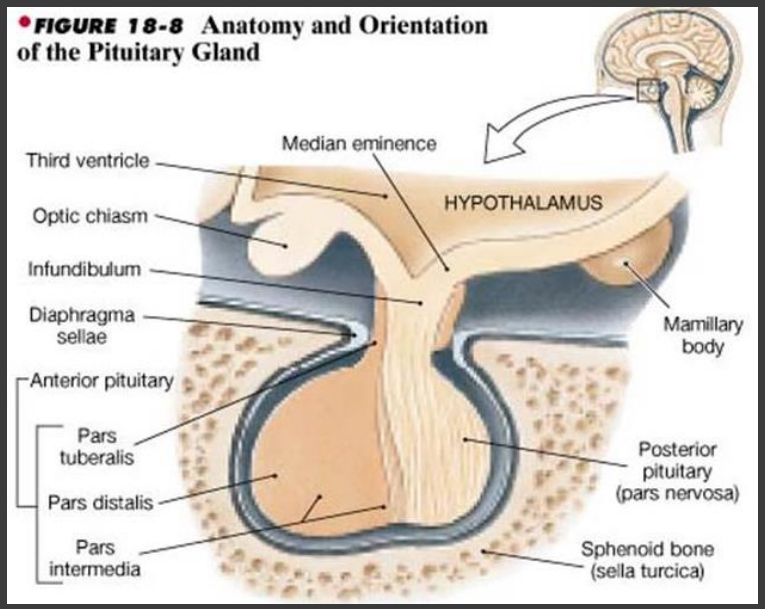
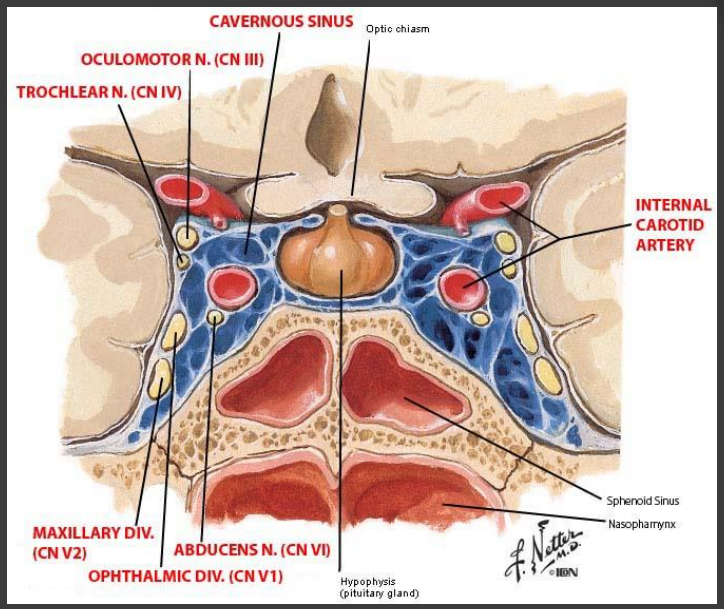
Rad-Path Conference: Pituitary region lesions

Sha-har Admoni
Tyler Janovitz
February 8, 2016

Please take a TurningPoint clicker!

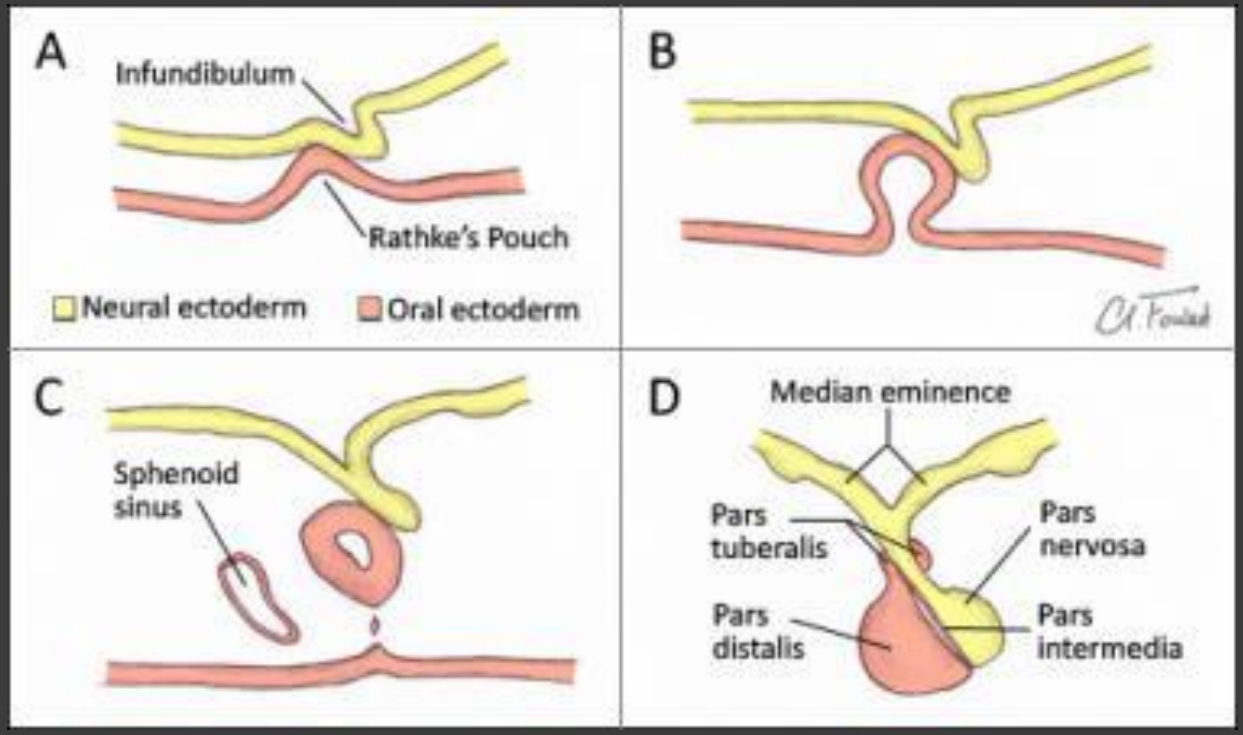
Anatomy





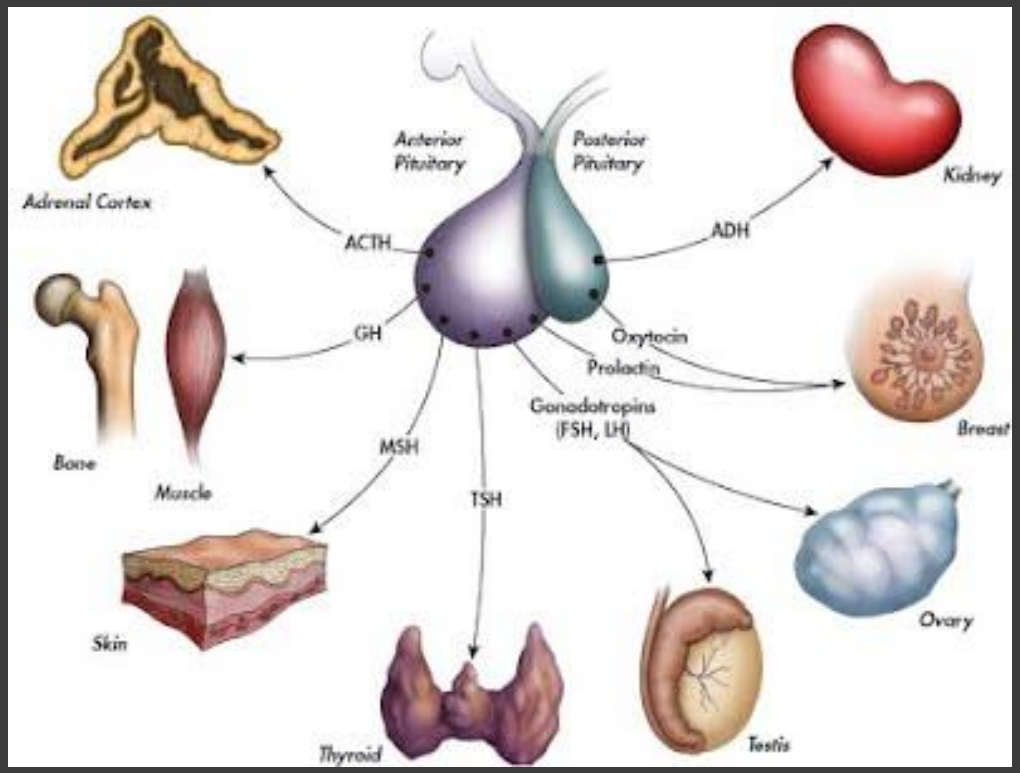
- The pituitary gland is made up of anterior and posterior lobes
- Sits in the sella turcica, a cup-shaped depression of the sphenoid bone
- Pituitary fossa is surrounded by the cavernous sinus

Embryology



- **Anterior pituitary** forms from Rathke's pouch, a superior invagination from the primitive oral ectoderm
  - Rathke's pouch closes off to form a vesicle that involutes
- **Posterior pituitary** originates from neural ectoderm, and migrates inferiorly from the hypothalamus

Physiology



- Anterior
  - Produces and secretes endocrine hormones
- Posterior
  - Composed of axons from the hypothalamus, which transport oxytocin and vasopressin

American College of Radiology
ACR Appropriateness Criteria®

Clinical Condition: Neuroendocrine Imaging

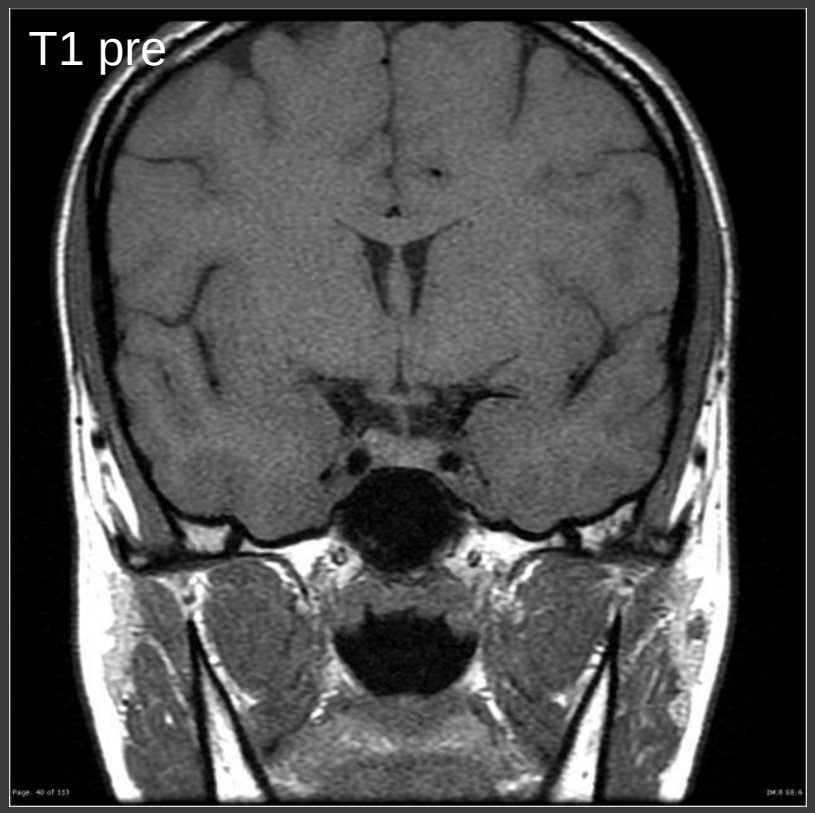
Variant 1: Hypopituitarism.

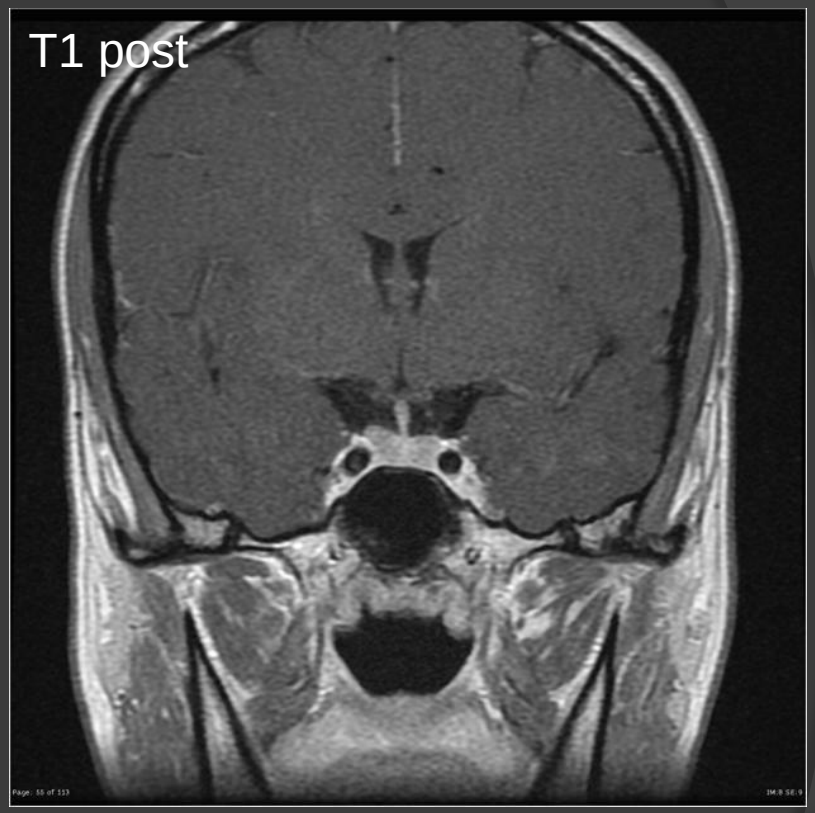
Radiologic Procedure	Rating	Comments	RRL*
MRI head without and with contrast	8	Multiplanar thin sellar imaging. See statement regarding contrast in text under "Anticipated Exceptions."	O
MRI head without contrast	7	Multiplanar thin sellar imaging.	O
CT head with contrast	5	Indicated if MRI is not available or contraindicated.	☢☢☢
CT head without contrast	4	Indicated if MRI is not available or contraindicated.	☢☢☢
CT head without and with contrast	4	Indicated if MRI is not available or contraindicated.	☢☢☢
MRA head without contrast	3	May be useful if vascular pathology is known or suspected.	O
MRA head without and with contrast	3	May be useful if vascular pathology is known or suspected.	O
CTA head with contrast	2	For surgical planning or vascular detail if MRI and MRA are contraindicated.	☢☢☢
X-ray tomography head	1		☢
X-ray sella	1		☢
Arteriography cerebral	1		☢☢☢
Venous sampling petrosal sinus	1		Varies

Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate

*Relative Radiation Level

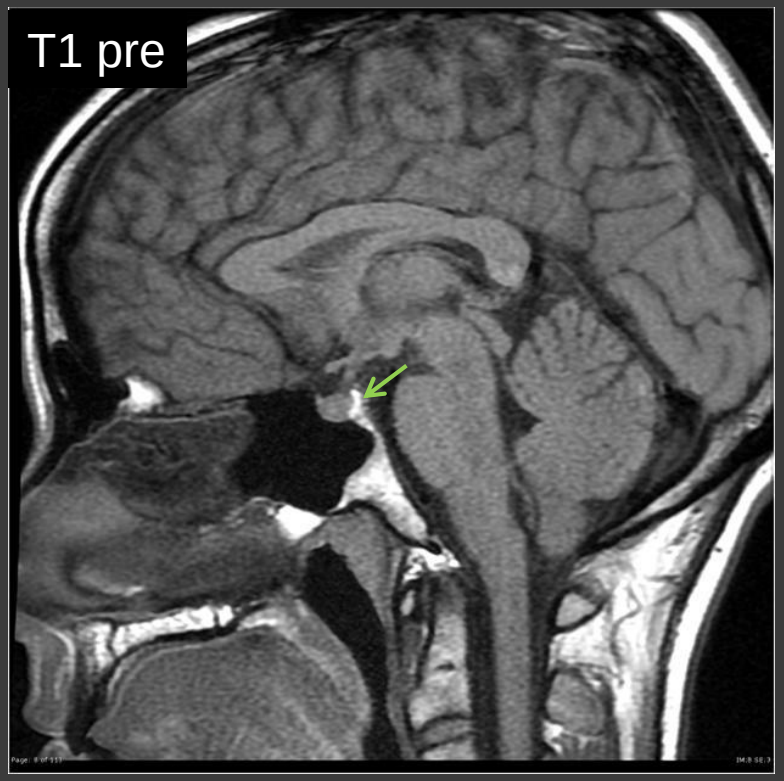
Normal imaging appearance

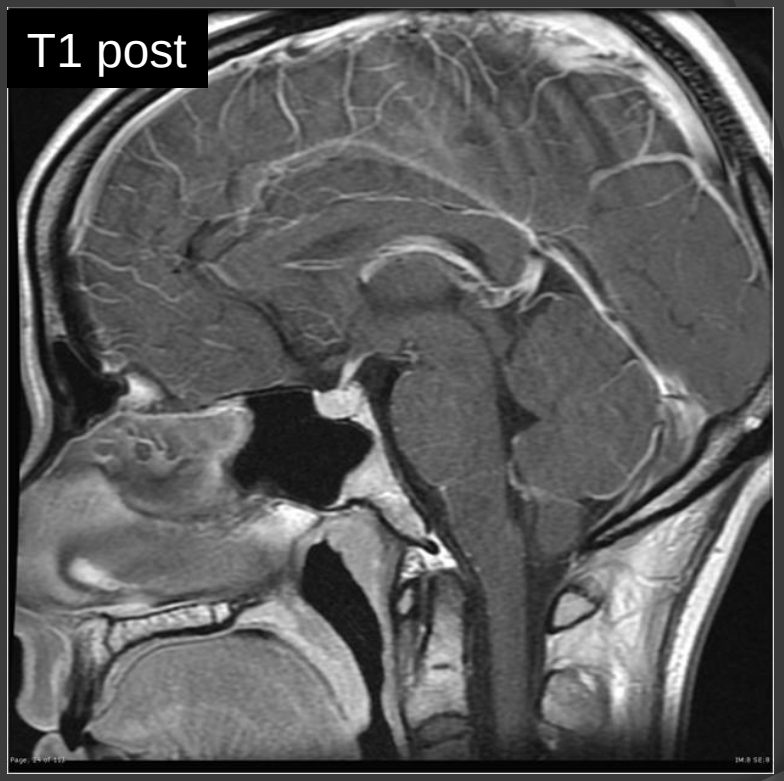




- Size varies based on age, sex, hormonal/pregnancy status
- Anterior pituitary is isointense to brain on T1WI and T2WI, and enhances promptly

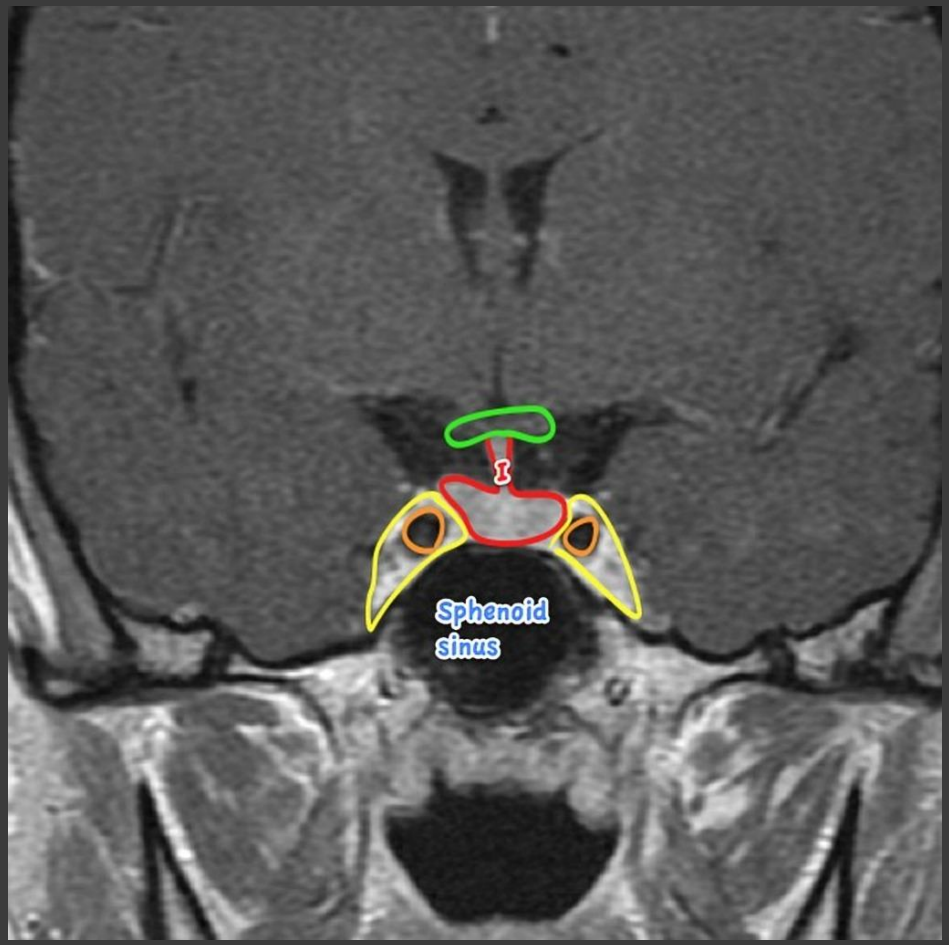
Normal imaging appearance





- Posterior pituitary is hyperintense on T1WI ("posterior pituitary bright spot"), best seen on sagittal images

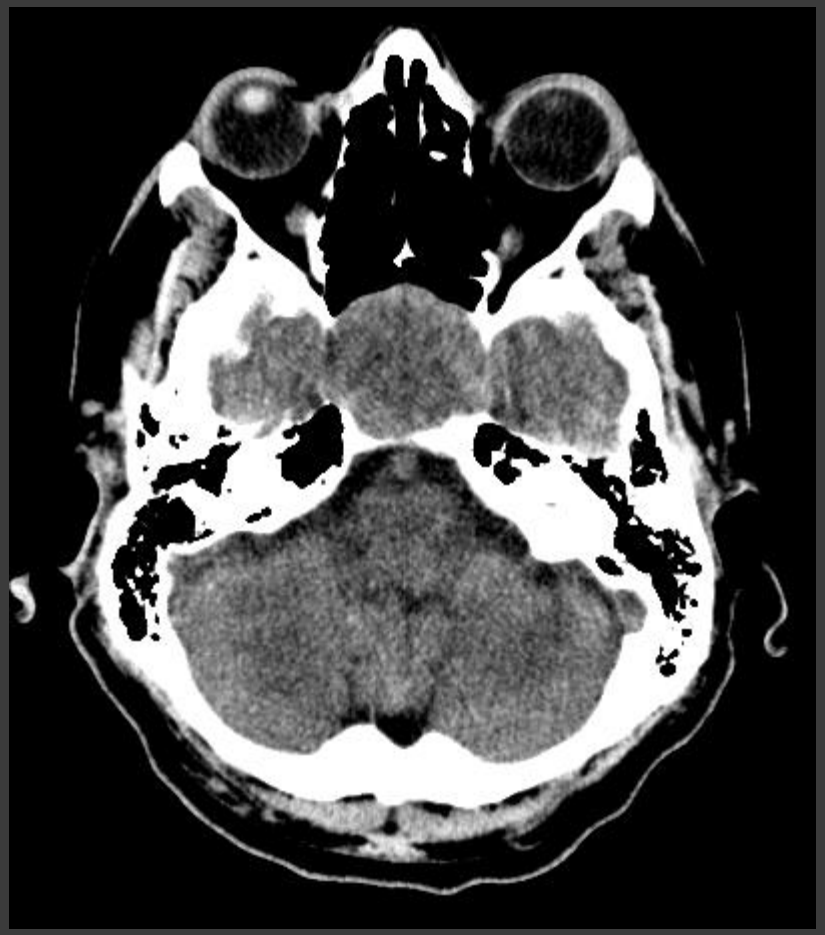
Pituitary region anatomy

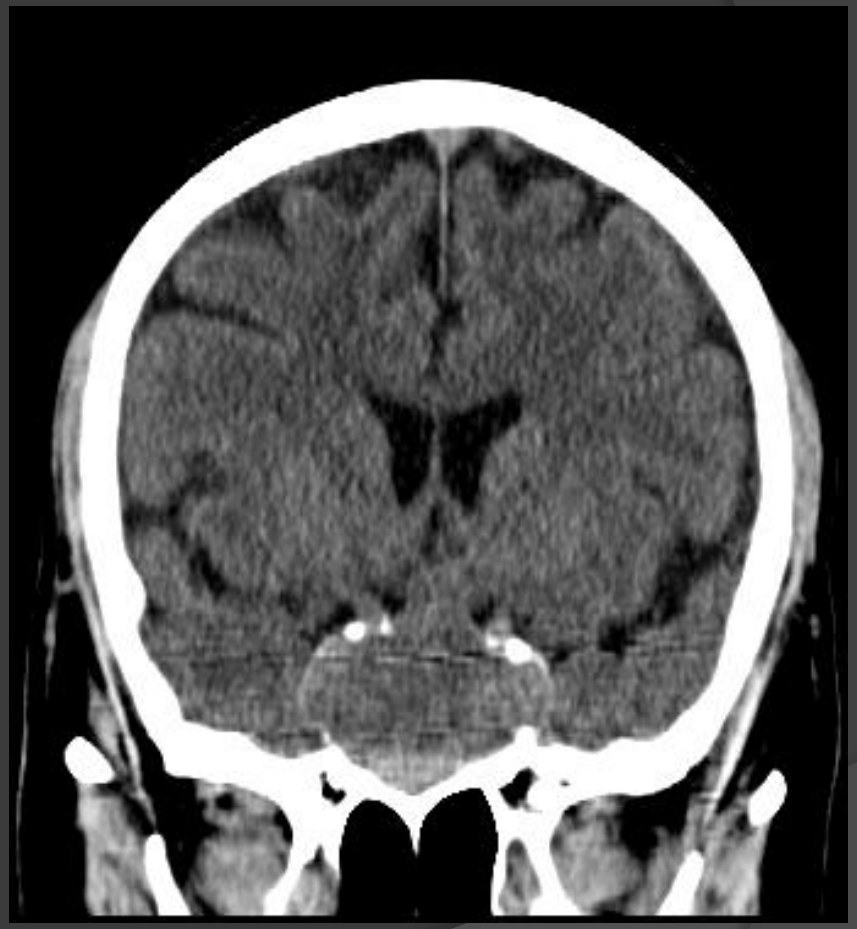


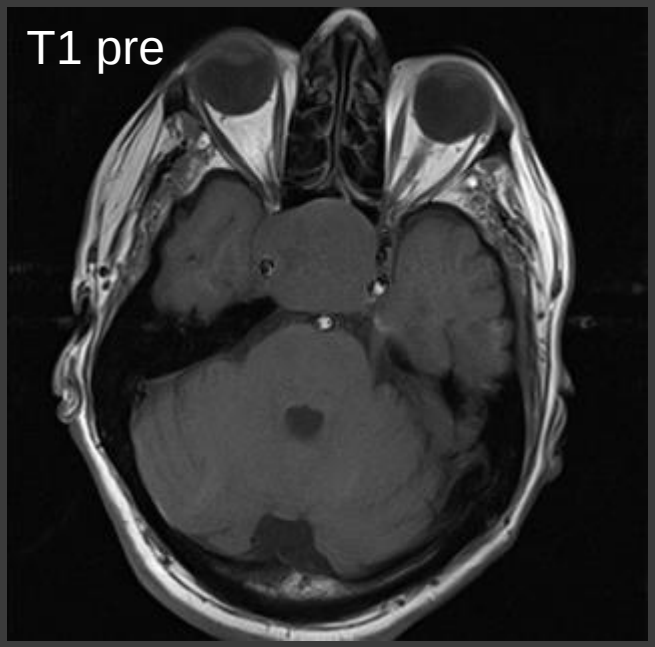
Pituitary gland and infundibulum
Optic chiasm
Cavernous sinus
Cavernous ICA

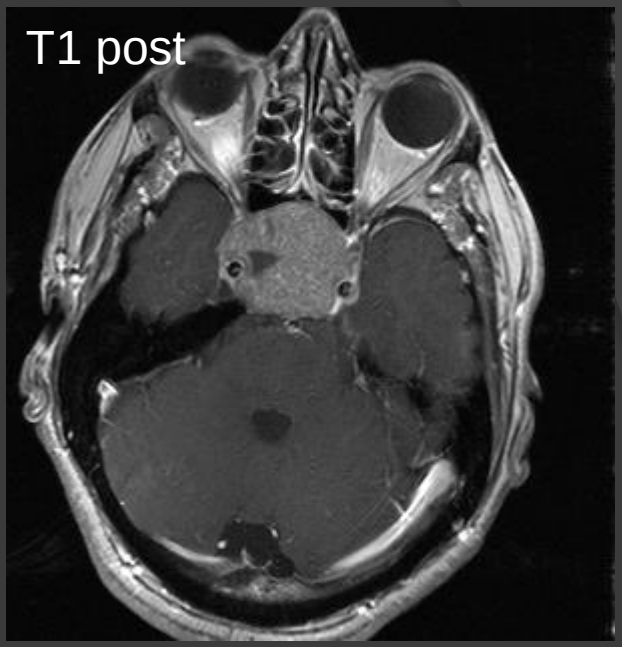
Case 1

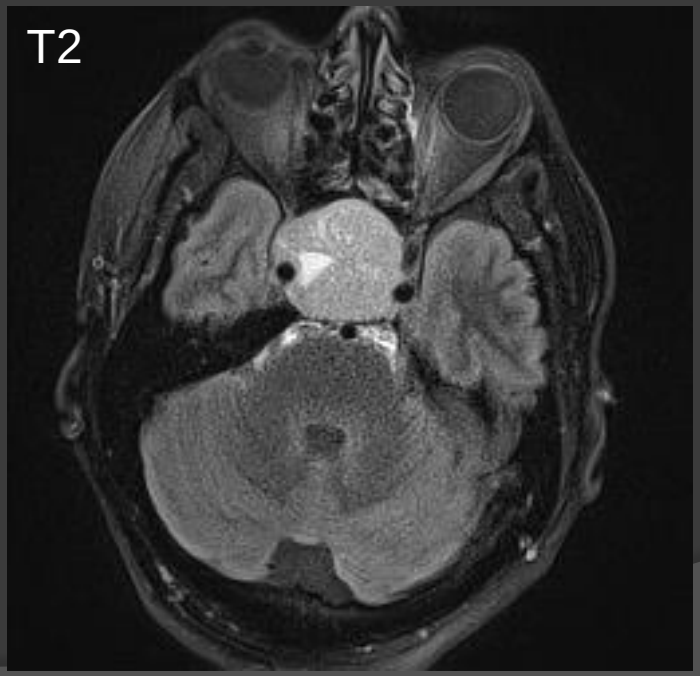
54 year old man with visual disturbance and hyponatremia.

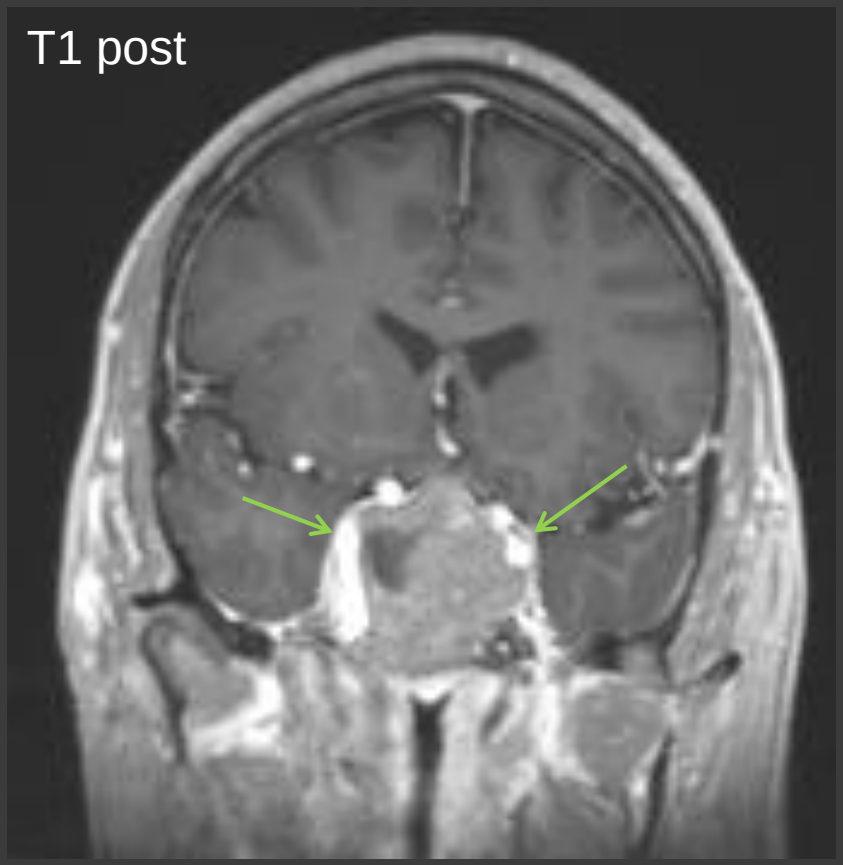


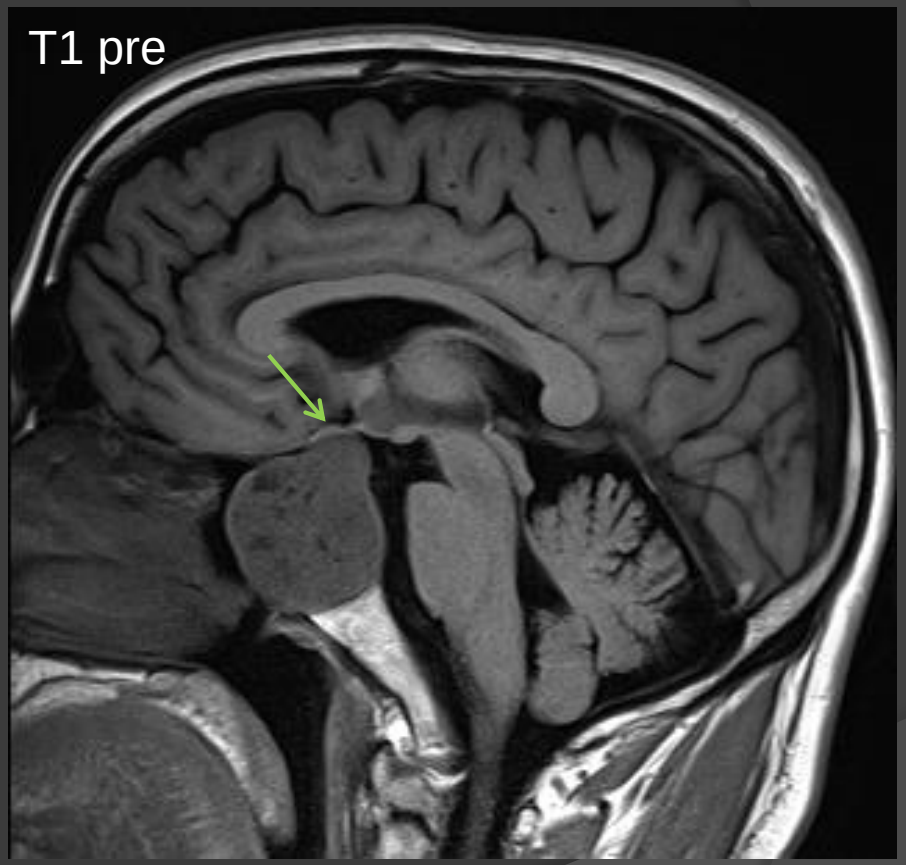



T1 pre


T1 post

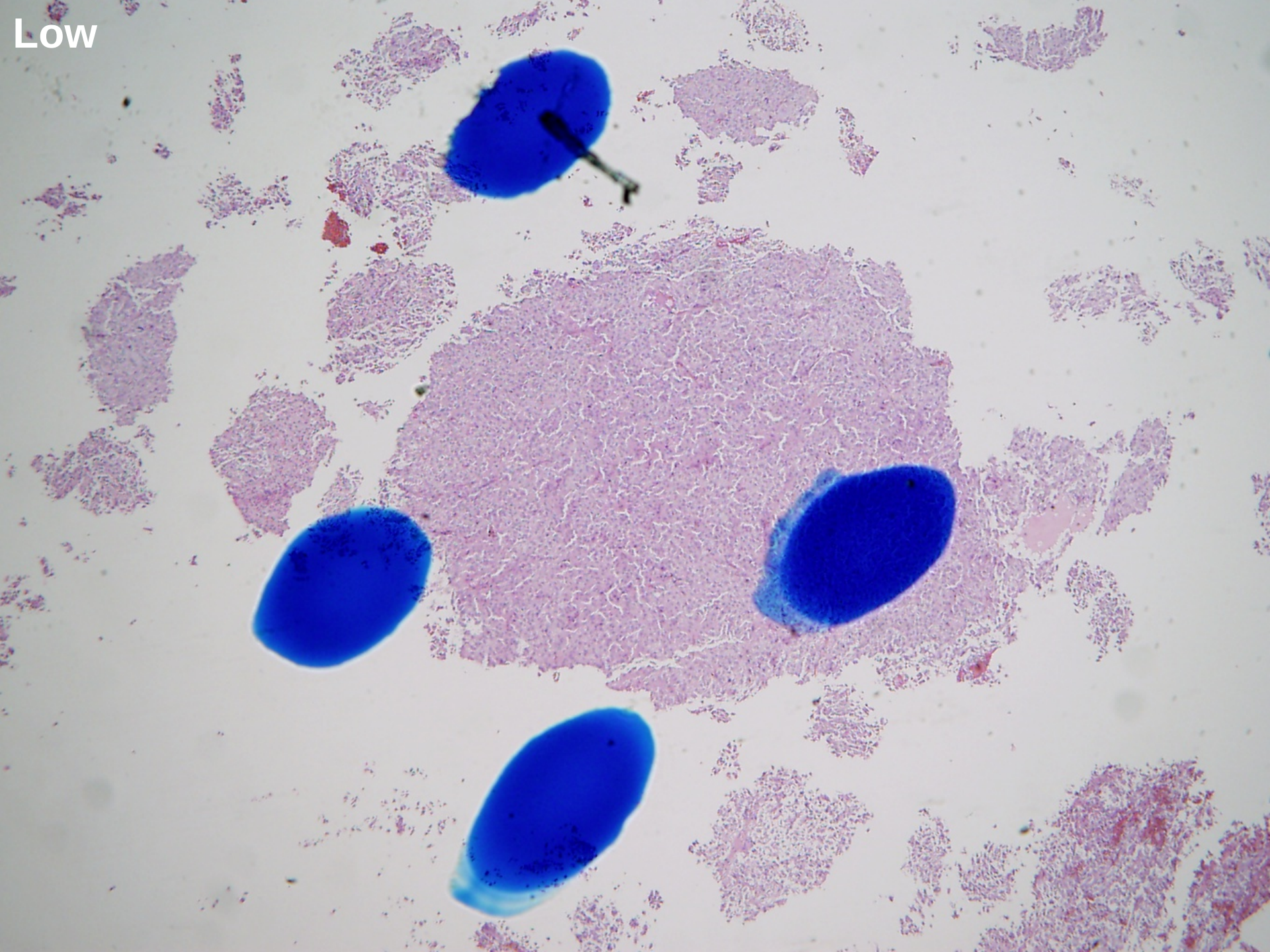

T2

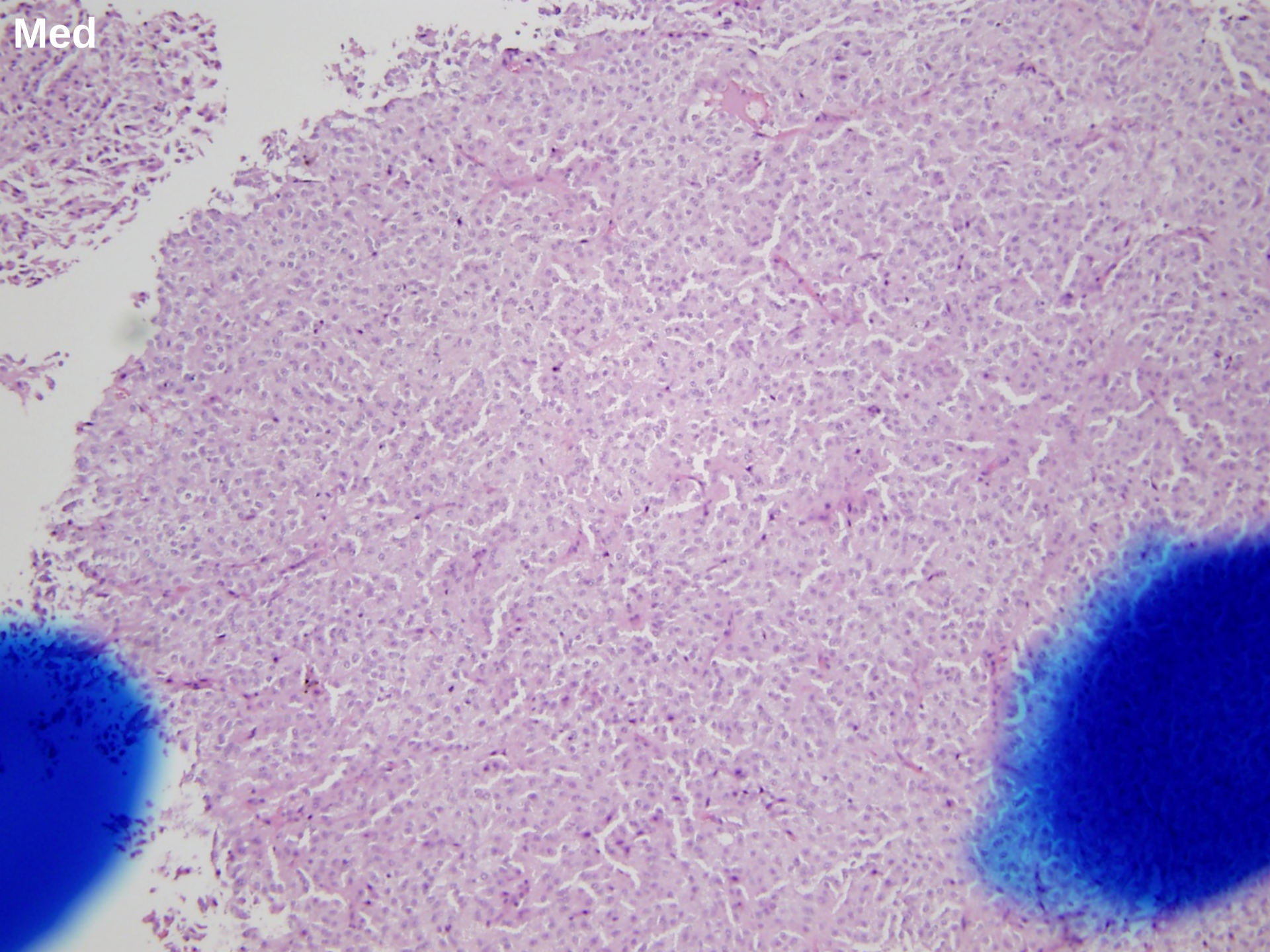

T1 post

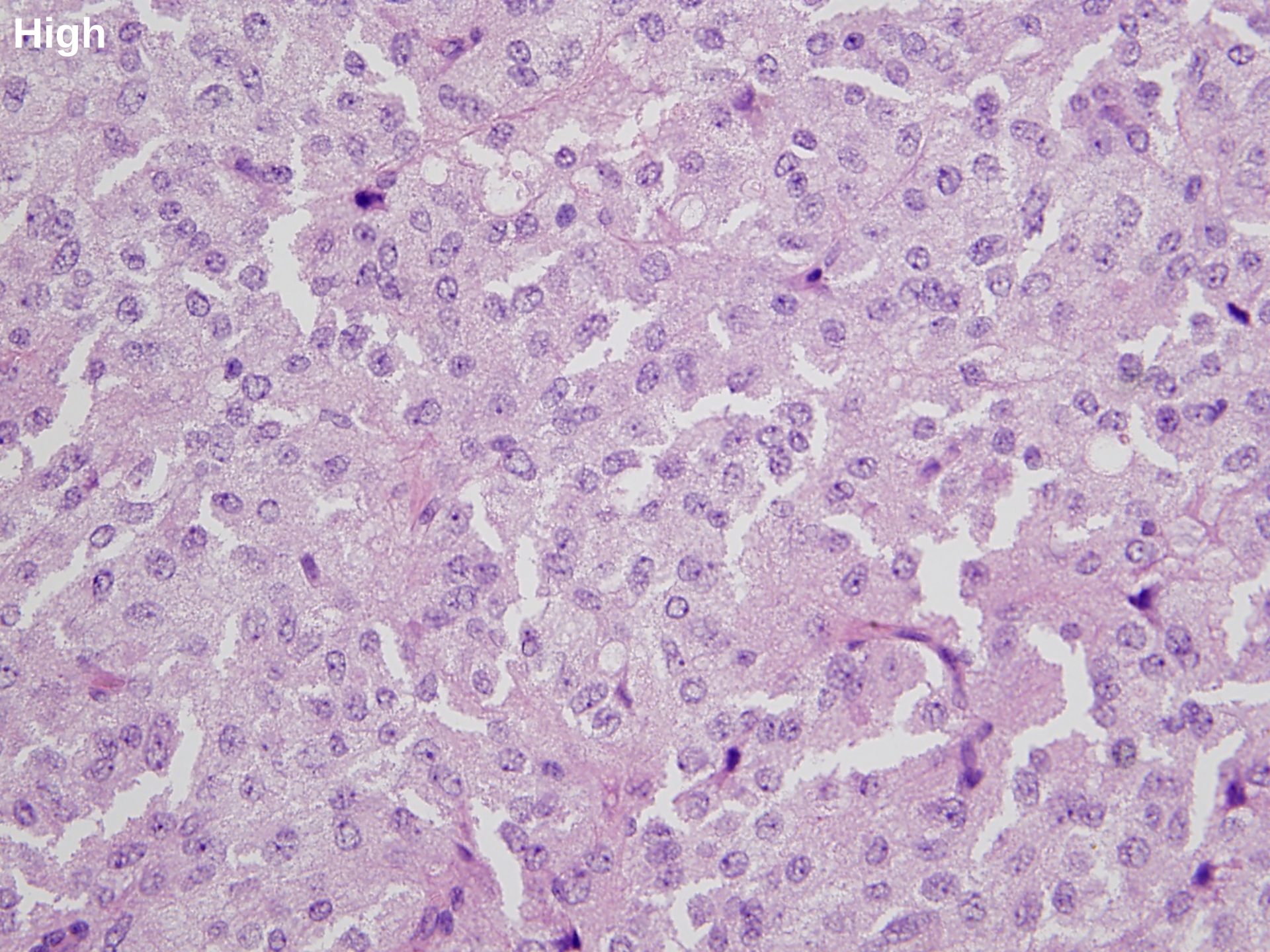

T1 pre

Pathology

BS-12-46336


Low


Med


High

Diagnosis

A-B. PITUITARY MASS:

PITUITARY ADENOMA
TYPICAL
W.H.O Tumor Type: Null Cell Adenoma (non-secretory)

The estimated MIB-1 proliferative index is <3%

Pituitary Adenoma

Sheets of round cells with spherical nuclei

Often classified by hormone production (growth hormone, prolactin, TSH, ACTH, FSH or alpha-glycoprotein subunit)

Most grow slowly and are considered benign

Pituitary adenoma: Clinical presentation

- Population prevalence of 0.1%, but autopsy prevalence of 15%
- Account for 30-50% of all pituitary region masses

Microadenoma <10 mm

Usually presents with hormonal imbalance

- 74% prolactin
- 15% growth hormone (acromegaly)
- 9% ACTH (Cushing's)
- 2% TSH (hyperthyroidism)
- Mixed

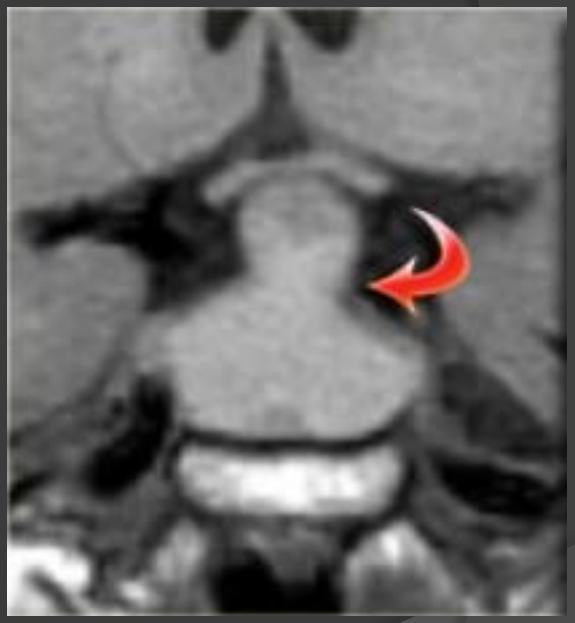
Macroadenoma >10 mm

Usually present with mass effect

- headache
- visual disturbances (bitemporal hemianopsia)
- cranial nerve palsy
- hypopituitarism (from compression)

Macroadenoma: Imaging characteristics on CT

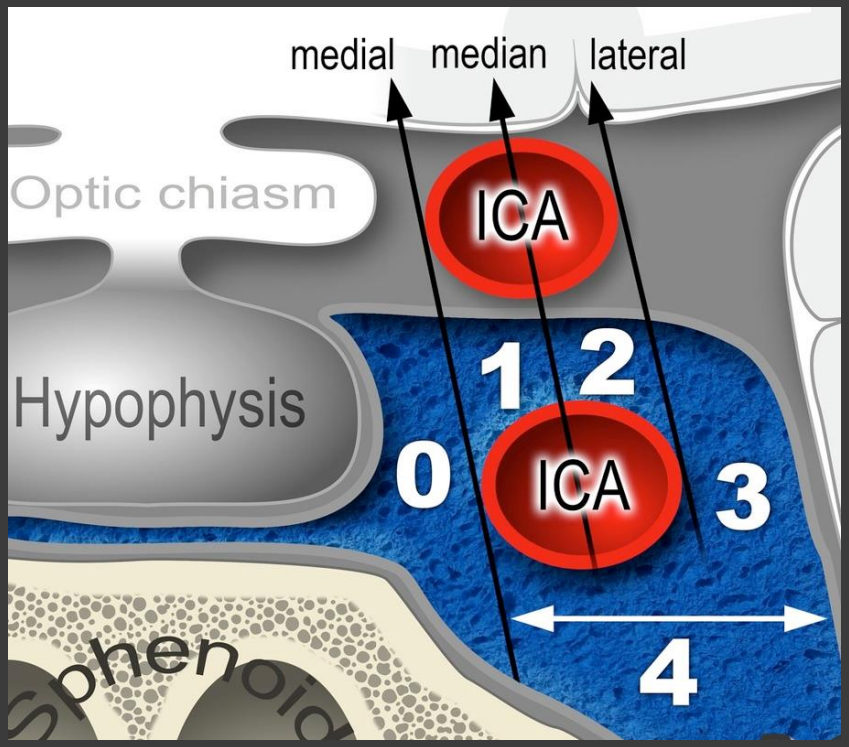
- Solid tumor with similar attenuation to brain (30-40 HU), moderate contrast enhancement
  - Indentation at the diaphragma sellae can give a **"snowman"** appearance
- May contain areas of cystic change, necrosis, or hemorrhage
- Calcification is rare



Macroadenoma: Imaging characteristics on MRI

- Isointense to gray matter on T1WI and T2WI
- Moderate to bright enhancement
- The **bony sella is often enlarged**
- Can cause optic chiasm compression
  - Can elevate and compress the central part of the chiasm as it grows superiorly out of the pituitary fossa
- Can cause cavernous sinus invasion
  - Can directly extend into the cavernous sinus
  - May cause cranial nerve compression (most commonly CN III, followed by CN VI)

Macroadenoma: Grading of cavernous sinus invasion

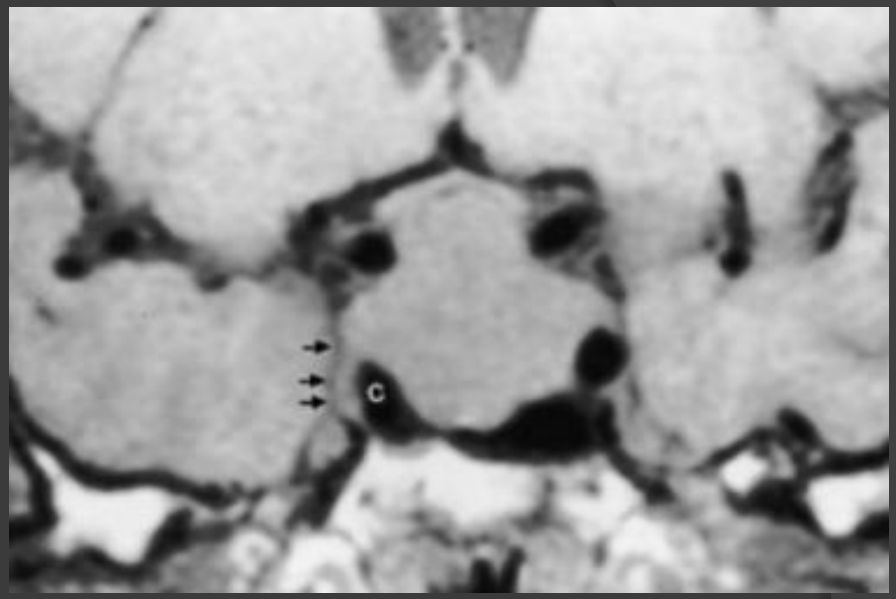


A rough guide:

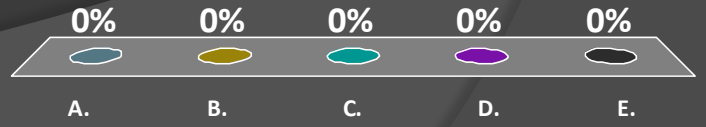
<90 degree encasement of cavernous ICA: unlikely cavernous sinus involvement

>180 degree encasement of cavernous ICA: almost certain cavernous sinus involvement

What is the most likely functional classification of the pituitary adenoma pictured?



A. Corticotropic (ACTH) cell adenoma
B. Growth hormone (GH) cell adenoma
C. Prolactin cell adenoma
D. Thyrotropin (TSH) cell adenoma
E. Non-secretory adenoma

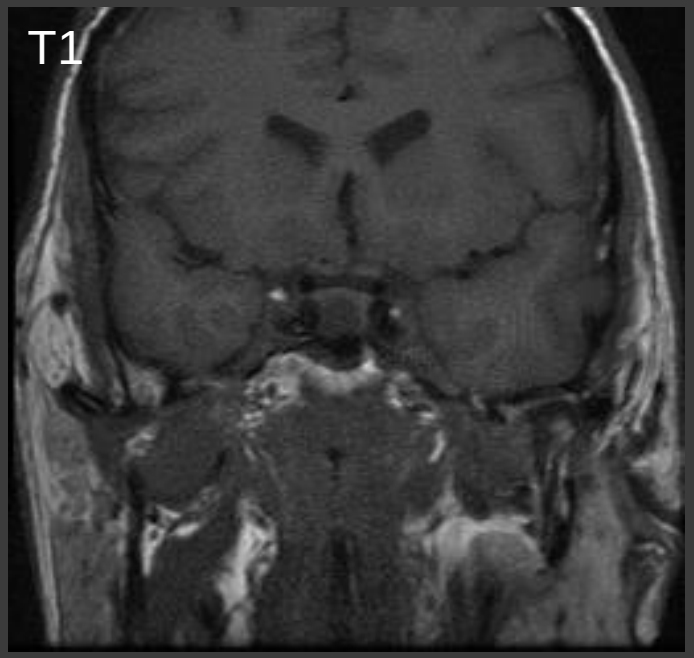


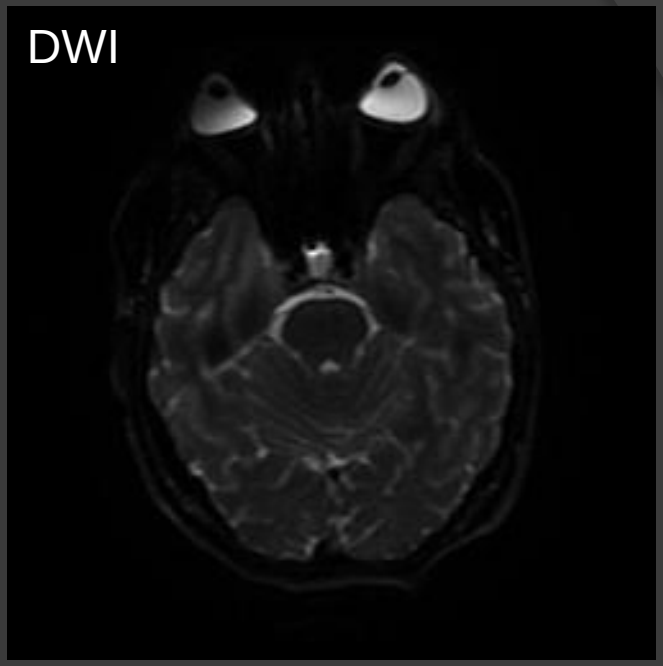
Macroadenoma: Treatment and prognosis

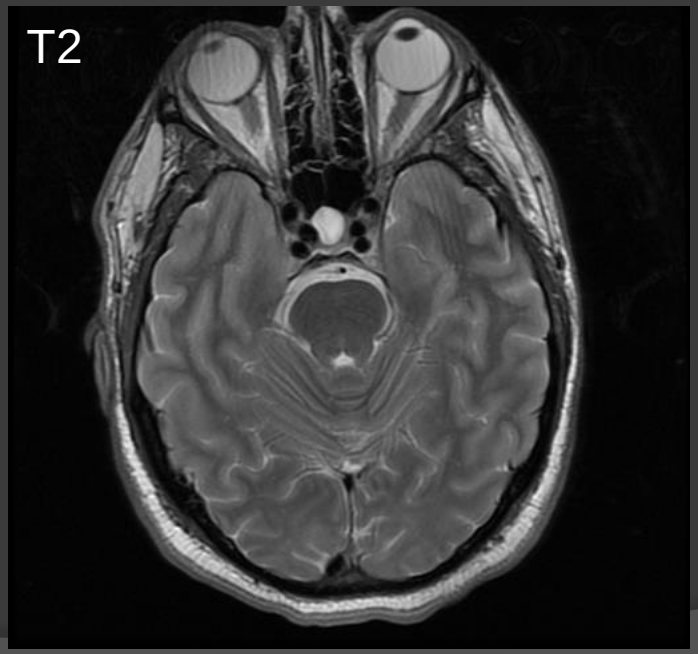
- Benign neoplasm, WHO grade I
- Symptoms related to mass effect usually necessitate surgical decompression
  - Typically a transsphenoidal approach, although craniotomy may be necessary for larger tumors
- Medical management may be appropriate for smaller prolactin- or GH-secreting tumors
  - Large tumors may bleed following medical therapy

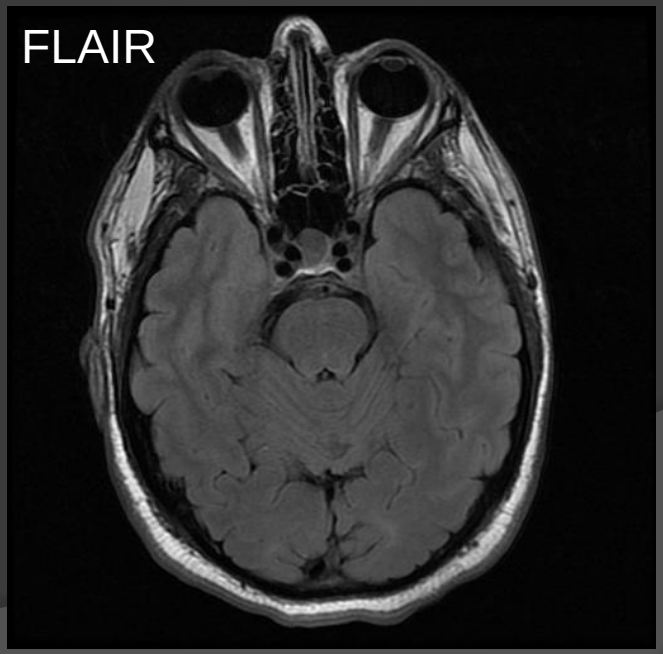
Case 2

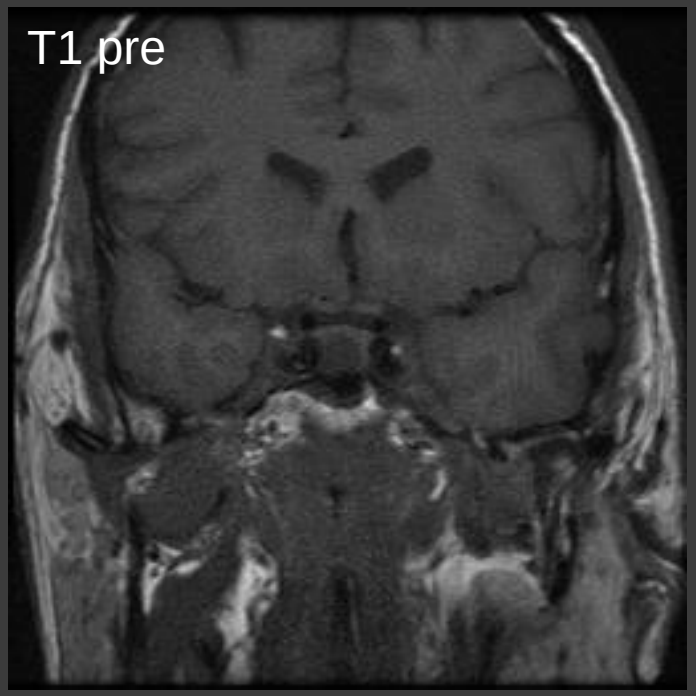
49 year old man with right-sided visual field deficit and temporal blurring.

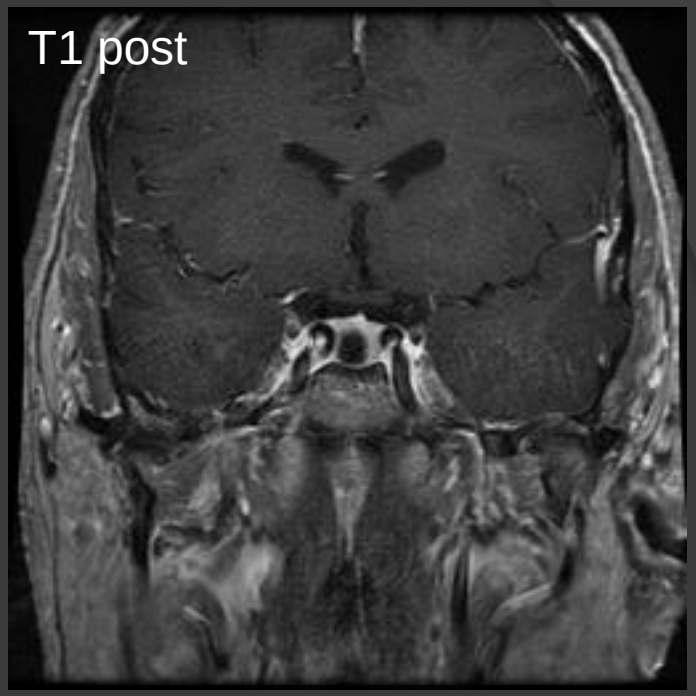

T1

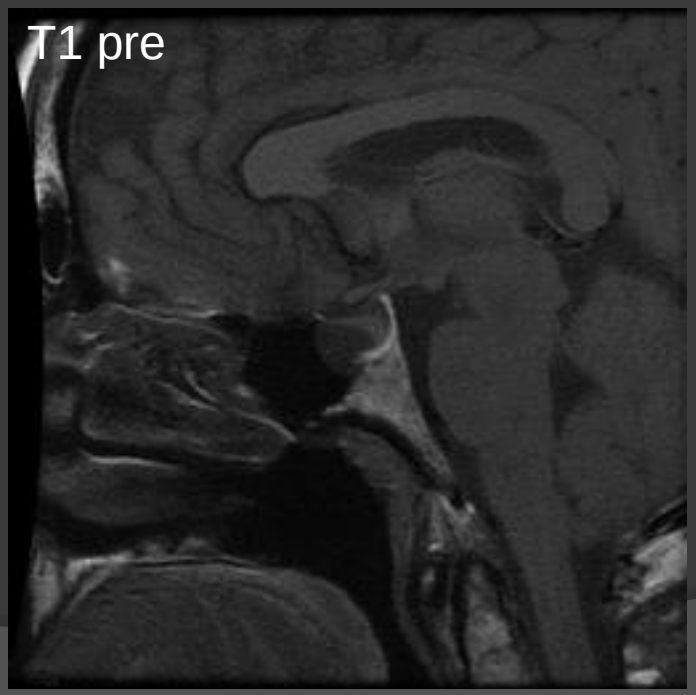

DWI

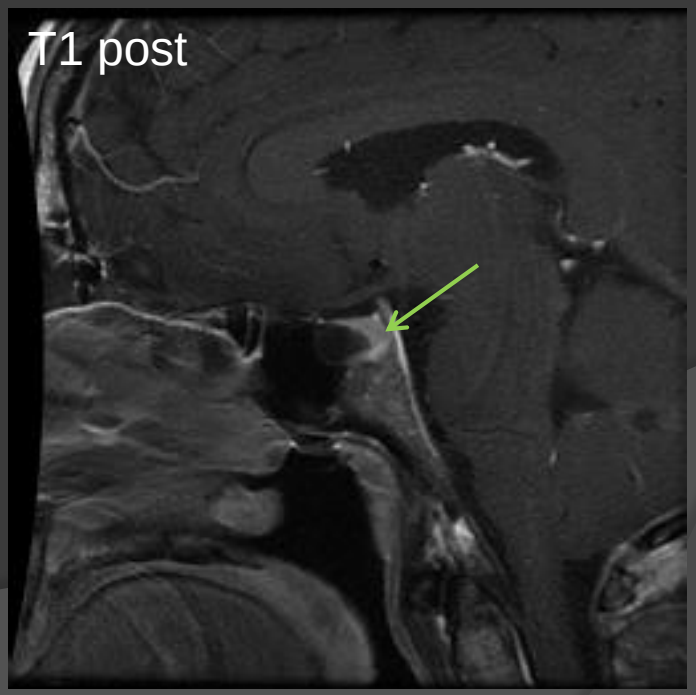

T2


FLAIR


T1 pre


T1 post

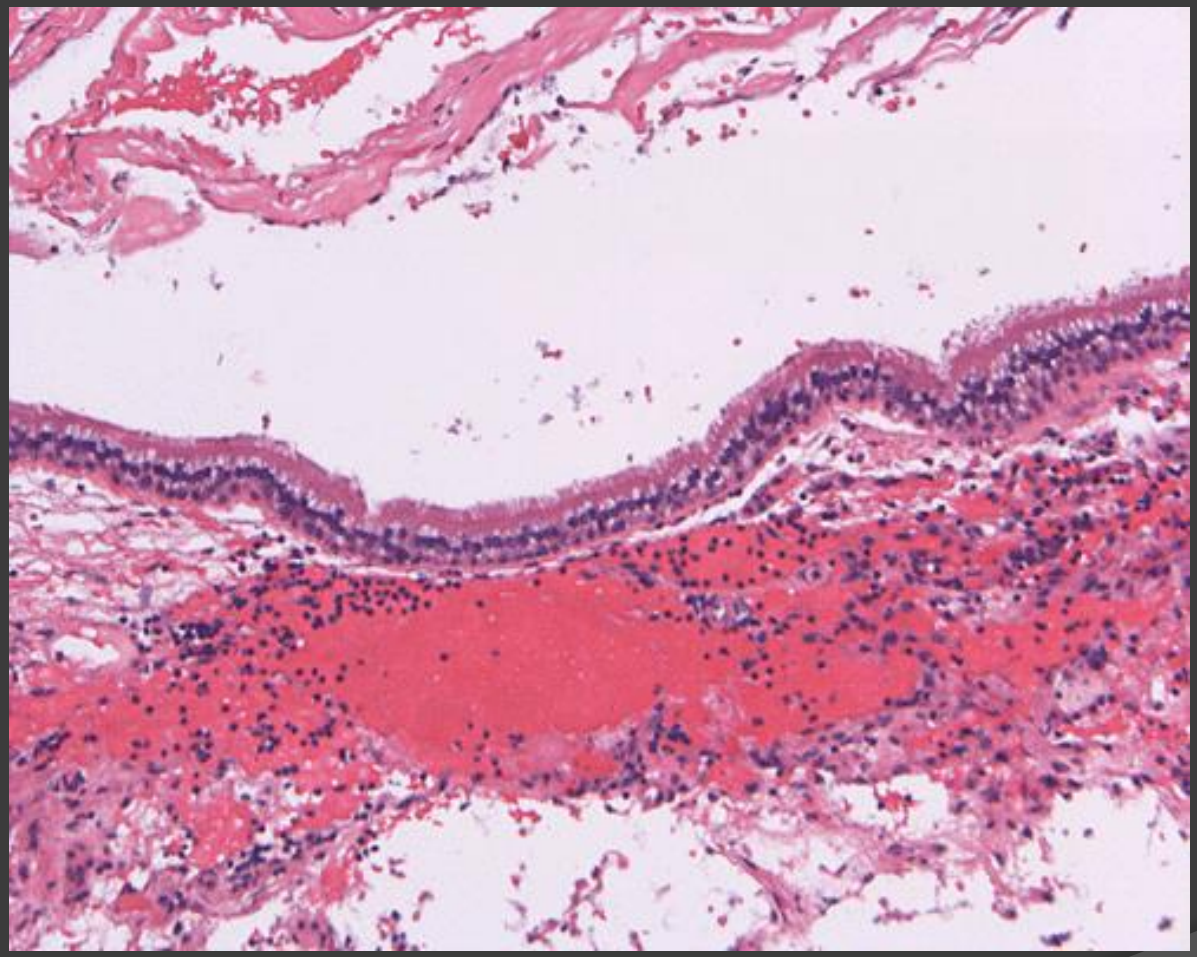

T1 pre


T1 post

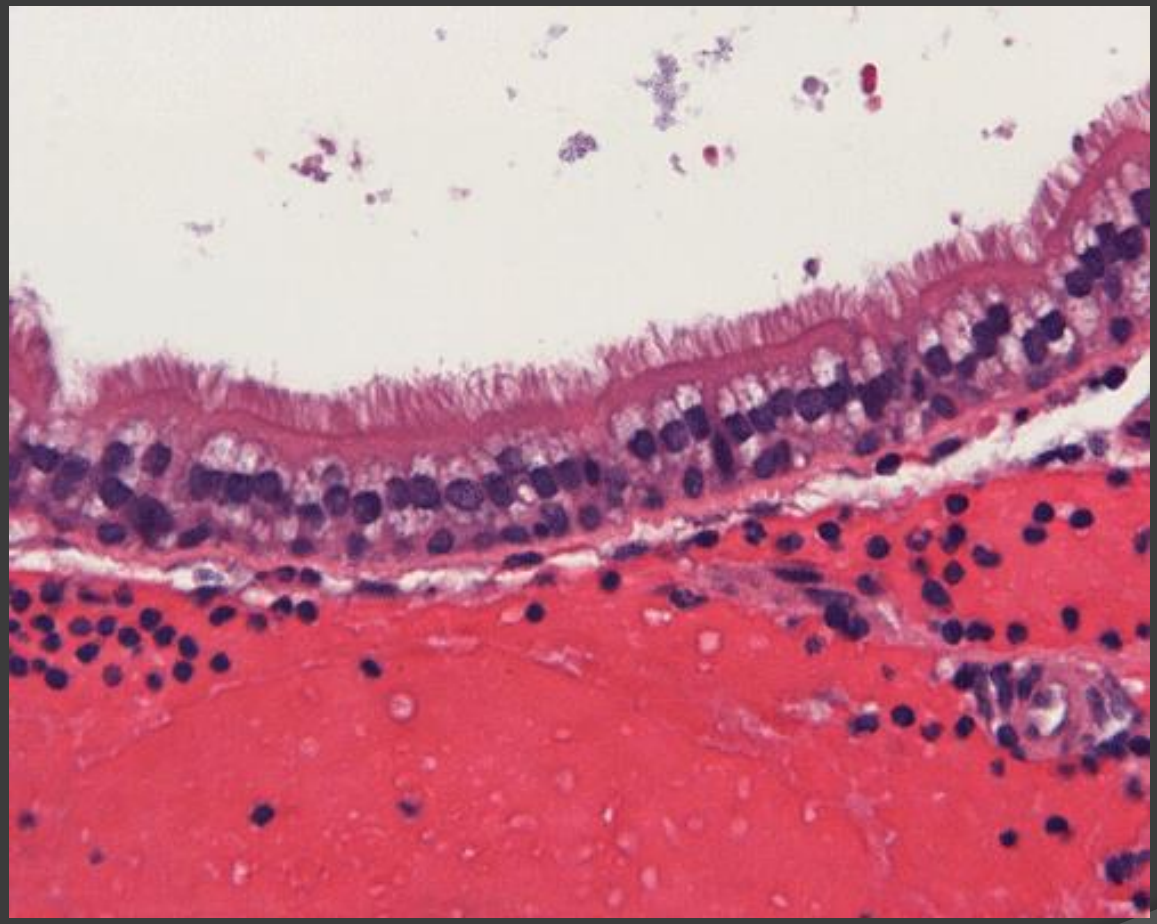
Pathology

BS-15-54588

Med



High



Diagnosis

A. SPECIMEN DESIGNATED: "DURA AND CYST WALL":

Fibrous tissue with associated cuboidal-to-columnar epithelium consistent with Rathke's cleft cyst

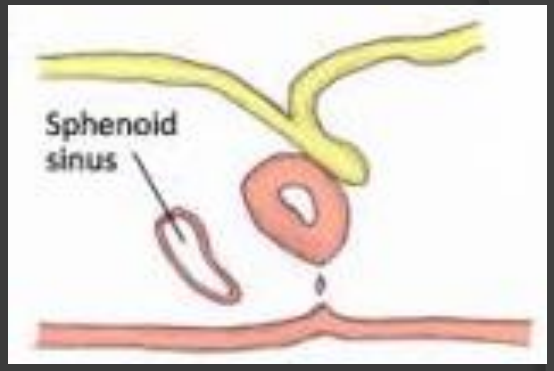
Rathke's Cleft Cyst

Lined by columnar ciliated epithelium with goblet cells

Variable fragments of pituitary tissue, squamous metaplasia, xanthogranulomatous reaction

Rathke's cleft cyst: Clinical presentation

- Caused by incomplete involution of Rathke's pouch, leaving behind a fluid-filled cleft

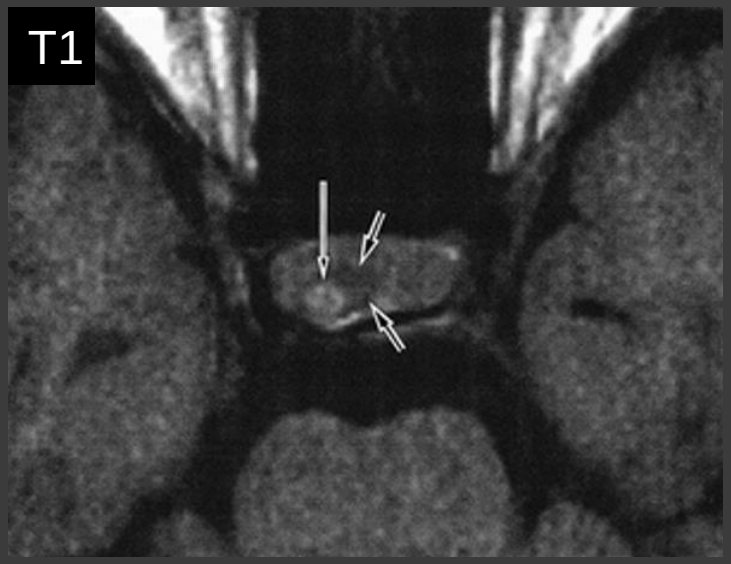


- Usually clinically asymptomatic or discovered incidentally (up to 22% incidence in autopsy studies)
- Most common in middle-aged adults, twice as common in women

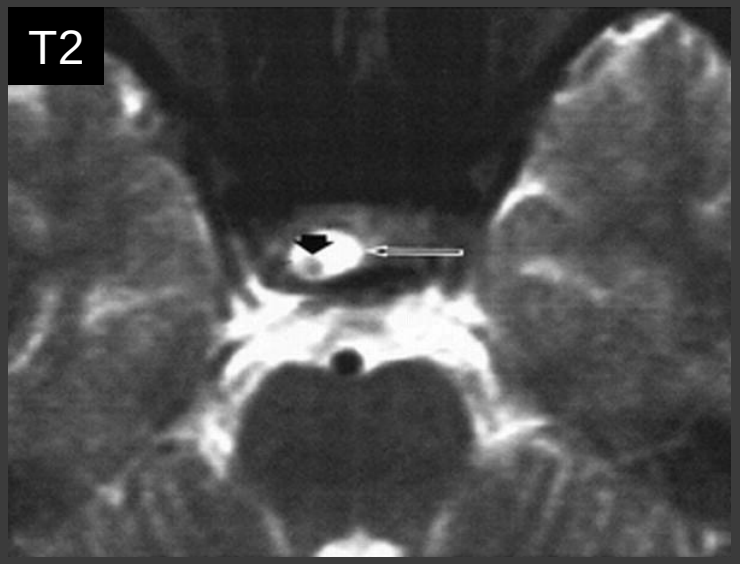
Rathke's cleft cyst: Imaging characteristics

- Seen as a well-defined, non-enhancing **midline cyst**
- May be limited to the pituitary gland (40%) but is more commonly seen with suprasellar extension (60%)
- MRI signal characteristics are dependent on the protein content of the cyst
  - T1WI: 50% isointense to CSF (low protein), 50% hyperintense (high protein)
  - High protein content can cause incomplete nulling of the intracystic fluid on FLAIR
- Claw sign: enhancing rim of compressed pituitary tissue around the cyst
- A small, **non-enhancing intracystic nodule** (75%) is pathognomonic

Rathke's cleft cyst: Intracystic nodule



Long arrow = intracystic nodule
Small arrows = cystic fluid
T1 hyperintense to surrounding fluid
Does not enhance



Short arrow = intracystic nodule
Long arrow = cystic fluid
T2 hypointense to surrounding fluid

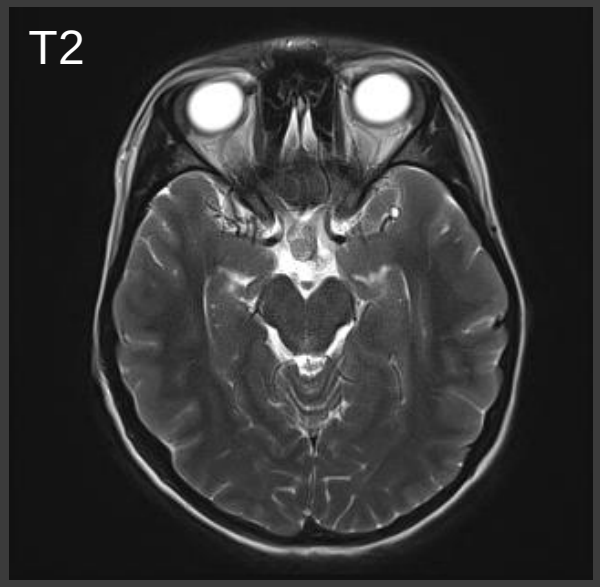
Byun, et al. *AJNR* 2000

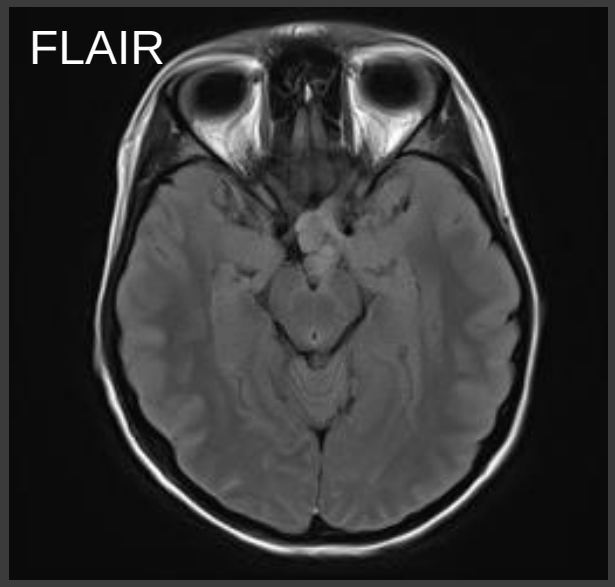
Rathke's cleft cyst: Treatment and prognosis

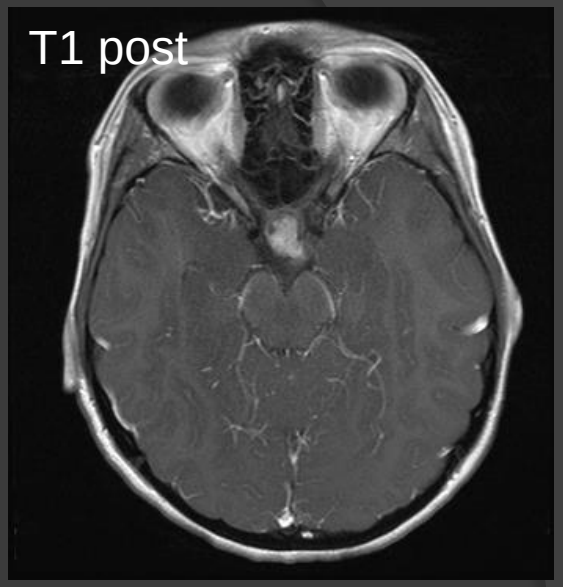
- Benign, non-neoplastic
- Surgical treatment for symptomatic cases only
- Some have been reported to resolve without therapy
- 18% recurrence rate after surgical resection

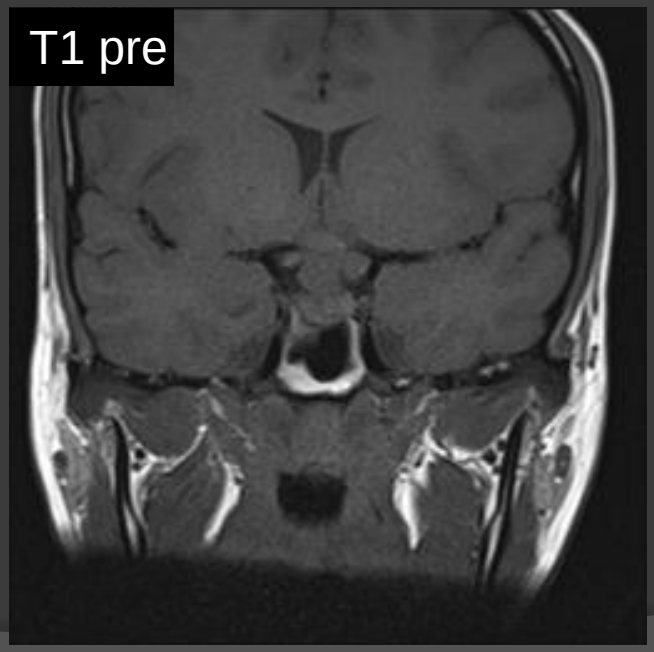
Case 3

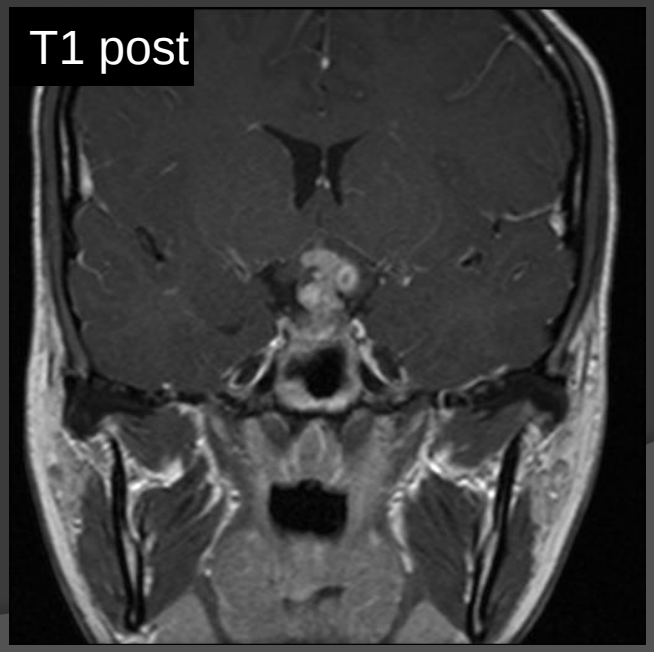
19 year old woman with chronic headaches and new eye pain, polyuria, and polydipsia.

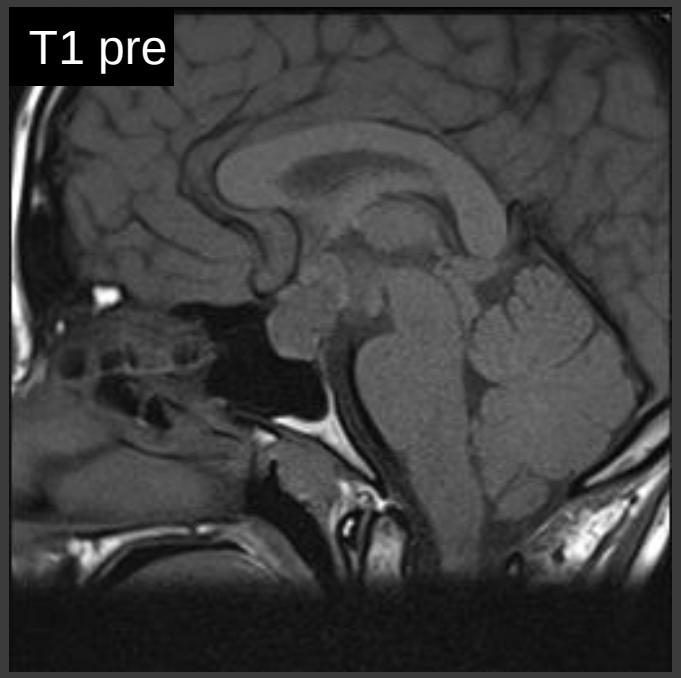

T2

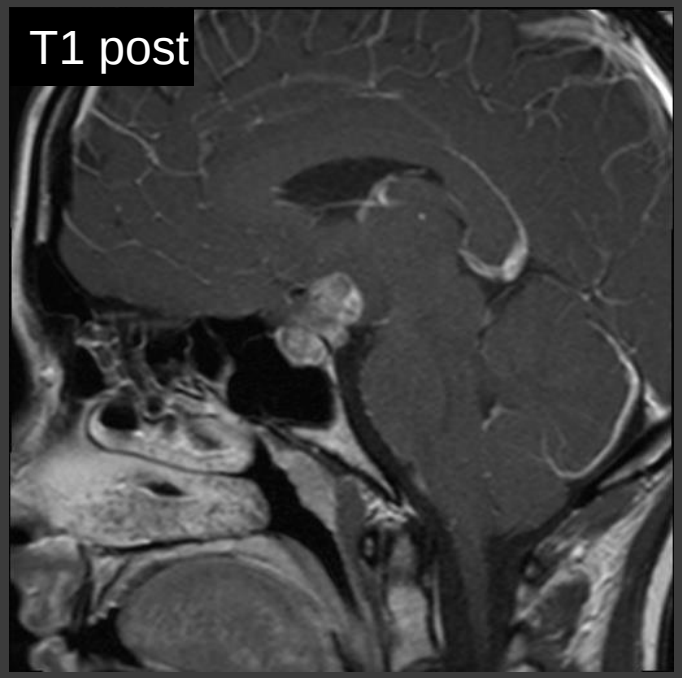

FLAIR

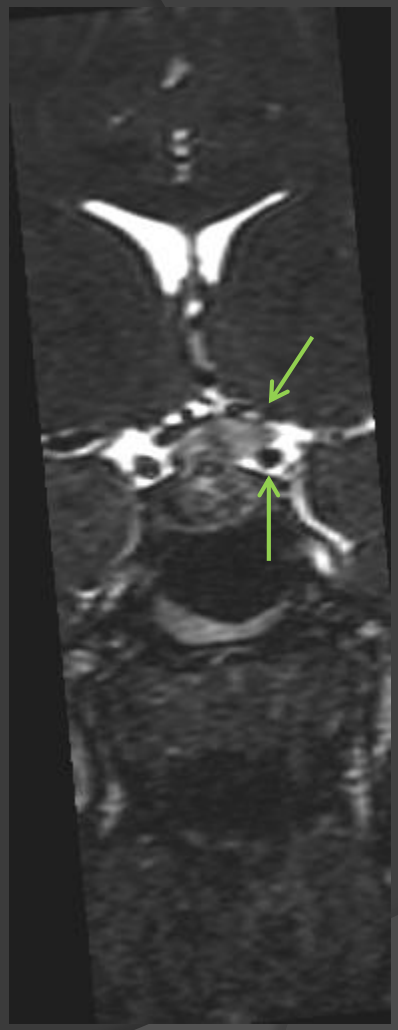

T1 post


T1 pre


T1 post

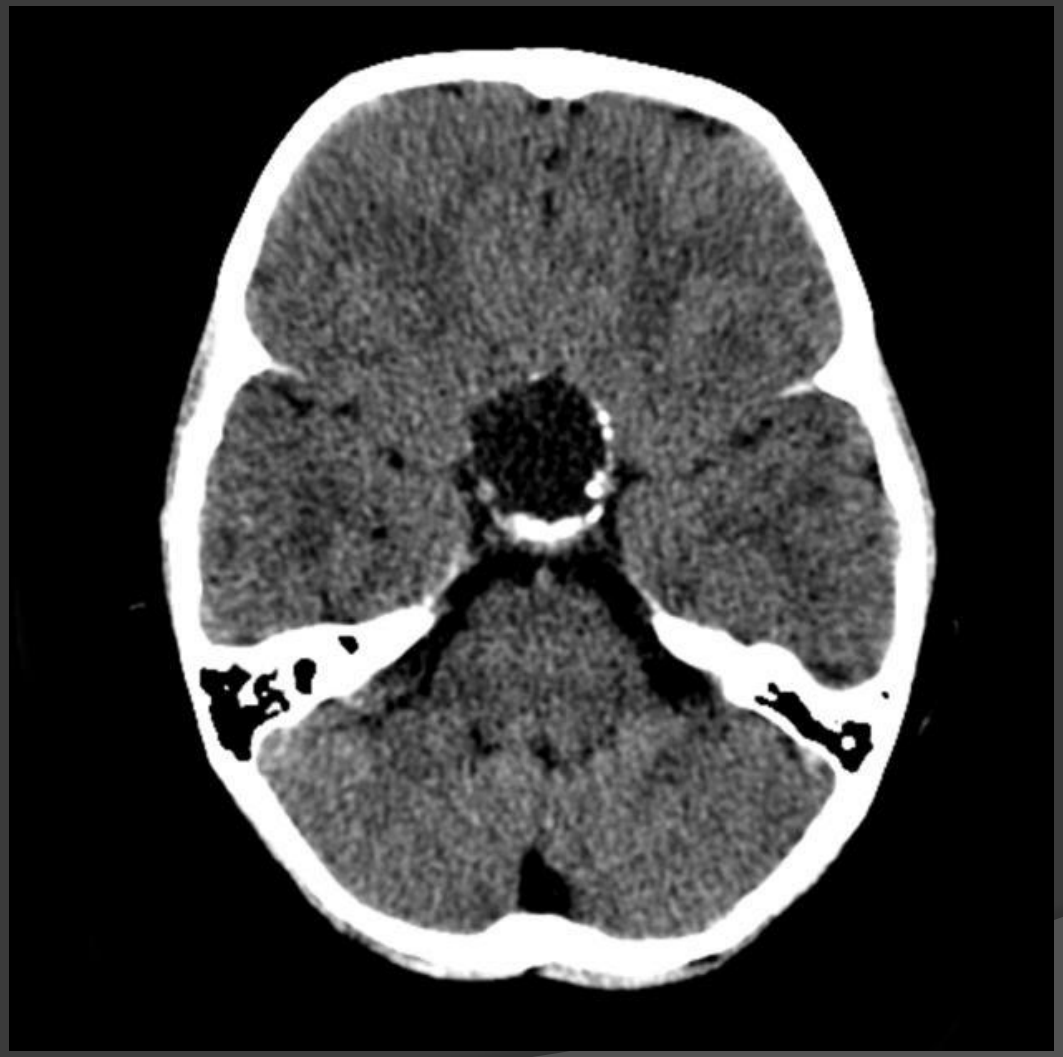






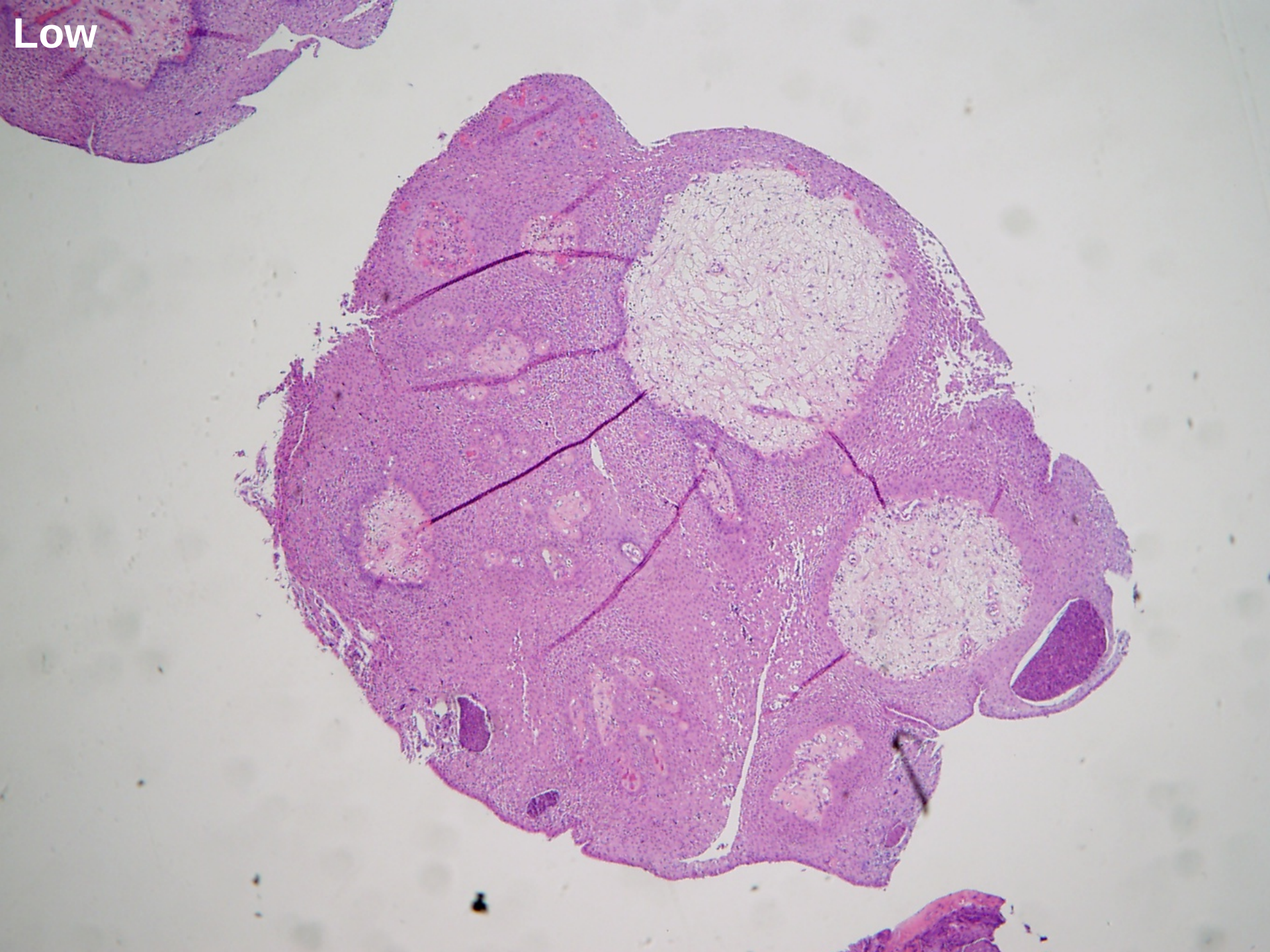
T2 coronal reformat

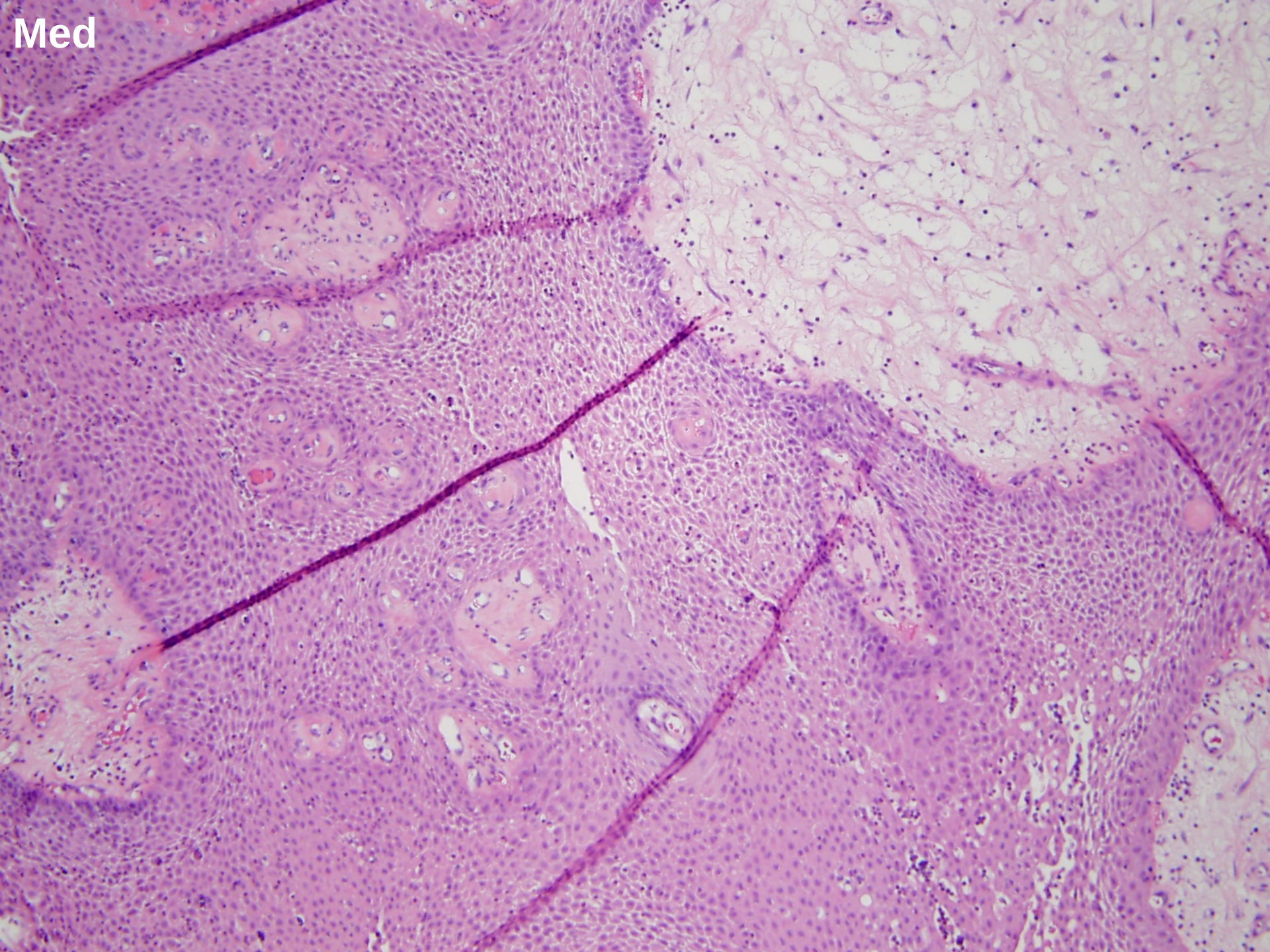
CT I- from a different patient

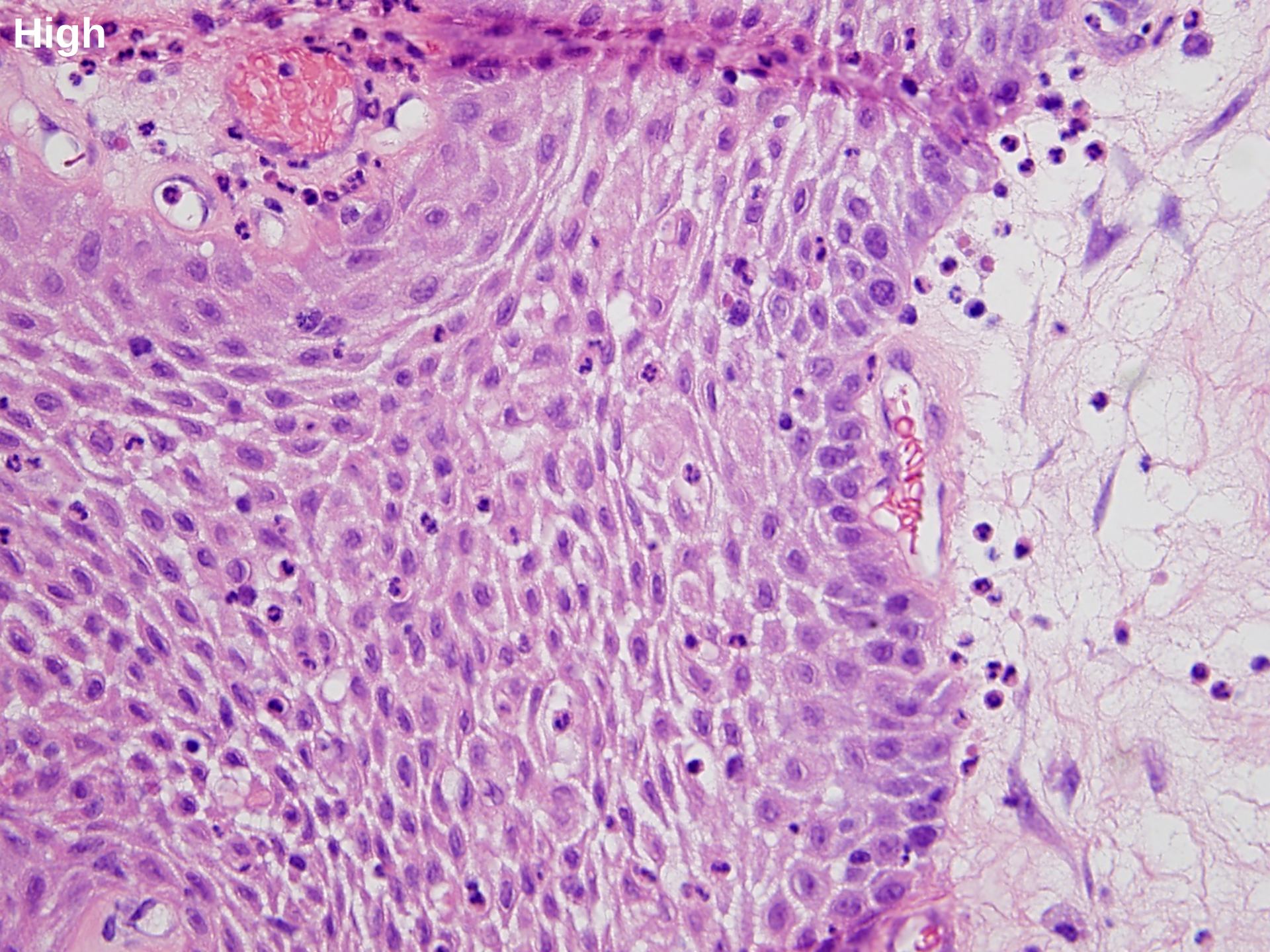


Pathology

BS-15-19512


Low


Med


High

Diagnosis

A. SPECIMEN DESIGNATED "PITUITARY TUMOR":

CRANIOPHARYNGIOMA, papillary type

Craniopharyngioma

Well-circumscribed

Composed of cores of fibrovascular stroma lined by well differentiated squamous epithelium

May separate to form pseudopapillae

Craniopharyngioma: Clinical presentation

- Most common suprasellar lesion of childhood
- Occurs in bimodal age distribution
  - 10-14 years
  - > 50 years
- Arises from squamous epithelial remnants of Rathke's pouch
- Clinical presentation is variable, but can include headache, visual symptoms, and hypopituitarism

Craniopharyngioma: Imaging characteristics

- 75% suprasellar and intrasellar
  - 20% suprasellar only
  - <5% intrasellar only
  - Rare intraventricular or ectopic
- Almost always separate from the pituitary
- Complex **cystic** mass with some **solid components**
  - Cysts T1WI hyperintense due to protein or blood products ("machine oil" on gross exam)
- Avid **enhancement** of solid elements and cyst walls
- Has potential for enamel production and almost always **calcifies**
- Can grow to distort the optic chiasm, encase vessels, or cause obstructive hydrocephalus

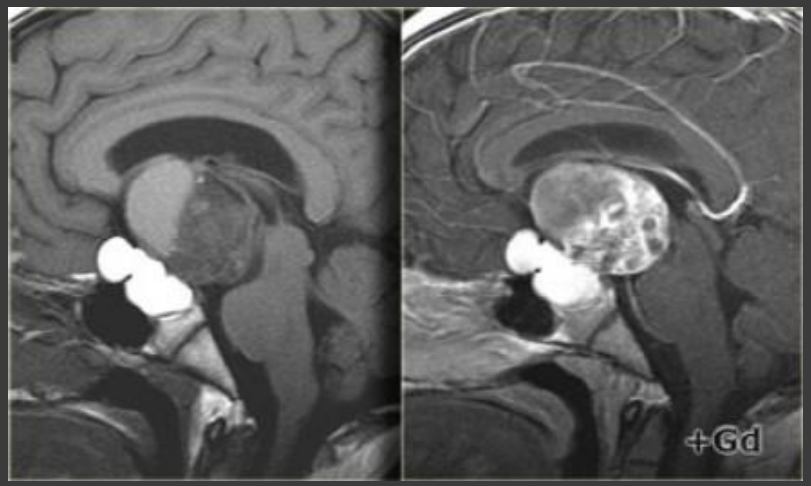
Craniopharyngioma vs. Rathke's cleft cyst

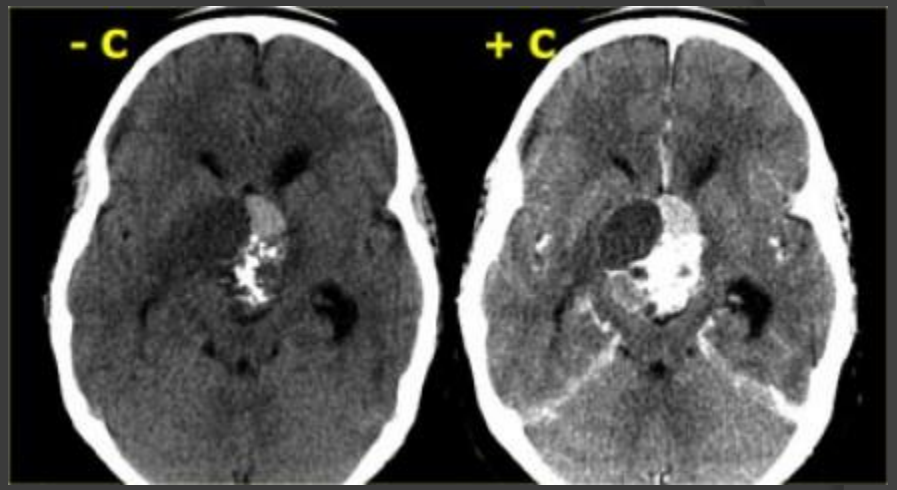
Craniopharyngioma	Rathke's cleft cyst
Remnant of embryologic Rathke's pouch	Remnant of embryologic Rathke's pouch
Squamous epithelium	Simple columnar or cuboidal epithelium
Almost always enhances	Does not enhance (except possibly rim)
Almost always calcifies	Does not calcify
Nearly always distinct from pituitary	Can be inseparable from pituitary

Craniopharyngioma: Treatment and prognosis

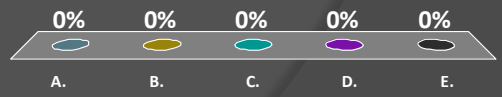
- Benign neoplasm, WHO grade I
- Most frequently treated with surgery plus radiotherapy
  - Transsphenoidal approach or craniotomy
- Local recurrence in up to 30% of patients

What is the most likely presentation for this patient with a craniopharyngioma?



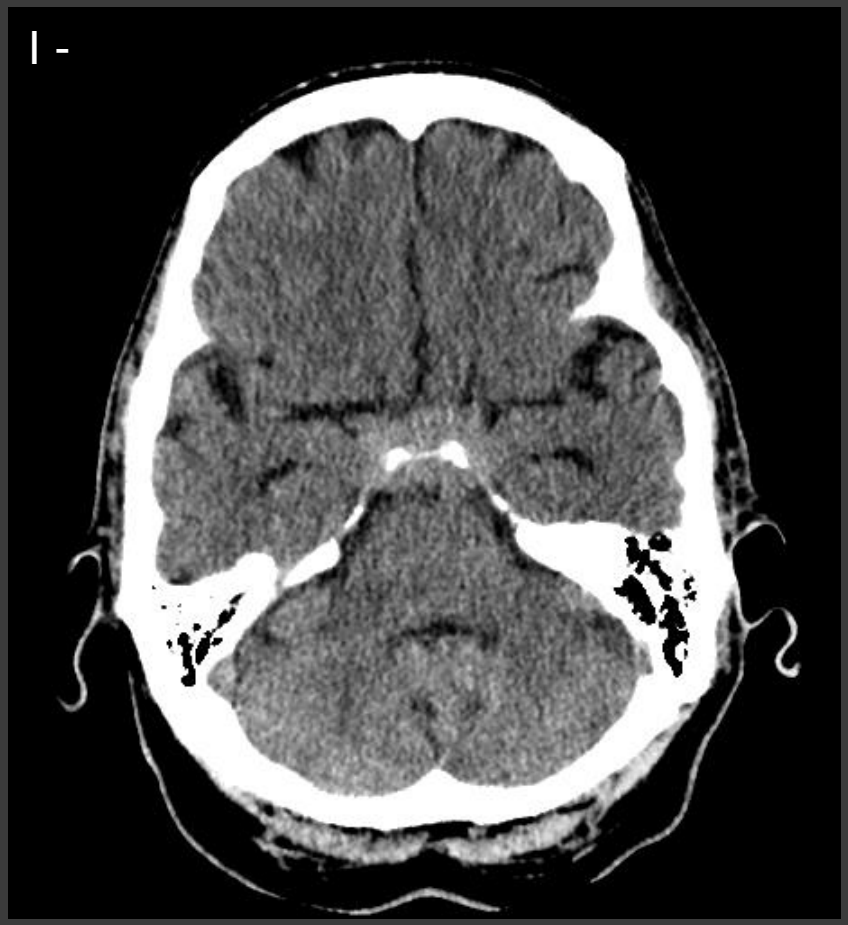


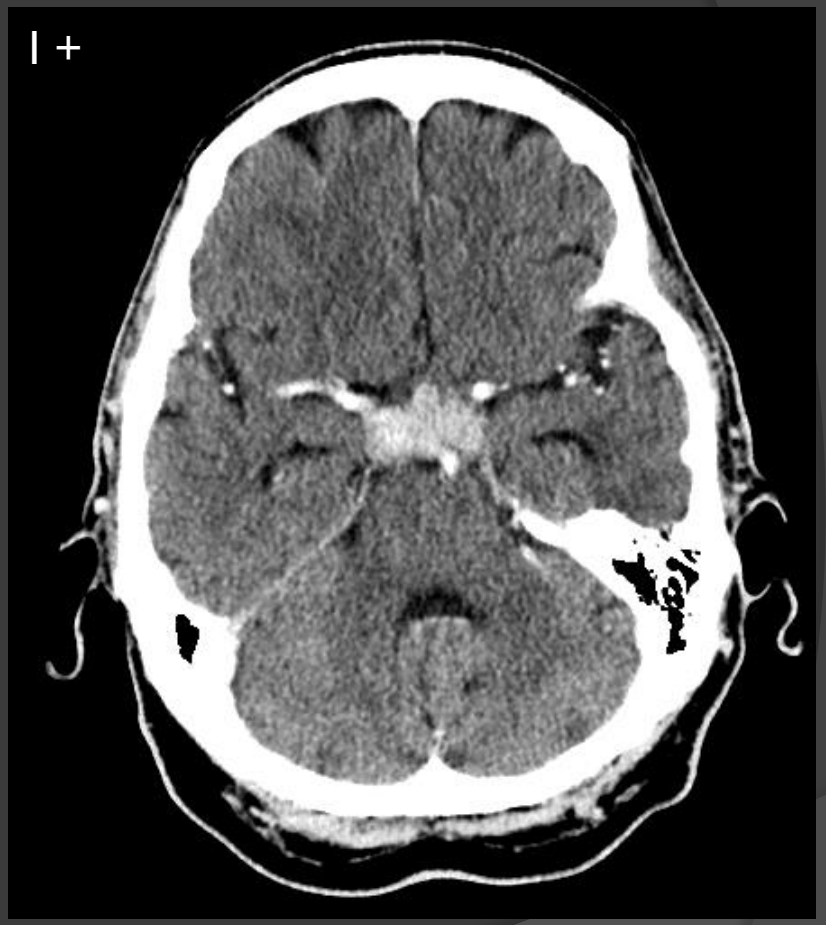
A. 10 year old girl with morning headaches and short stature
B. 14 year old boy with bitemporal hemianopsia, weight loss, and excessive sweating
C. 29 year old woman with amenorrhea and infertility
D. 55 year old man with trouble fitting on his wedding ring and hat
E. 82 year old asymptomatic woman

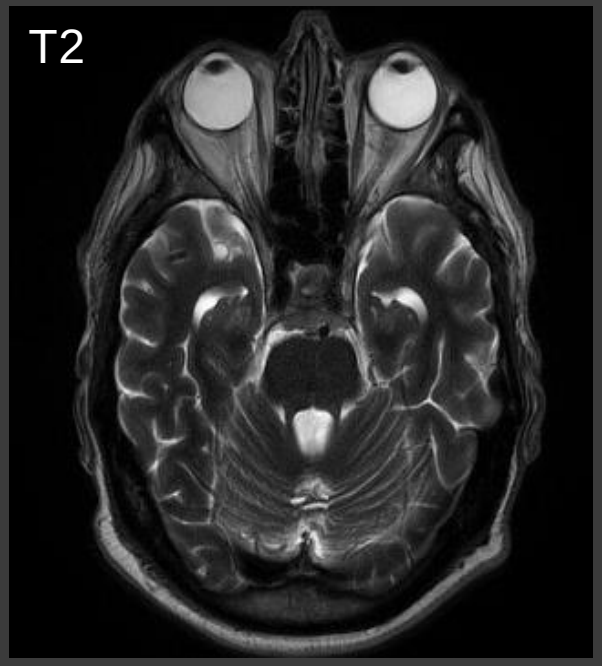


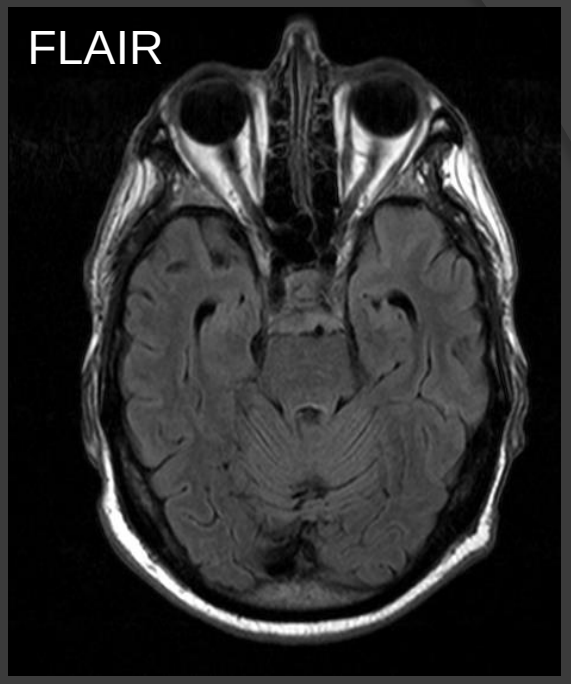
Case 4

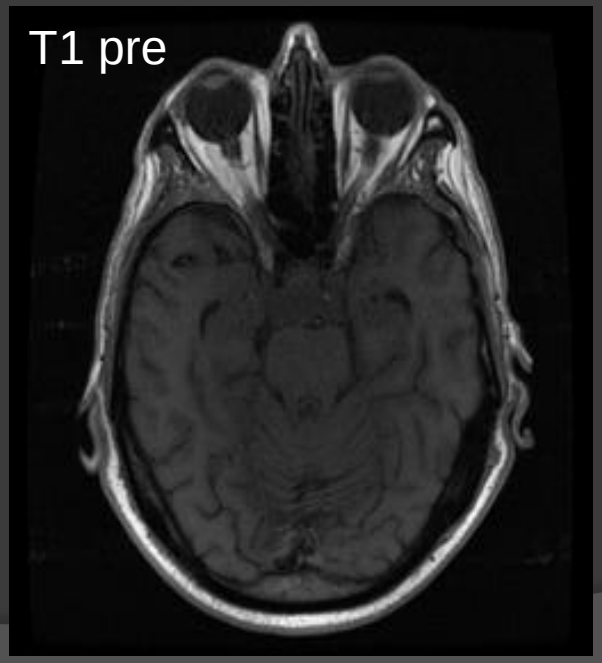
60 year old man with acute headache, awakening him from sleep. History of metastatic carcinoid tumor.

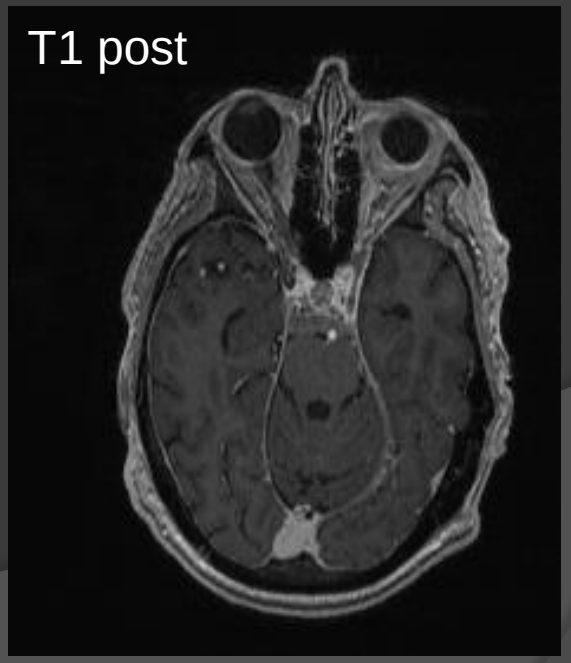

I -

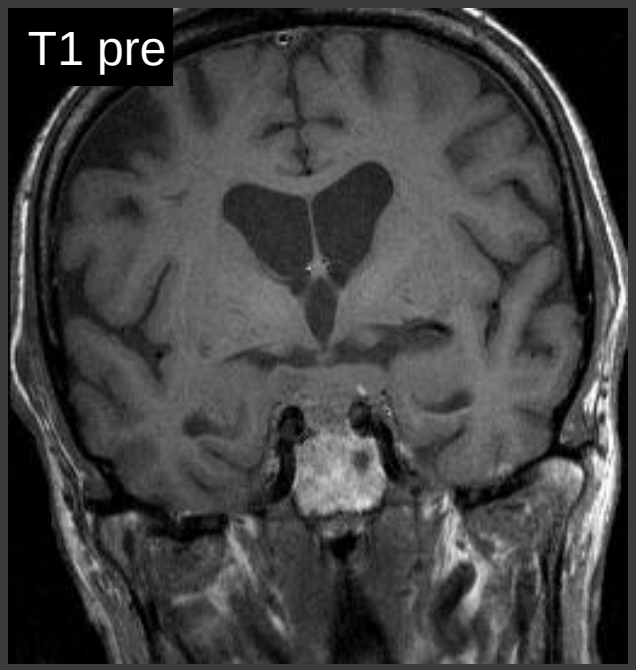

I +

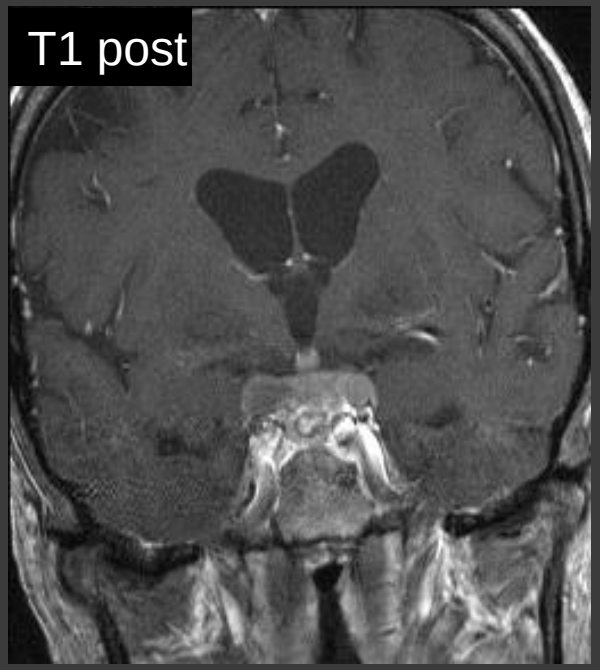

T2

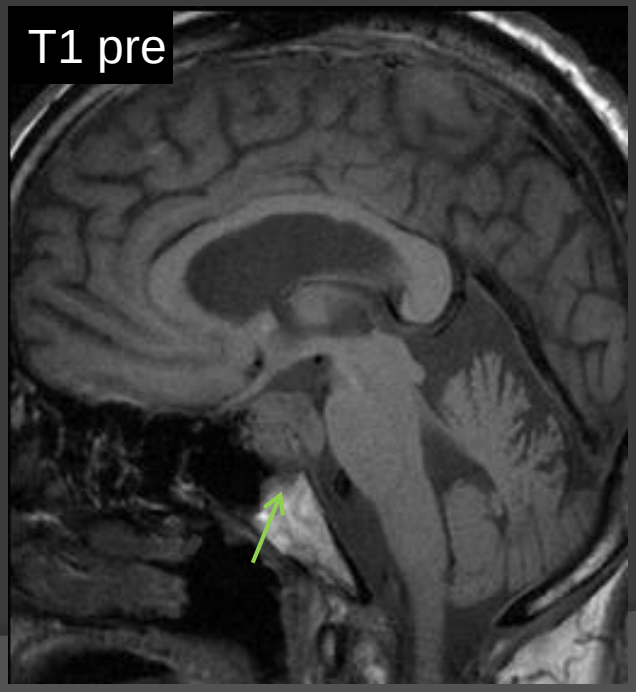

FLAIR

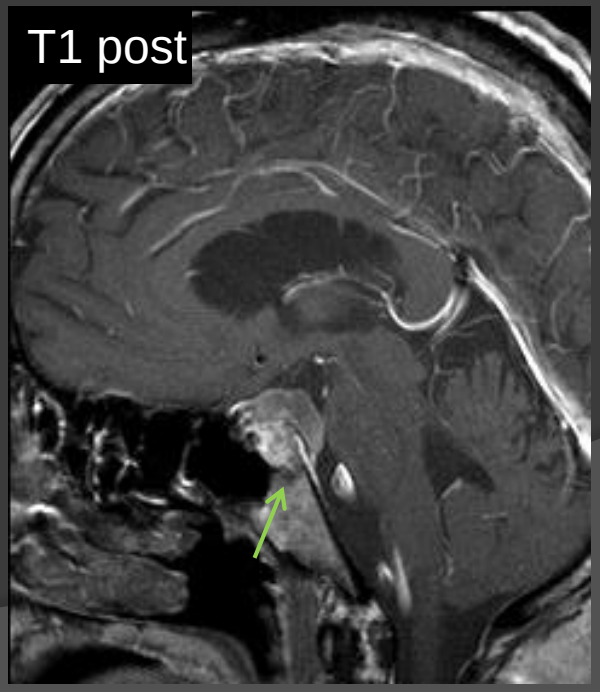

T1 pre


T1 post


T1 pre

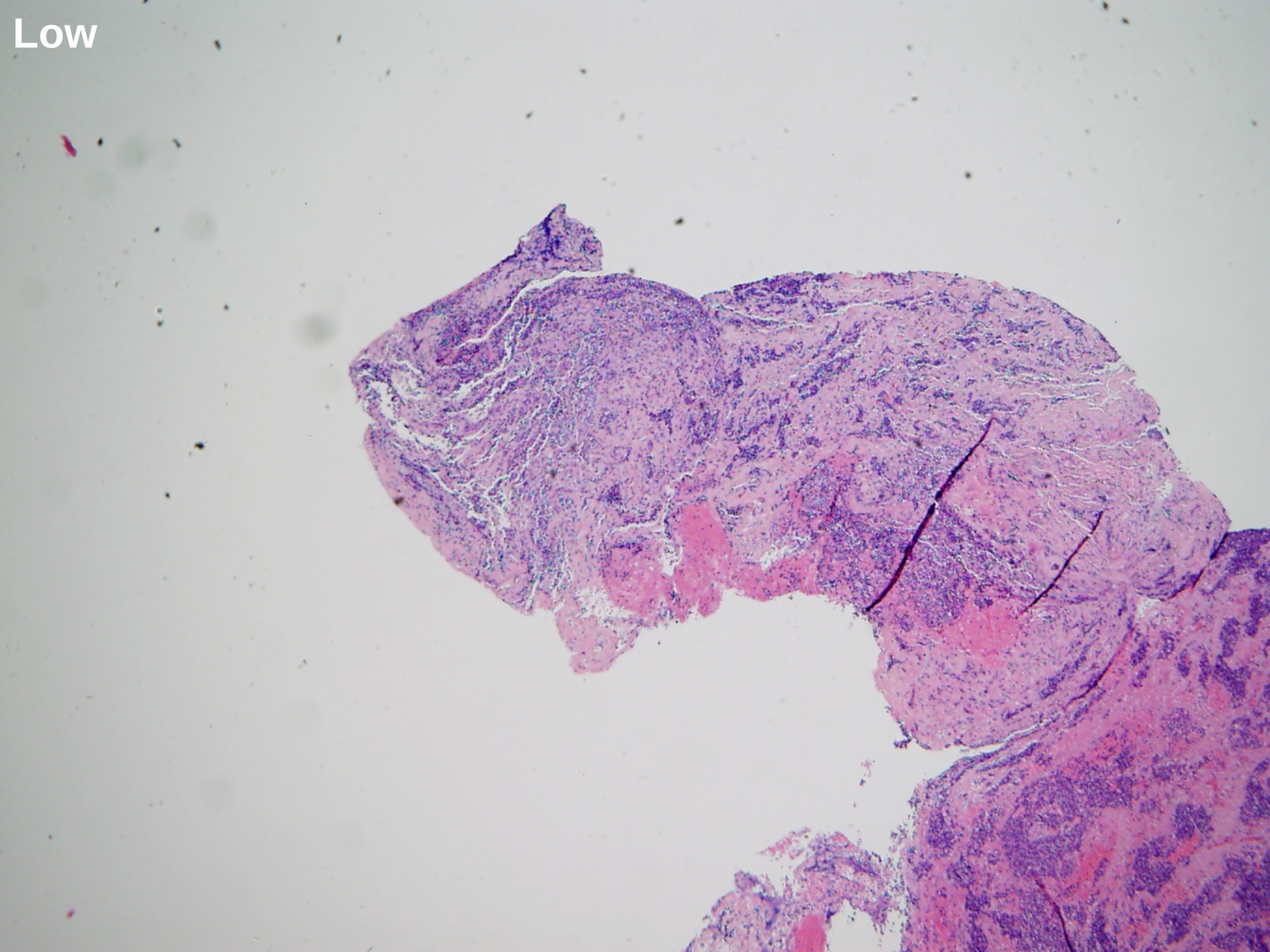

T1 post

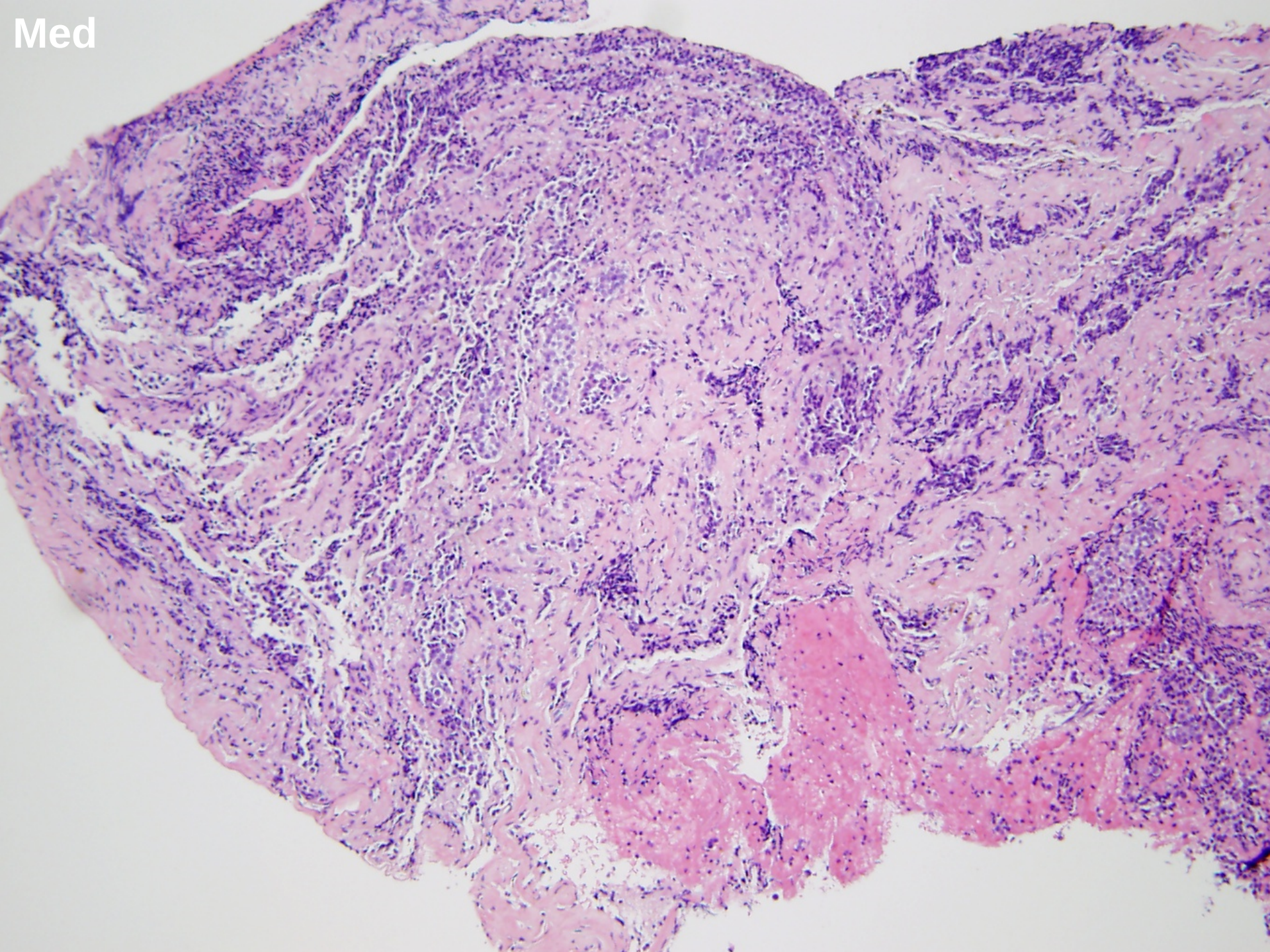

T1 pre

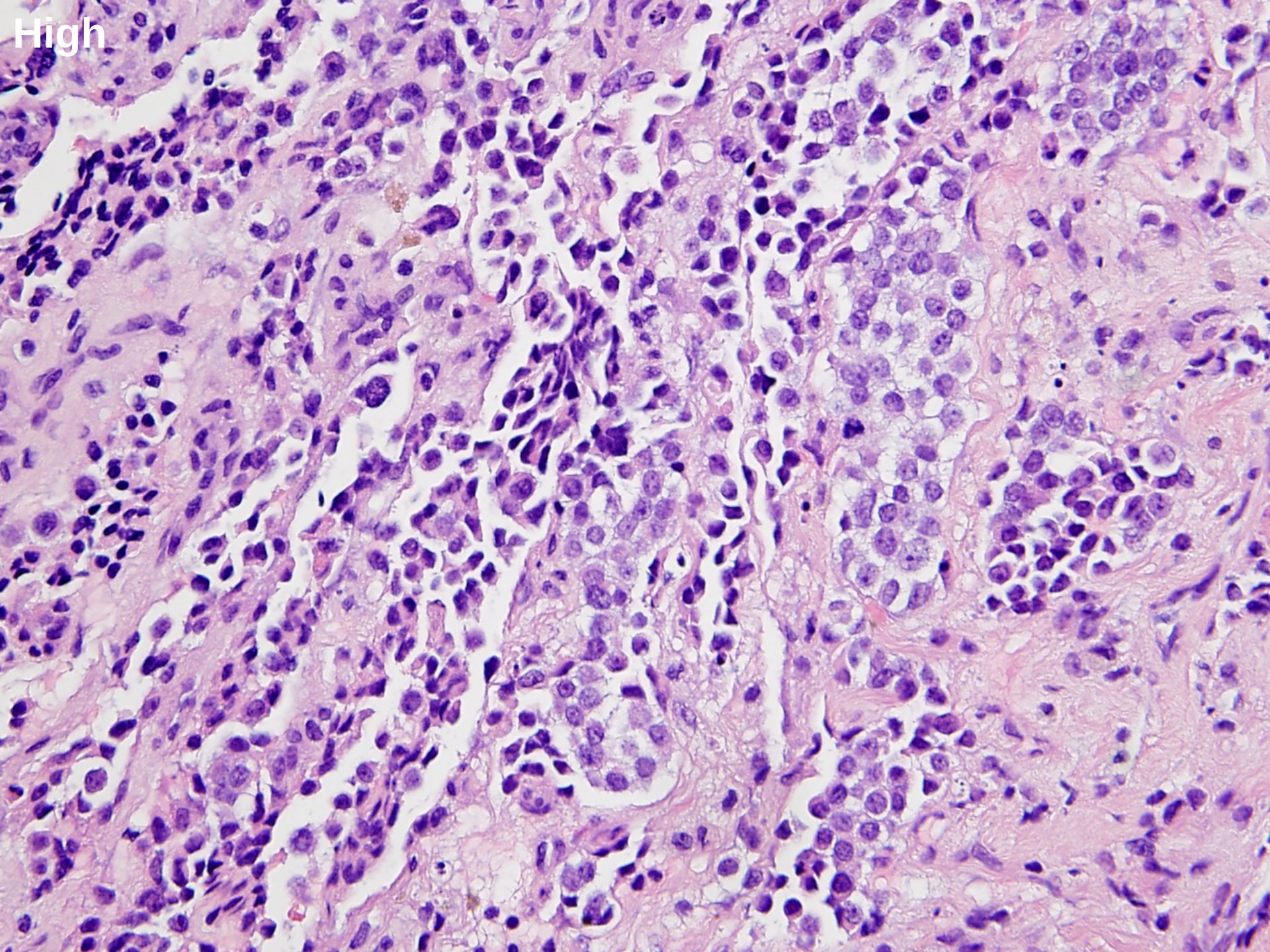

T1 post

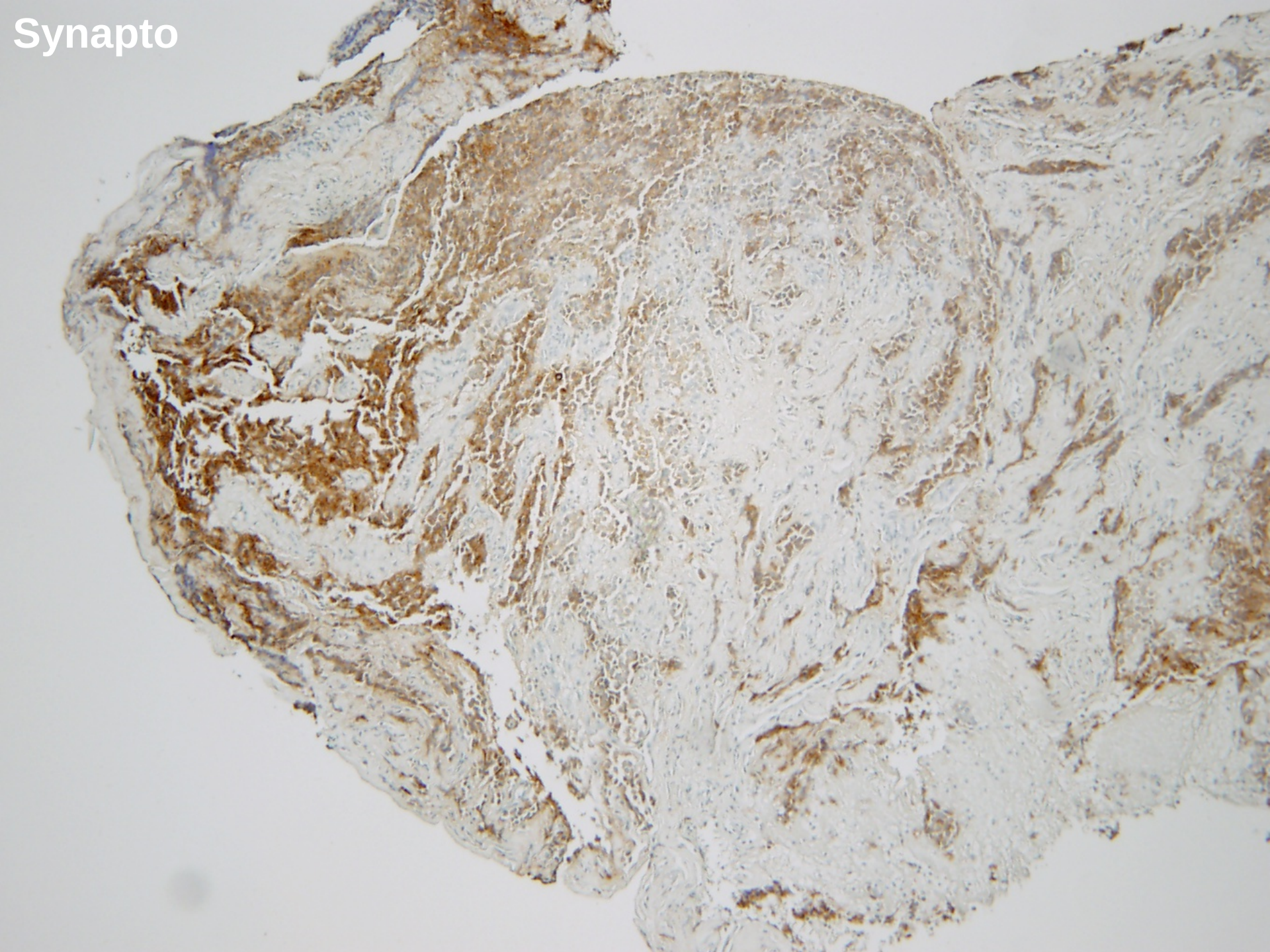
Pathology

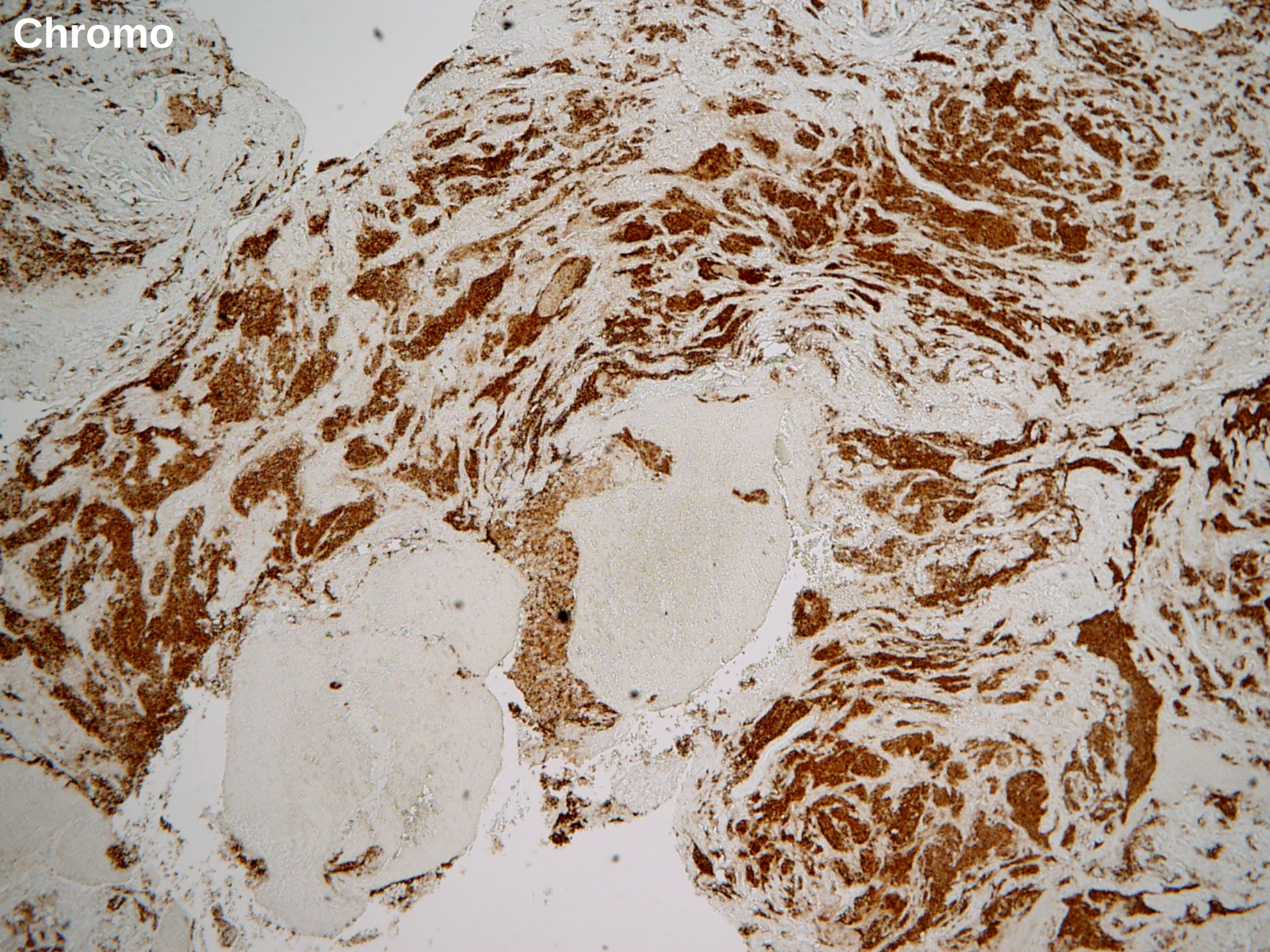
BS-13-31954

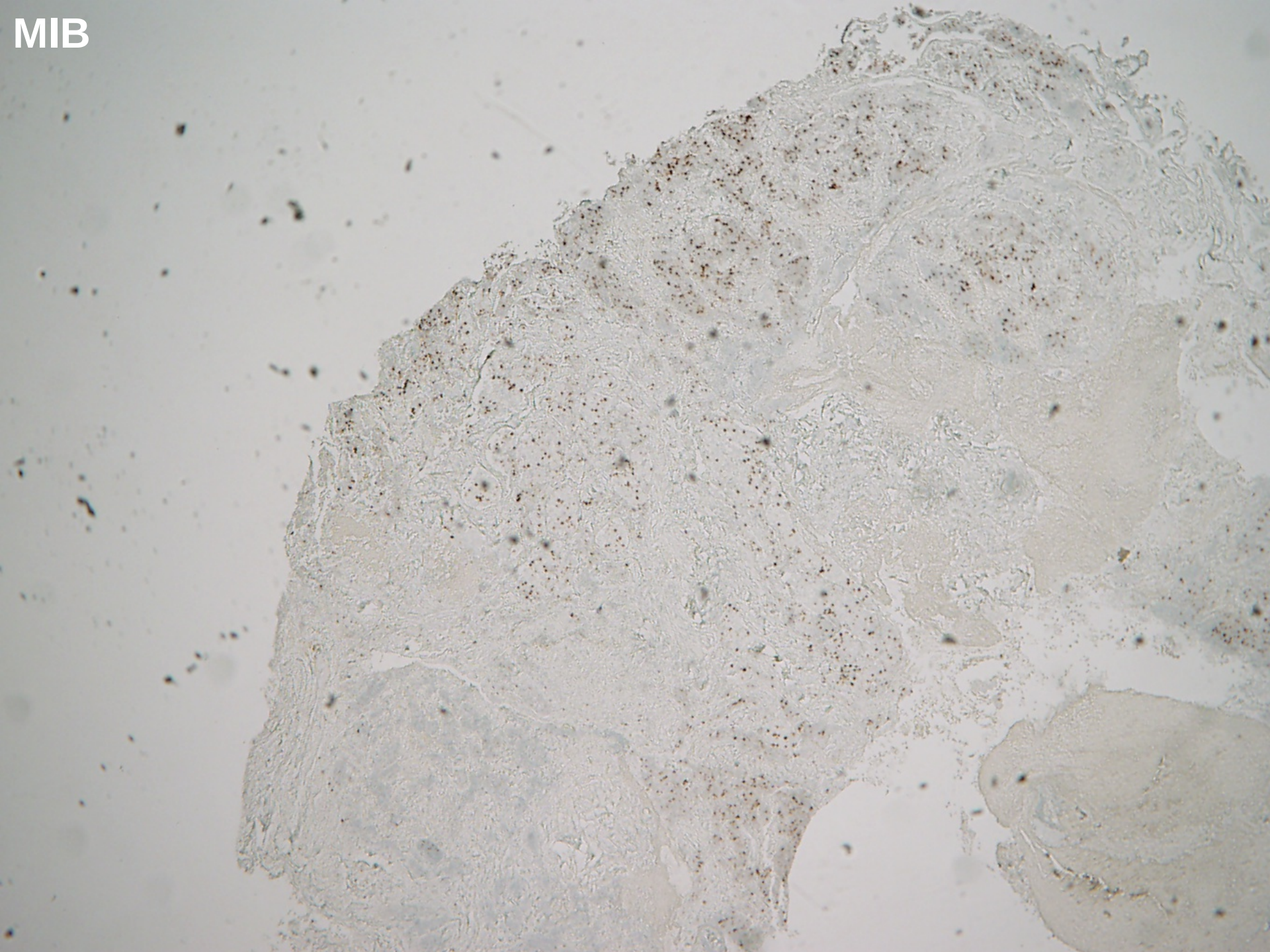

Low


Med


High


Synapto


Chromo


MIB

Diagnosis

C. SPECIMEN LABELED "3 SELLAR MASS":

Metastatic carcinoma, compatible with metastasis from the patient's known neuroendocrine carcinoma

Mitotic count is 1/10hpf. Formal proliferation index is 20.77%

Positive - Chromogranin, Synaptophysin
Negative - Pituitary hormones, P53, TTF1, CDX2, PAX8, PDX1
Note - all pituitary hormones are positively expressed in the residual pituitary gland.

Neuroendocrine Carcinoma

Pleomorphism, nuclear molding, hyperchromatic nuclei

Eccentric nuclei, coarsely stippled (salt-and-pepper) chromatin, finely granular cytoplasm

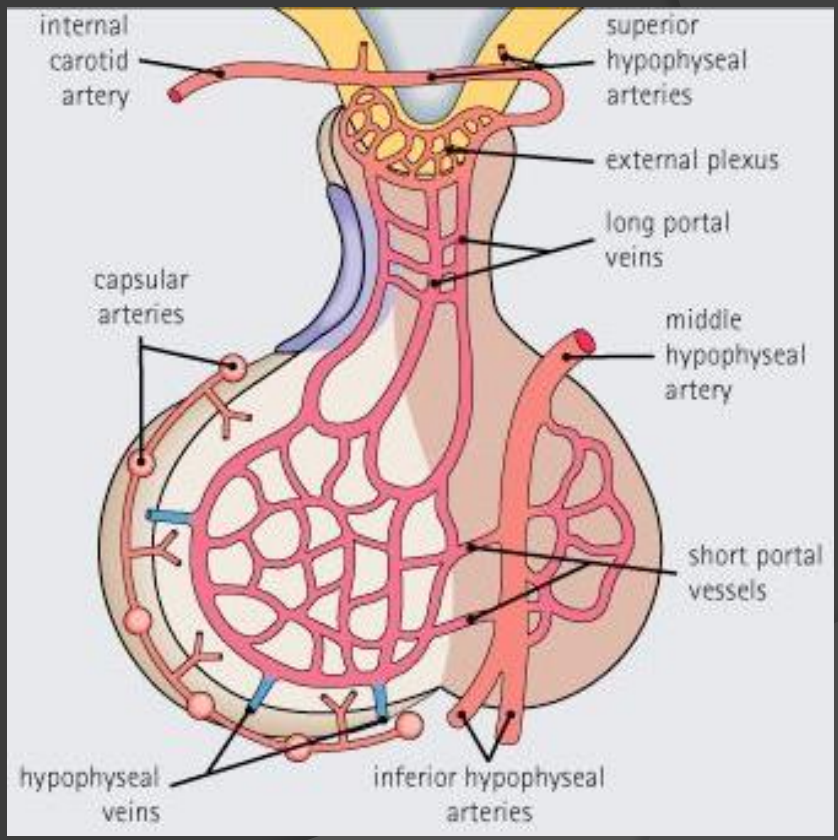
Mitoses, crush artifact, necrosis (apoptotic figures)

Neuroendocrine markers: synaptophysin, chromogranin

Mitotic rate and Ki-67 index determine grade

Metastasis: Clinical presentation

- Breast cancer is by far the most common lesion to metastasize to the parasellar region
  - Lung is most common in men
- Can present with mass effect or hormonal dysfunction
- Favors **posterior lobe** of the pituitary gland



Metastasis: Imaging characteristics

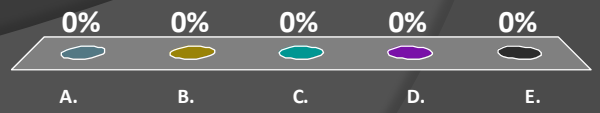
- Large mass arising from the pituitary fossa
  - Can look like macroadenoma, except:
    - Do not affect the size of the fossa (growth in a short period)
    - Cause **bony destruction** rather than remodeling
    - **Dural thickening**
    - Irregular edges
- Infundibular lesion
  - Nodular thickening and enhancement
- Posterior pituitary bright spot may be absent

Metastasis: Treatment and prognosis

- Surgical decompression to treat symptoms or to establish definitive diagnosis if uncertain
- Whole brain radiotherapy
- Typically poor prognosis (mean survival 6 months)

Which of the following is more commonly associated with pituitary metastasis than with pituitary macroadenoma?

A. Enlargement of the pituitary fossa
B. Diabetes insipidus
C. Acromegaly
D. Smooth mass margins
E. MEN1 gene mutation



Pituitary Region Mass Mnemonic

S

A

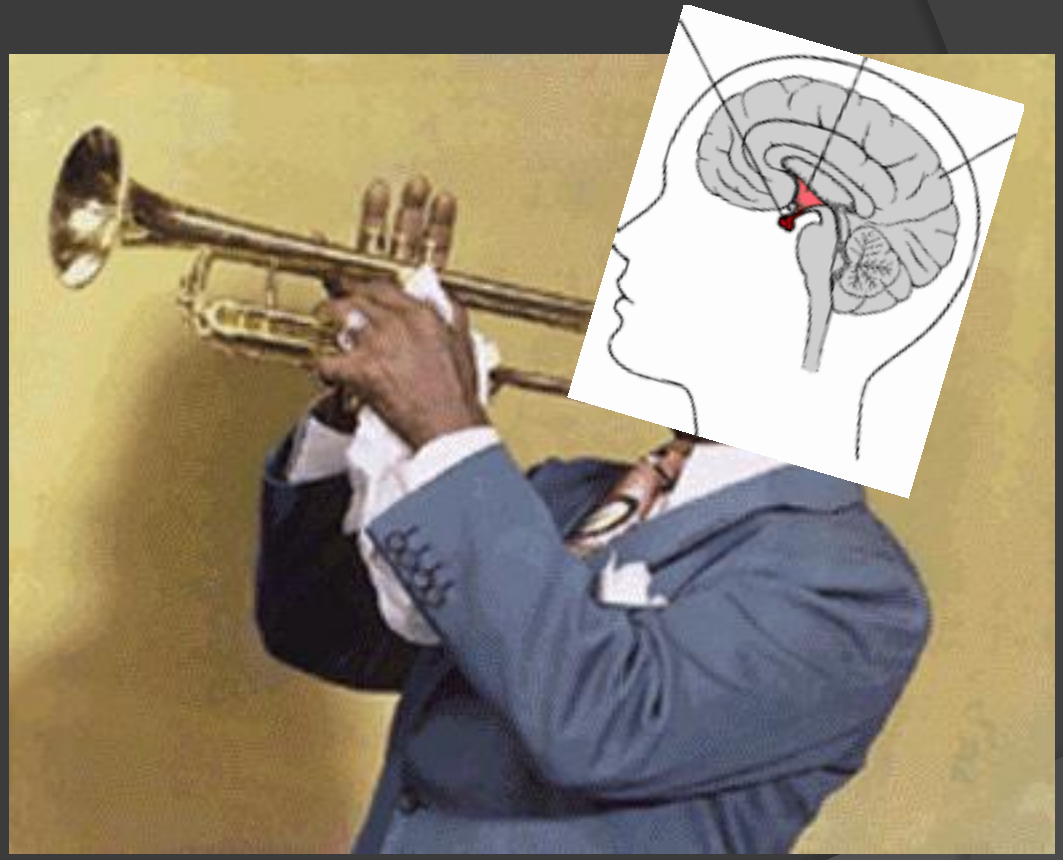
T

C

H

M

O



Mnemonic!

Sarcoidosis / Sellar tumor (adenoma)

Aneurysm

Teratoma (dermoid cyst) / Tuberculosis

Craniopharyngioma / Cleft cyst (Rathke's) / Chordoma

Hypothalamic glioma (adults) / Hypothalamic hamartoma (children) / Histiocytosis

Meningioma / Metastasis

Optic nerve glioma

Summary

- The preferred imaging modality for pituitary lesions is MRI with thin cuts through the sella
- Pituitary macroadenomas are >10 mm and slow growing, leading to “snowman” appearance and expansion of the fossa
- Rathke’s cleft cysts are midline cysts which can be proteinaceous; a non-enhancing intracystic nodule is pathognomonic
- Craniopharyngiomas are complex cysts which are typically calcified and enhance
- Pituitary metastases are most often seen in breast cancer and favor the posterior fossa
- SATCHMO is a mnemonic for pituitary region masses

Thank you!

- Tyler Janovitz
- Jeff Guenette
- Billy Wrobel
- Angela Giardino