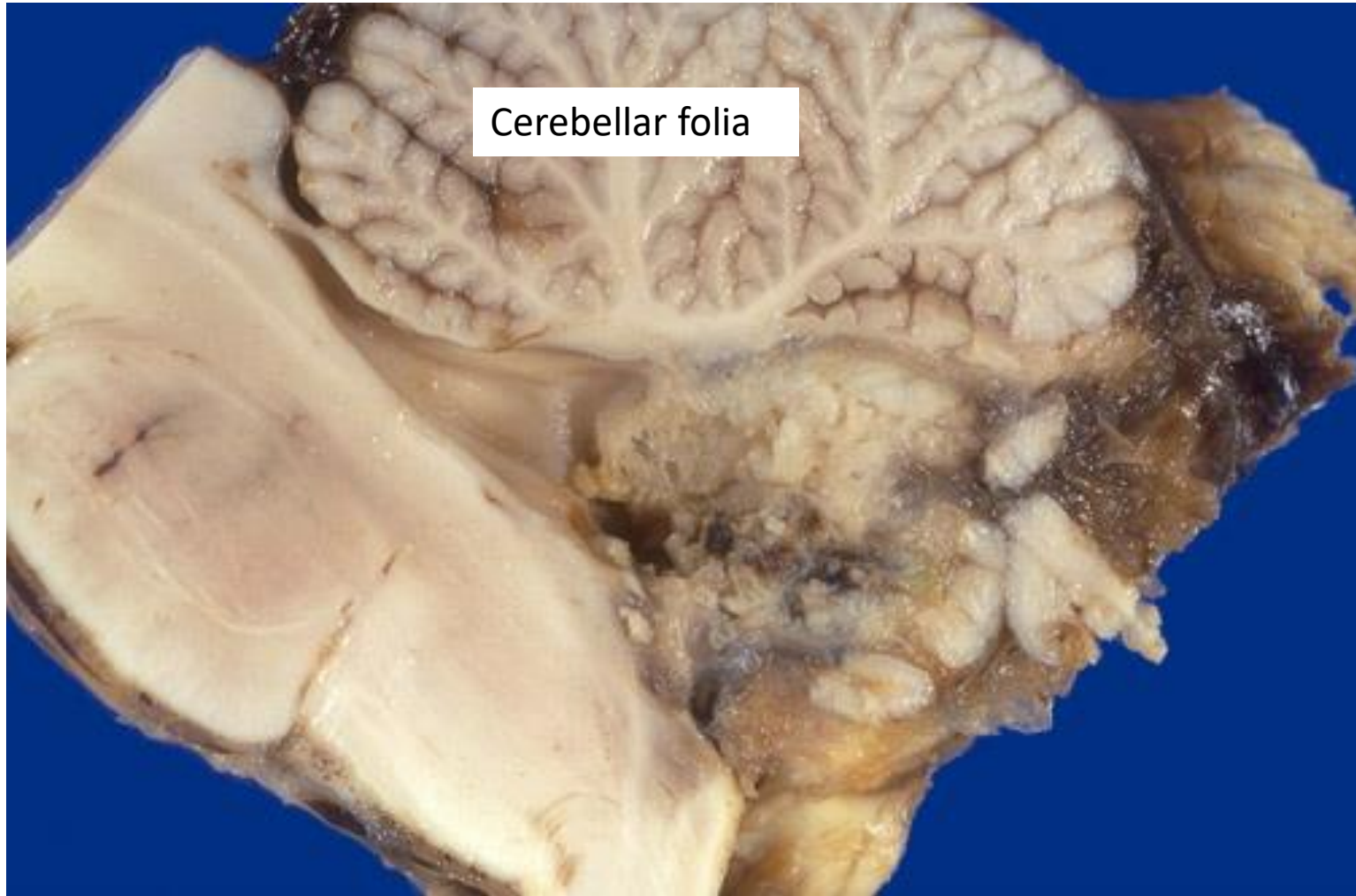
# Medulloblastoma

- Macroscopy
  - Often well circumscribed, grey and friable
  - Some show haemorrhages
  - may extend to the cerebellar folia
  - may involve the leptomeninges

# Medulloblastoma - CT scan

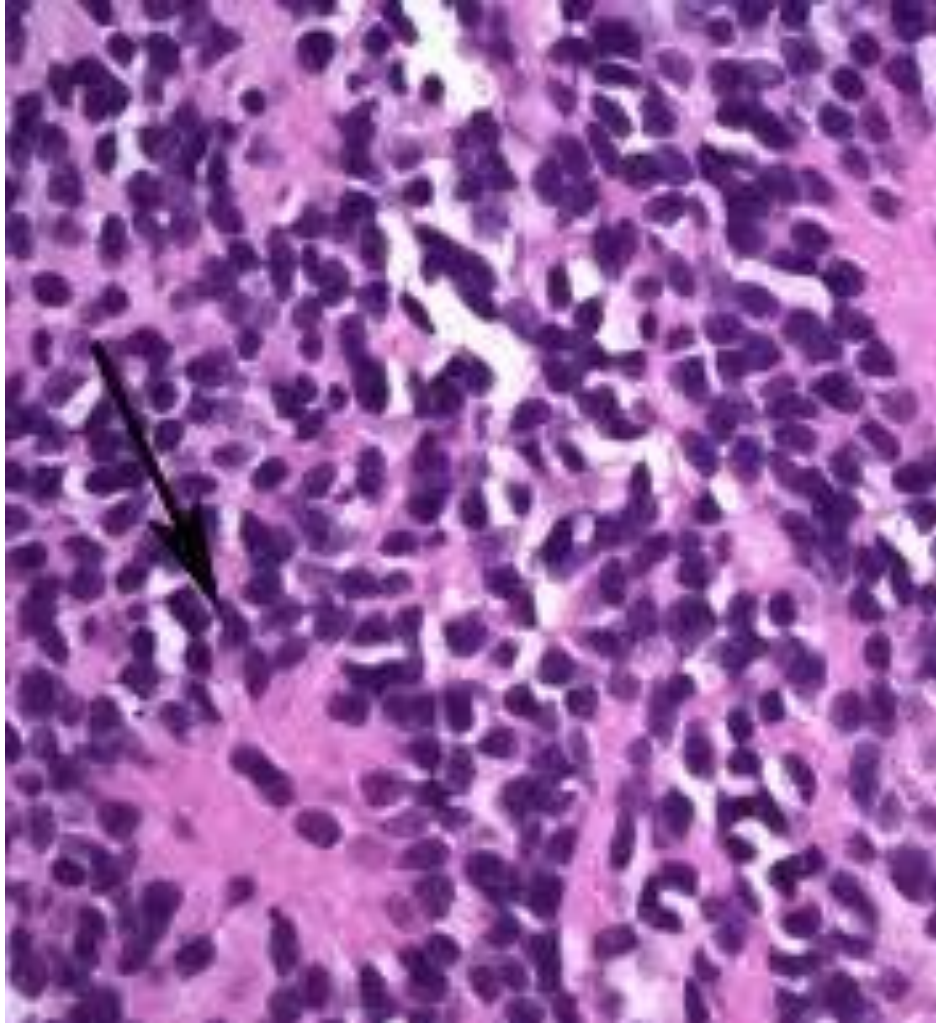


# Medulloblastoma - Macroscopy





# Medulloblastoma - Microscopy



## Microscopy

- Highly cellular sheets of round to oval anaplastic cells

# Other tumours of the CNS

- Meninges - Meningioma
- Hemopoietic tissue - Lymphoma
- Germ cell tumours
- Sellar region tumours - Craniopharyngioma
- Peripheral nerves - Schwannoma, neurofibroma

# Meningioma

- Predominantly benign tumors of adults
- Female > male
- Usually attached to the dura
- Usually solitary tumours
- Arise from the meningotheelial (arachnoid) cells
- Slow growing tumours
- Clinical features due to compression

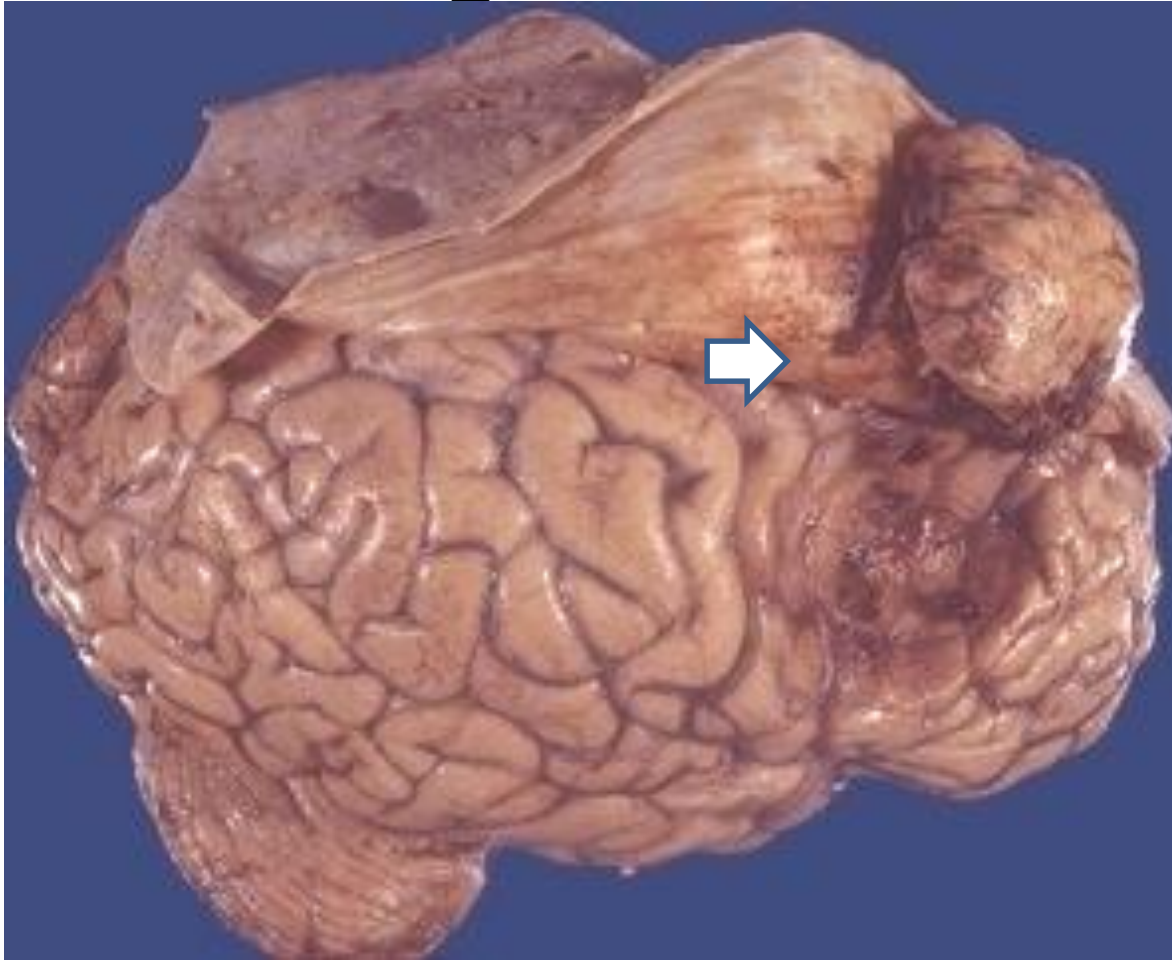
# Meningioma

- Site - Parasagittal sinus  
Olfactory groove  
Sphenoidal ridges  
tentorium cerebelli etc.
- Spinal meningioma may cause cord compression

# Meningioma

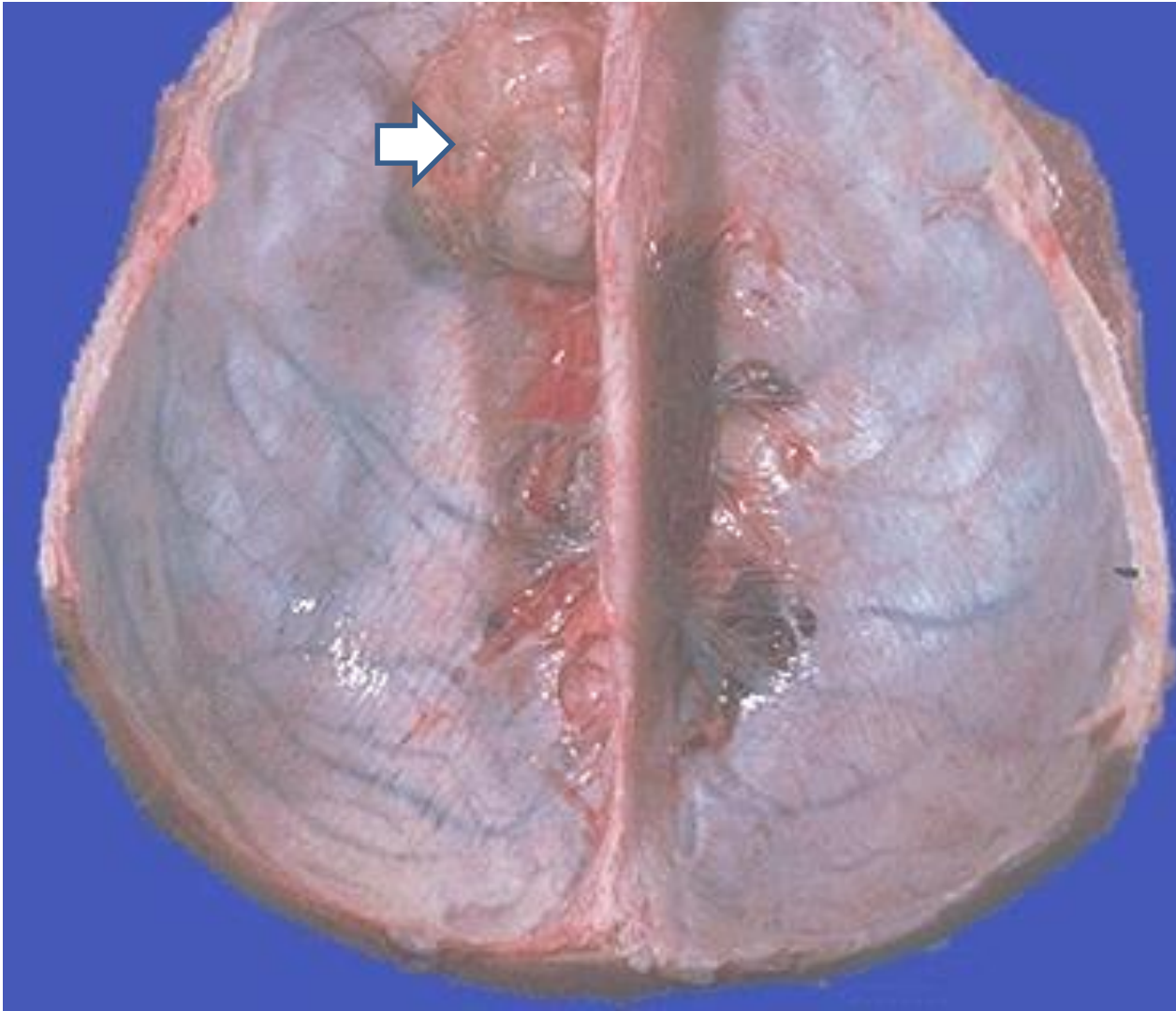
- Depending on the cellular atypia , mitosis , necrosis different types are recognized
- Atypical meningioma - High recurrence
- Anaplastic meningioma - Highly aggressive

# Meningioma - Macroscopy



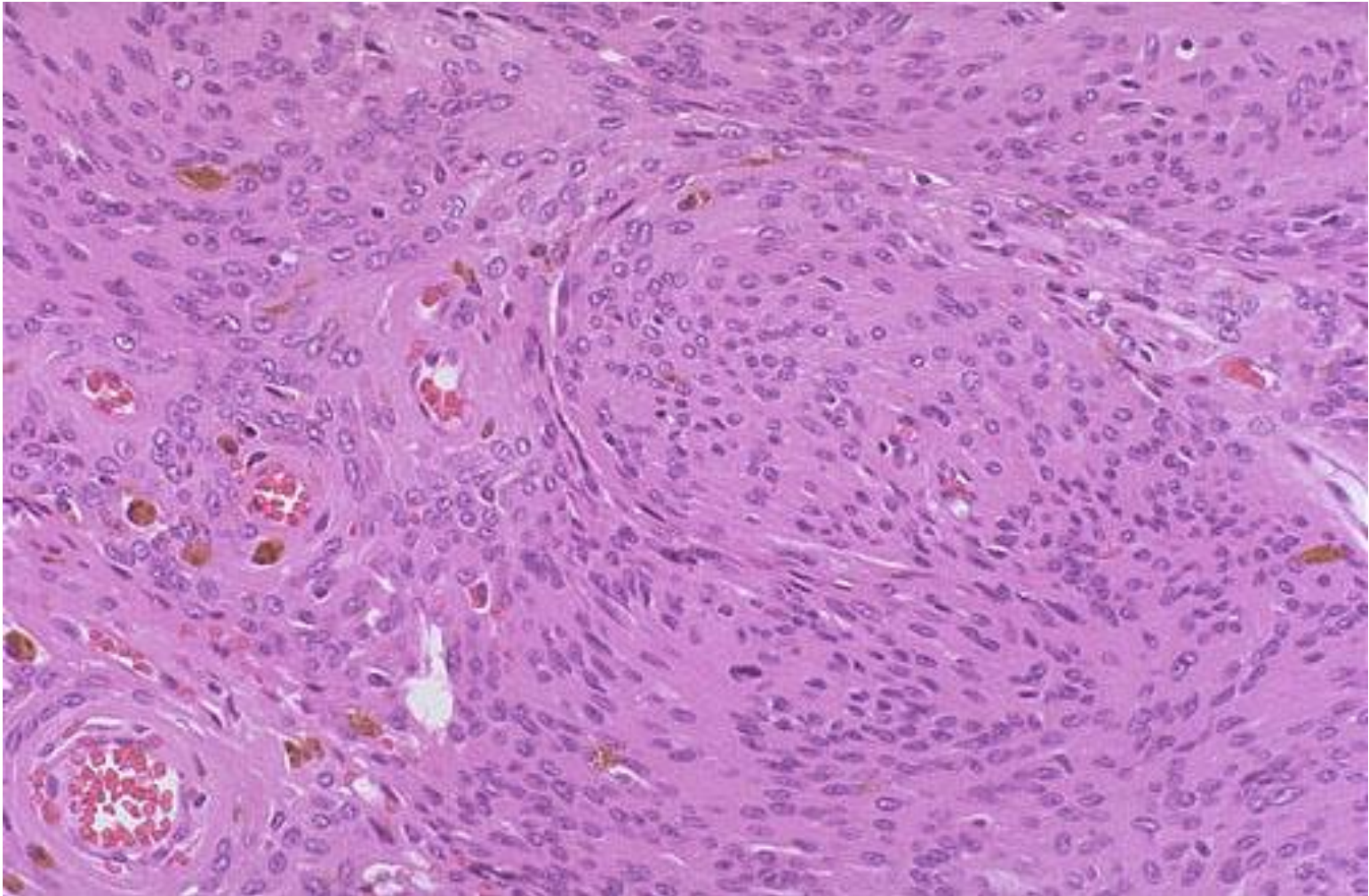
- Note:
- Well defined, smooth, solid, lobulated tumour
  - Adherent to the dura
  - Compression of brain matter

# Meningioma - Macroscopy





# Meningioma - microscopy



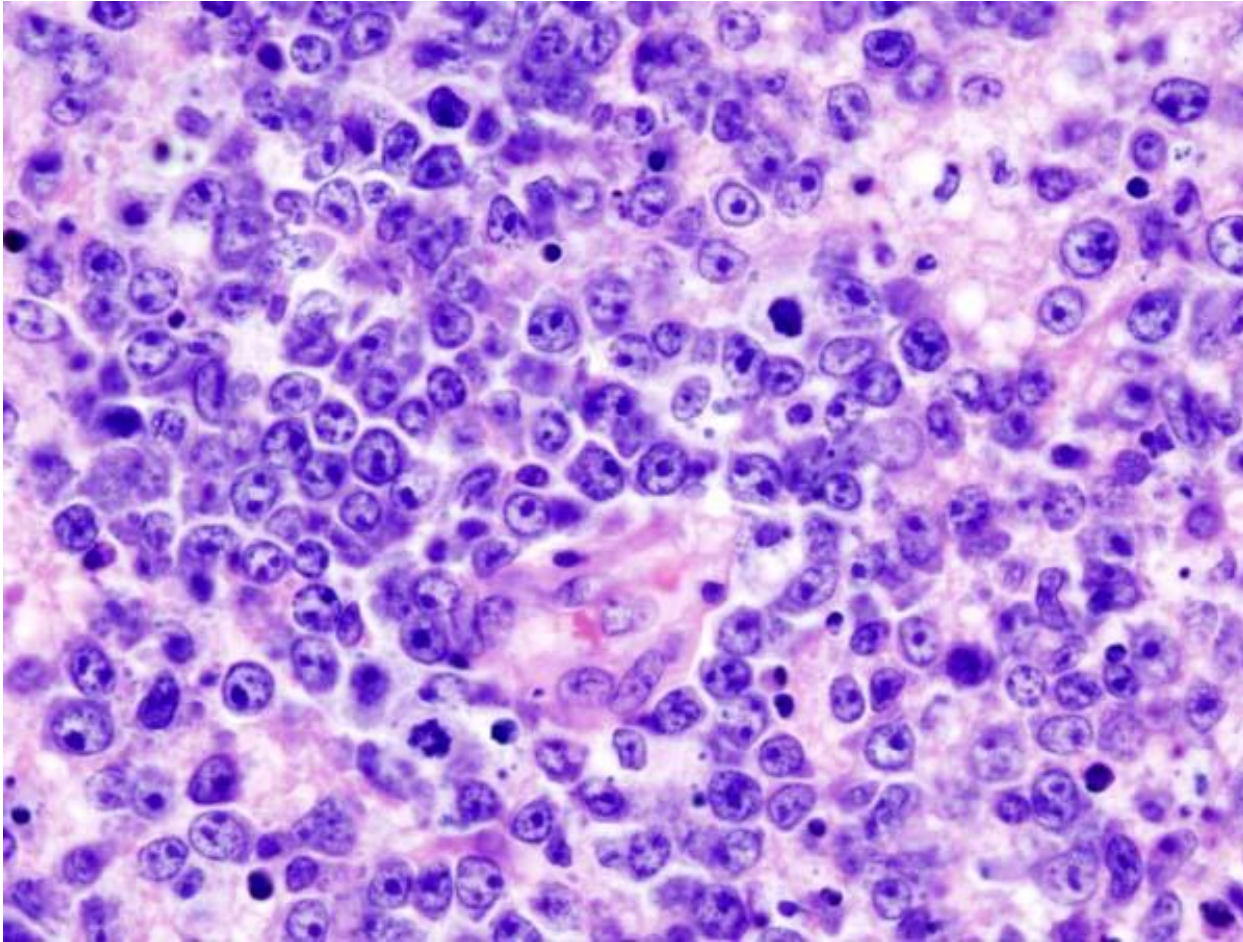
Note: Uniform tumour cells with oval nuclei



# CNS - Lymphoma

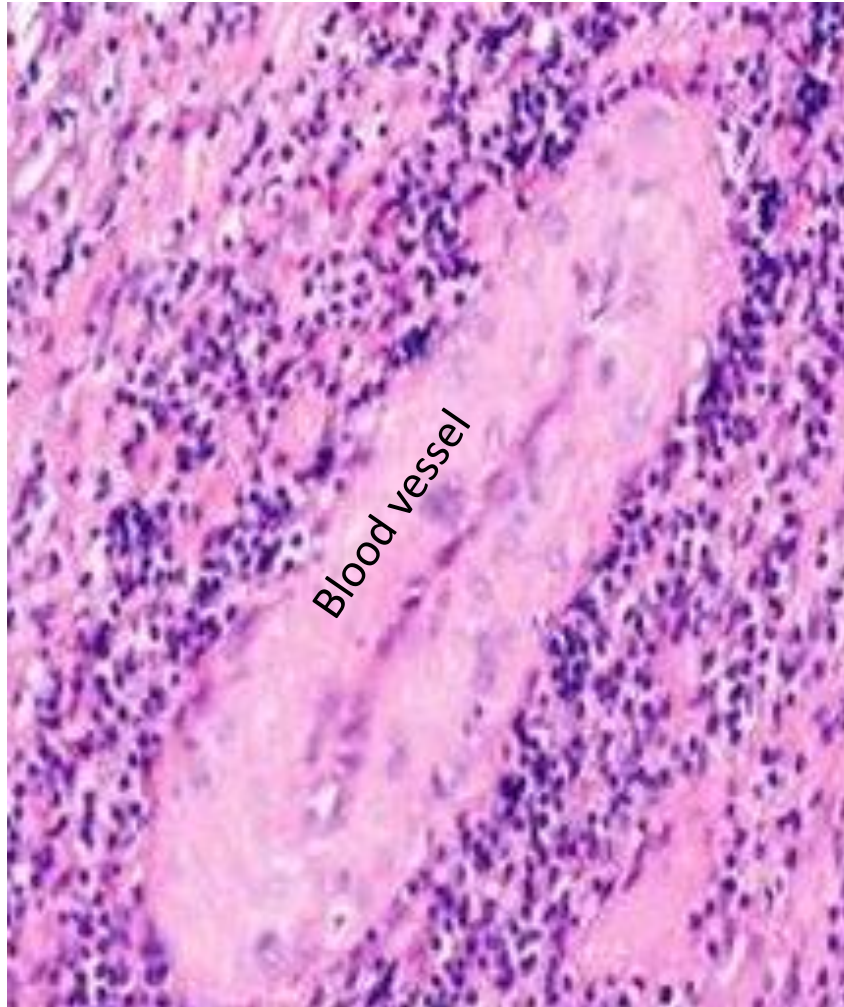
- Uncommon
- Multiple nodules involving the brain parenchyma
- Nodal and extranodal spread is rare
- Immunosuppressed patients - Organ transplant  
- AIDS
- EBV associated
- High grade NHL- B cell
- Poor prognosis

# Lymphoma - Microscopy



Note: Sheets of atypical lymphoid cells

# CNS - Lymphoma



Note: Atypical lymphocytes around a blood vessel (angiocentric)

# Nerve sheath tumours

## Schwannoma

- Encapsulated
- Cerebello pontine angle - 8<sup>th</sup> cranial nerve  
acoustic neuroma

## Neurofibroma

- Non-encapsulated
- Dorsal nerve roots of the spinal cord

# Schwannoma - Macroscopy





# Acoustic neuroma



# Metastatic tumours

- Commoner than primary tumours in adults
- Commonly carcinomas metastases from
  - Breast
  - Lung
  - Kidney
  - Colon
  - Melanoma
  - Lymphoma
  - Sarcoma
  - Germ cell tumours

# Metastatic deposits - Macroscopy



eg. Malignant melanoma

Note: Multiple , discrete nodules in the grey – white matter interphase

# CNS tumours - Prognosis

- Histological type
- Tumour margins
- Site
- Size
- Age

# Raised ICP



# Raised ICP - Objectives

- Causes of raised ICP
- Effects of raised ICP

Upper lid  
of left eye

Arachnoid matter

Cerebral cortex covered by  
pia matter

Dura matter

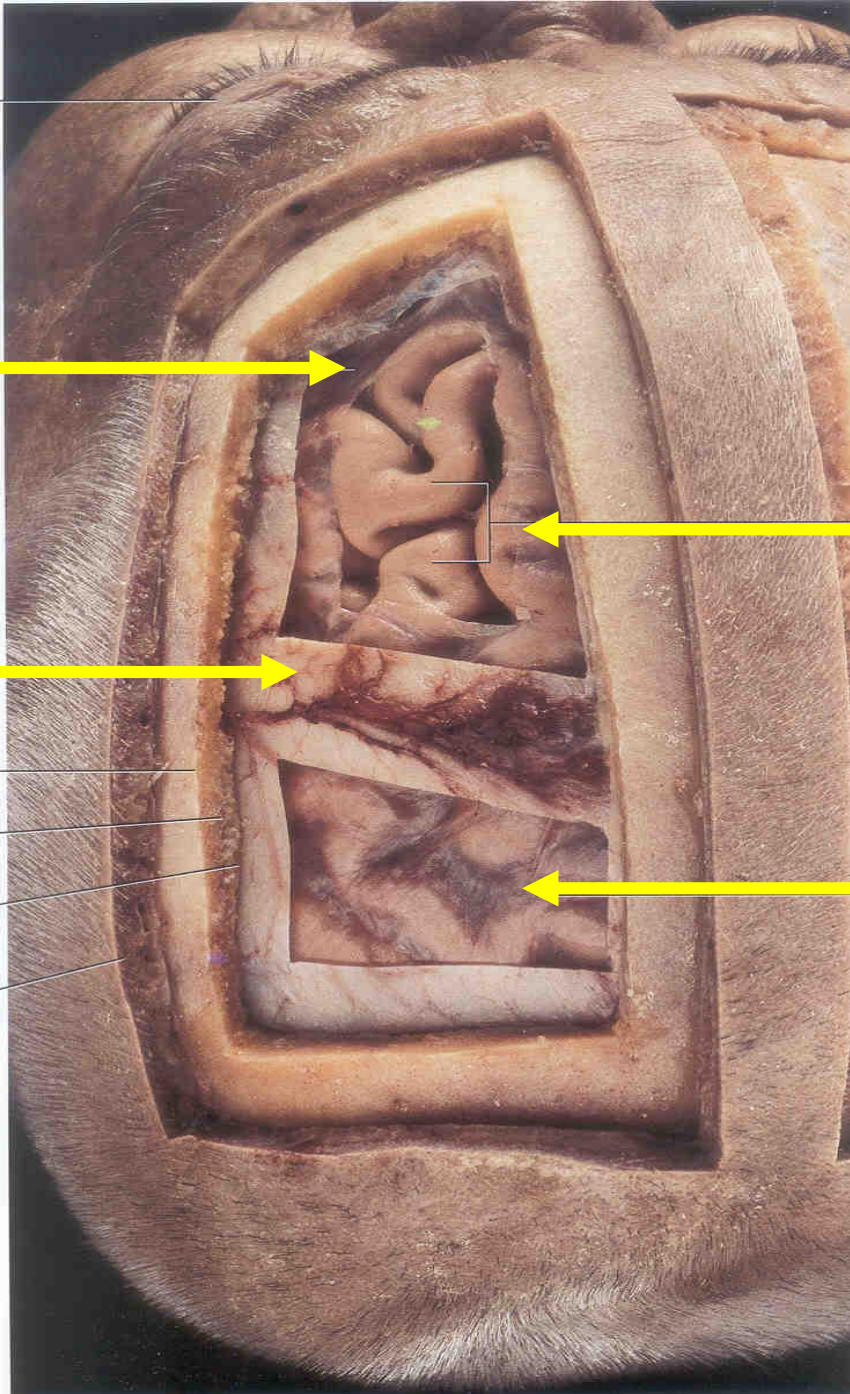
Outer table  
of skull

Diploë of skull

Inner table  
of skull

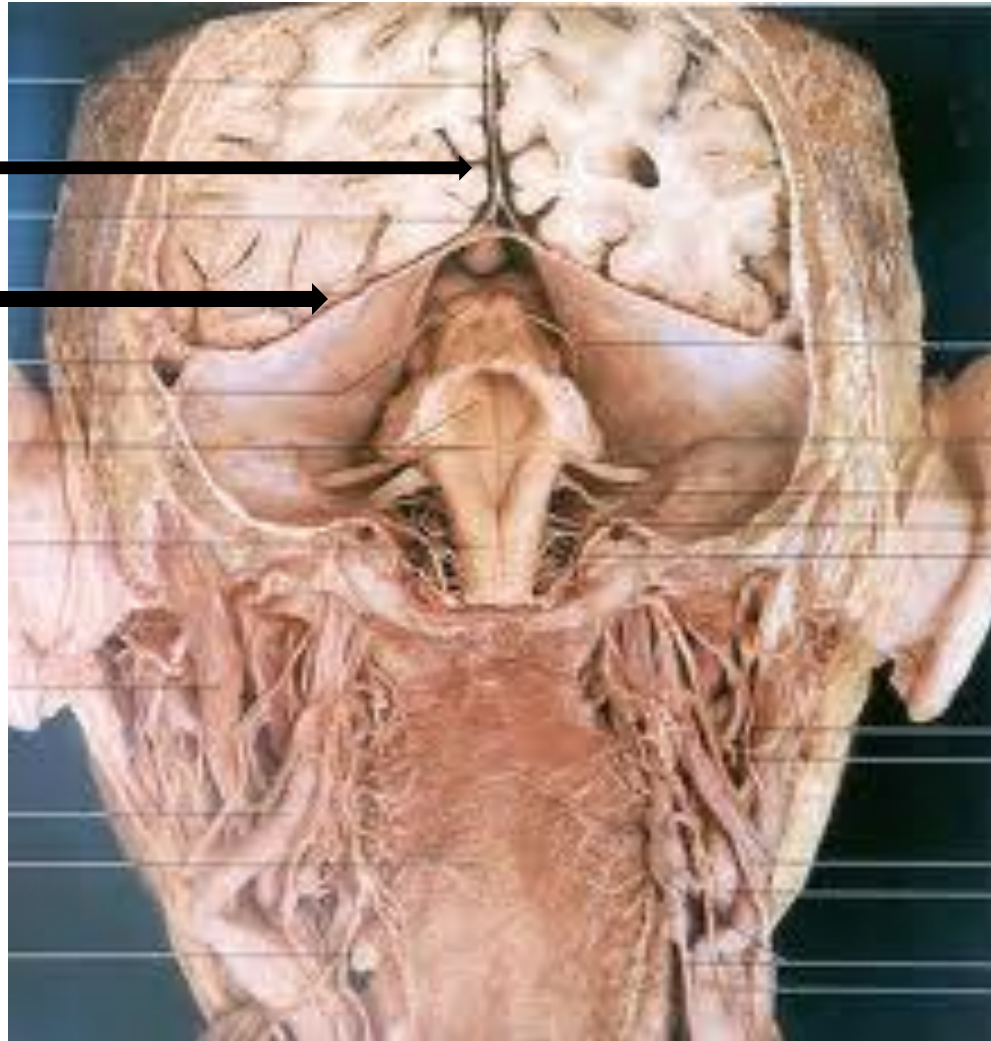
Skin and dense  
connective tissue

Arachnoid matter



Falx cerebri

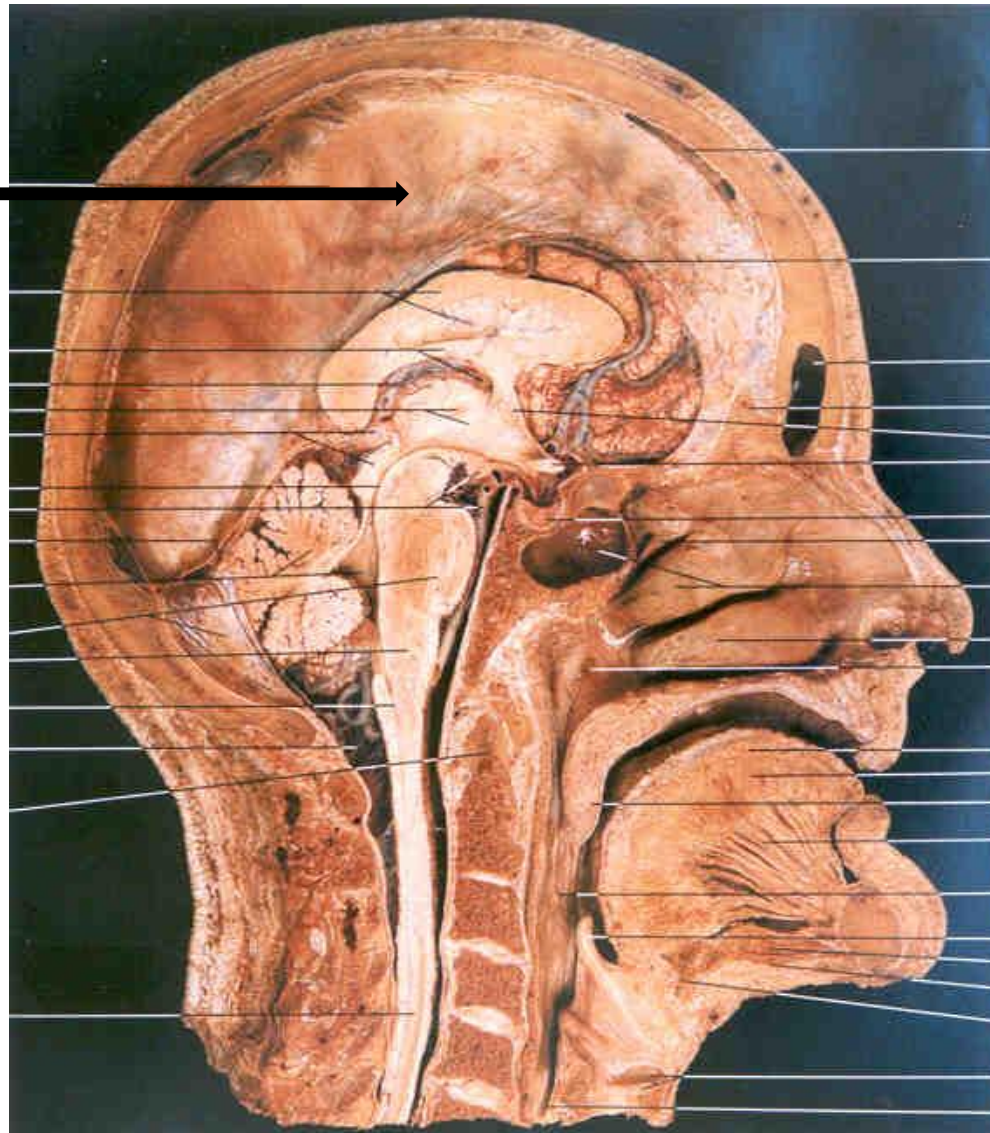
Tentorium cerebri



Posterior view

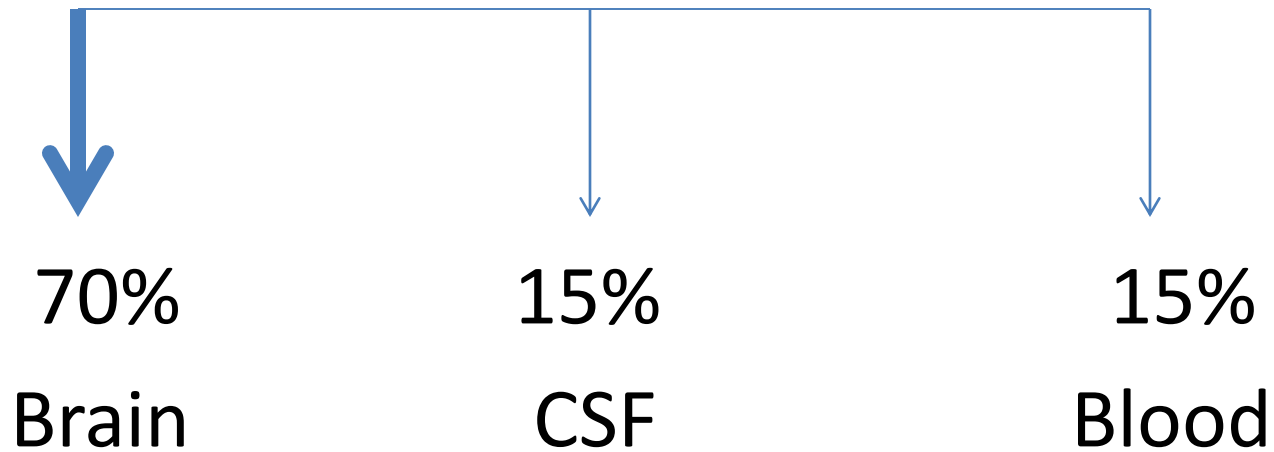


Falx cerebri



Lateral view

# Intracranial contents





# Raised intracranial pressure

## 1) Space occupying lesions

- Neoplasm

- Abscess

- Haemorrhages

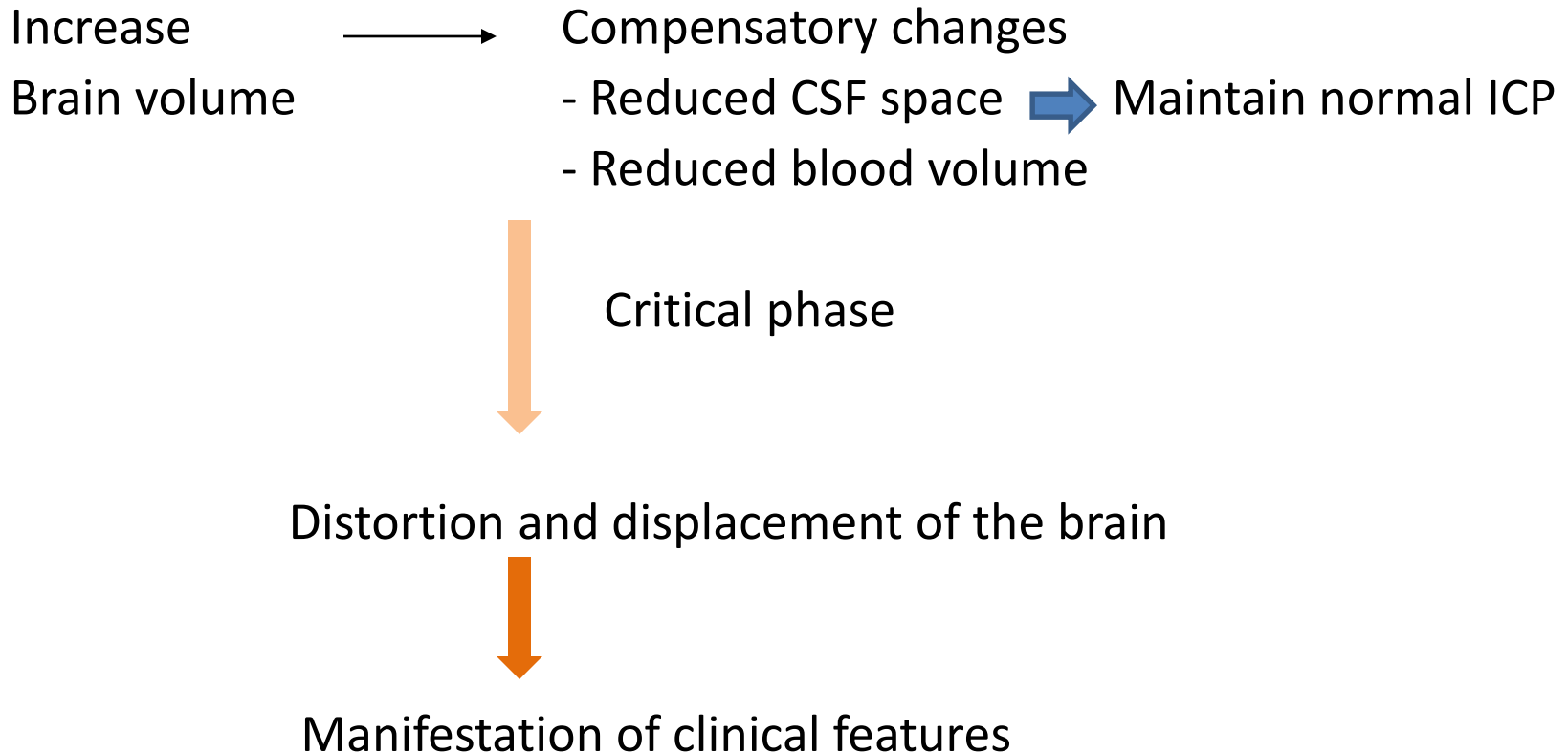
## 2) Oedema – Diffuse / focal

- Infarction

- Infections

## 3) Obstructive hydrocephalus

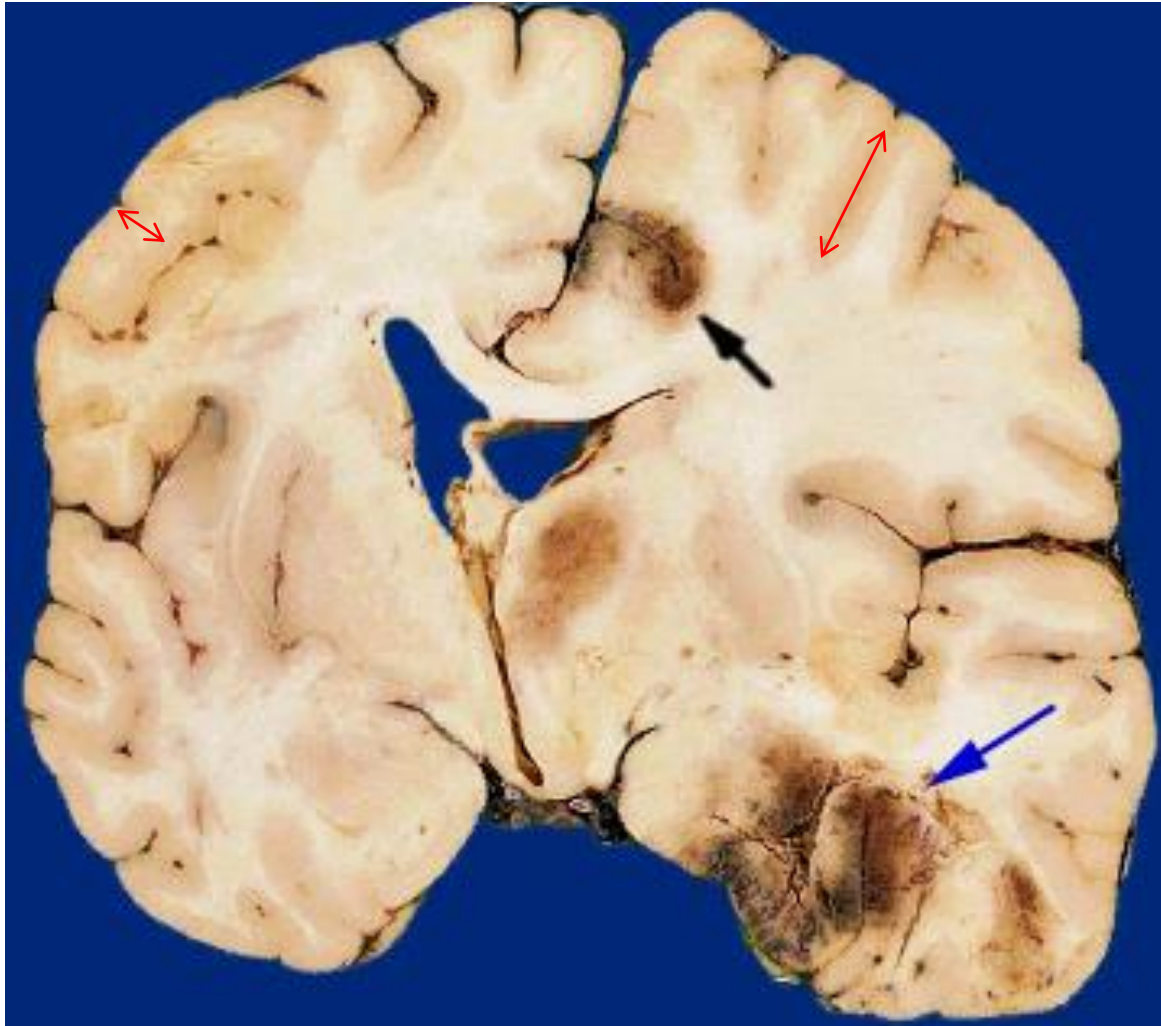
# Raised intracranial pressure



# Effects of raised intracranial pressure

- Narrowing of sulci
- Flattening of gyri
- Surface of the brain looks dry
- Compression of the ipsilateral lateral ventricle
- Shift of midline structures
- Internal brain herniation
  - Transtentorial hernia
  - Subfalcine hernia
  - Tonsillar hernia

# Macroscopy

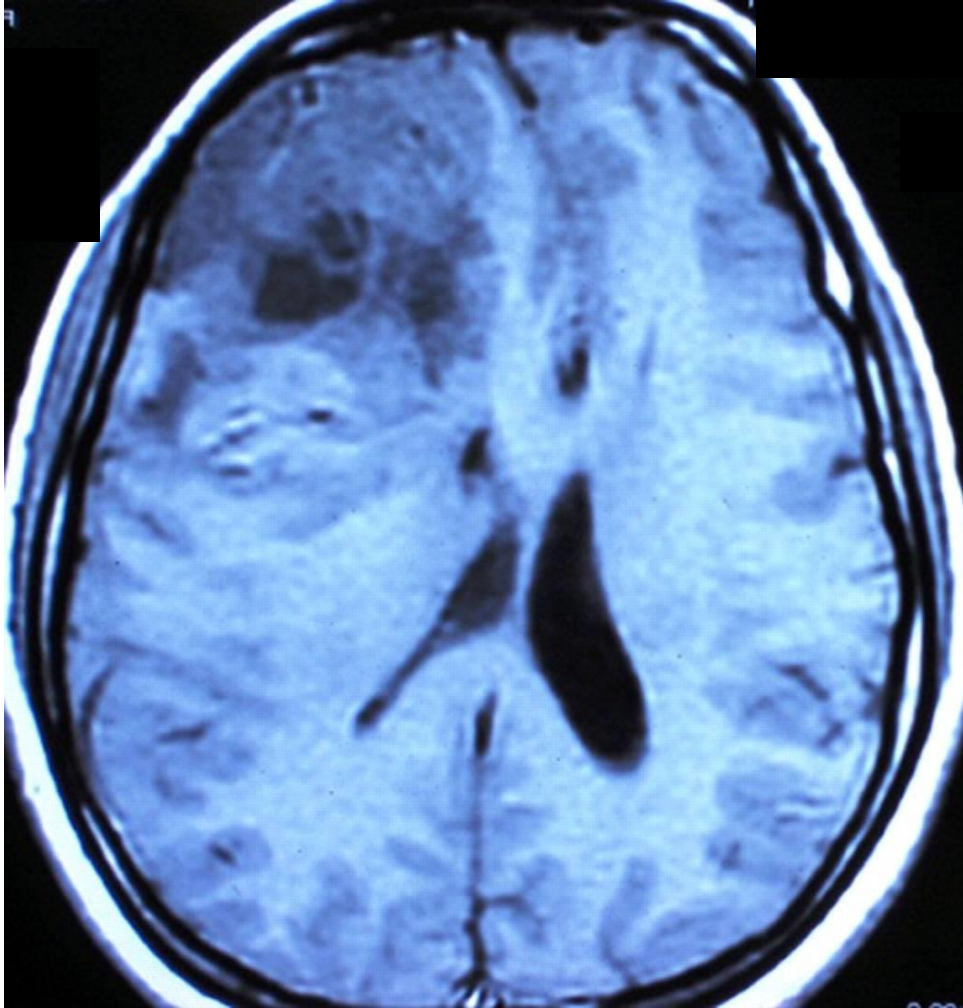


Note:

Narrowing of sulci

Flattening of Gyri

# Brain CT scan



Note: shift of midline structures



# Macroscopy

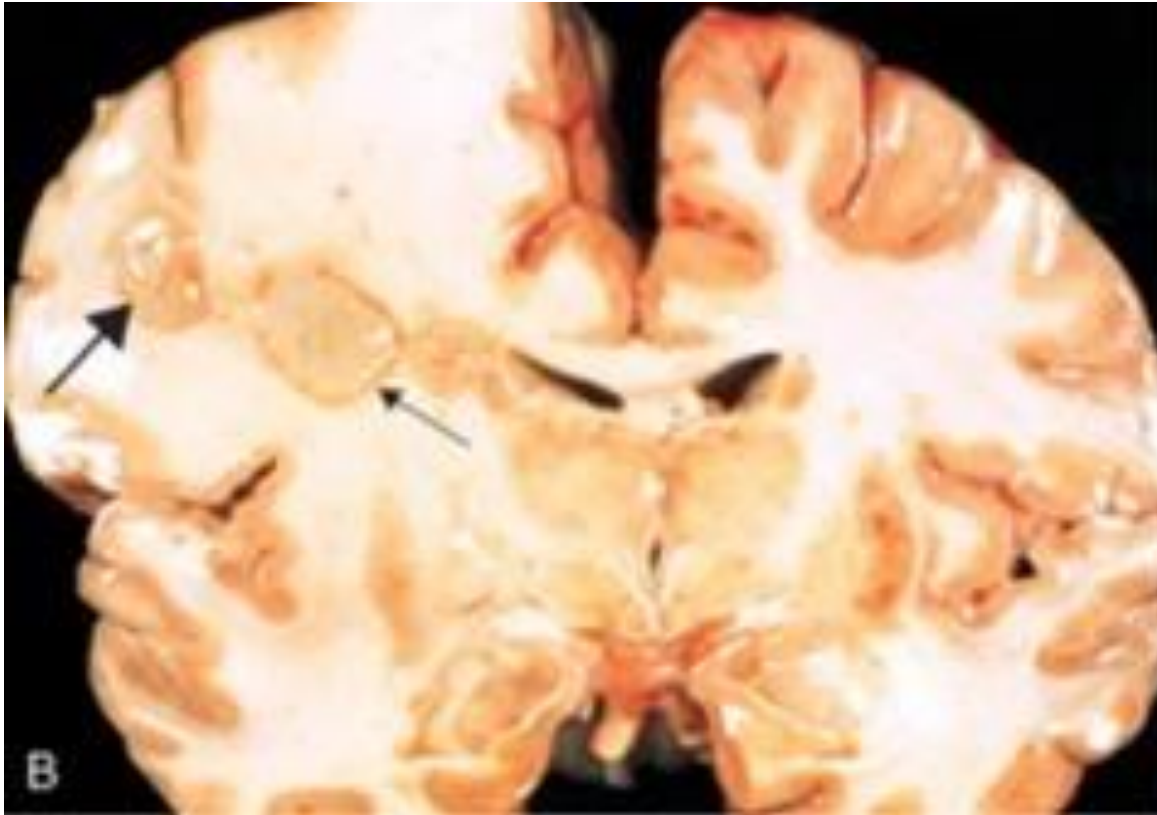


Note:

Shift of midline structures

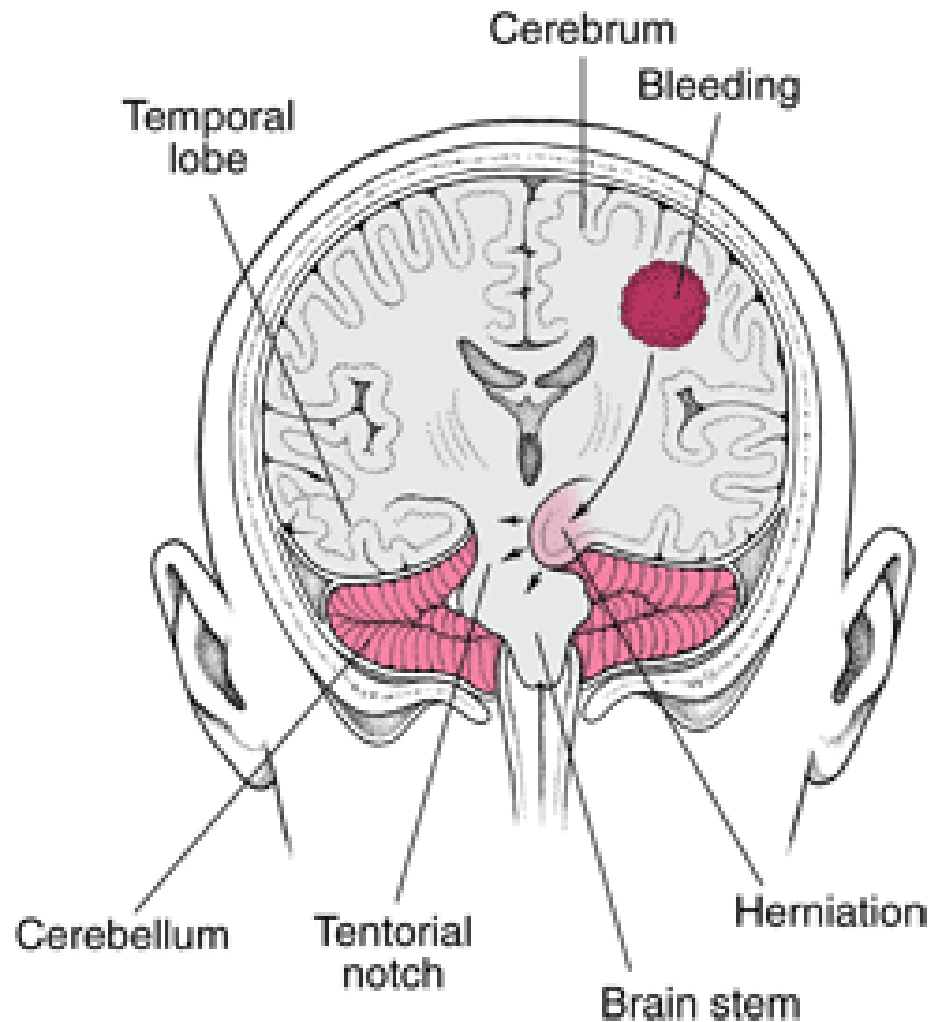
- interventricular septum
- anterior cerebral arteries
- third ventricle

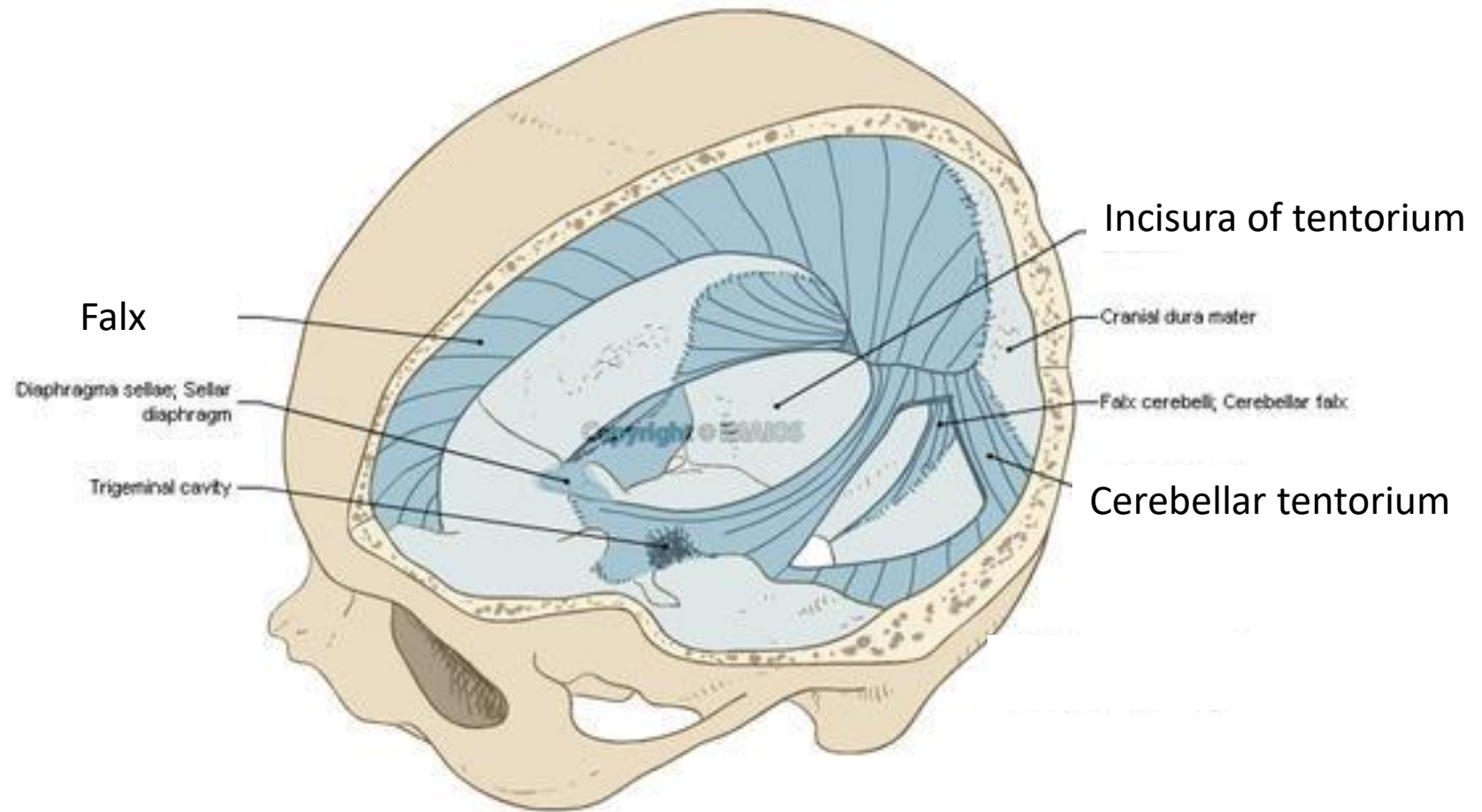
# Macroscopy



Multiple brain abscesses

# Herniation of brain tissue





# Skull and dura

# Transtentorial herniation

- Ipsilateral 3<sup>rd</sup> nerve compression
  - Fixed dilated pupils
- Ipsilateral 6<sup>th</sup> nerve compression
  - Lateral rectus palsy
- Brain stem compression and haemorrhages  
(duret haemorrhages)
  - Cardiorespiratory failure and death

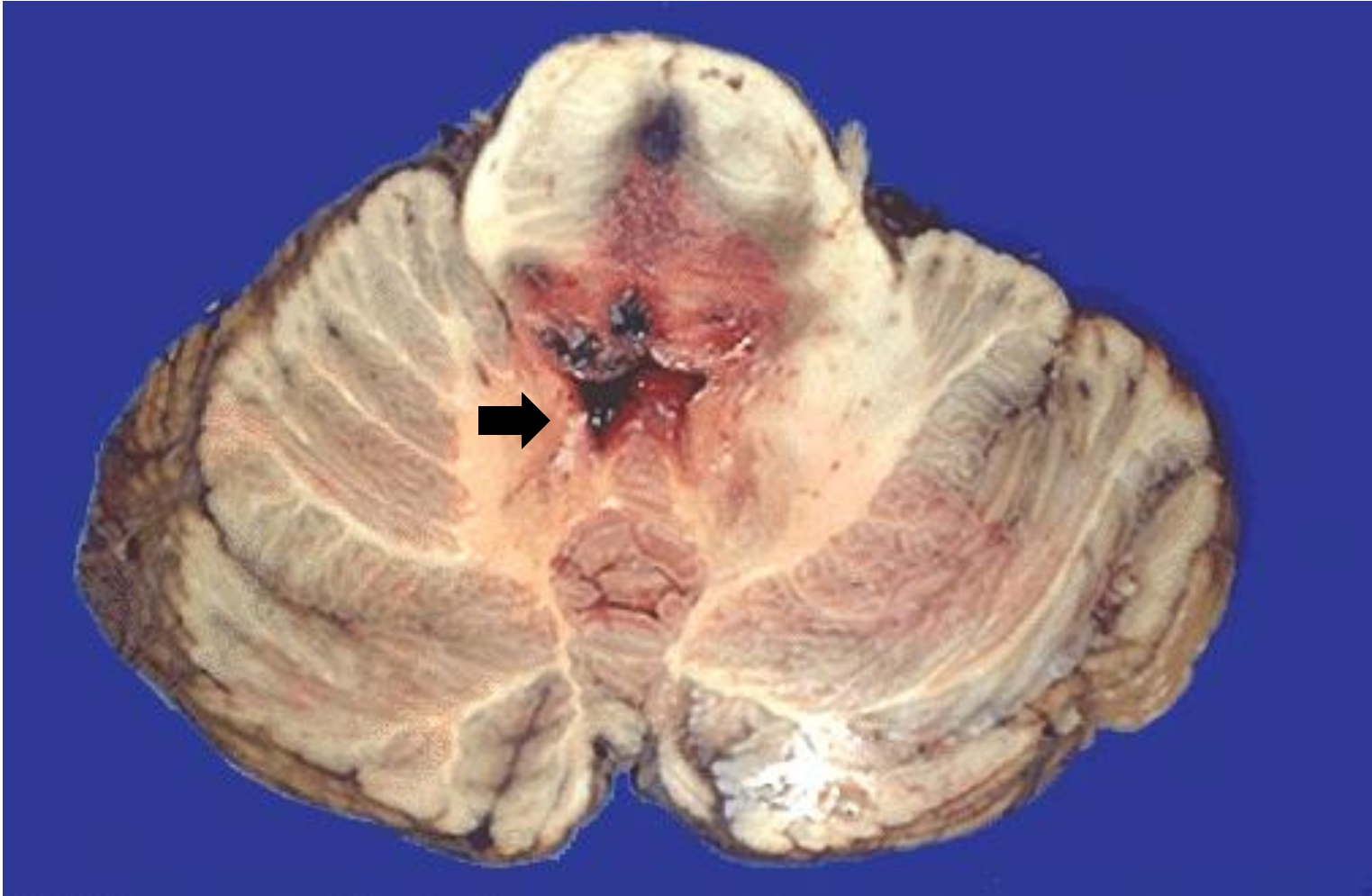


# Transtentorial herniation

- Caused by mass effect in the cerebrum which pushes the supratentorial brain through the incisura to the posterior fossa
- Occurs when the brain traverses across the tentorium at the level of the incisura
- Medial part of the ipsilateral temporal lobe is squeezed through the tentorial opening
- Midbrain is pushed against the contralateral rigid edge of the tentorium - often sufficient to produce a groove on the surface of the mid brain



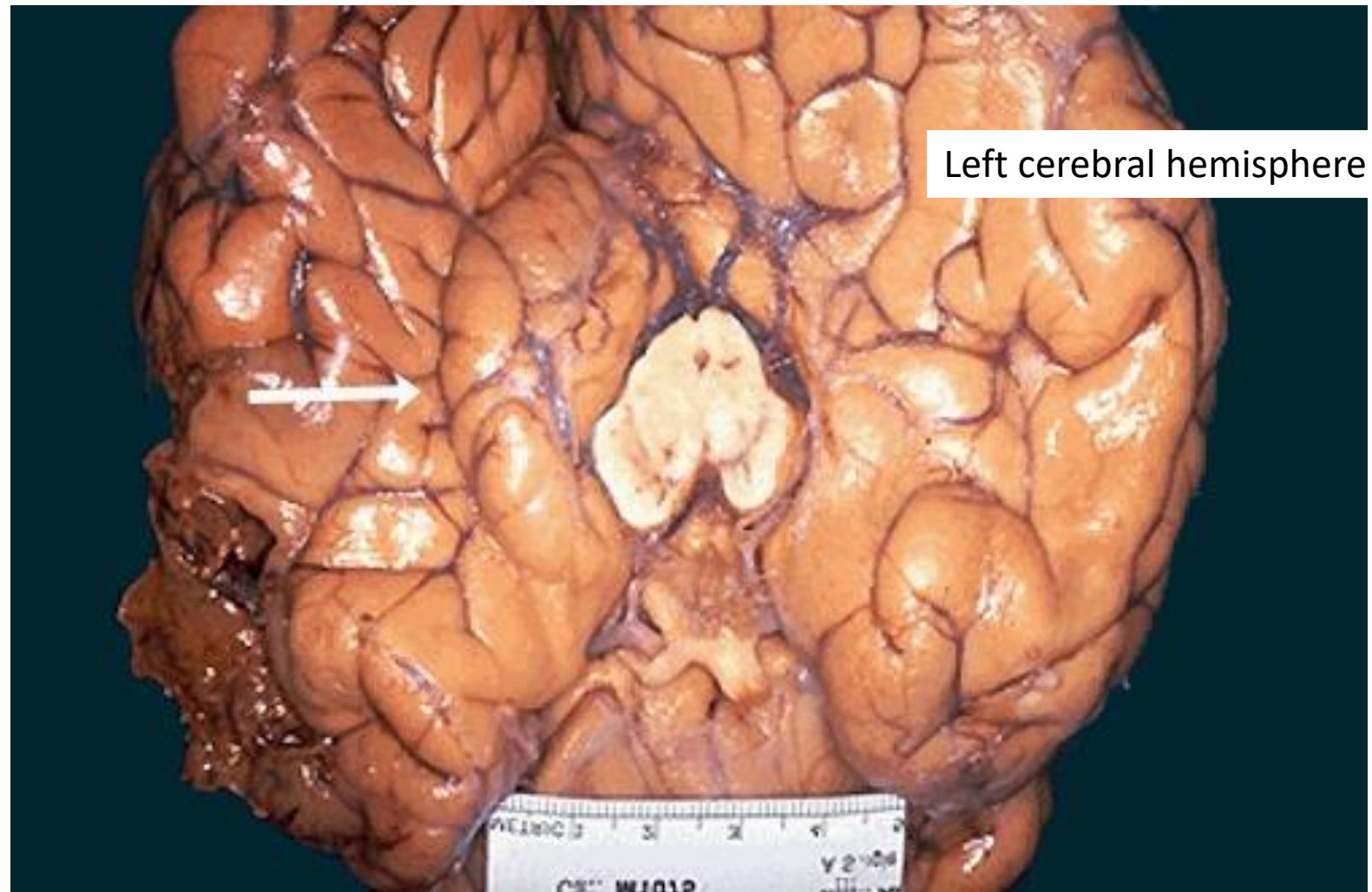
# Duret haemorrhages



Haemorrhages in pons and medulla during rapid herniation of brain

# Transtentorial herniation

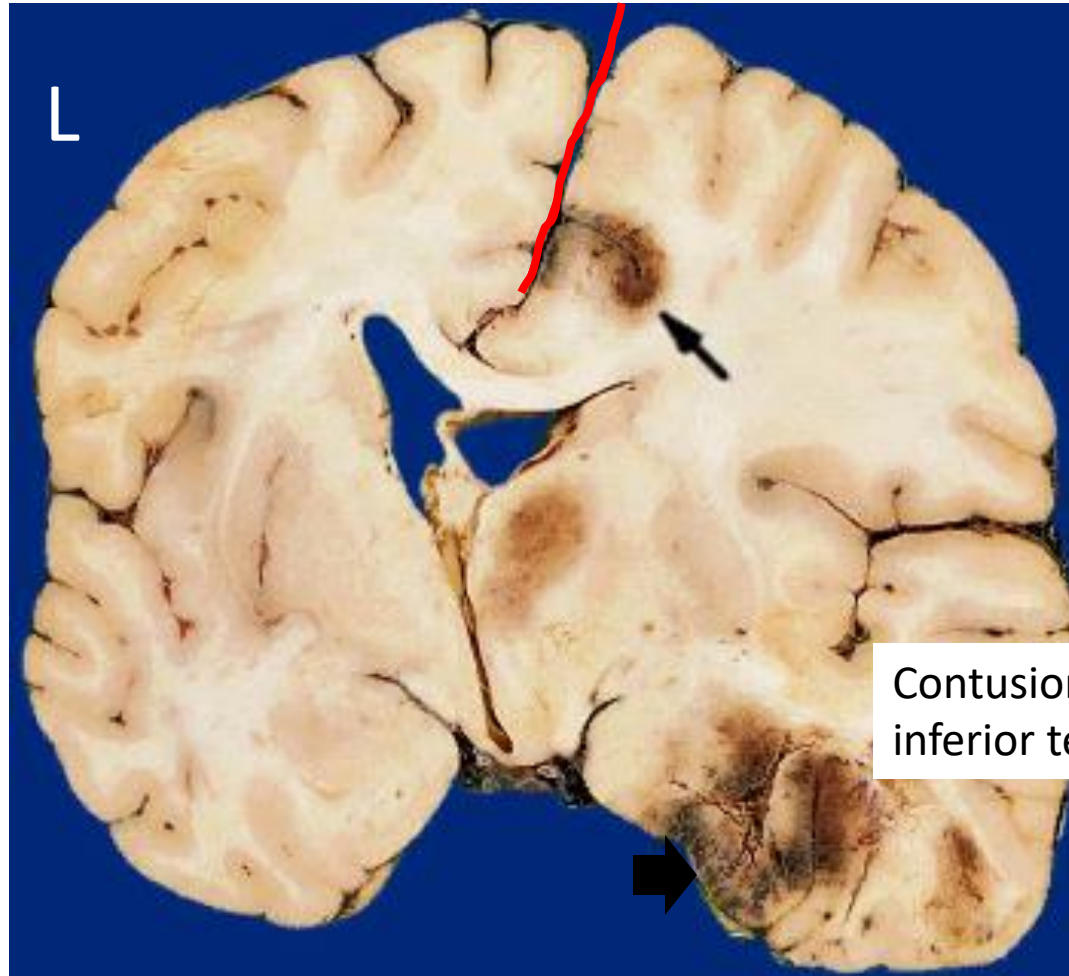
- Kernohan notch
  - Displace and compress the midbrain against the contralateral ridge of the edge of the tentorium.
  - Formation of a groove and haemorrhage on the contralateral midbrain



Left cerebral hemisphere

- Note :
- Swollen left cerebral hemisphere
  - Herniation of the uncus of the hippocampus through the tentorium
  - Groove on the mid brain (white arrow)

# Subfalcine herniation



Note:

- Diffuse oedema of the brain
- Narrow sulci and flattened gyri (L)
- Herniation of cingulate gyrus
- Secondary haemorrhagic infarct
- Midline shift

Contusion of the  
inferior temporal lobe

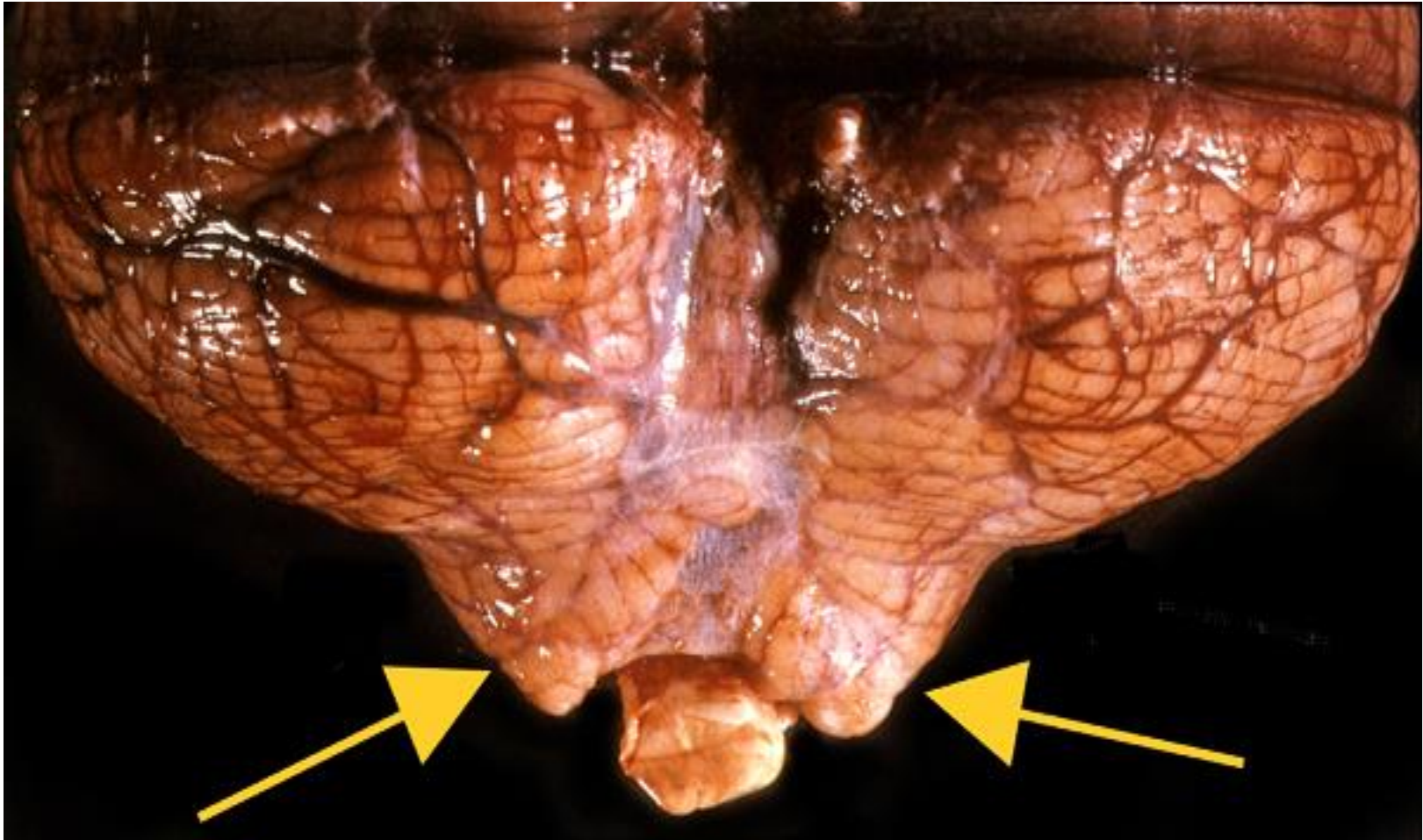
Cingulate gyrus herniates through the free edge of the falx

# Tonsilar herniation

- Brain matter herniates through the foramen magnum
- Common with infratentorial expansile lesions
- Brain stem compression and haemorrhages
  - Cardiorespiratory failure and death
- Can be dramatic if a lumbar puncture is done with an undiagnosed mass lesion



# Tonsilar herniation



Note: Grooved cerebellar tonsils by compression against the foramen magnum (arrows)



# Summary

- Classification of CNS tumours
- Morphology of common tumours of the CNS
- Causes of raised ICP
- Effects of raised ICP

# Reading assignments

- Clinical manifestations of CNS neoplasm
- Clinical manifestations of raised ICP
- Cerebral oedema
- Hydrocephalus