

Tumors of the Central Nervous System II

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12/22/2024

Neuronal Tumors

01

Far less common than gliomas.

Tumors composed of cells with neuronal features. Typically, lower-grade lesions, often present with seizure

02

03

Composed of cells that express neuronal markers; synaptophysin & neurofilaments.

Examples: Gangliogliomas, central neurocytoma

04

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

01

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

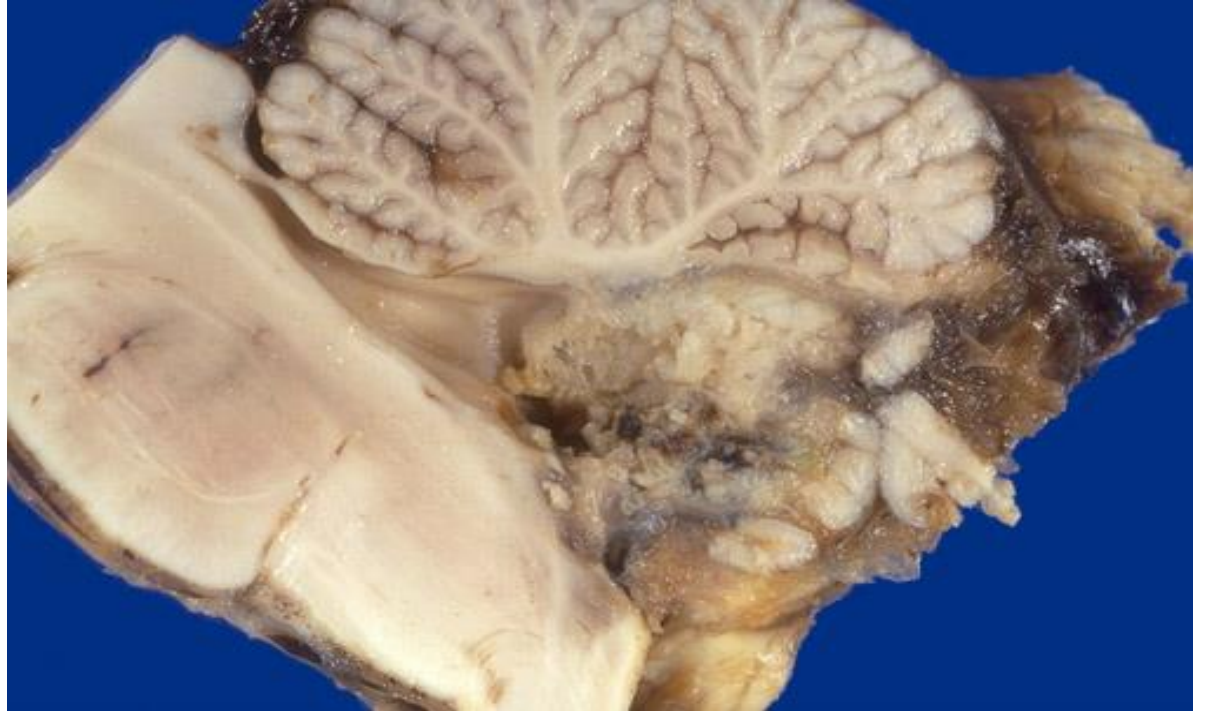
Prognosis

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

06

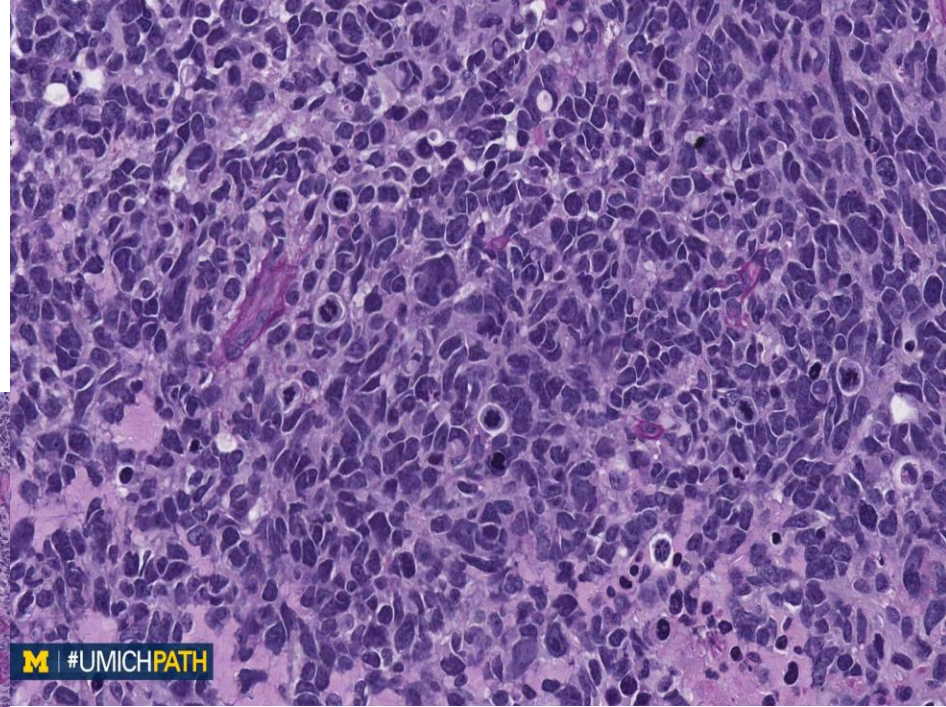
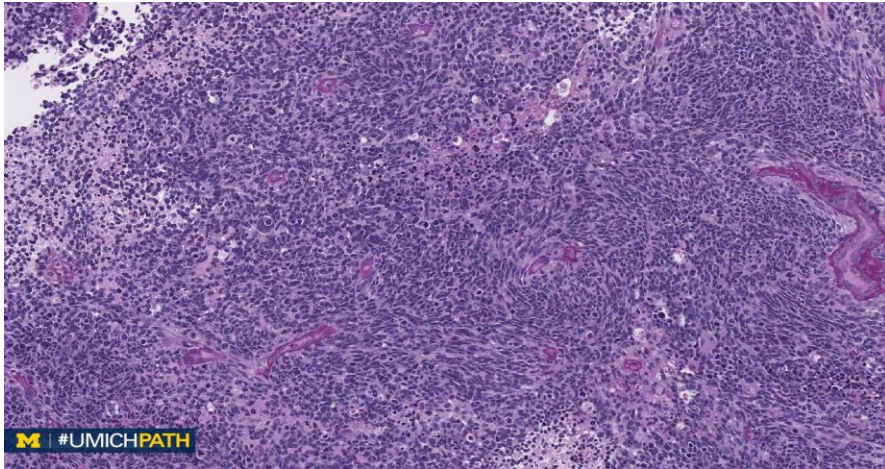
Medulloblastoma - Gross

often well circumscribed, gray, & friable. Maybe Extending to the surface of the cerebellar folia & leptomeninges



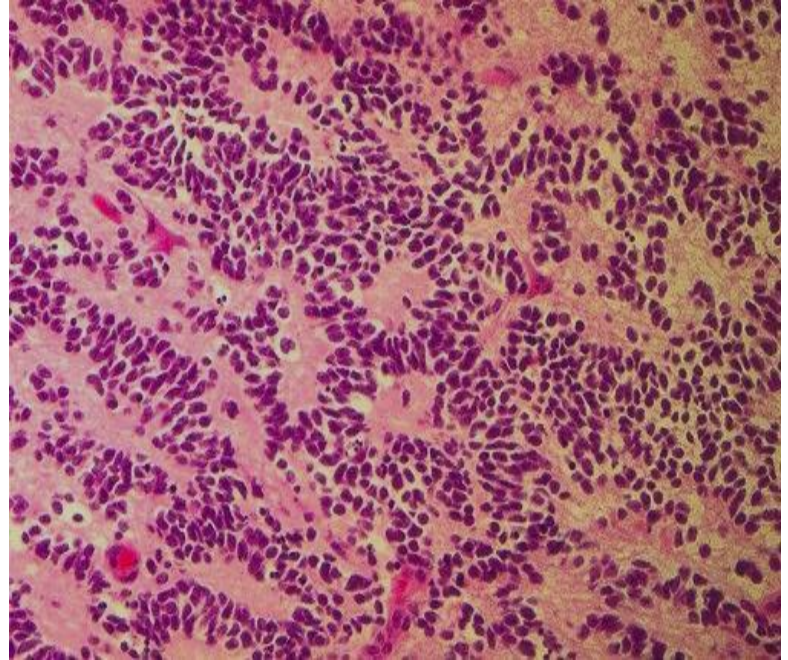
Medulloblastoma - Morphology

- 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.
- 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

Primary Central Nervous System Lymphoma

Type

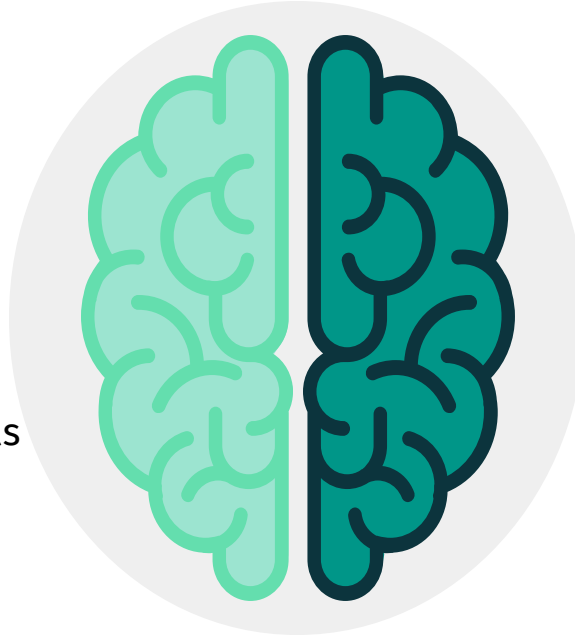
Occurring mostly as diffuse large B-cell lymphomas

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.

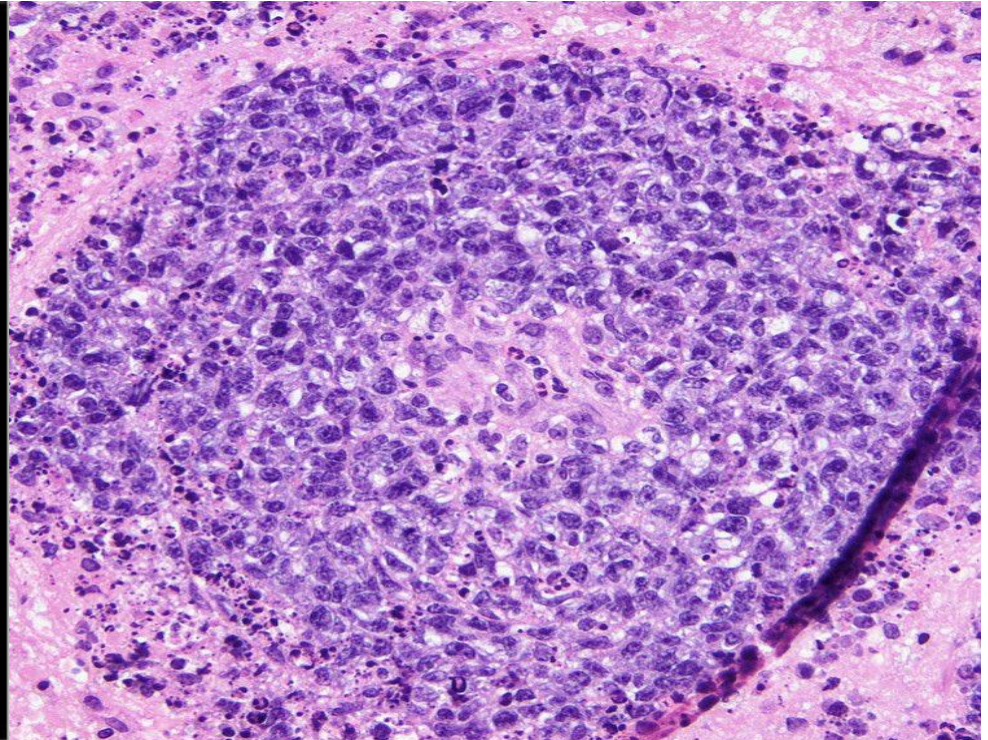
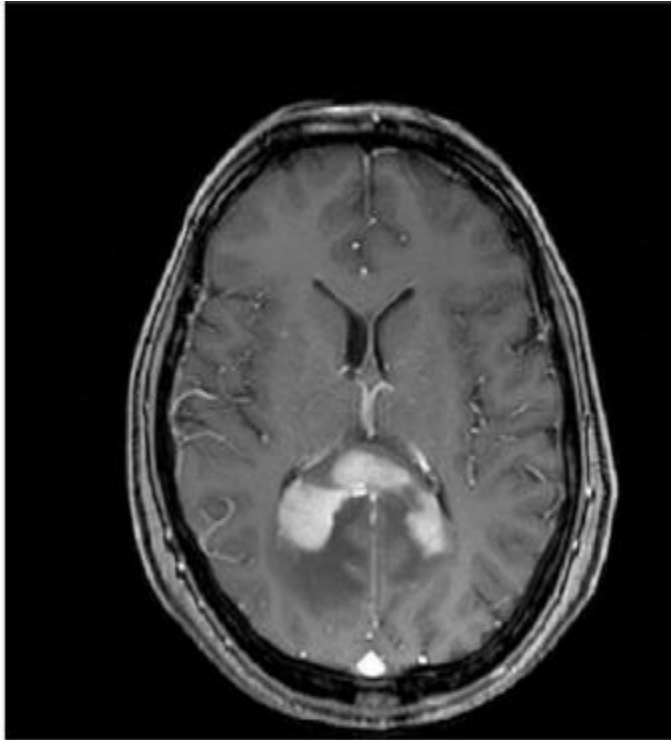
Microscopic

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.

Primary Central Nervous System Lymphoma



Meningiomas

Age

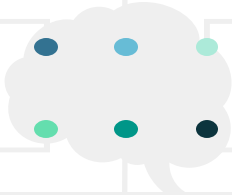
Adults

Location

Attached to the dura, along any of the external surfaces of the brain or in ventricular system

Genetics

Loss-of-function mutations in the NF2 tumor suppressor gene on chromosome 22.



Behavior

Predominantly benign; Most are easily separable from brain. Some are infiltrative, (ass. Recurrence).

Gross

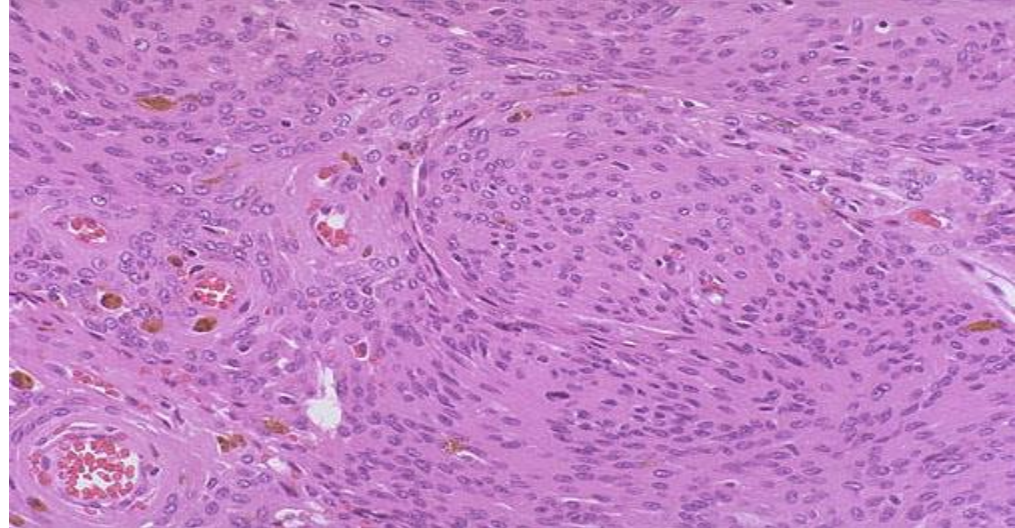
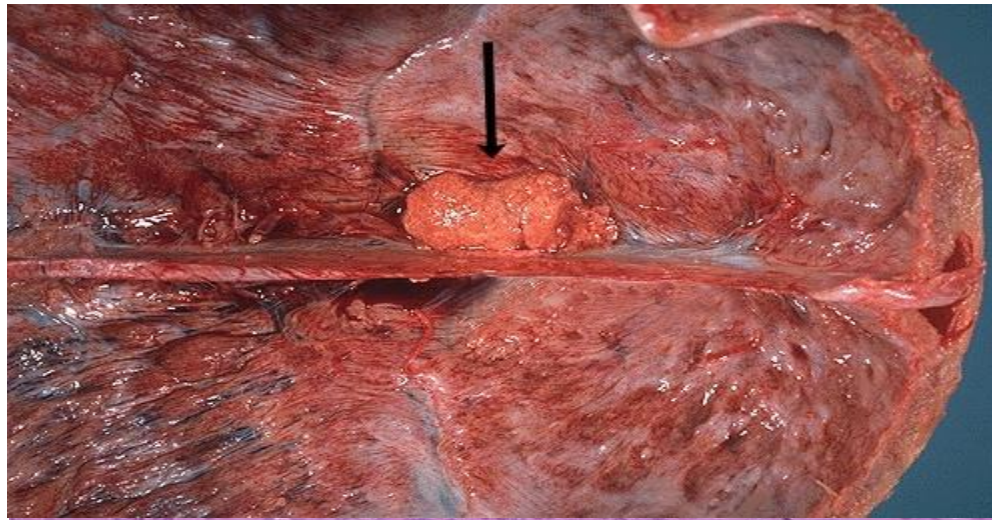
often cystic; if solid, it may be well circumscribed. Rarely infiltrative

Prognosis

Determined by: size & location, & histologic grade

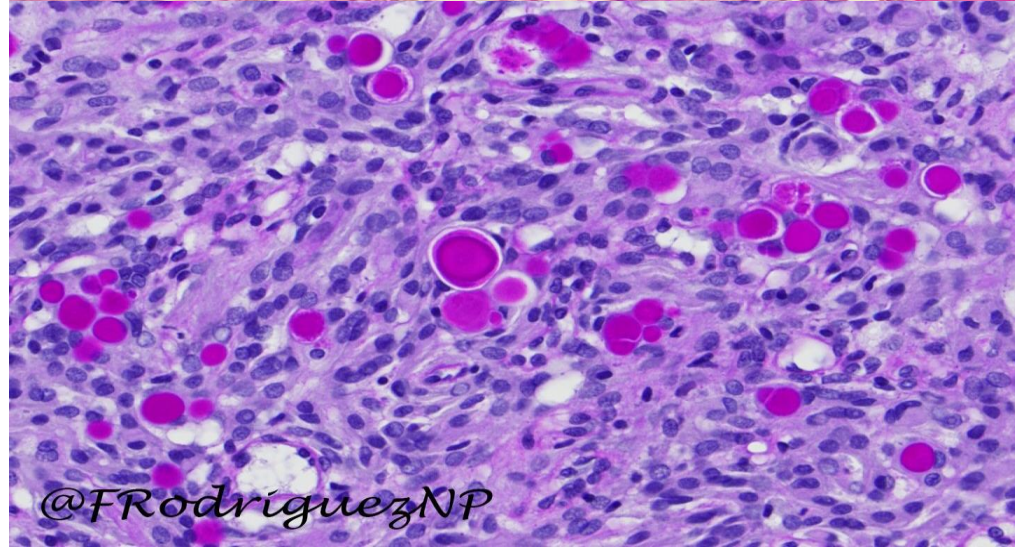
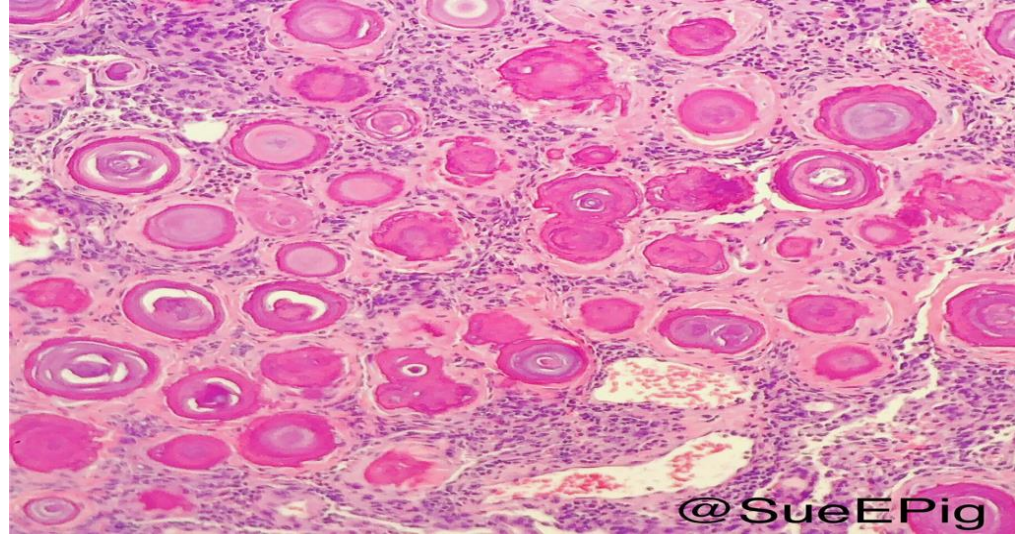
Meningiomas - WHO grade I

- 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 **meningotheial**; named for whorled, tight clusters of cells without visible cell membranes



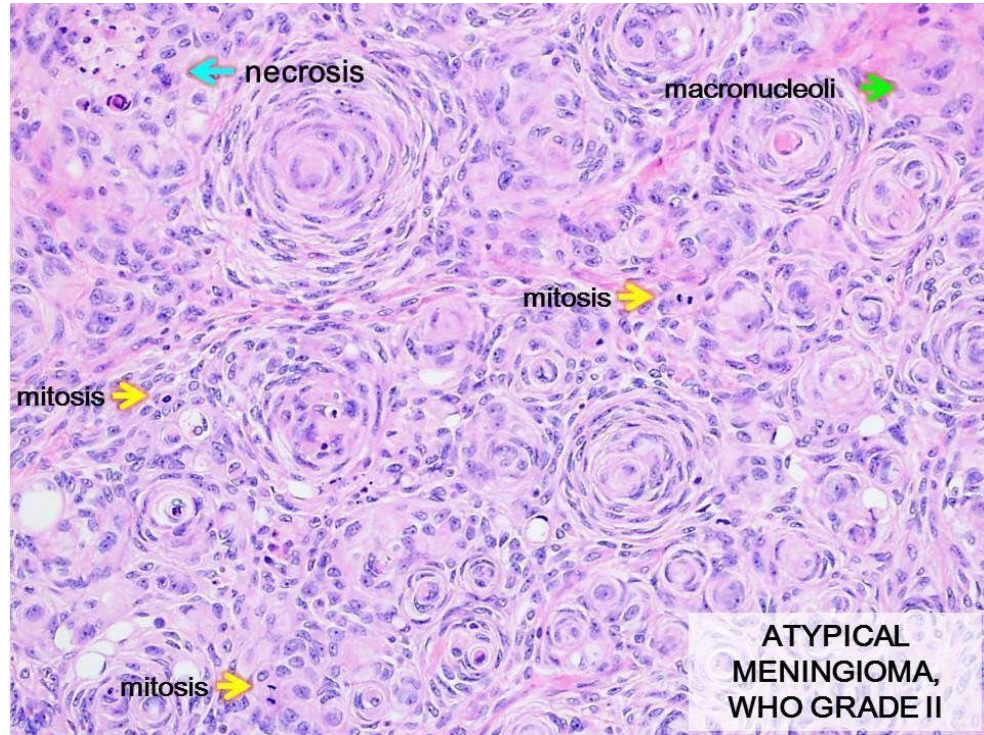
Meningiomas - WHO grade I

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



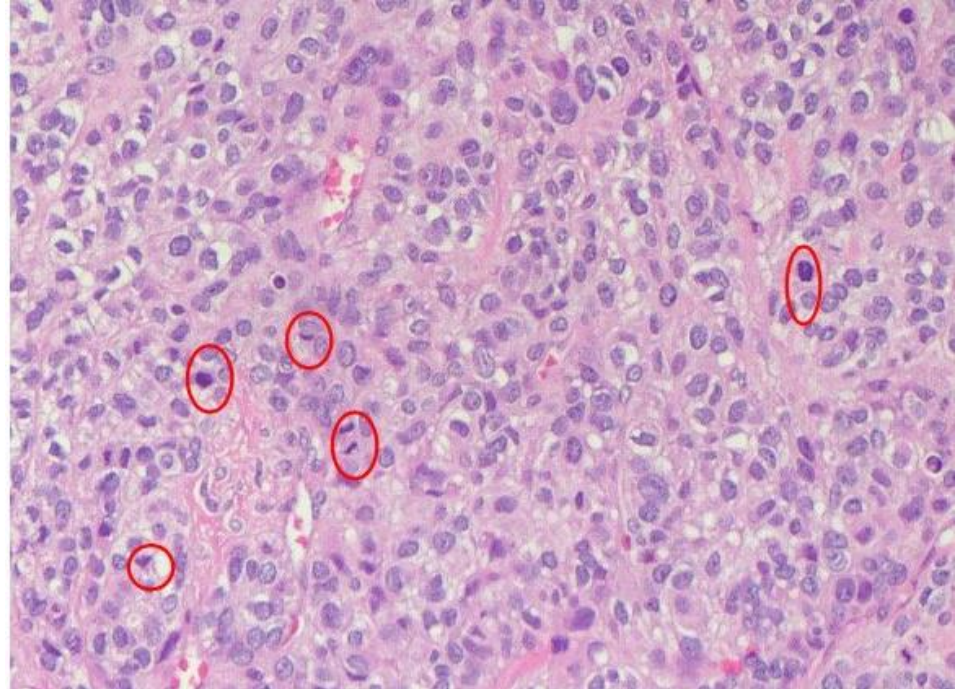
Meningiomas-WHO grade 2

- 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

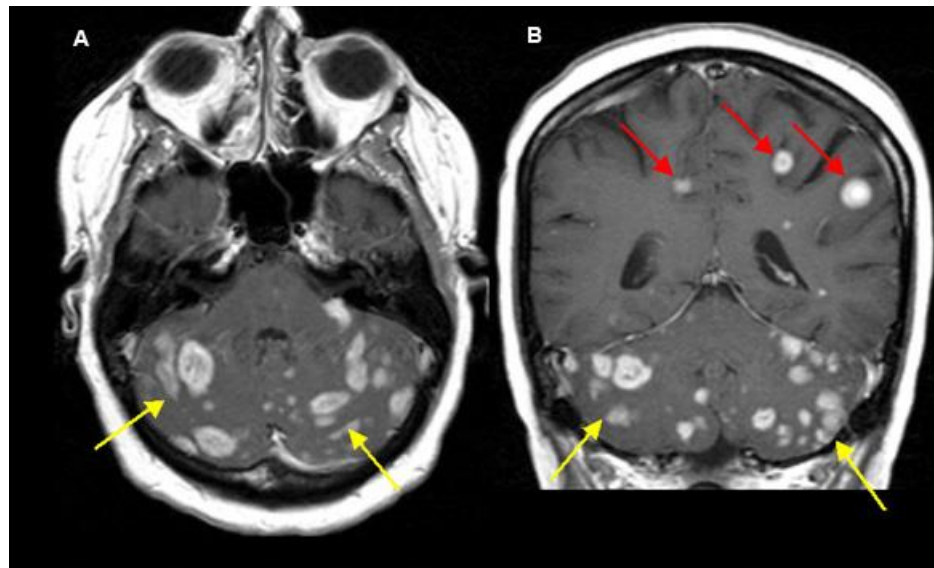
- Highly aggressive tumors that resemble a high-grade sarcoma or carcinoma morphologically.
- Mitotic rates are typically much higher than in grade 2 meningiomas.



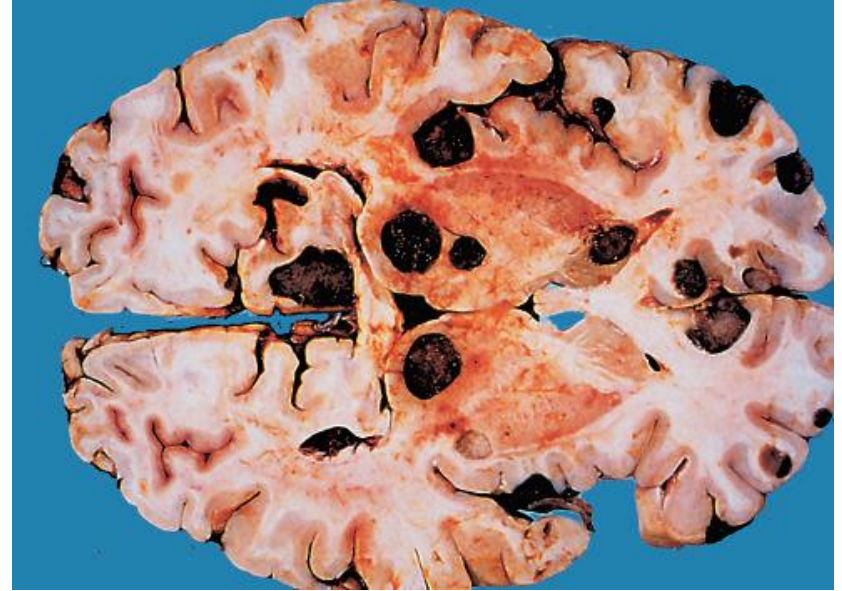
Metastatic Tumors

- 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

Metastatic Tumors



Metastatic Tumors



“We have to endure the discordance between imagination and fact. It is better to say, “I am suffering,” than to say, “This landscape is ugly.”
— Simone Weil