

# Tumors of the Central Nervous System-2



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# Neuronal Tumors



Far less common than gliomas.



Typically, lower-grade lesions, often present with seizure. Composed of cells that express neuronal markers; synaptophysin & neurofilaments.



Tumors composed of cells with neuronal features.

Examples: Gangliogliomas, central neurocytoma

## Embryonal(Primitive) Neoplasms



Tumors with Primitive “small round cell” appearance that is reminiscent of normal progenitor cells encountered in the developing CNS.



The most common is the medulloblastoma, accounting for 20% of pediatric brain tumors

# Medulloblastoma

## Age

Occurs predominantly in children

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## Location

**Exclusively** in the cerebellum:  
Children, often midline.  
Adults: more lateral tumors

02

## Genetics

**P53** mutant or **SHH** activated worst prognosis.  
**WNT** pathway activation: best prognosis.

03

## Gross

Often well circumscribed, gray, & friable.

04

## WHO grade (IV)

Highly malignant tumor, dismal prognosis if untreated.  
But exquisitely radiosensitive.

05

## Prognosis

With total excision, chemotherapy & irradiation → 5-year survival rate ~ 75%

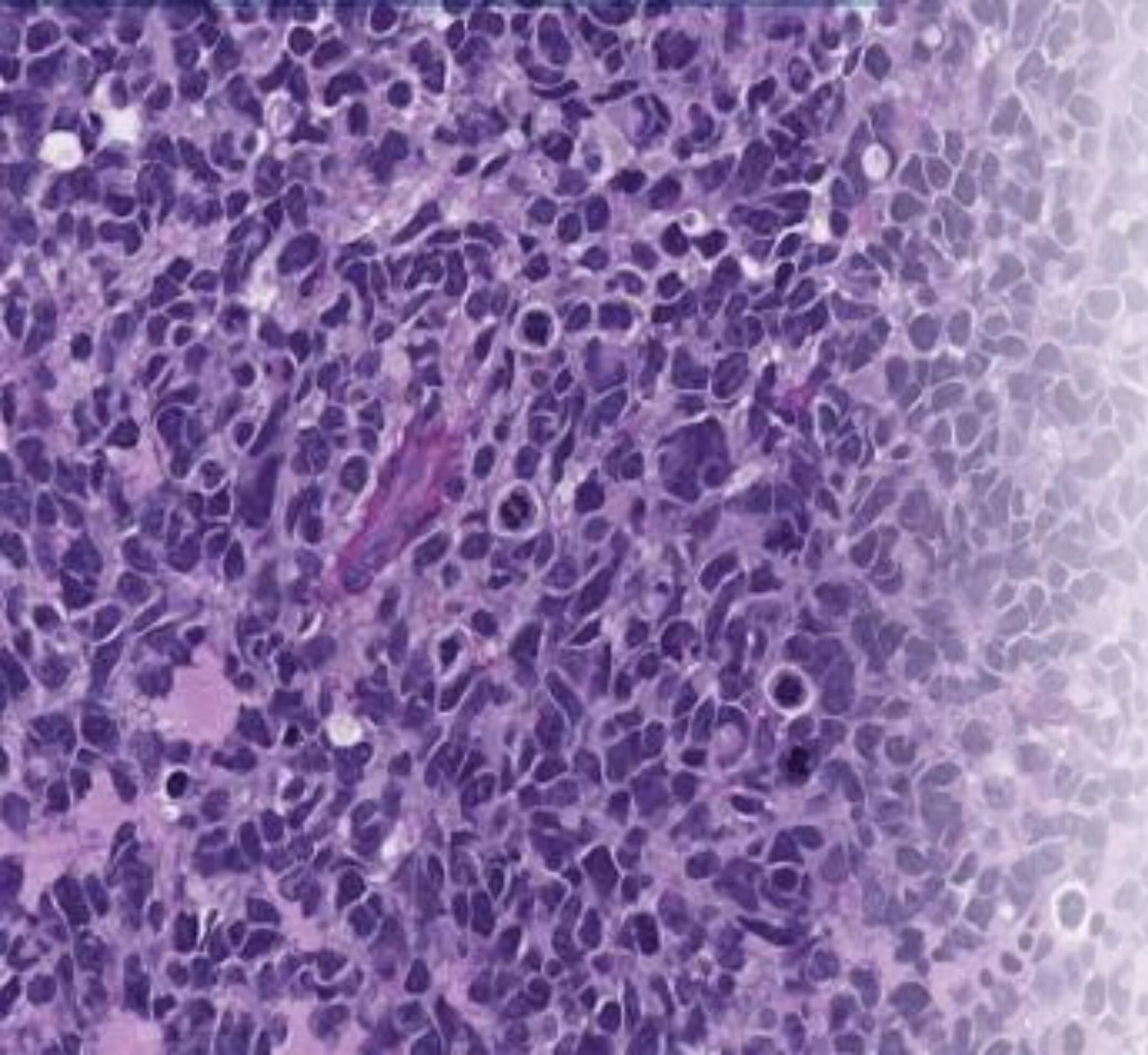
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## Medulloblastoma- Gross

often well circumscribed, gray, & friable.

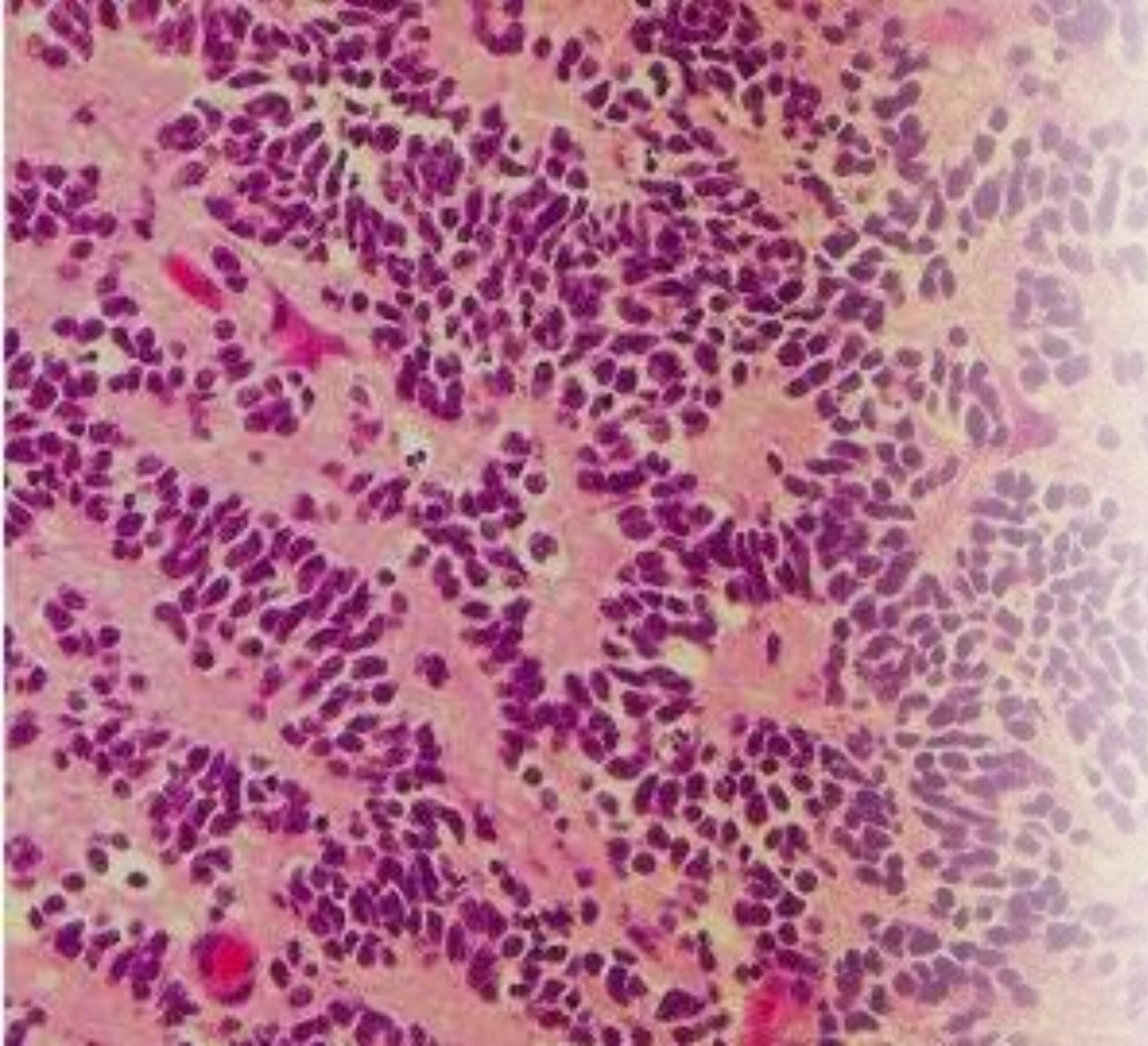
Maybe Extending to the surface of the cerebellar folia & leptomeninges



## Medulloblastoma- Morphology

Densely cellular tumor → with sheets of anaplastic (“small blue cells”), with little cytoplasm and hyperchromatic nuclei.

Mitoses are abundant.



# Medulloblastoma- Morphology

- Often, focal neuronal differentiation is seen in the form of rosettes.
- Resemble the rosettes encountered in neuroblastomas
- Primitive tumor cells surrounding central neuropil (delicate pink material formed by neuronal processes).
- Called Homer Wright Rosettes



# Clinical presentation and consequences

- Acute symptoms of posterior fossa tumors in children → result of increased intracranial pressure (mass effect & obstructive hydrocephalus) → headaches, nausea, emesis and cranial neuropathies. Ataxias are also frequent.
  - Radiotherapy is usually avoided in patients younger than three years, due to the deleterious impact on the developing brain.
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# Posterior fossa syndrome (PFS)

- Acute consequence in ~ 25% of children after tumor resection in the posterior fossa; postoperative mutism, ataxia, hypotonia, emotional lability and behavioral symptoms. Recovery of PFS is slow & often incomplete → long-term symptoms: reading deficits, lower intellectual ability, psychosocial complaints, & lower quality of life in general
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# Primary Central Nervous System Lymphoma

## Type

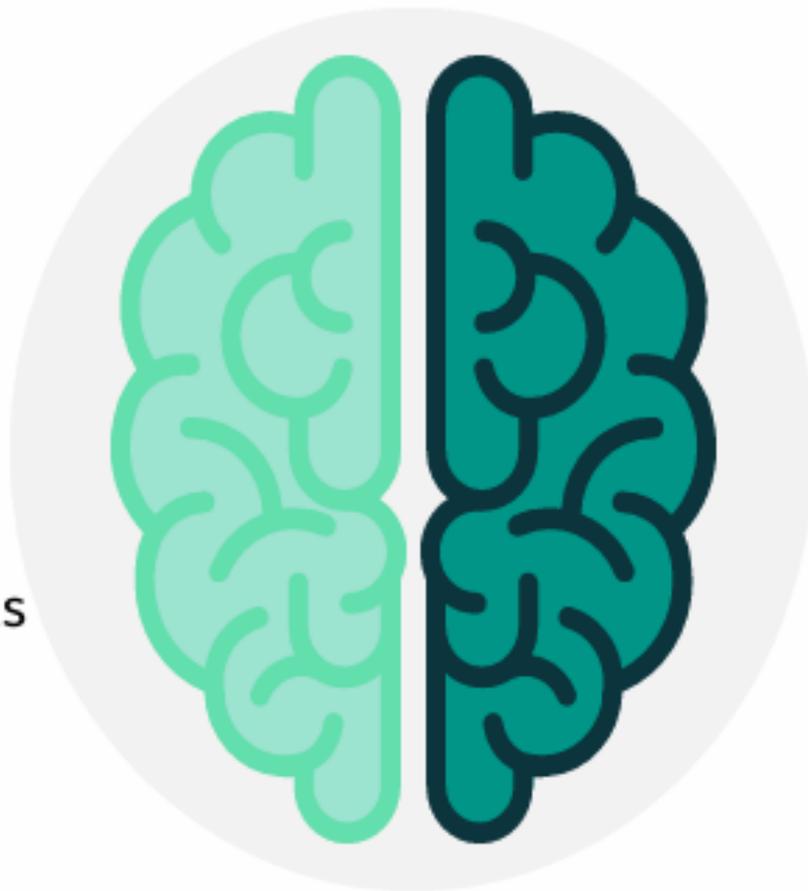
Occurring mostly as diffuse large B-cell lymphomas

## Epidemiology

1% of intracranial tumors. It is the most common CNS neoplasm in immunosuppressed individuals

## Prognosis

An aggressive disease with a poor response to chemotherapy compared with peripheral lymphomas.



## Gross/radio

Multiple tumor nodules within the brain parenchyma. Periventricular spread is common.

## Microscopic

Malignant lymphoid cells accumulate around blood vessels & infiltrate the surrounding brain parenchyma.

## Spread

Spreading outside the brain happens rarely. Peripheral lymphoma rarely spreads to the brain.



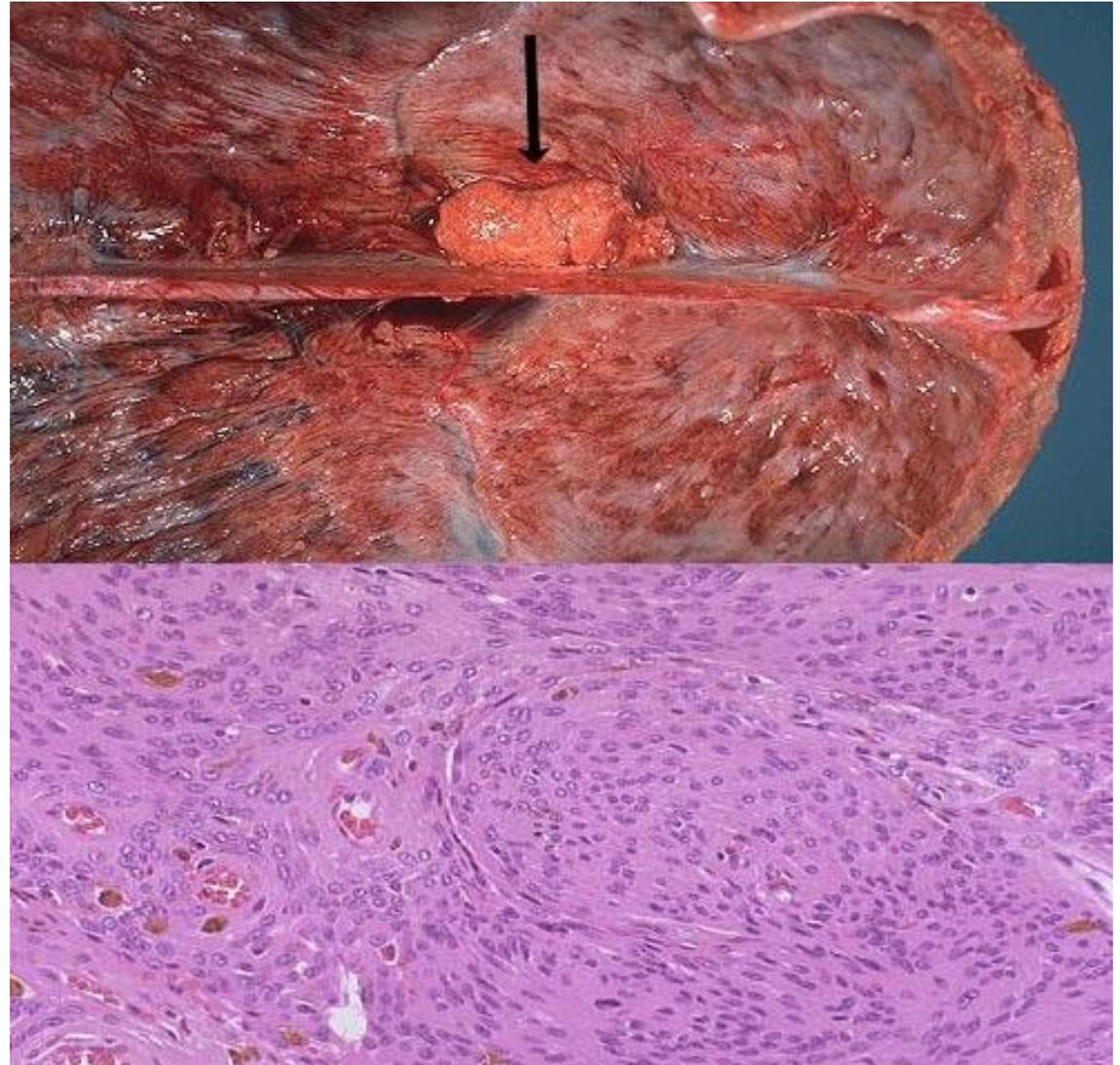
# Meningiomas

- Genetics: Loss-of-function mutations in the NF2 tumor suppressor gene on chromosome 22.
  - Age: Adults.
  - Location: Attached to the dura. along any of the external surfaces of the brain or in ventricular system.
  - Behaviors: Predominantly benign; Most are easily separable from brain. Some are infiltrative, (ass. Recurrence).
  - Gross: often cystic; if solid, it may be well circumscribed. Rarely infiltrative
  - Prognosis: Determined by: size, location, & histologic grade.
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# Meningiomas - WHO grade I

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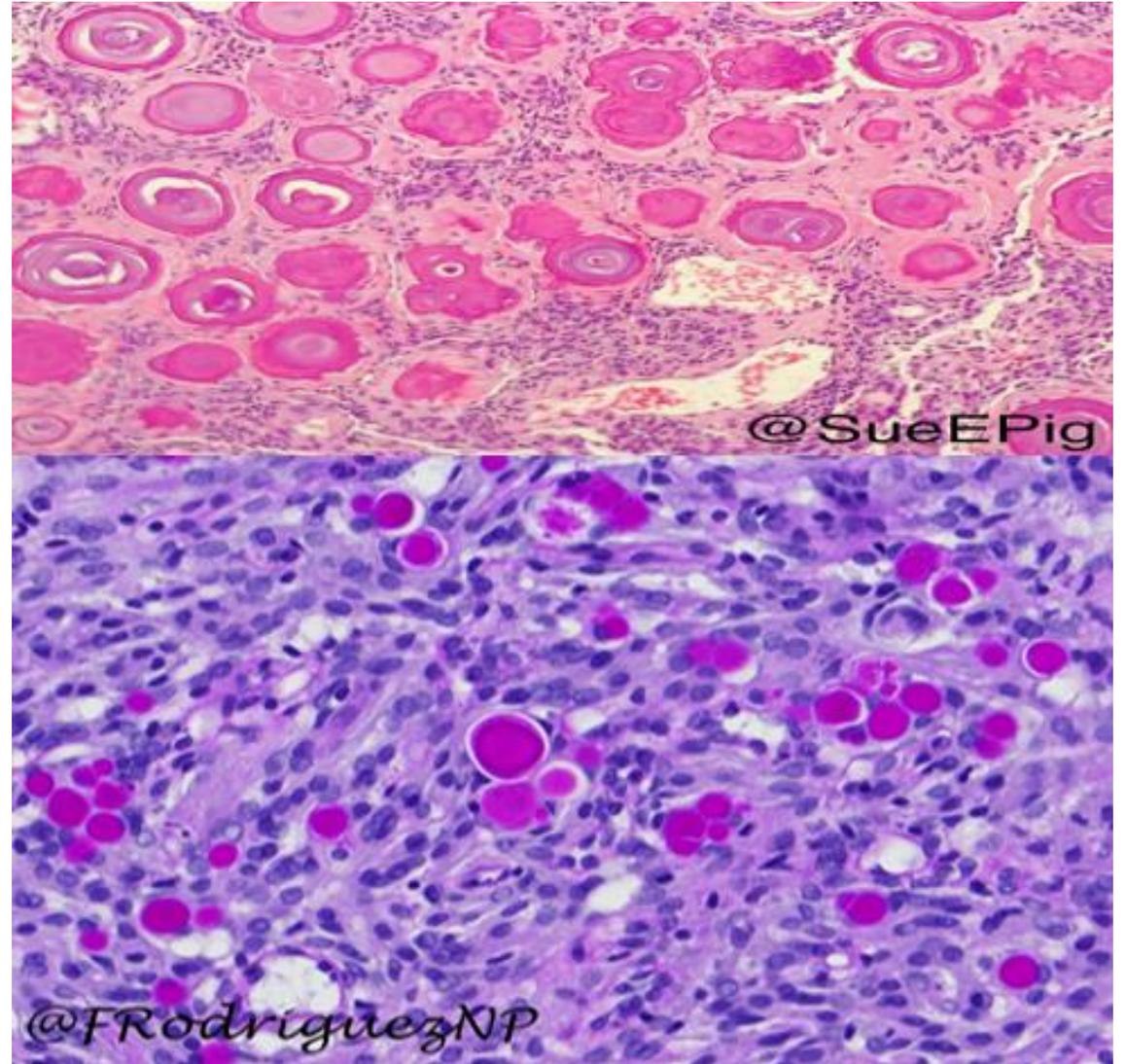
- Well-defined include masses that may compress the brain but no invasion.
- Extension into the overlying bone may be present.
- Variable histologic patterns include, most common meningothelial; named for whorled, tight clusters of cells without visible cell membranes



# Meningiomas - WHO grade I

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- Other variants:
  1. fibroblastic, with elongated cells & abundant collagen
  2. transitional, with features of the meningothelial & fibroblastic
  3. psammomatous, with numerous psammomabodies.
  4. Secretory, with glandlike spaces containing PAS-positive eosinophilic material

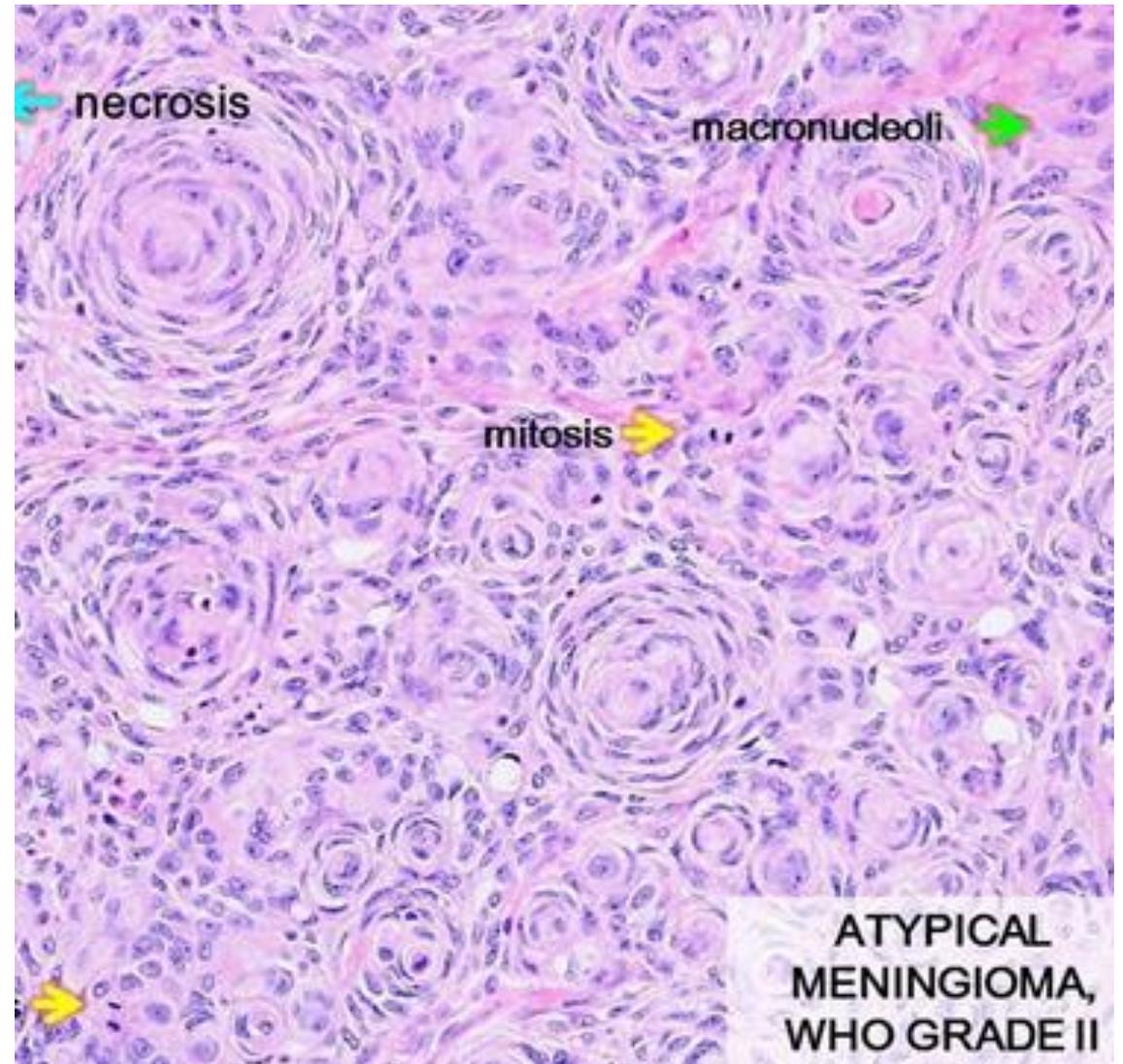


# Meningiomas-WHO grade 2

- Features include:

1. Increased mitotic rate.
2. Prominent nucleoli.
3. Increased cellularity.
4. High nucleus-to-cytoplasm ratio.
5. Necrosis.

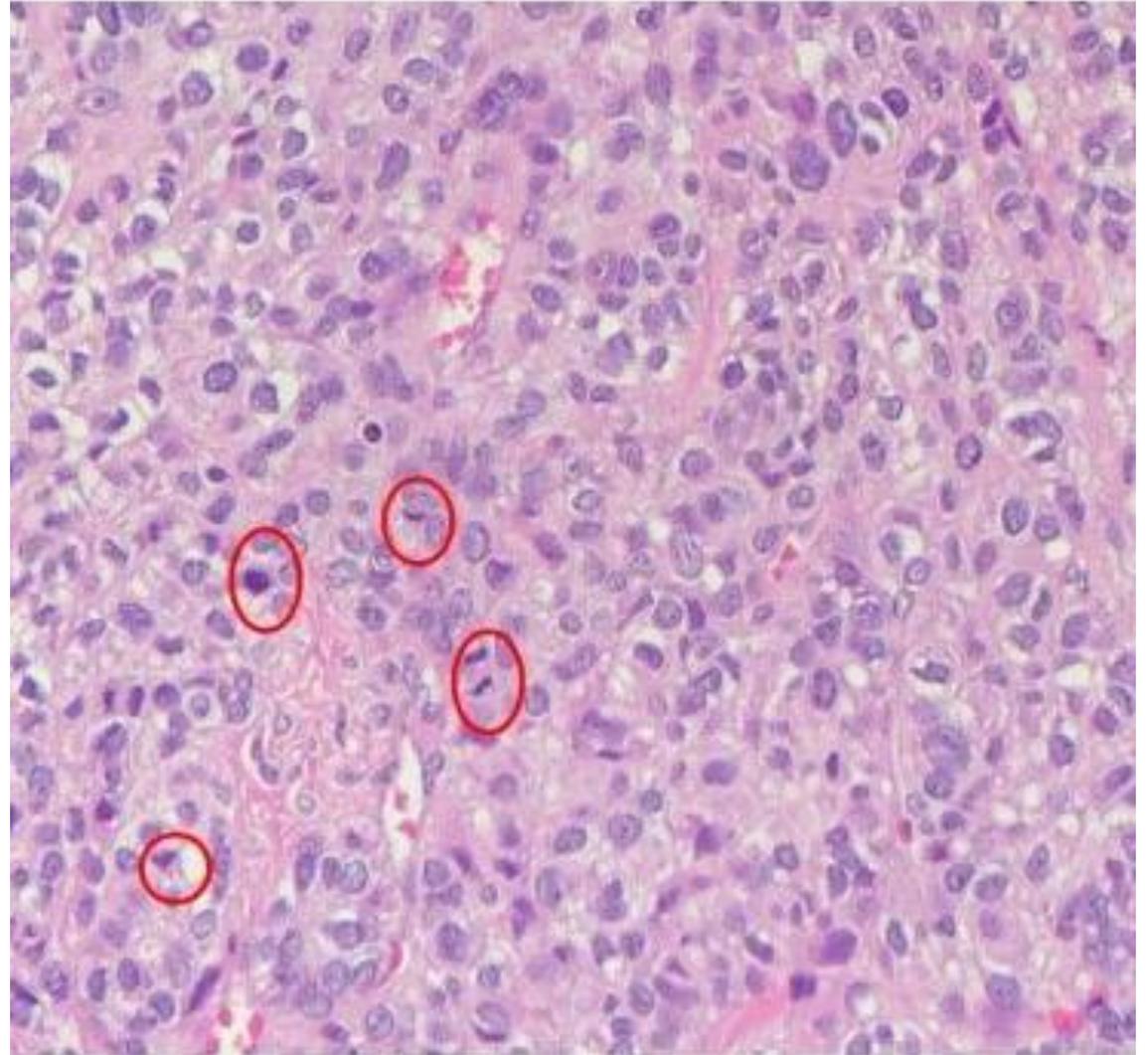
These tumors demonstrate more aggressive local growth and a higher rate of recurrence.



# Meningiomas - WHO grade 3

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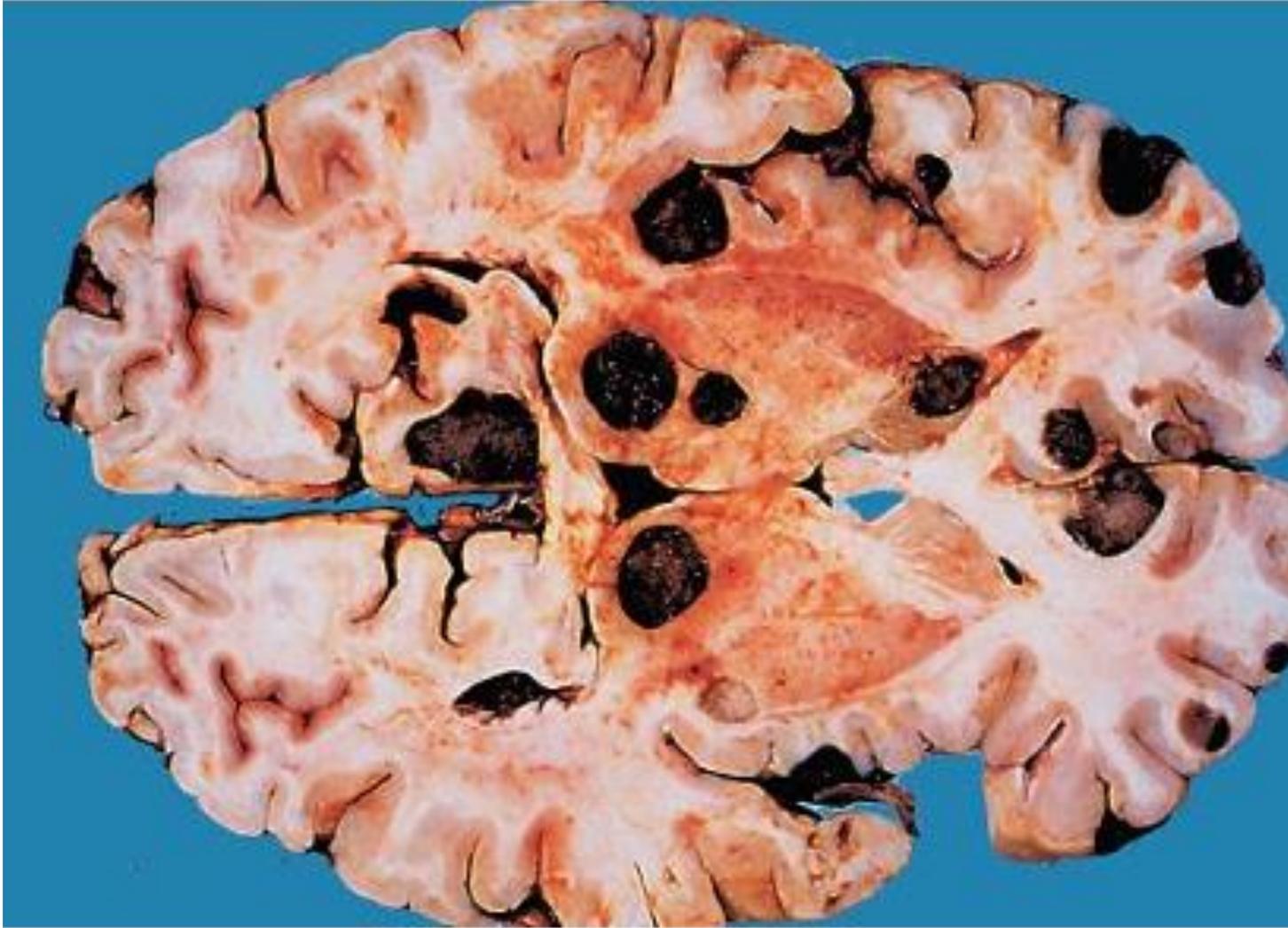
- Highly aggressive tumors that resemble a high-grade sarcoma or carcinoma morphologically.
- Mitotic rates are typically much higher than in grade 2 meningiomas.



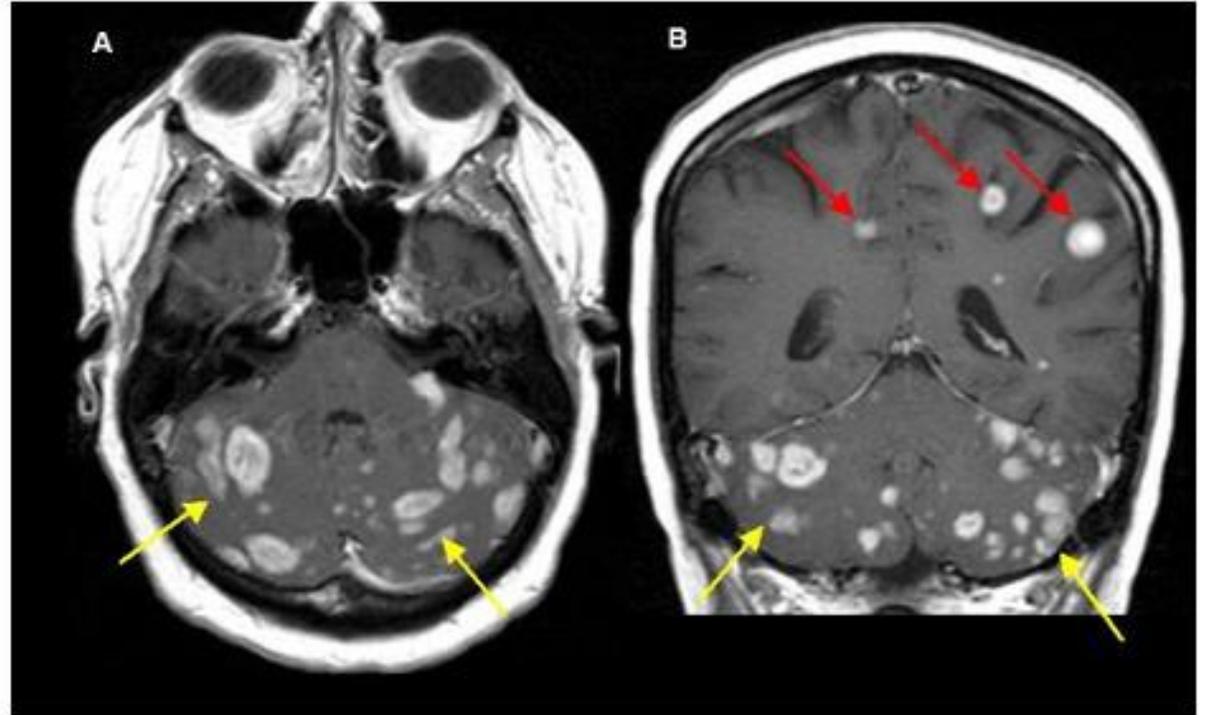
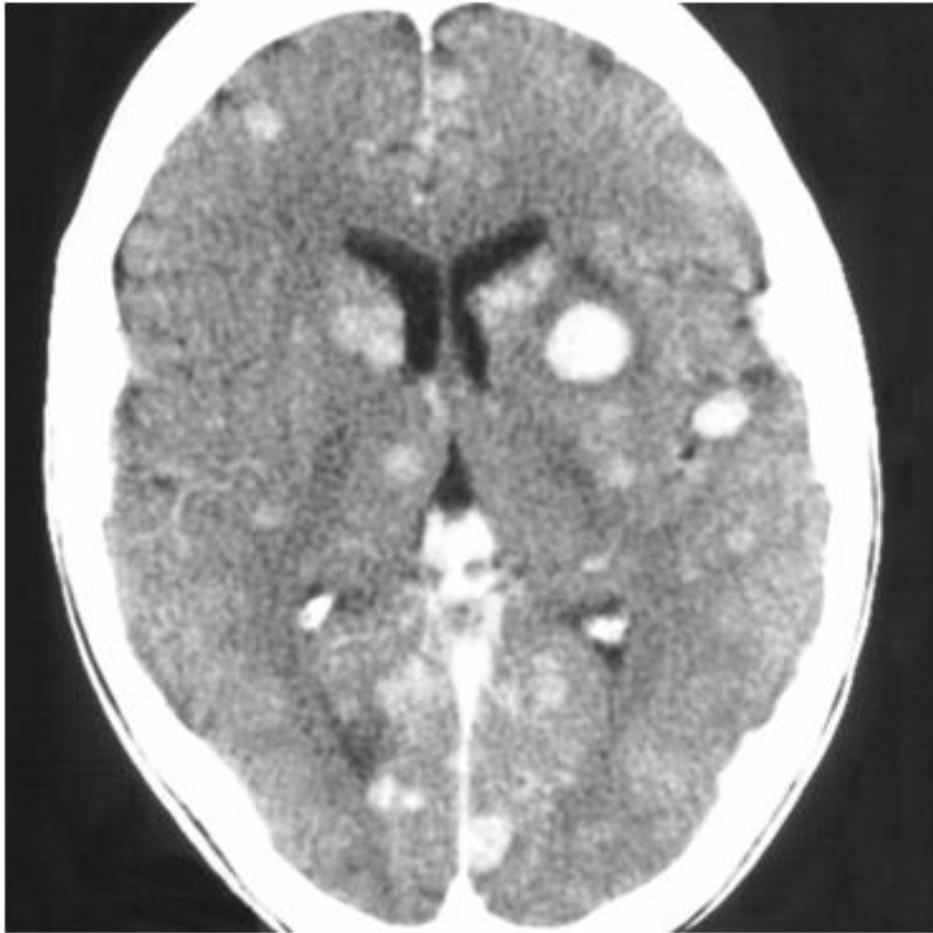


# Metastatic Tumors

- Metastatic lesions, mostly carcinomas.
  - Over half of intracranial tumors.
  - The most common primary sites are lung, breast, kidney, colon, and skin (melanoma).
  - The boundary between Tumor and brain parenchyma is sharp at the gross and microscopic levels
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## Metastatic Tumors



Metastatic Tumors

THANK YOU

