Rad Path

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Clinical Condition: **Hearing Loss and/or Vertigo**

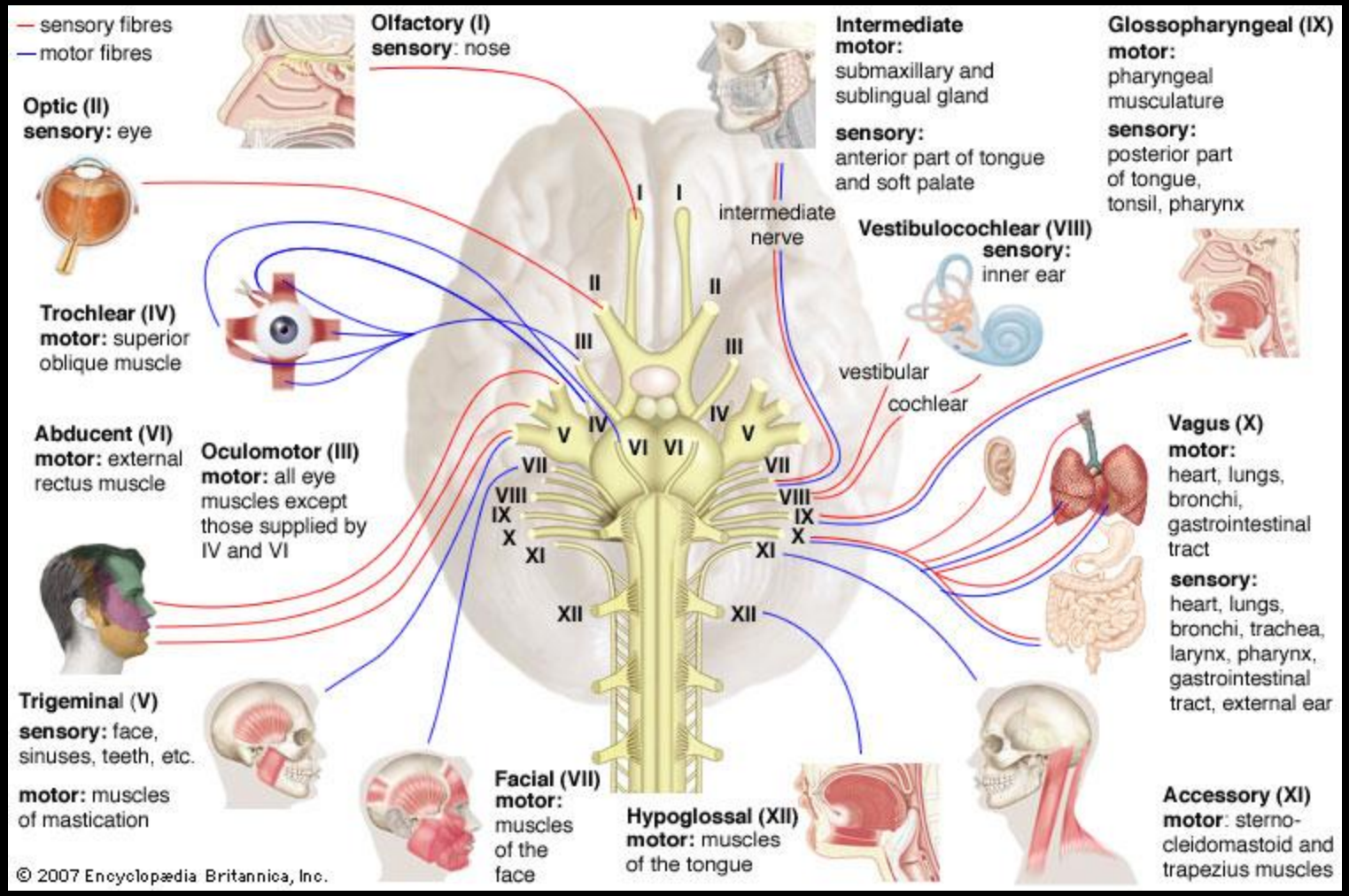
Variant 3: **Sensorineural hearing loss.**

Radiologic Procedure	Rating	Comments	RRL*
MRI head and internal auditory canal without and with contrast	9	See statement regarding contrast in text under "Anticipated Exceptions."	O
MRI head and internal auditory canal without contrast	7	If contrast cannot be administered, CISS sequences are needed.	O
CT temporal bone without contrast	6		☢☢☢
CT temporal bone with contrast	4		☢☢☢
CT head without contrast	3		☢☢☢
CT head with contrast	3		☢☢☢
CT head without and with contrast	3		☢☢☢
CT temporal bone without and with contrast	1		☢☢☢
CTA head with contrast	1		☢☢☢
MR venography head without contrast	1		O
MRA head without and with contrast	1		O
MRA head without contrast	1		O
Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			*Relative Radiation Level

Variant 4: **Mixed conductive and sensorineural hearing loss.**

Radiologic Procedure	Rating	Comments	RRL*
MRI head and internal auditory canal without and with contrast	8	See statement regarding contrast in text under "Anticipated Exceptions."	O
CT temporal bone without contrast	8	If contrast cannot be administered, CISS sequences are needed.	☢☢☢
MRI head and internal auditory canal without contrast	7		O
CT temporal bone with contrast	3		☢☢☢
CT head without contrast	3		☢☢☢
CT head with contrast	2		☢☢☢
CT head without and with contrast	2		☢☢☢
CT temporal bone without and with contrast	1		☢☢☢
CTA head with contrast	1		☢☢☢
MR venography head without contrast	1		O
MRA head without and with contrast	1		O
MRA head without contrast	1		O
Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			*Relative Radiation Level

Anatomy Review


sensory fibres
motor fibres
Olfactory (I)
sensory: nose
Optic (II)
sensory: eye
Trochlear (IV)
motor: superior oblique muscle
Abducent (VI)
motor: external rectus muscle
Oculomotor (III)
motor: all eye muscles except those supplied by IV and VI
Trigeminal (V)
sensory: face, sinuses, teeth, etc.
motor: muscles of mastication
Facial (VII)
motor: muscles of the face
Hypoglossal (XII)
motor: muscles of the tongue
Intermediate
motor: submaxillary and sublingual gland
sensory: anterior part of tongue and soft palate
intermediate nerve
Vestibulocochlear (VIII)
sensory: inner ear
vestibular
cochlear
Glossopharyngeal (IX)
motor: pharyngeal musculature
sensory: posterior part of tongue, tonsil, pharynx
Vagus (X)
motor: heart, lungs, bronchi, gastrointestinal tract
sensory: heart, lungs, bronchi, trachea, larynx, pharynx, gastrointestinal tract, external ear
Accessory (XI)
motor: sterno-cleidomastoid and trapezius muscles
I
II
III
IV
V
VI
VII
VIII
IX
X
XI
XII
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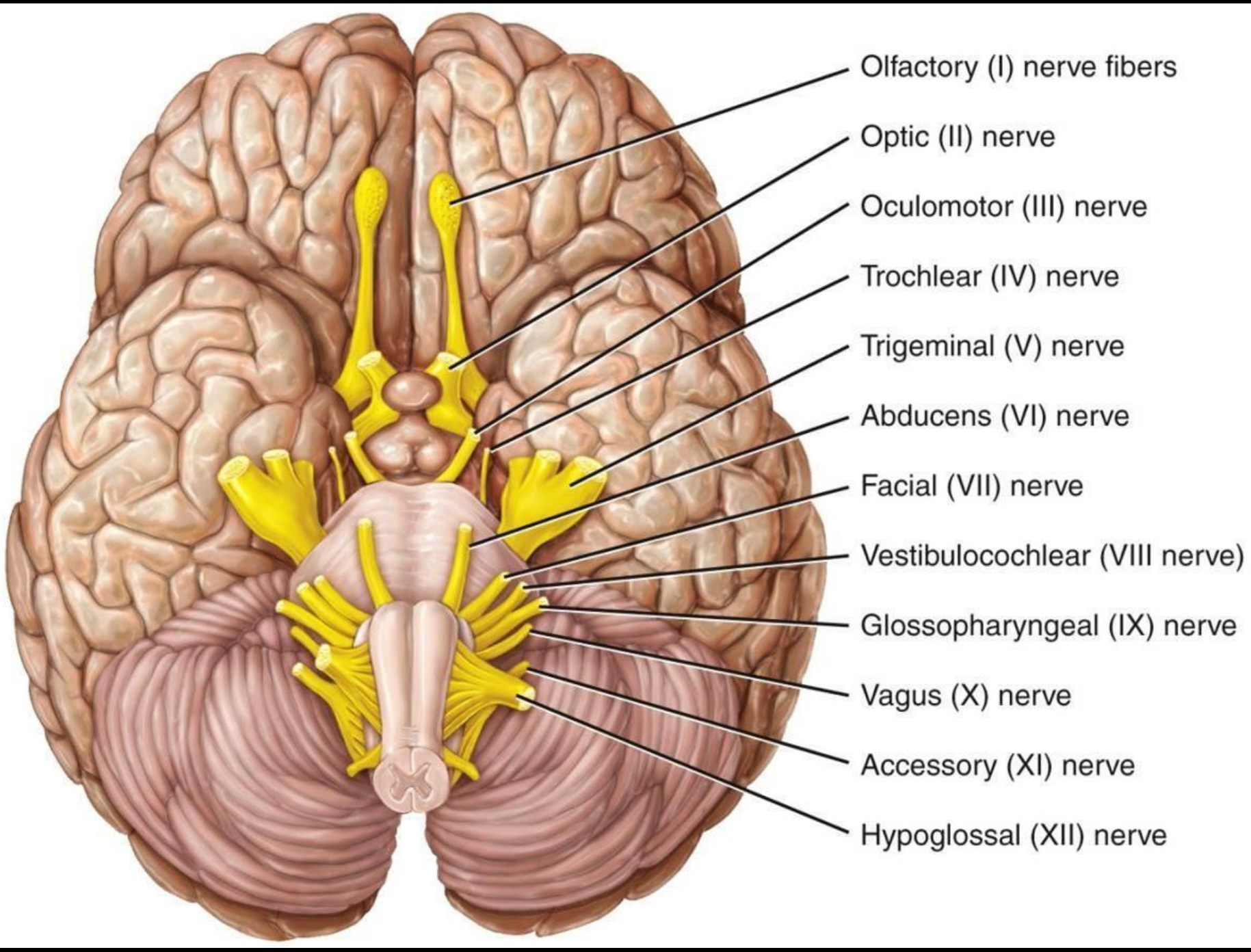
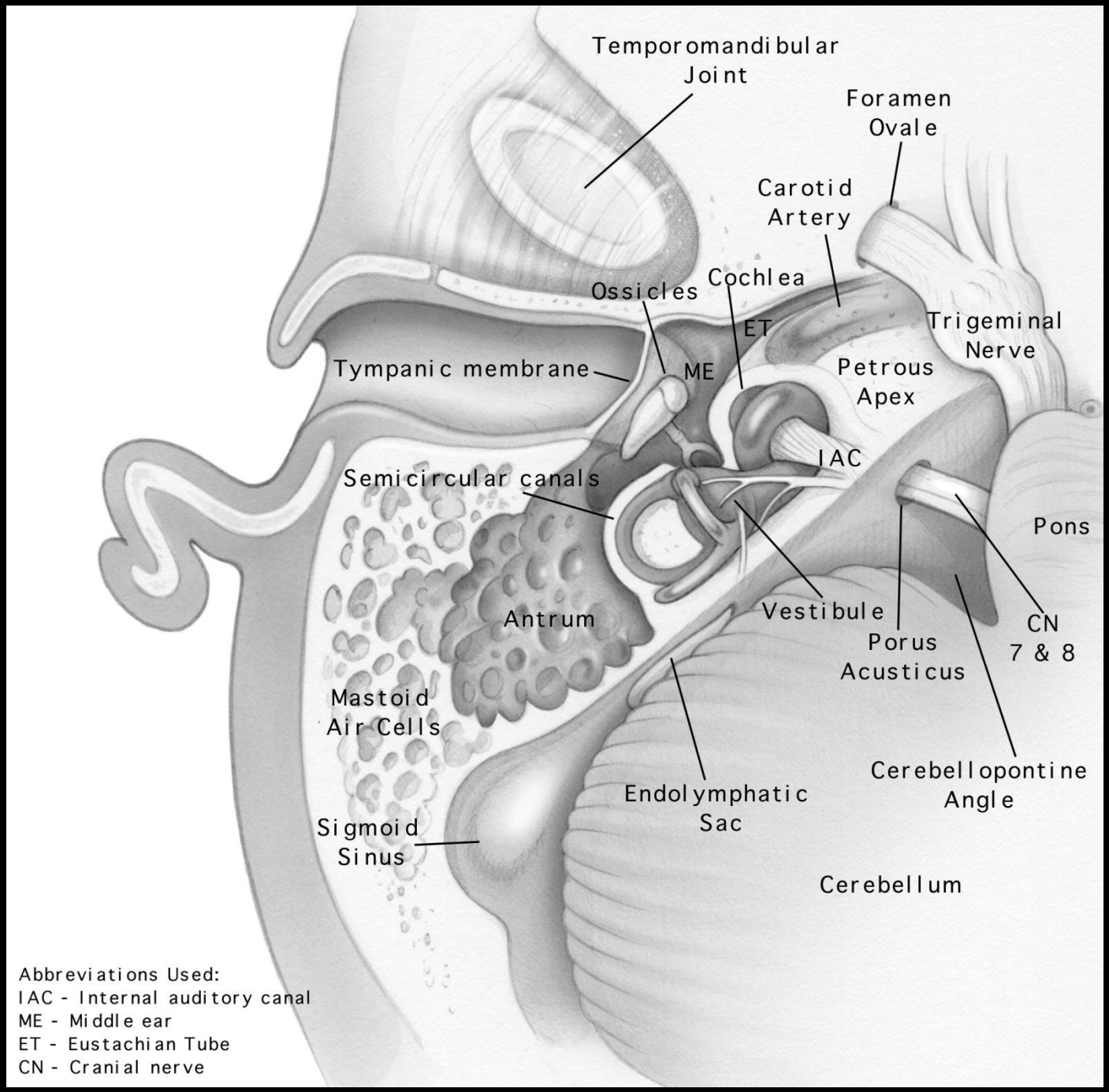
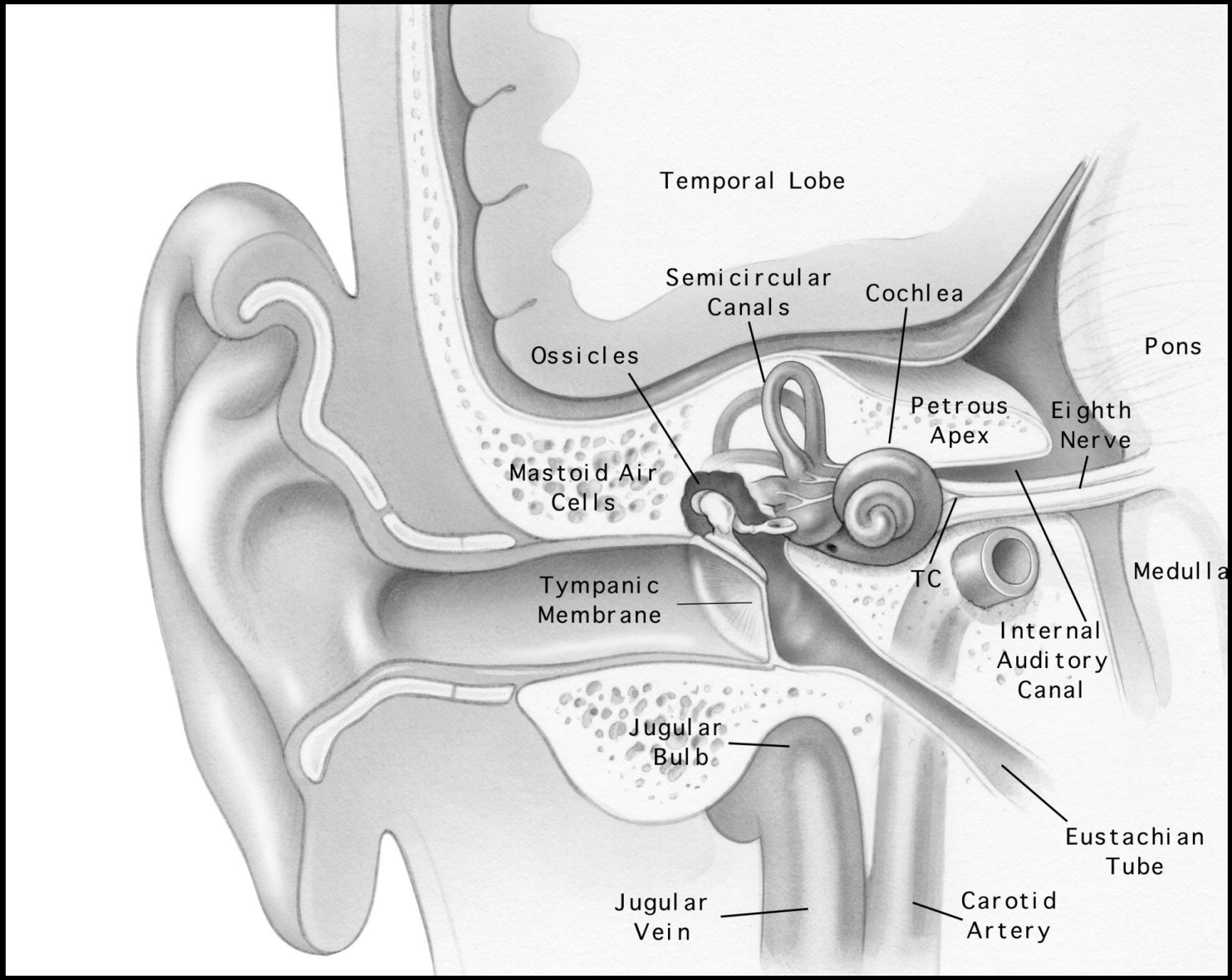

Olfactory (I) nerve fibers
Optic (II) nerve
Oculomotor (III) nerve
Trochlear (IV) nerve
Trigeminal (V) nerve
Abducens (VI) nerve
Facial (VII) nerve
Vestibulocochlear (VIII nerve)
Glossopharyngeal (IX) nerve
Vagus (X) nerve
Accessory (XI) nerve
Hypoglossal (XII) nerve

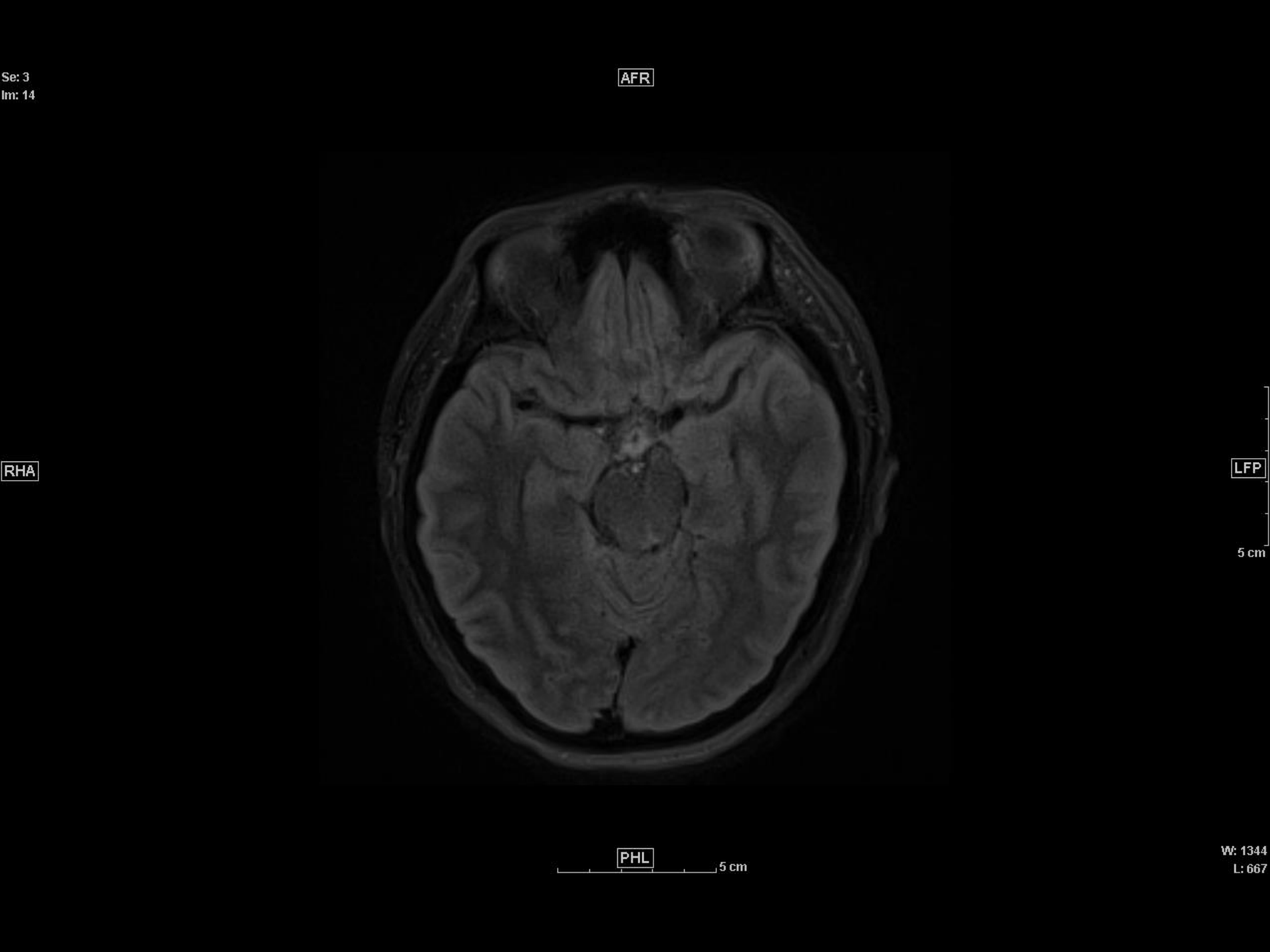

Temporomandibular Joint
Foramen Ovale
Carotid Artery
Ossicles
Cochlea
ET
Trigeminal Nerve
Tympanic membrane
ME
Petrous Apex
IAC
Semicircular canals
Pons
Antrum
Vestibule
CN 7 & 8
Porus Acusticus
Mastoid Air Cells
Cerebellopontine Angle
Endolymphatic Sac
Sigmoid Sinus
Cerebellum
Abbreviations Used:
IAC - Internal auditory canal
ME - Middle ear
ET - Eustachian Tube
CN - Cranial nerve


Temporal Lobe
Semicircular Canals
Cochlea
Pons
Ossicles
Petrous Apex
Eighth Nerve
Mastoid Air Cells
TC
Medulla
Tympanic Membrane
Internal Auditory Canal
Jugular Bulb
Eustachian Tube
Jugular Vein
Carotid Artery

Case 1

40 Female

- Vertigo x 3 weeks
- Feeling of tipping to right while walking
- Numbness tip of right tongue/lips x 5 years
- Numbness now extending to face and ear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RHA
LFP
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PHL
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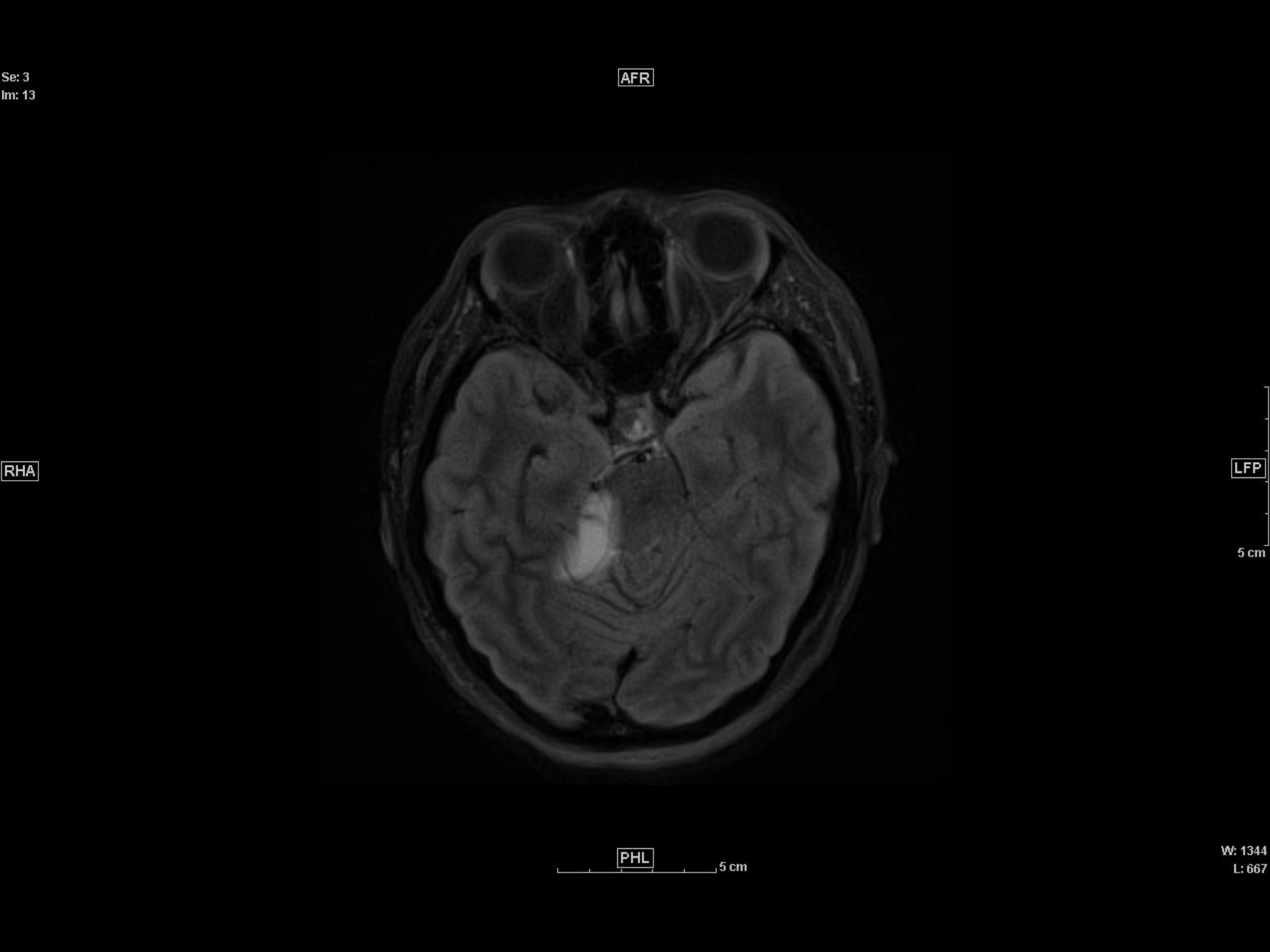
L: 667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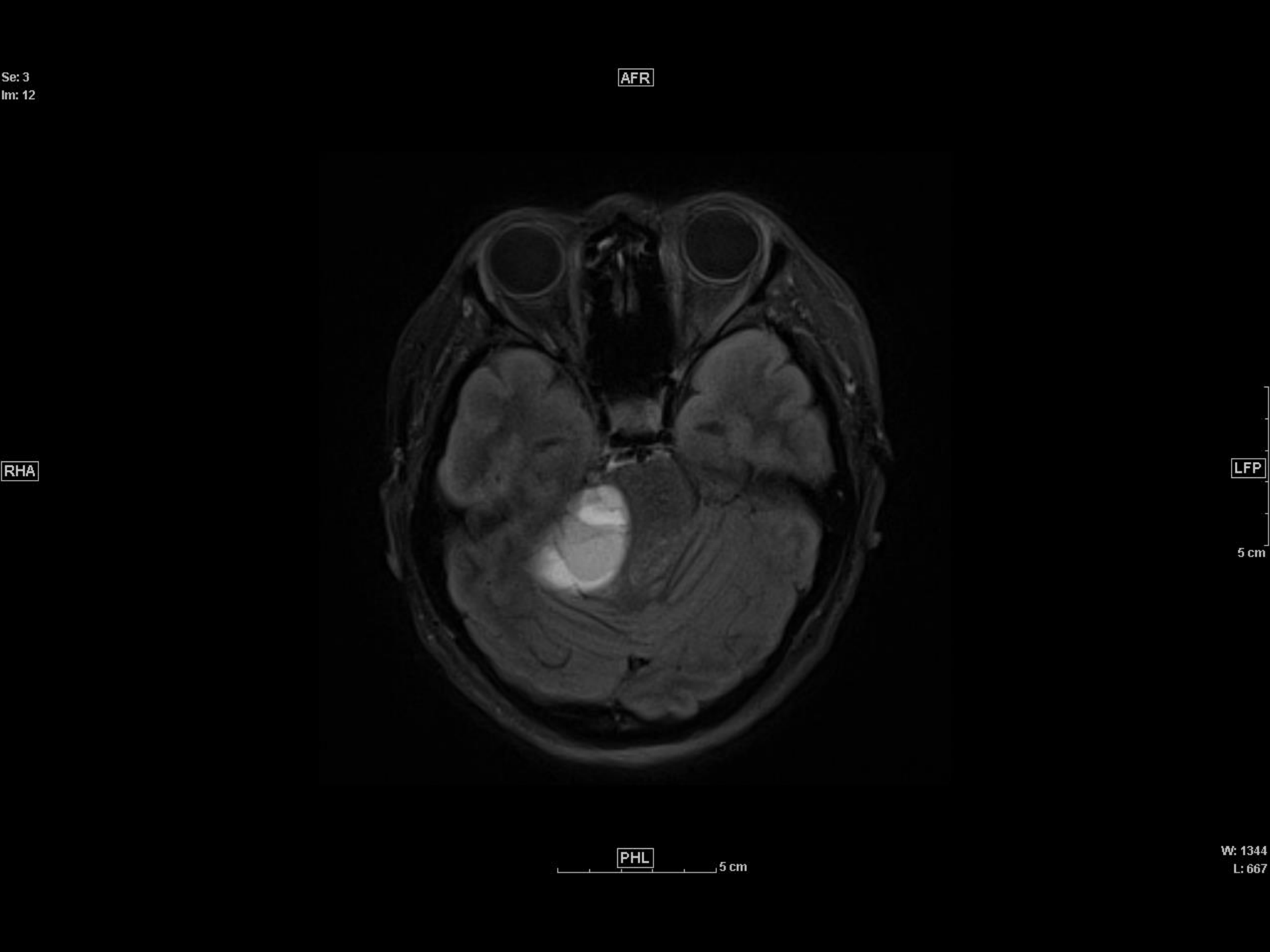
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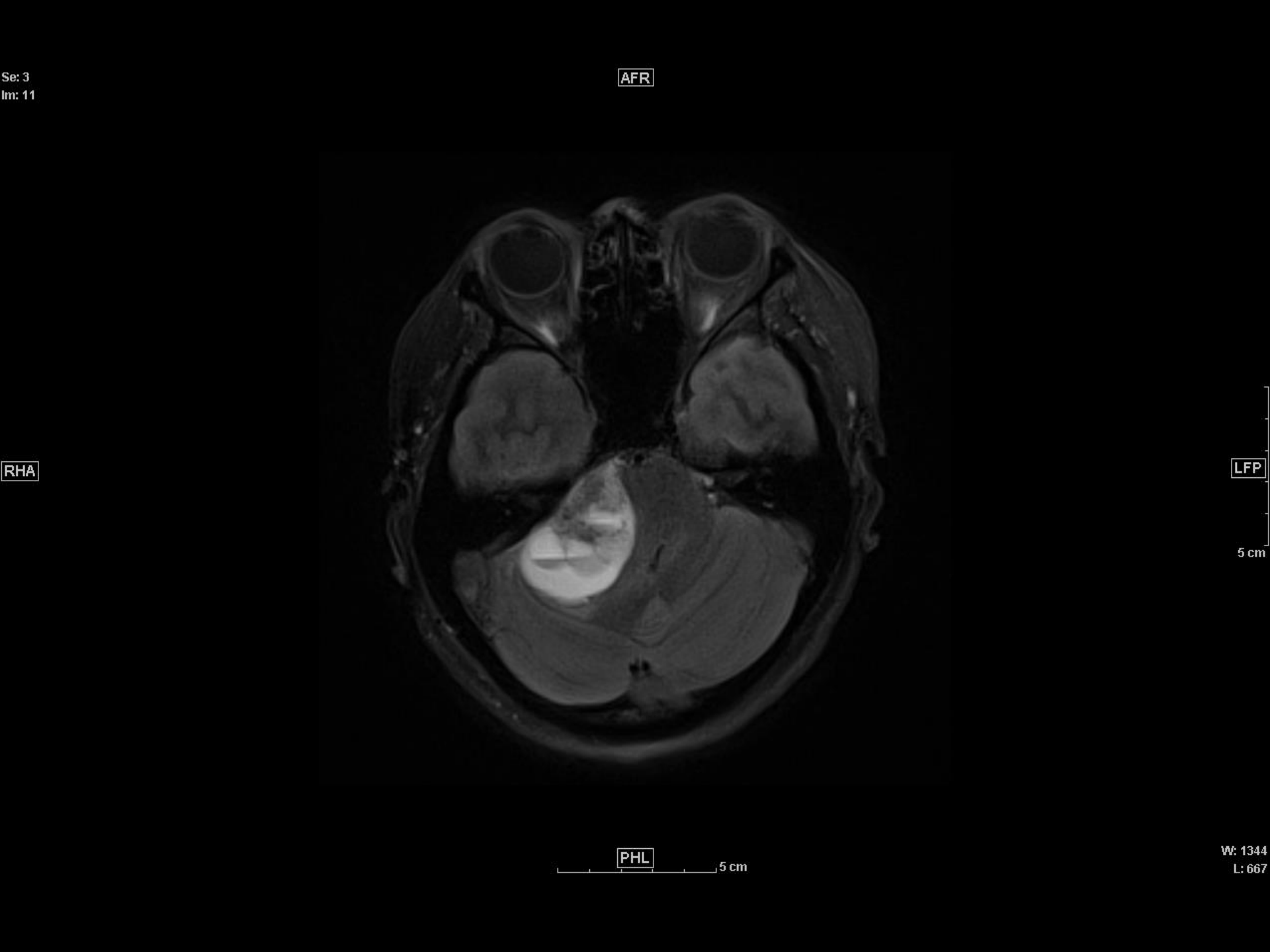
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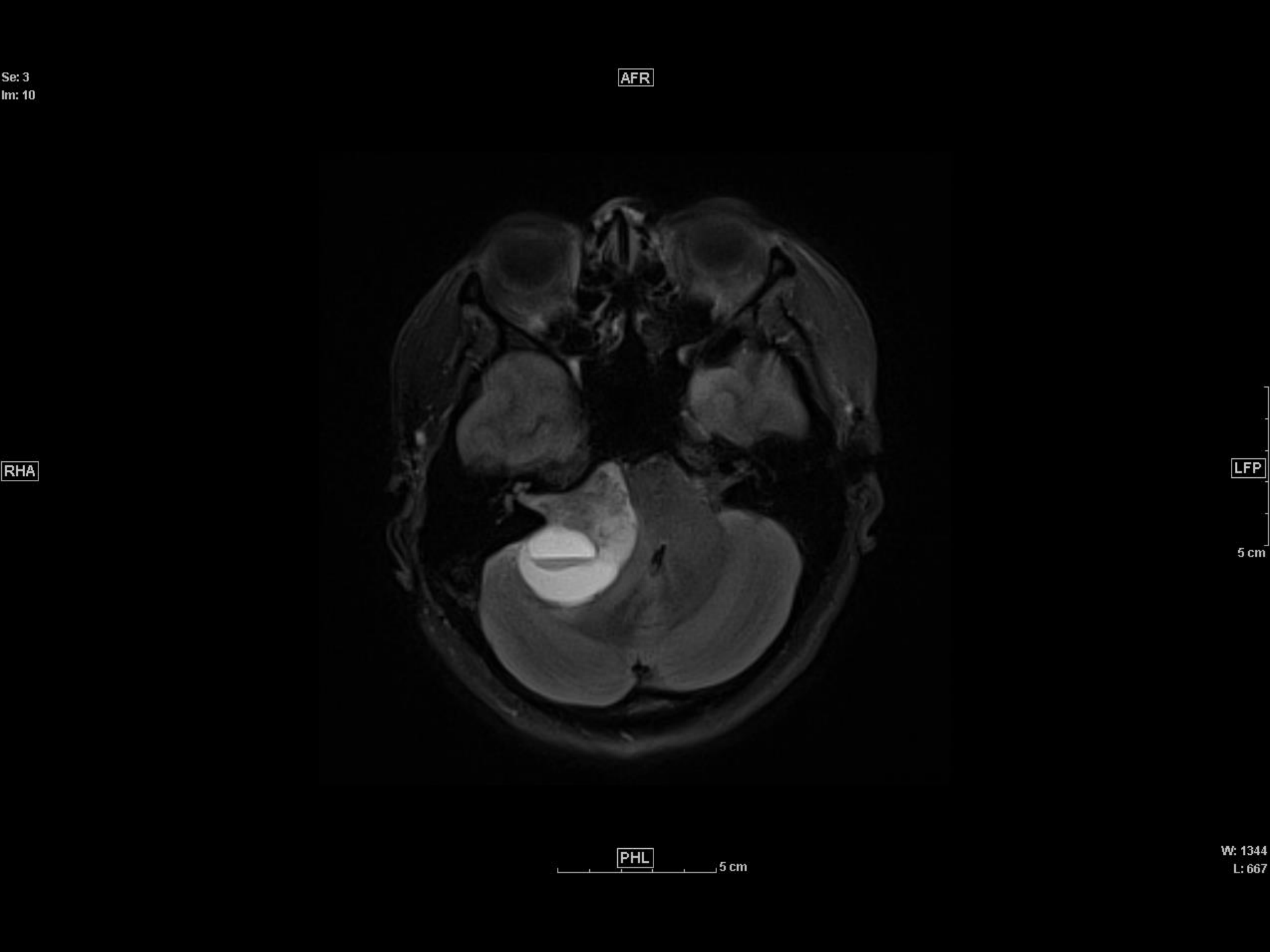
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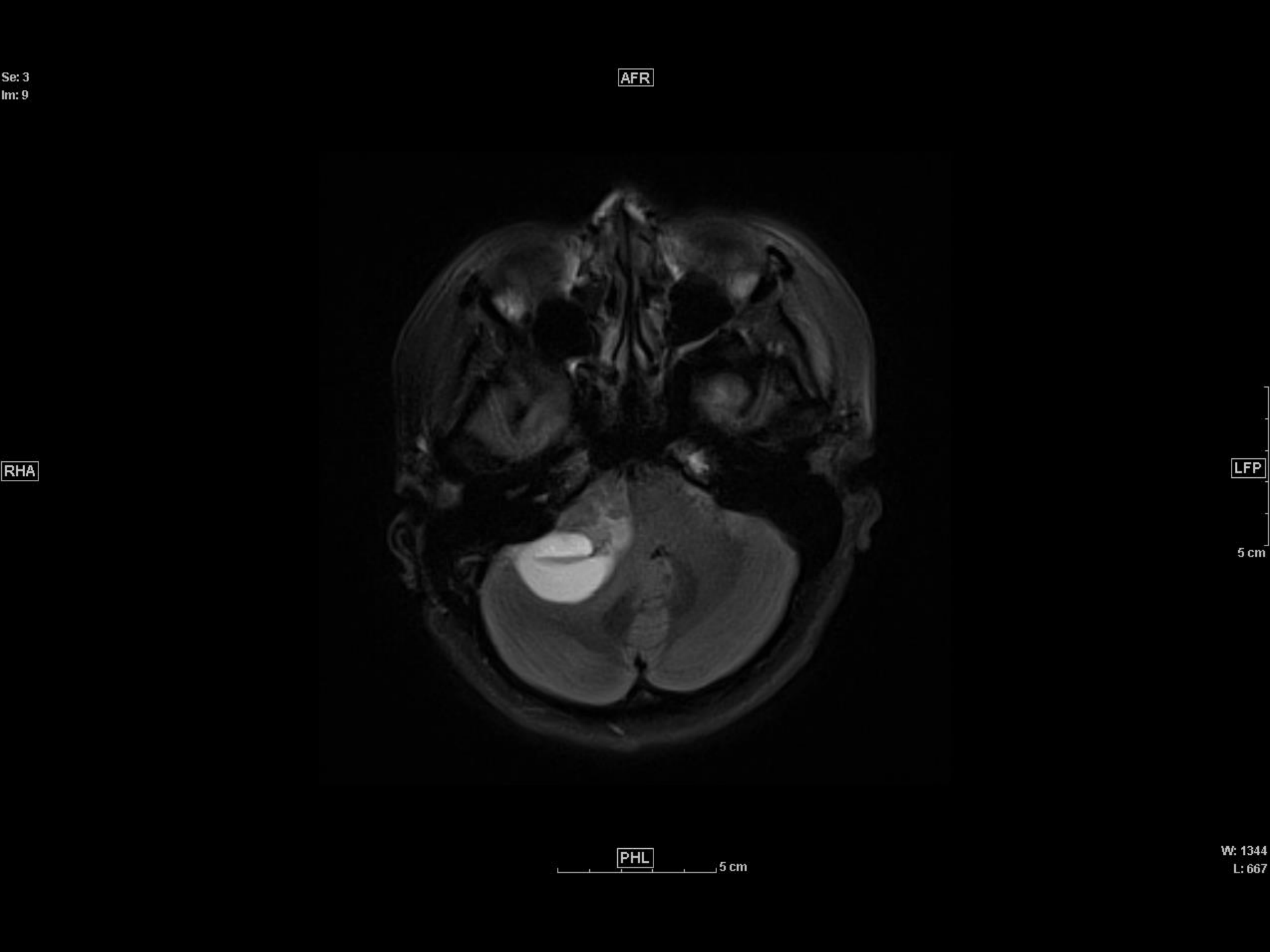
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Im: 9
AFR
RHA
LFP
5 cm
PHL
5 cm
W: 1344
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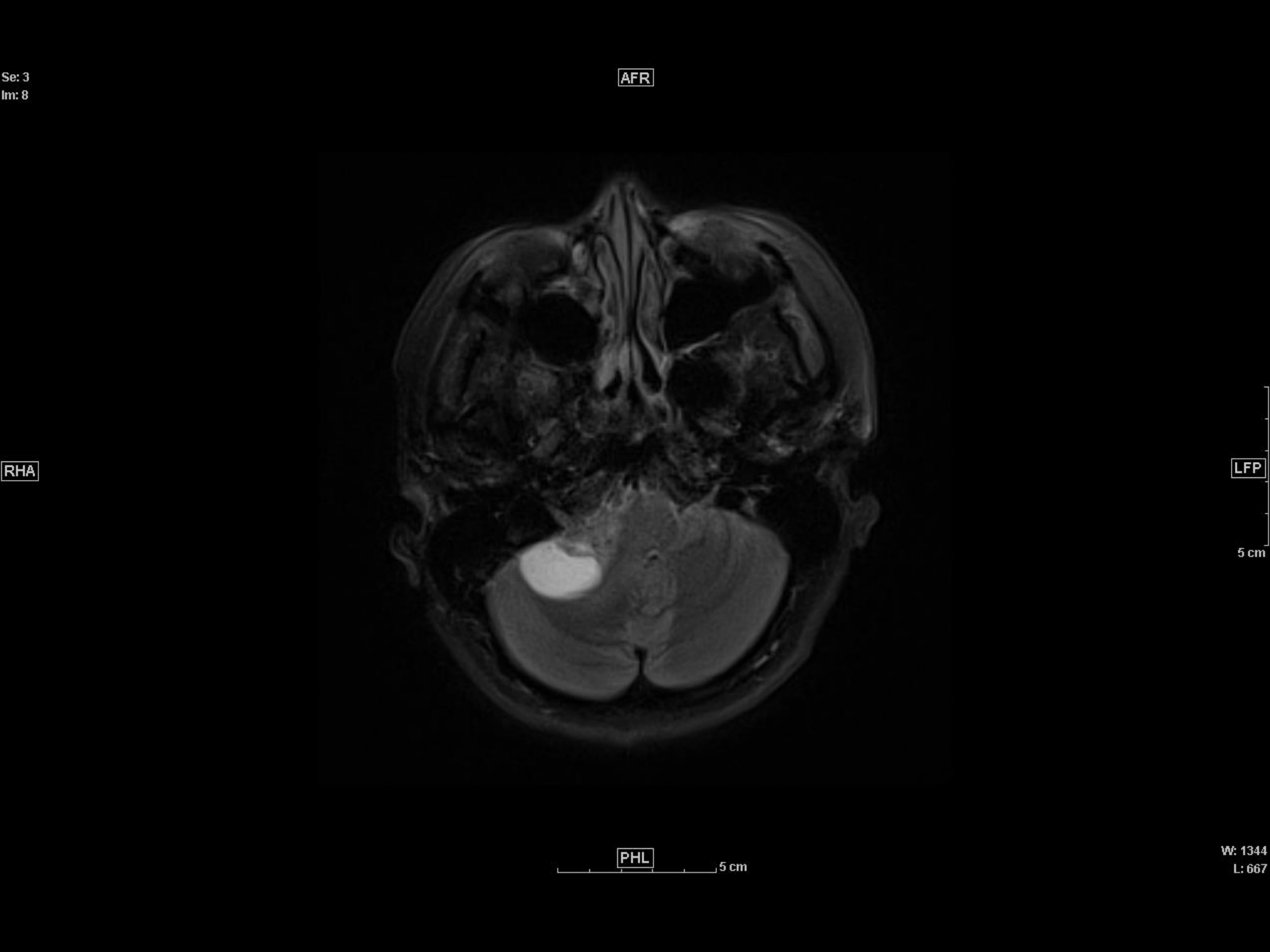
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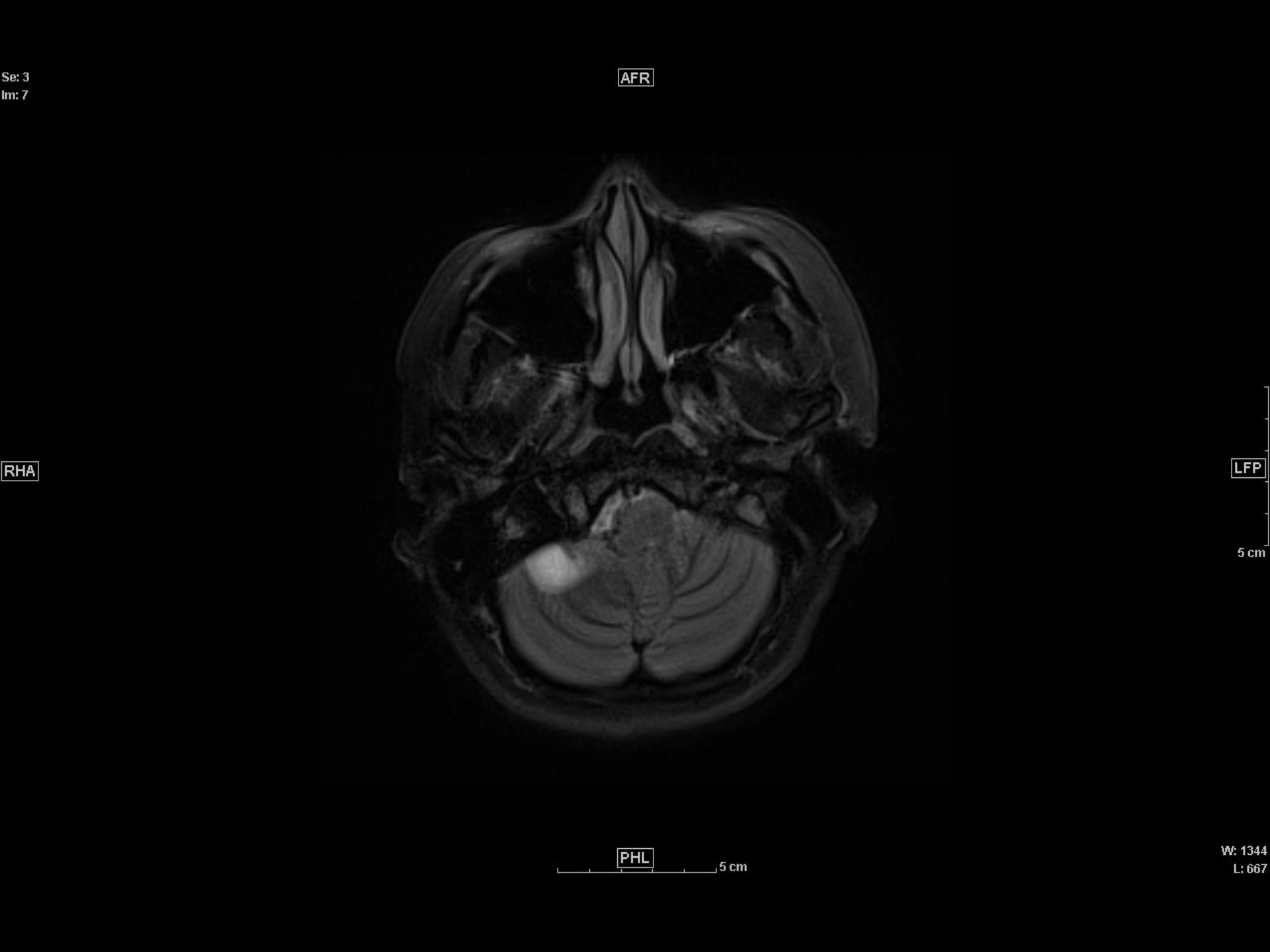
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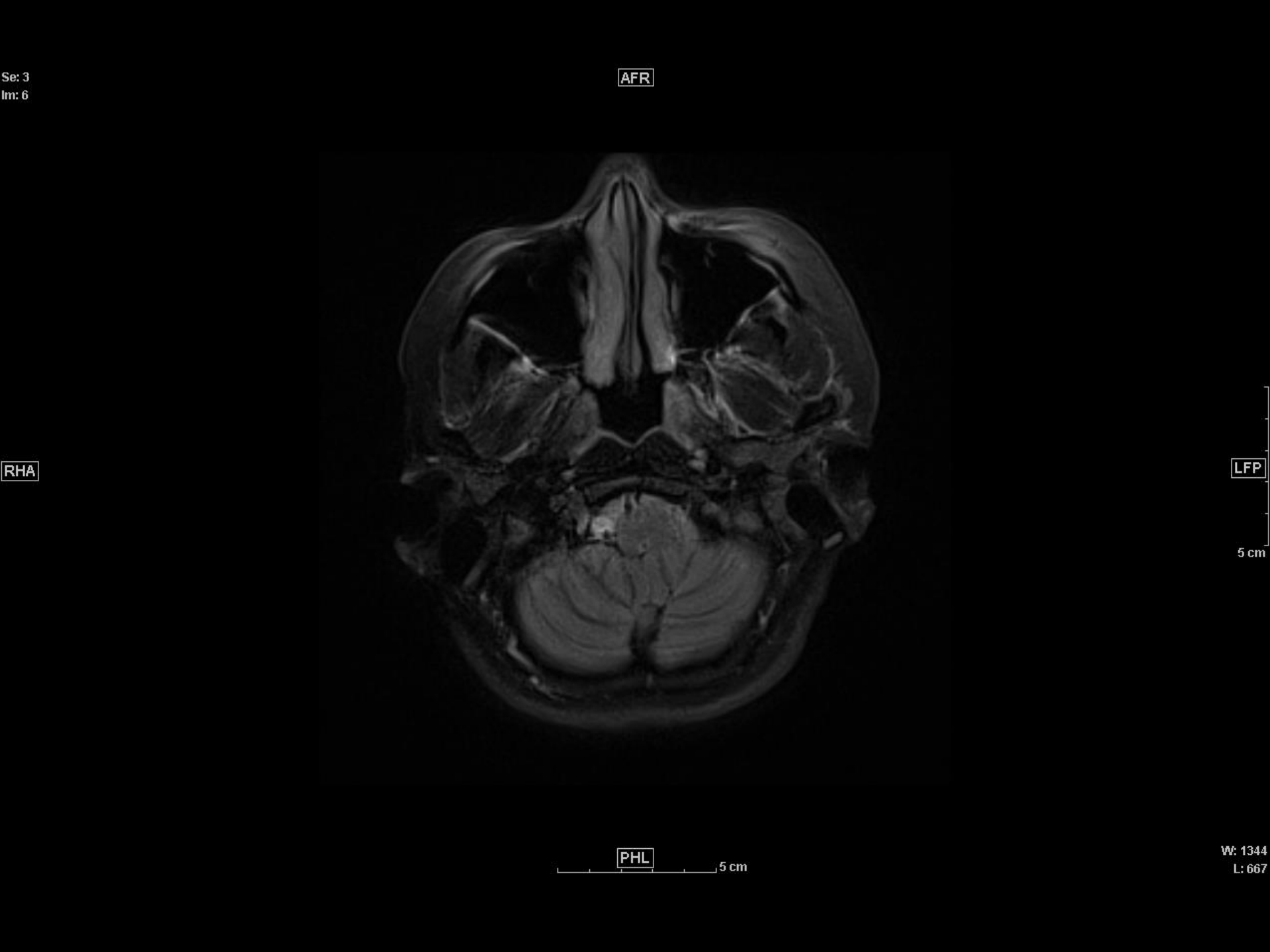
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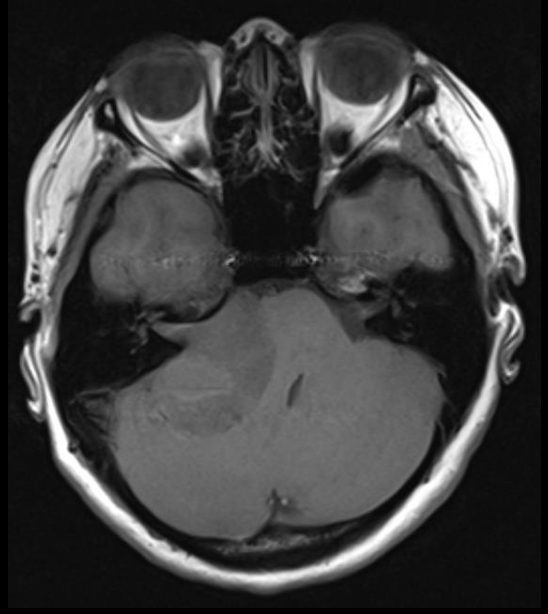

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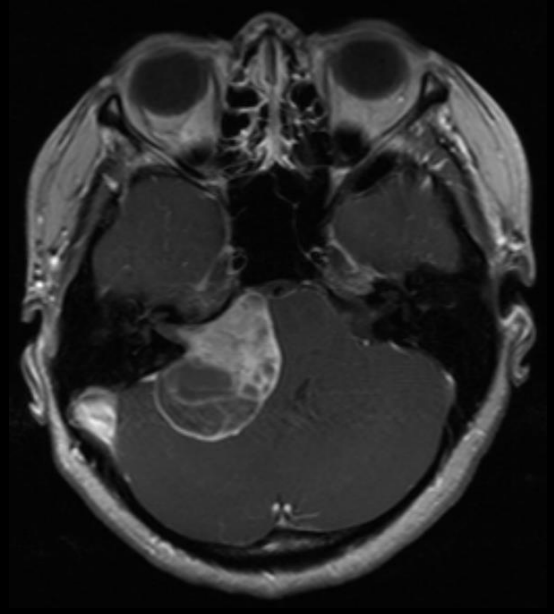

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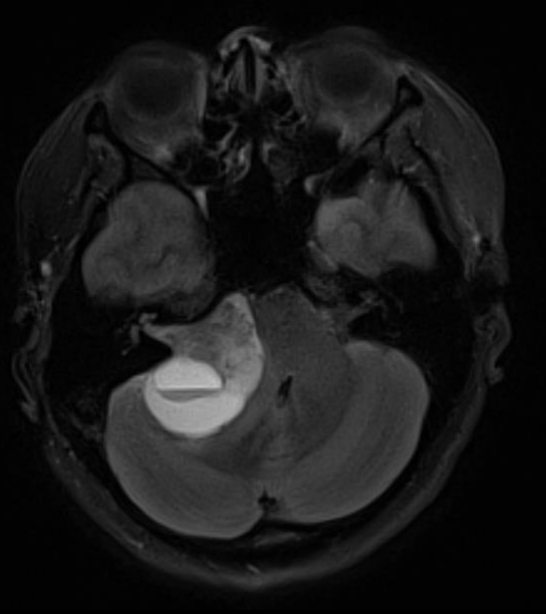

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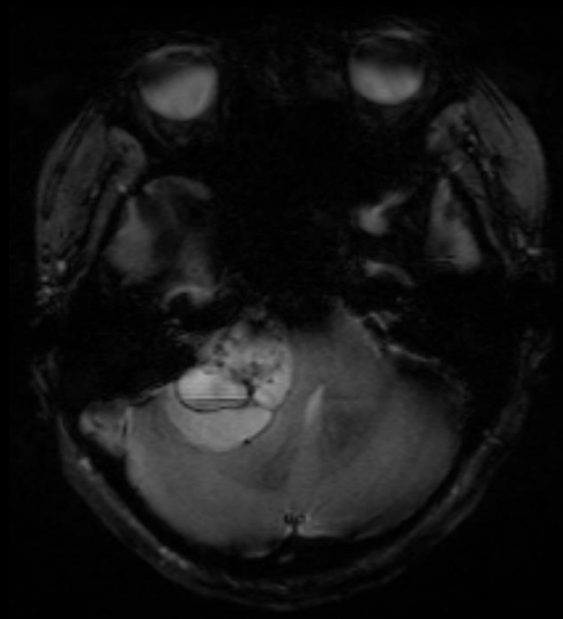
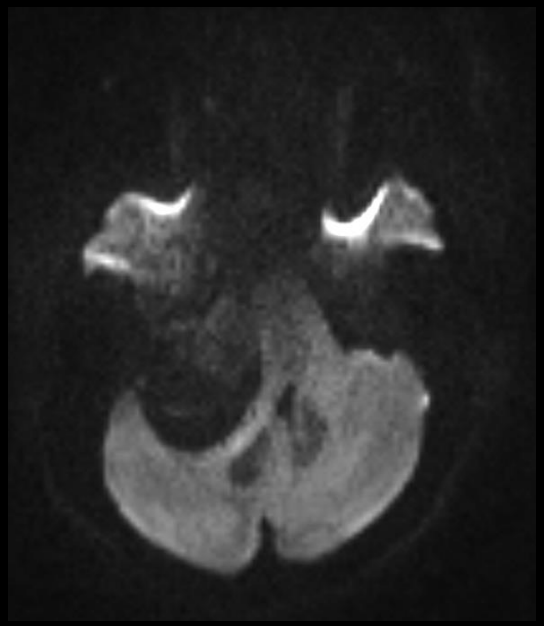


GRE



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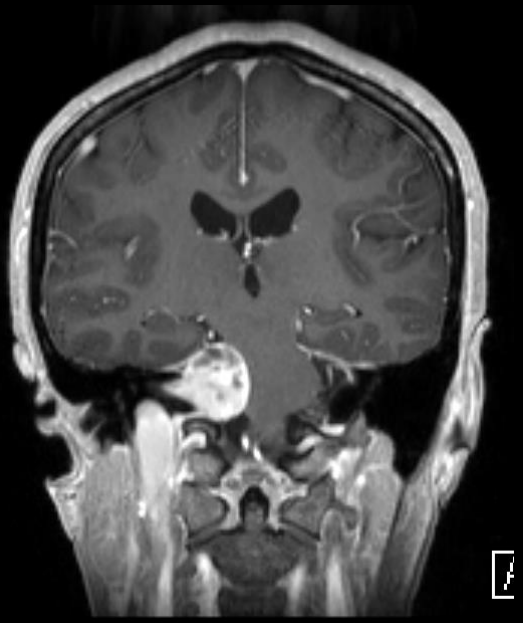

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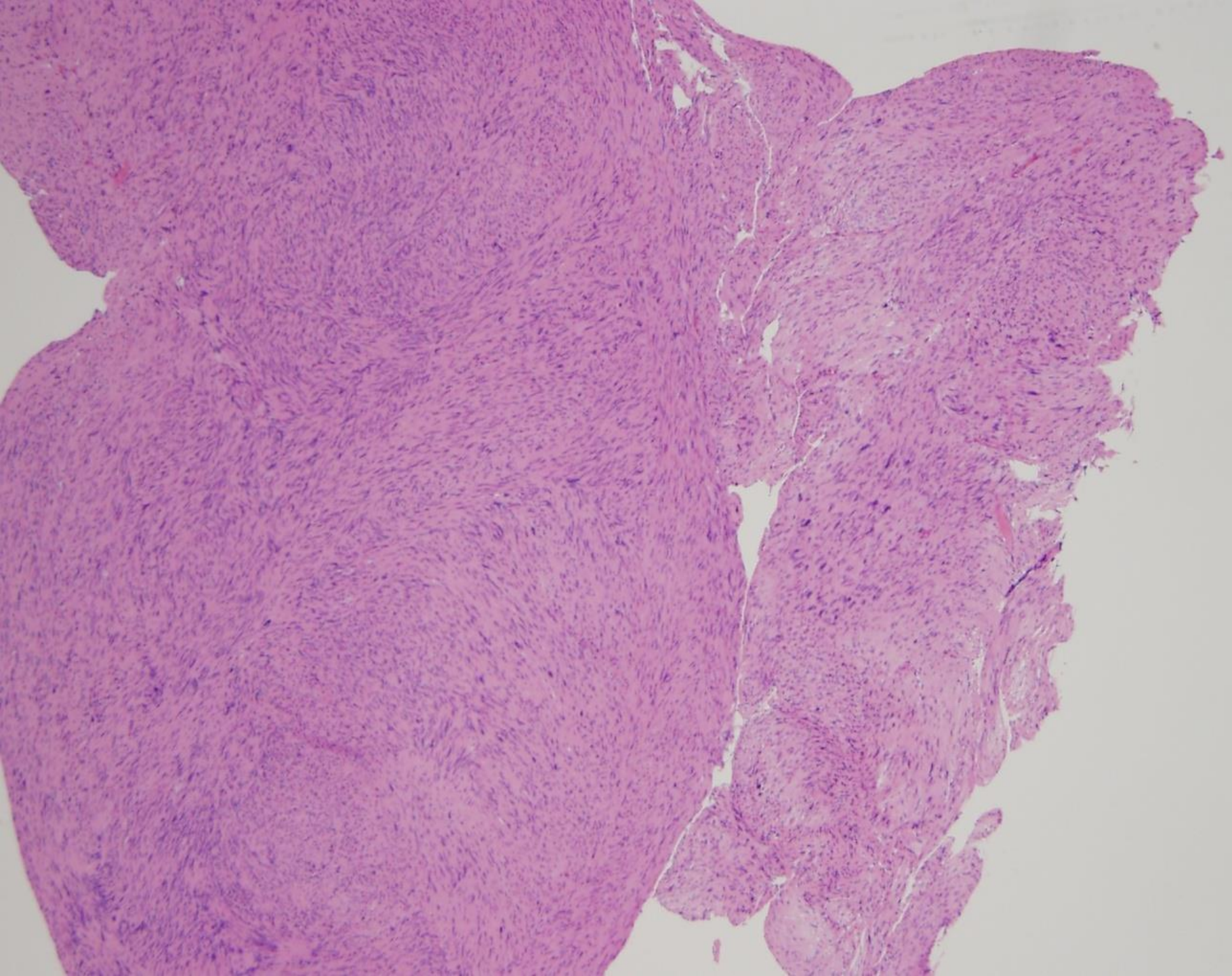
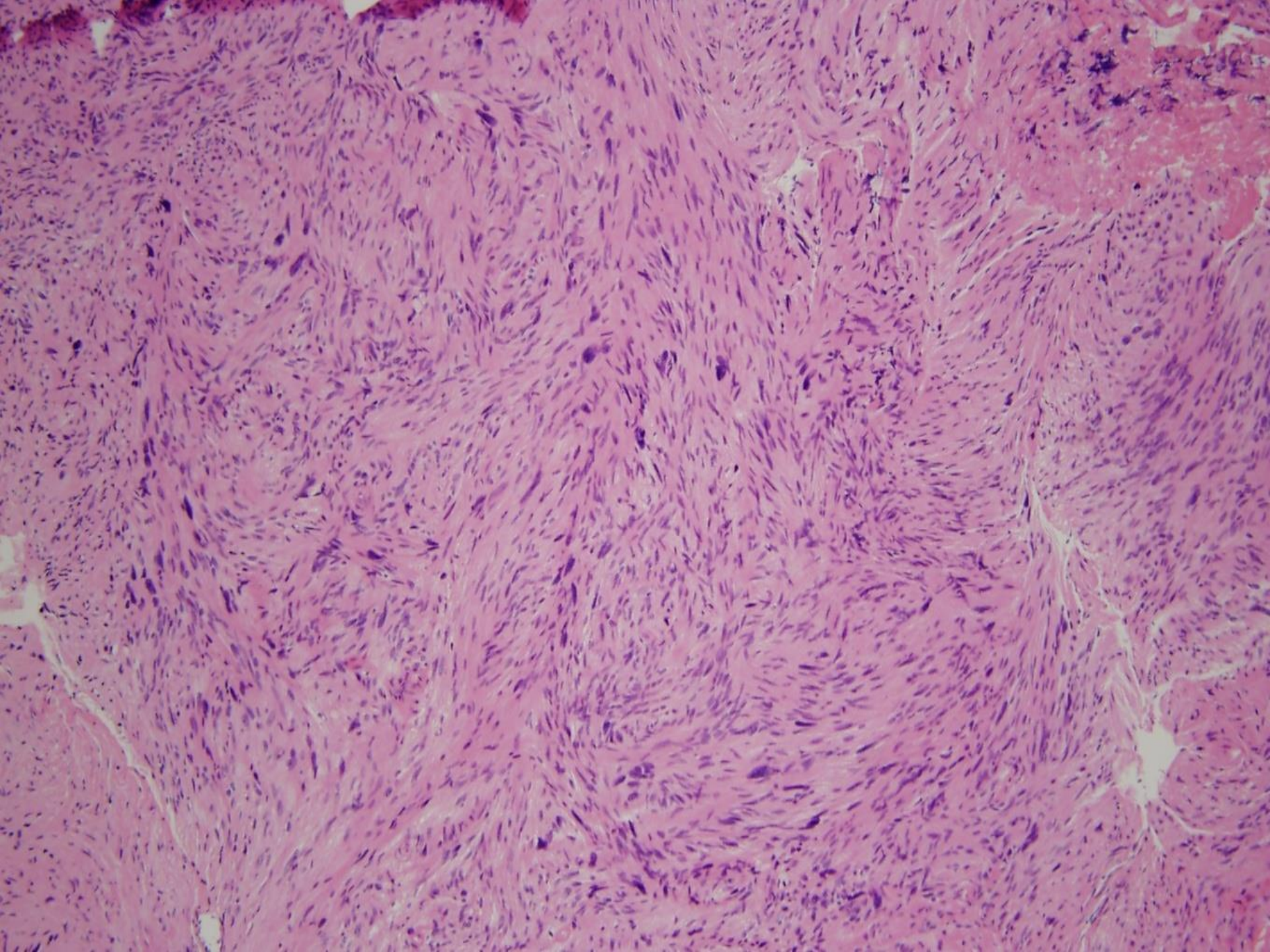
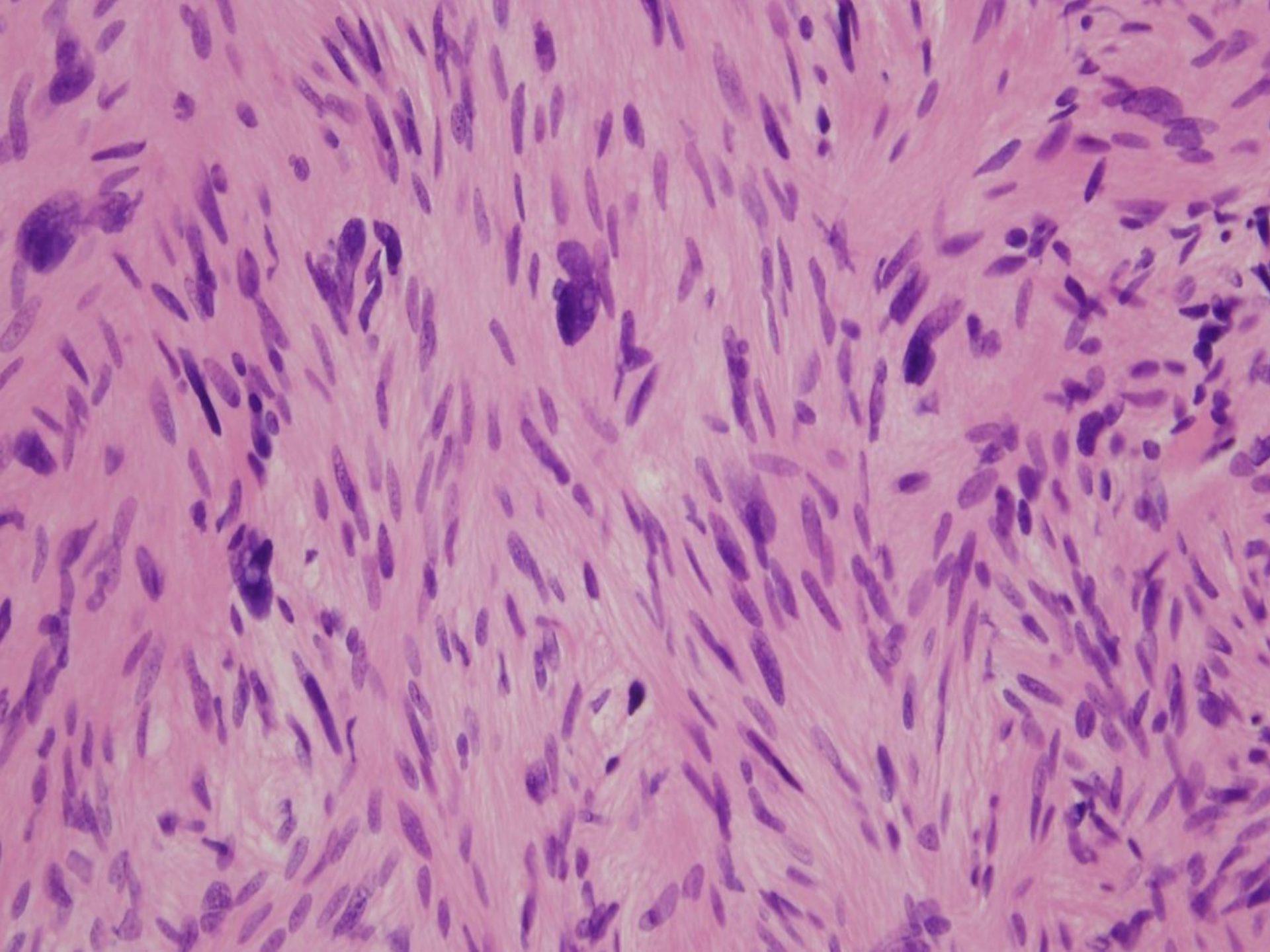






SPECIMEN DESIGNATED: "RIGHT CP ANGLE TUMOR":

-RESIDUAL/RECURRENT SCHWANNOMA.

Schwannoma

- Neural crest origin
- Antoni A (cellular fascicular) and Antoni B (myxoid vacuolated) regions
- Spindled cells with ill defined cytoplasm, dense chromatin
- No axons or mitotic figures
- Degenerative changes (ancient change): nuclear pleomorphism, xanthomatous change and vascular hyalinization are common

Vestibular Schwannoma

- T1 isointense with brain
- T2 “filling defect” in CSF
- FLAIR ↑ cochlear signal, ↑ protein
- GRE microhemorrhage low signal foci
- T1 C+ enhance, 15% with low signal cysts
- Rare dural tail

- Benign tumor arising from vestibular portion of CN8 at glial-Schwann cell junction

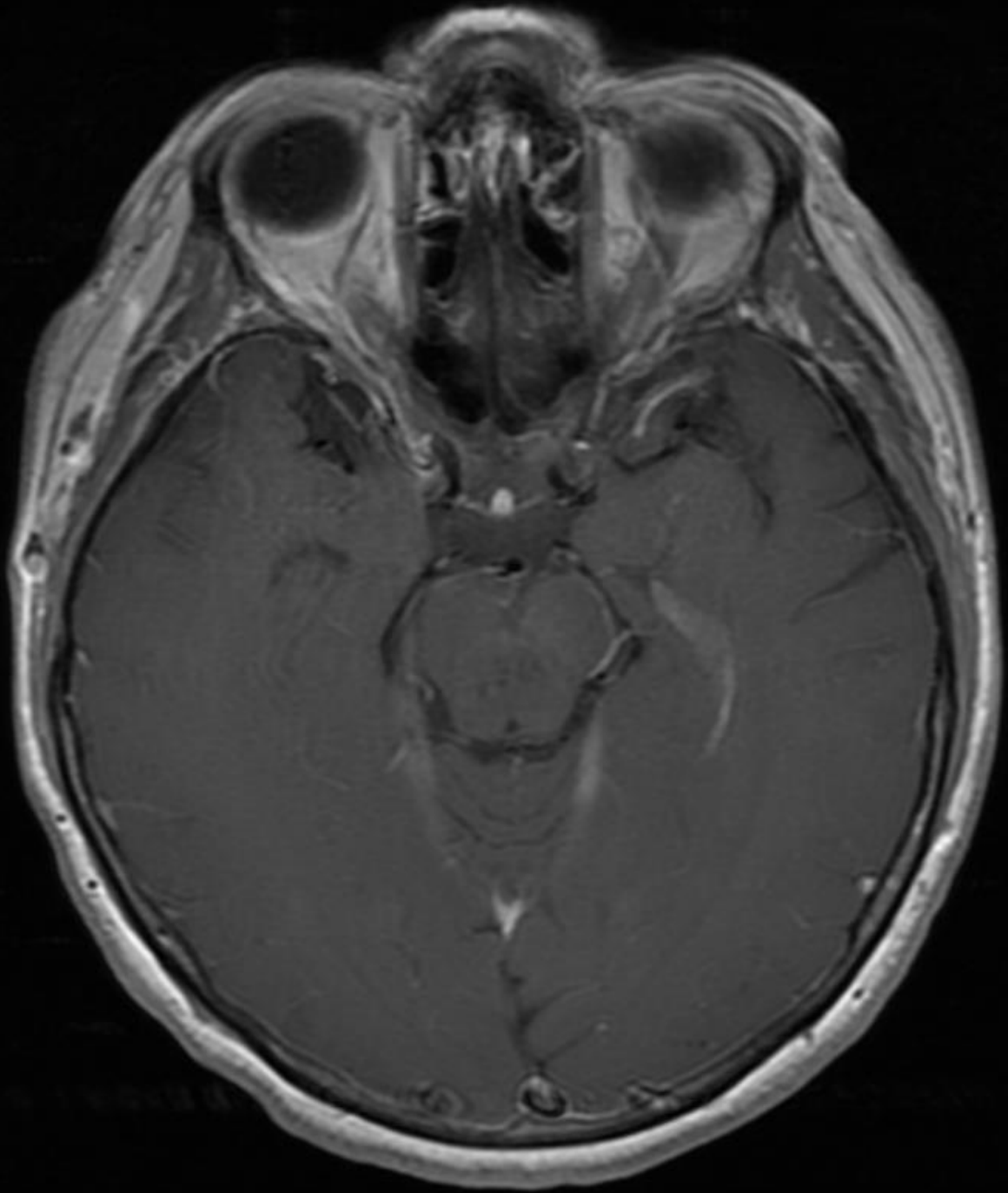
Vestibular Schwannoma

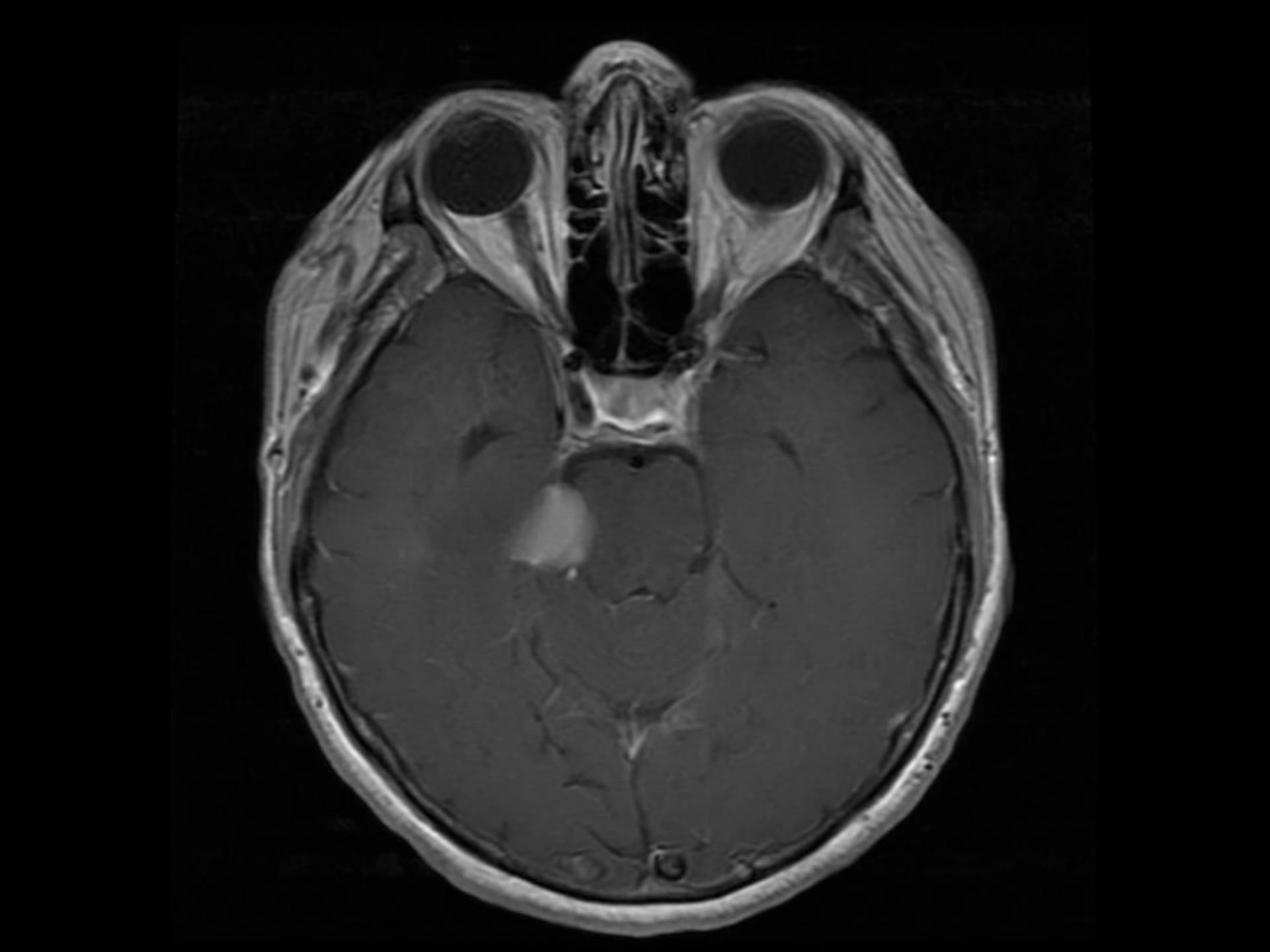
- Most common lesion unilateral SNHL (> 90%)
- Most common CPA-IAC mass (85-90%)
- 2nd most common extraaxial neoplasm adults
- 60% of VS are slow growing (< 1 mm/year)
- Surgical removal of VS will not restore any hearing already lost
- Stereotactic radiosurgery 1st treatment

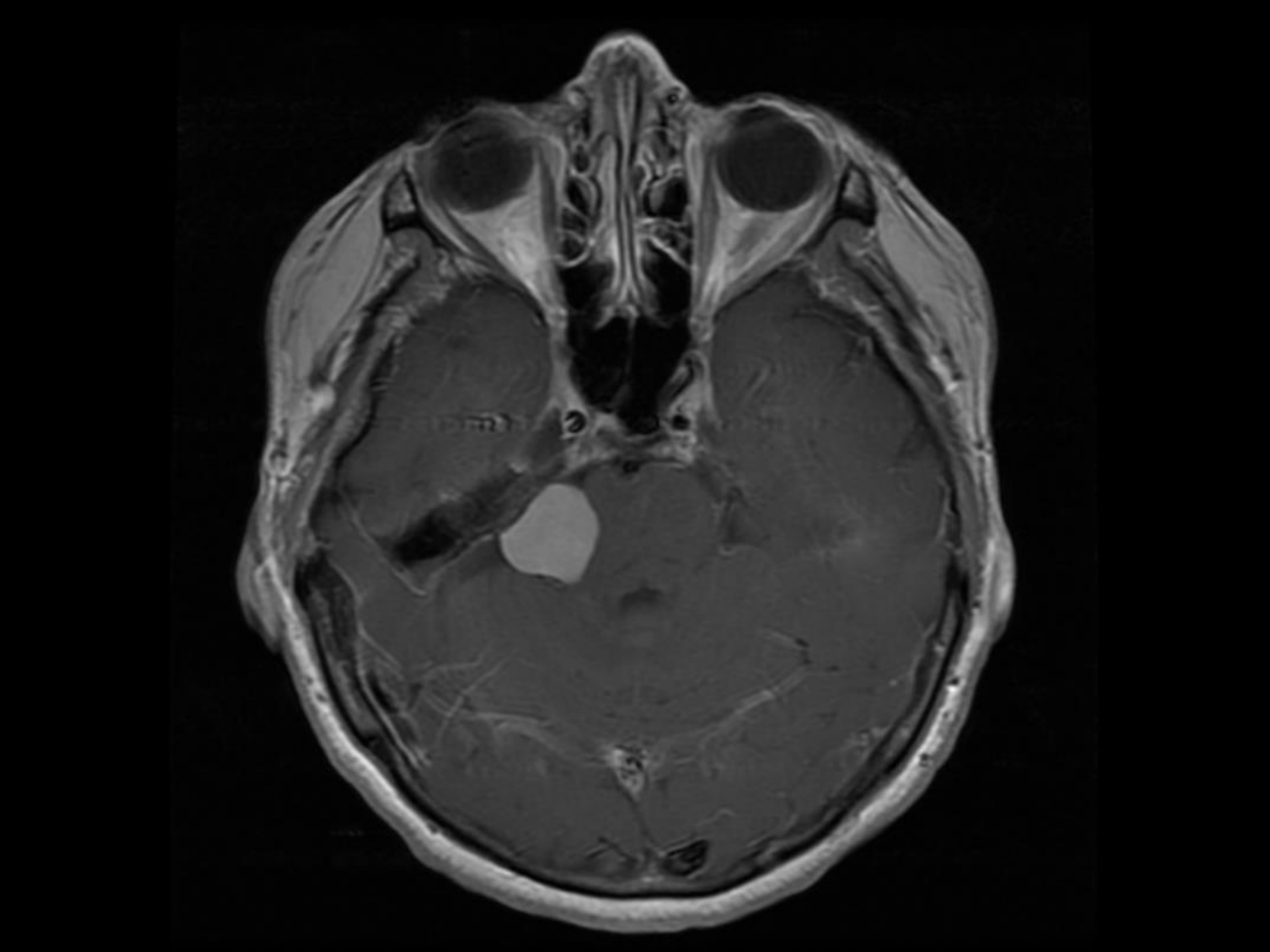
Case 2

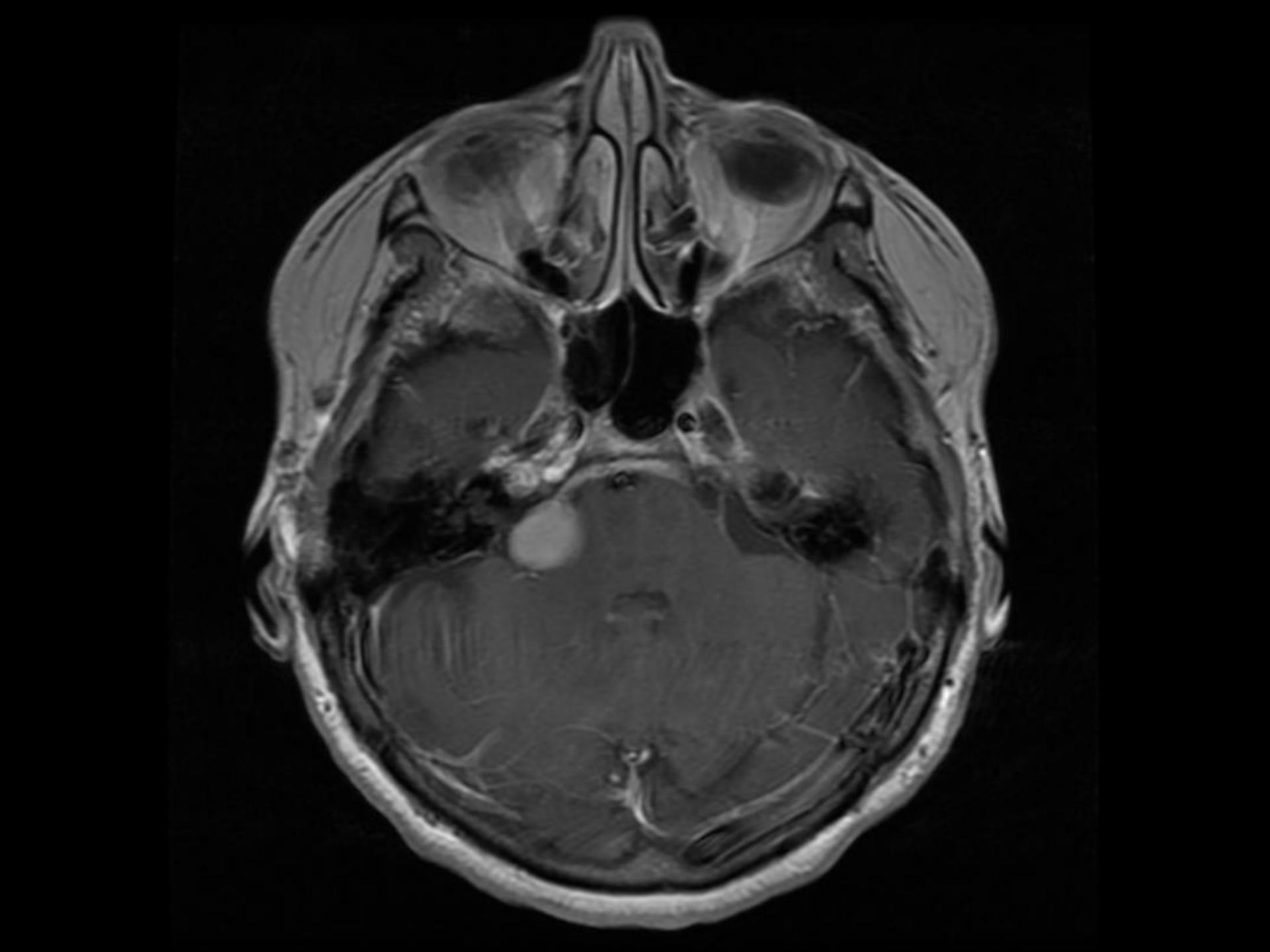
65 Female

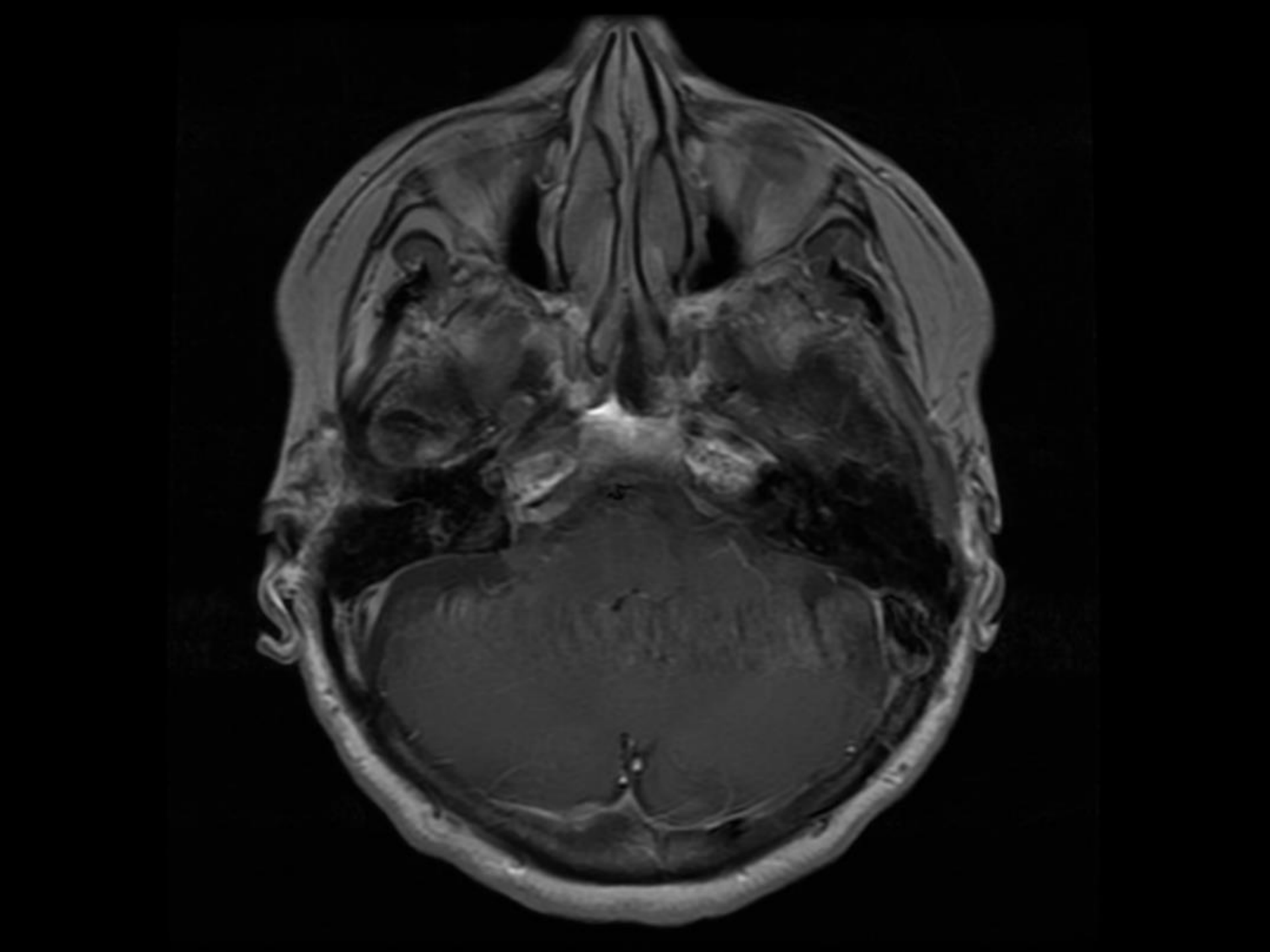
- Dizziness
- Facial Pain
- Lethargy
- All for some time…

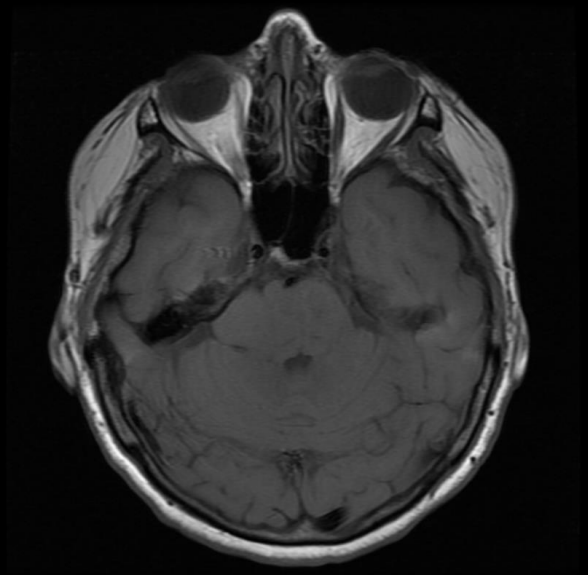












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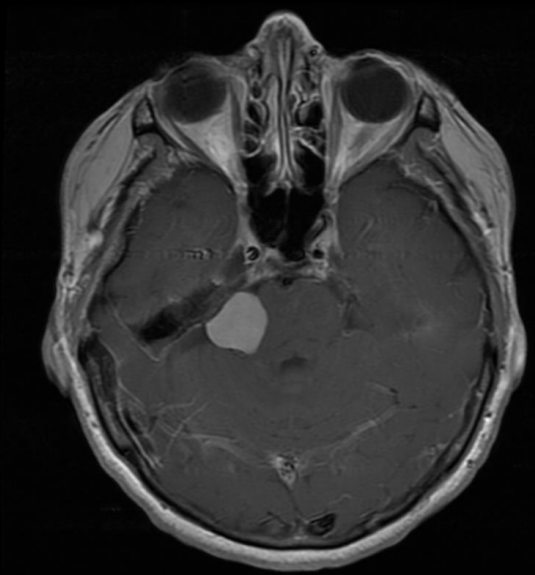

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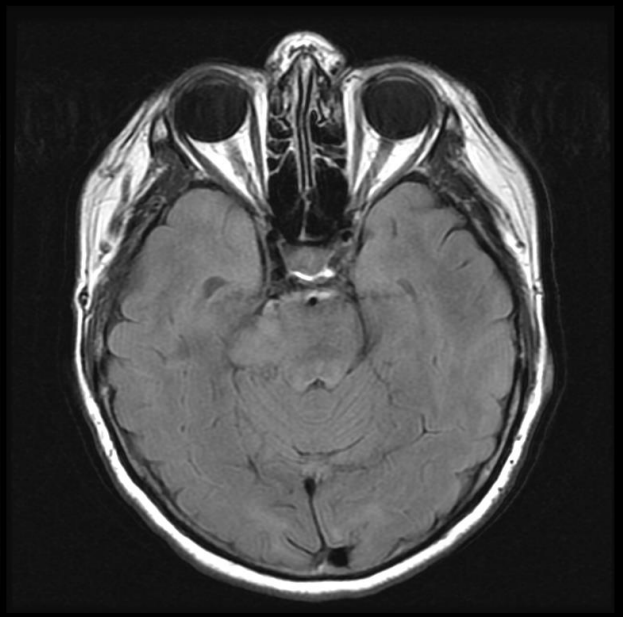

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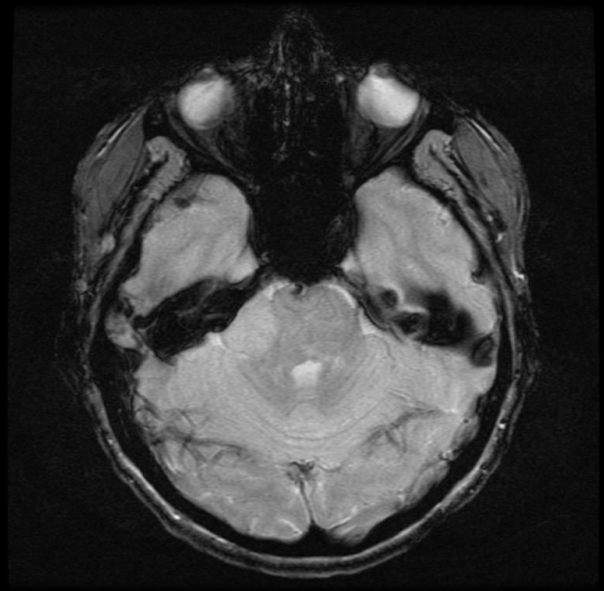

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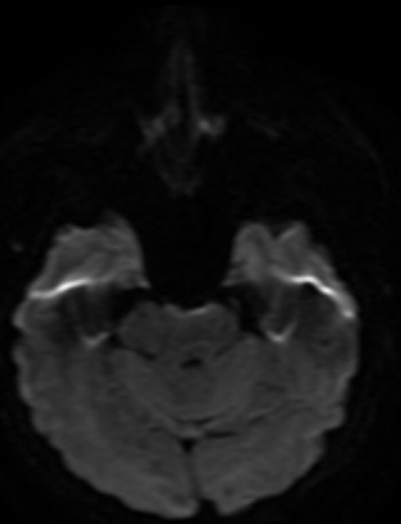

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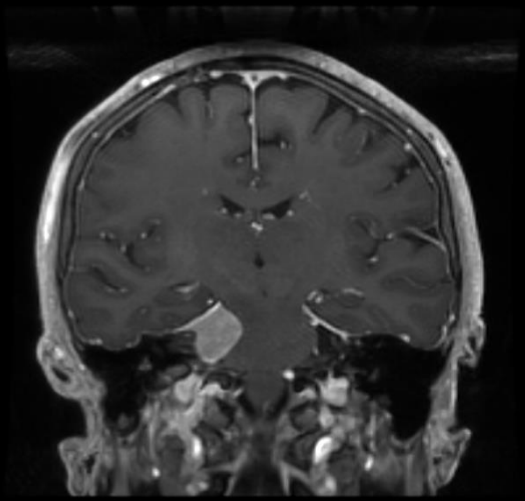

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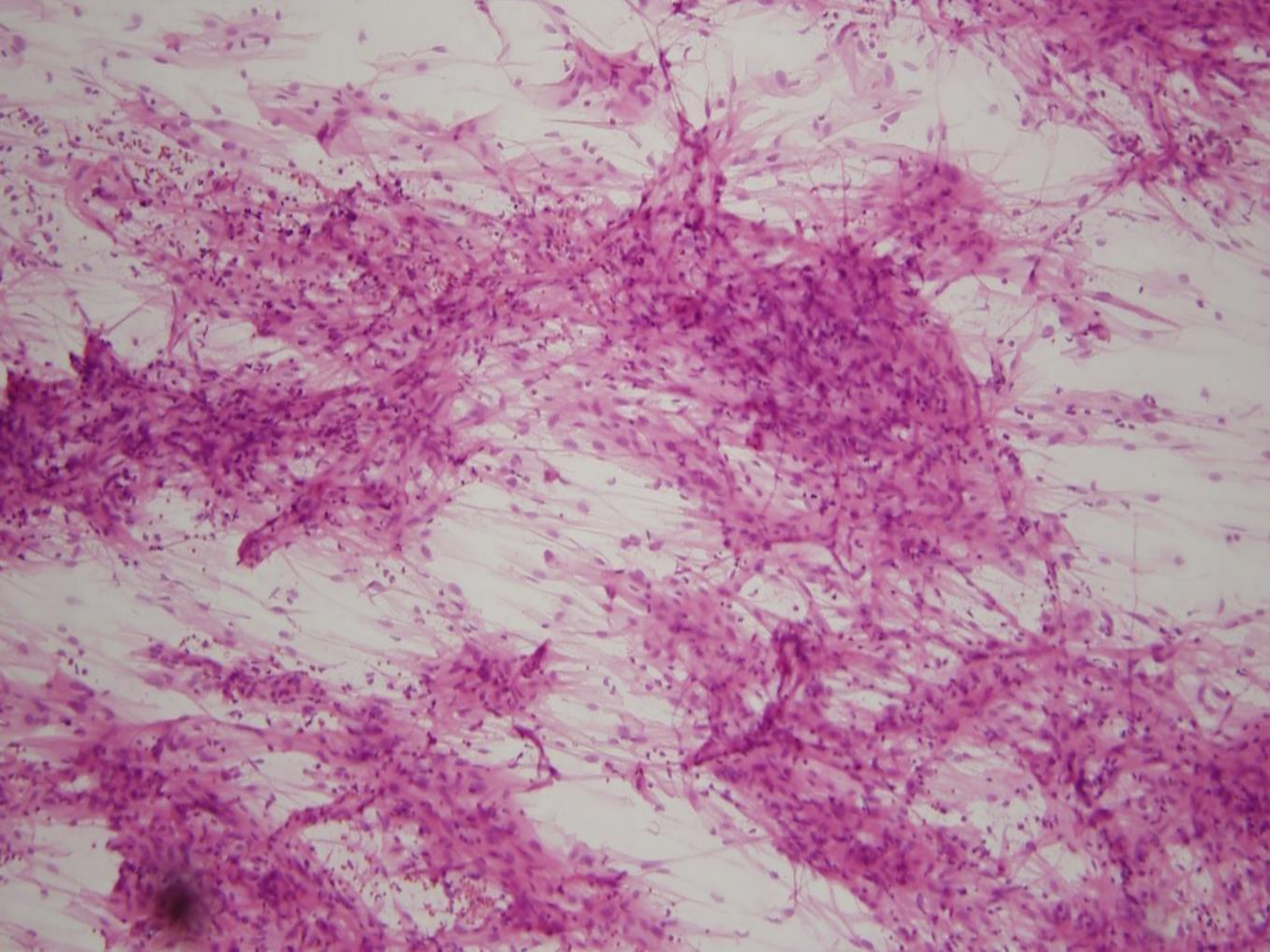
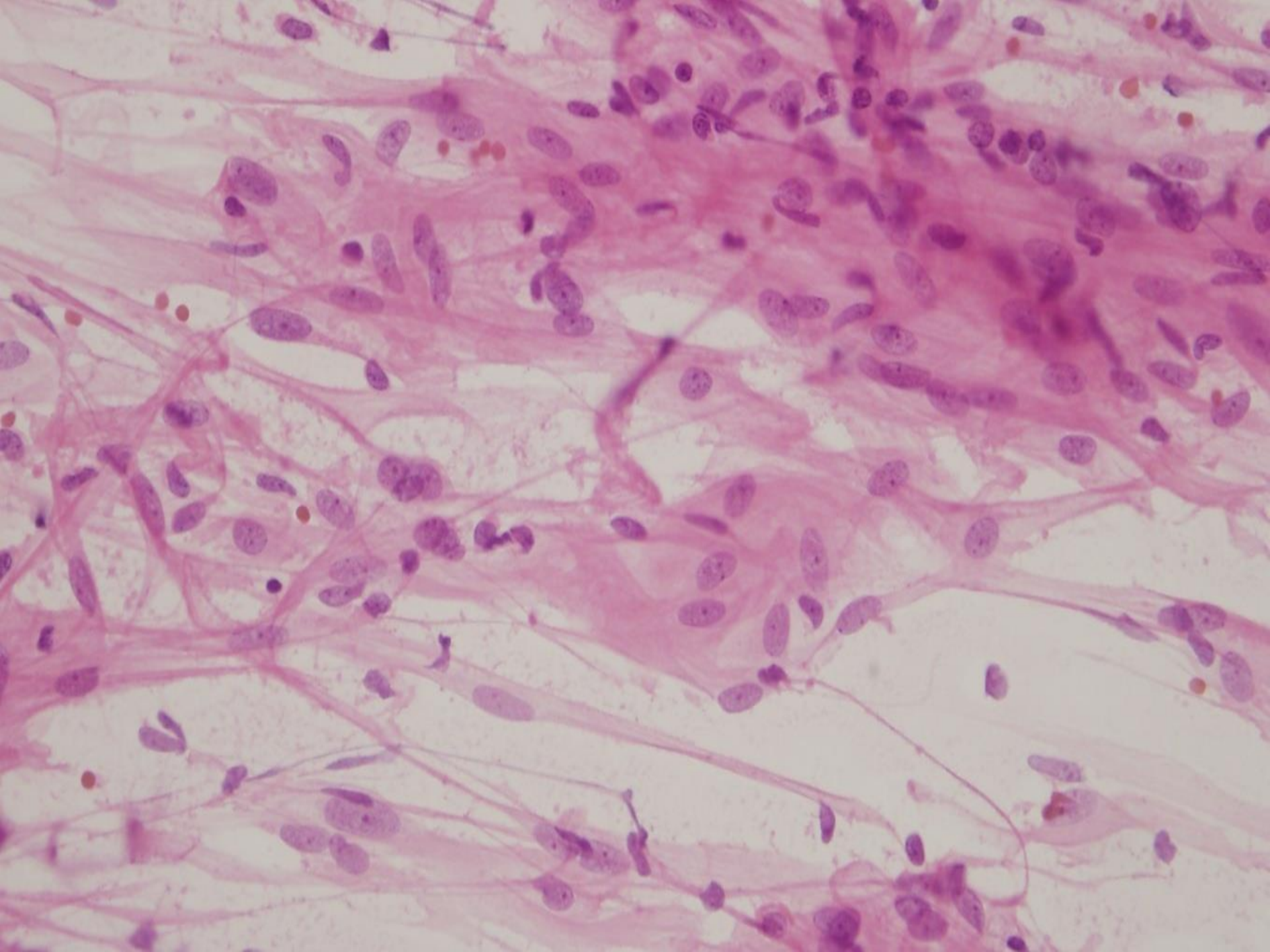
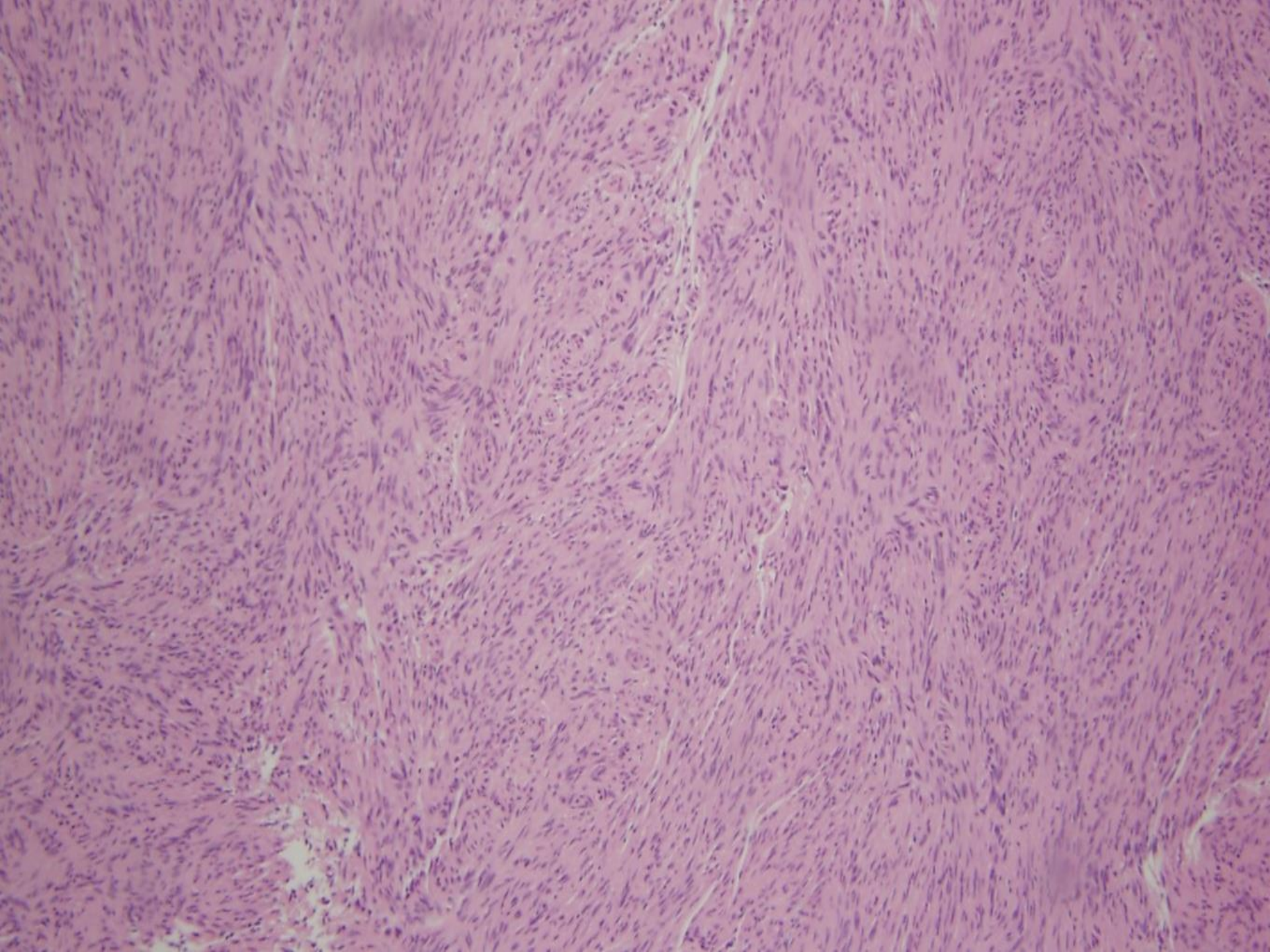
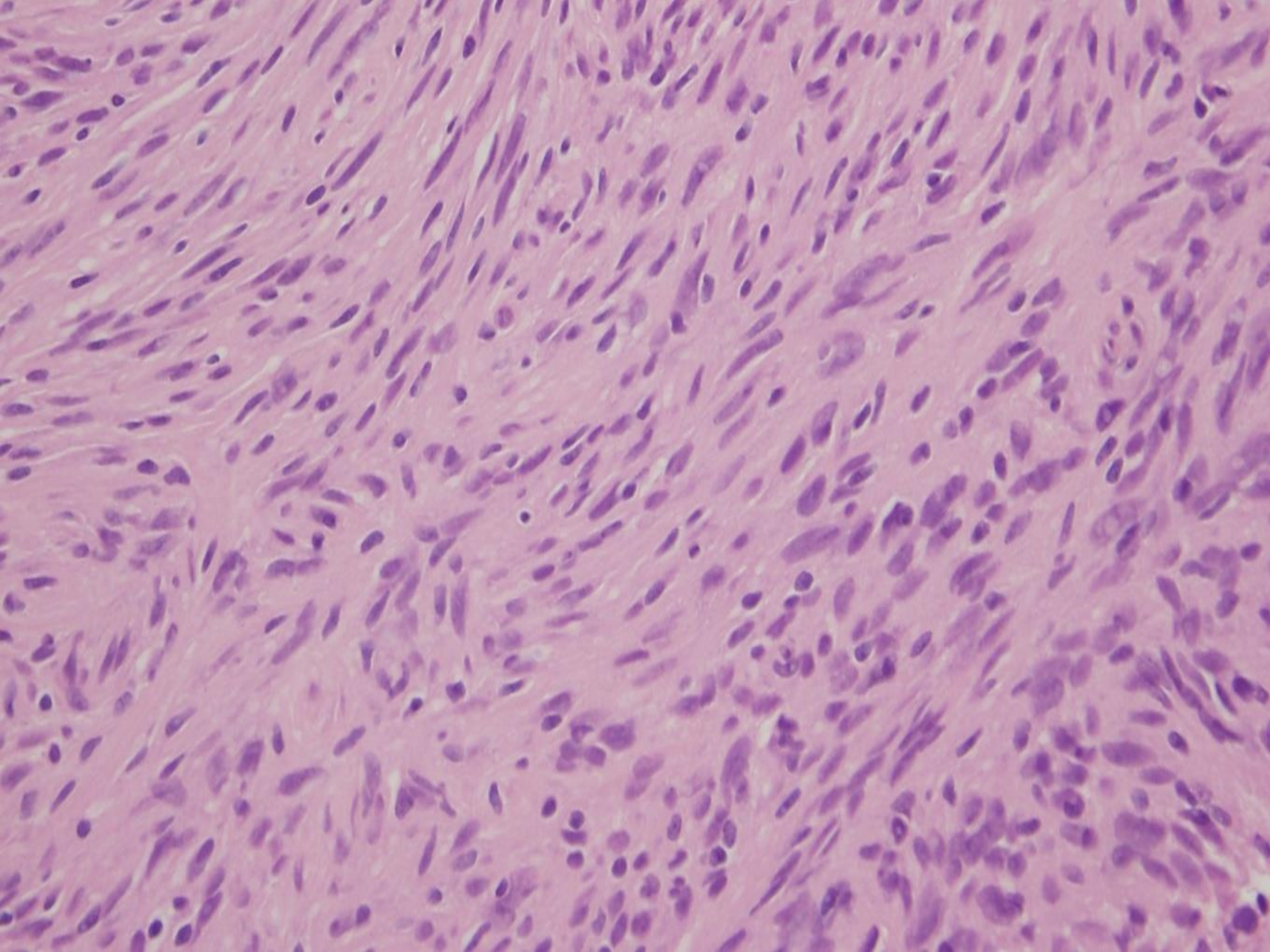




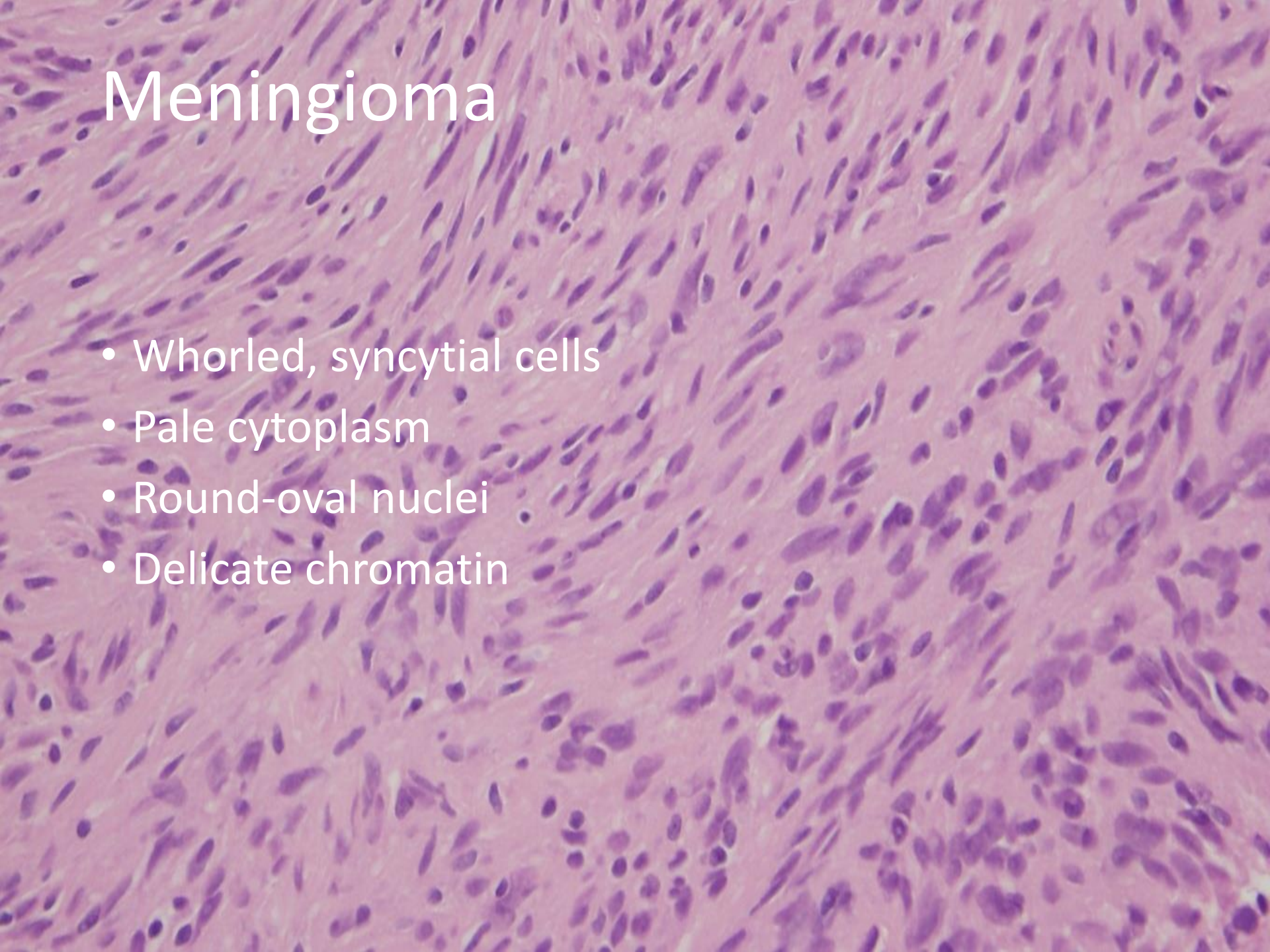




SPECIMEN LABELED "RIGHT SKULL BASE TUMOR“:

MENINGIOMA, W.H.O. Grade I (ICD-0 9530/0).

Meningioma



- Whorled, syncytial cells
- Pale cytoplasm
- Round-oval nuclei
- Delicate chromatin

Meningioma

- T1 iso/hypointense to gray matter
- T2 variable, 50% iso, 35-40% hyper
- DWI not helpful
- T1 C+ intense/homogenous enhancement
- CSF cleft sign
- Dural tail

- May grow during pregnancy (estrogen)
- Typically benign
- Arise from arachnoid cap cells

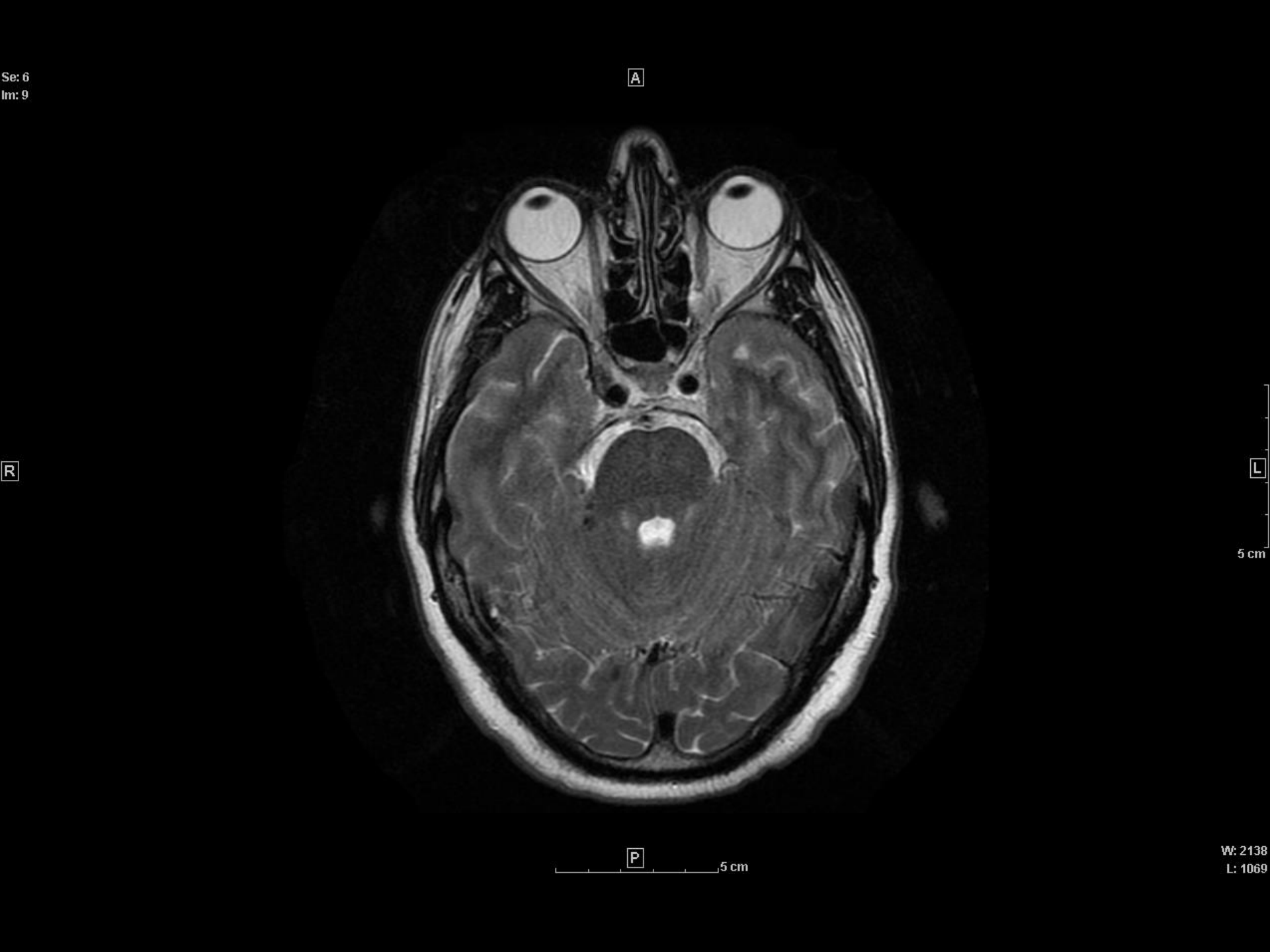
Meningioma

- Slow-growing benign tumor
- Relatively high recurrence rate due to difficulty of complete excision
- Prognosis relates to surgical outcome
- Hearing usually preserved at preop level
- Aggressive surgery advocated because bone invasion hard to see at surgery

Case 3

46 Male

- Sense vibration skull/head x 5 months
- Also some ringing in right ear
- Increased frequency/intensity x 2 months
- Hearing loss right ear on exam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R
L
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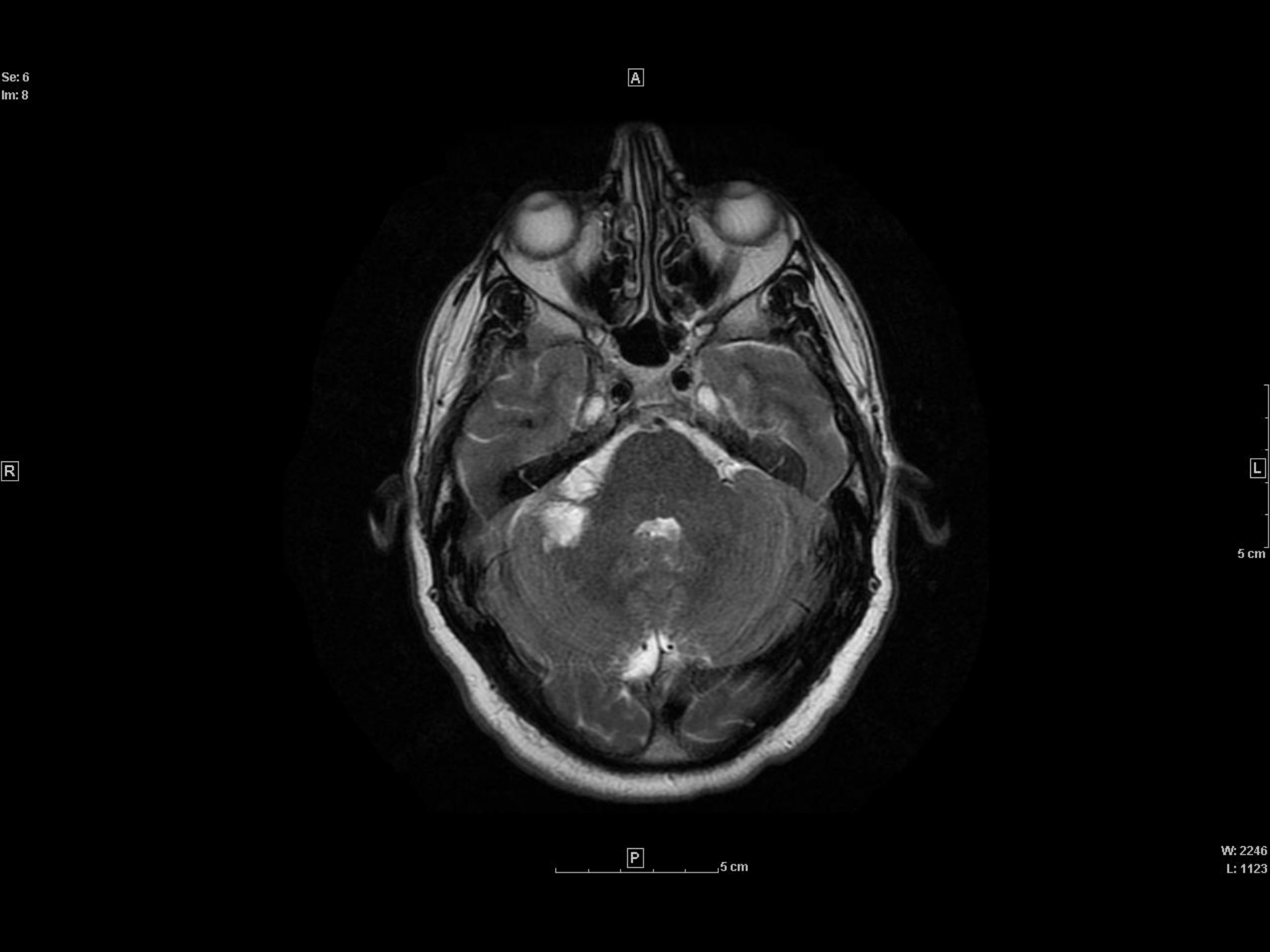
L: 1069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L
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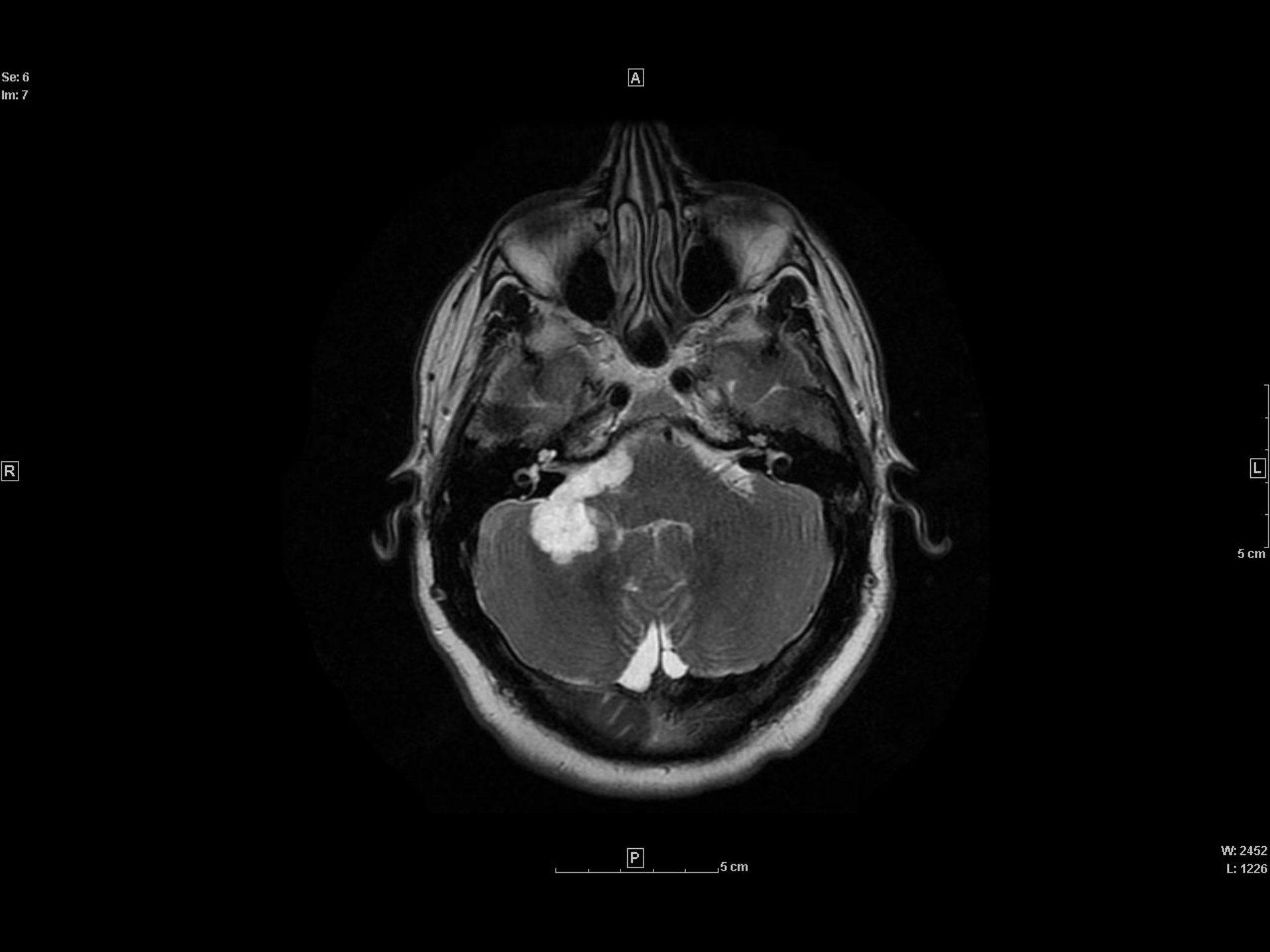
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Im: 7
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R
L
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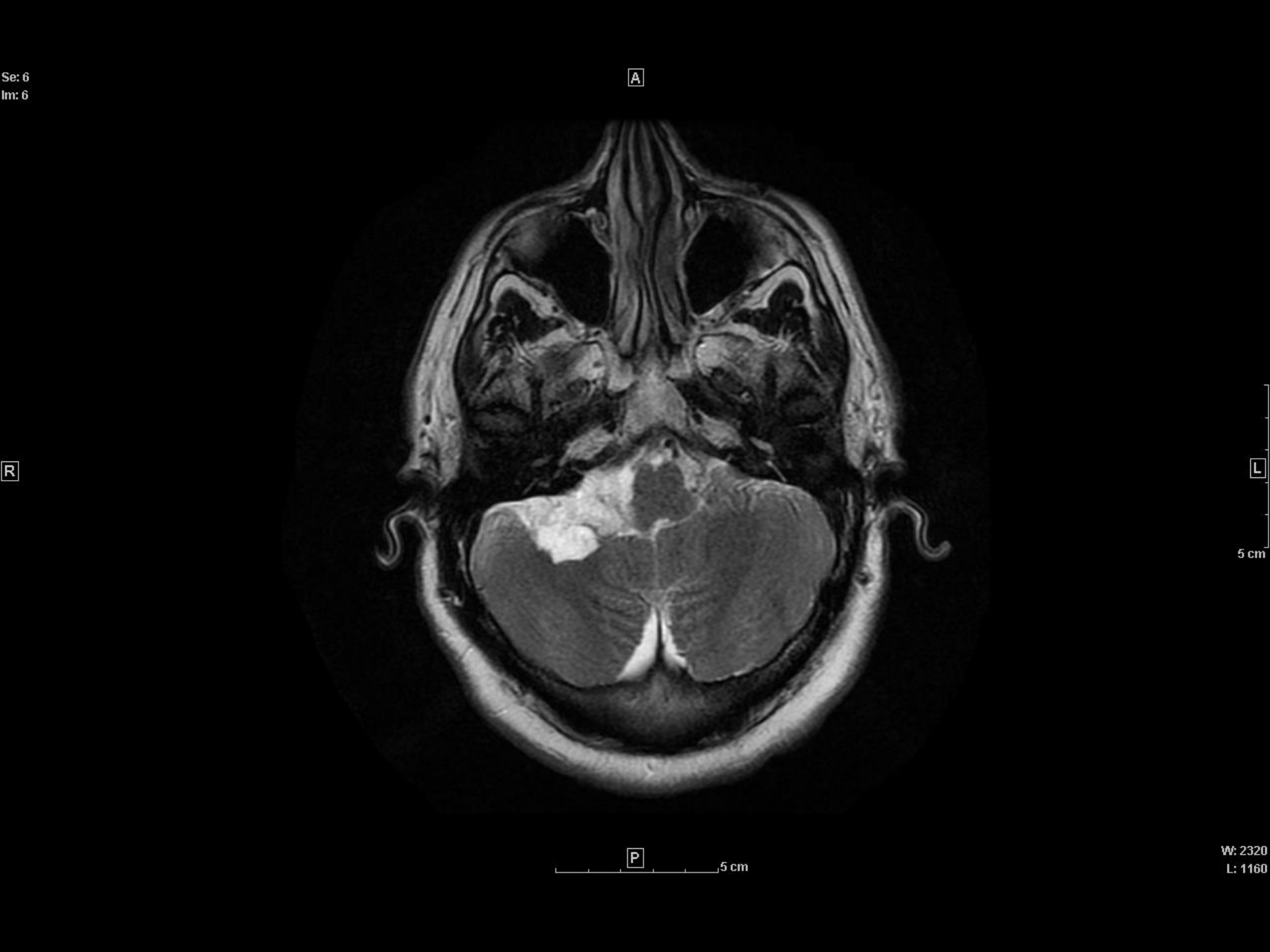
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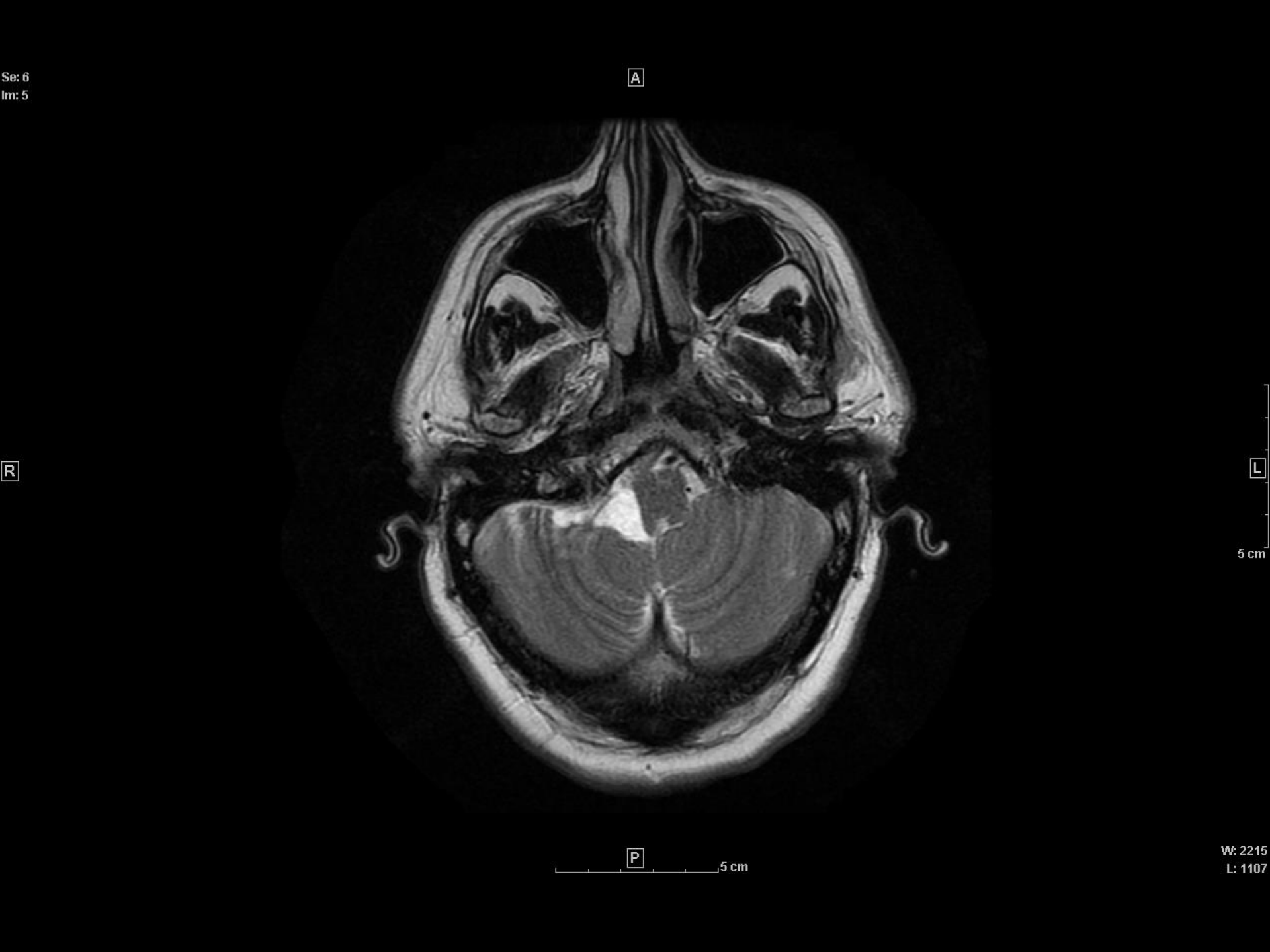
Im: 6
A
R
L
5 cm
P
5 cm
W: 2320
L: 1160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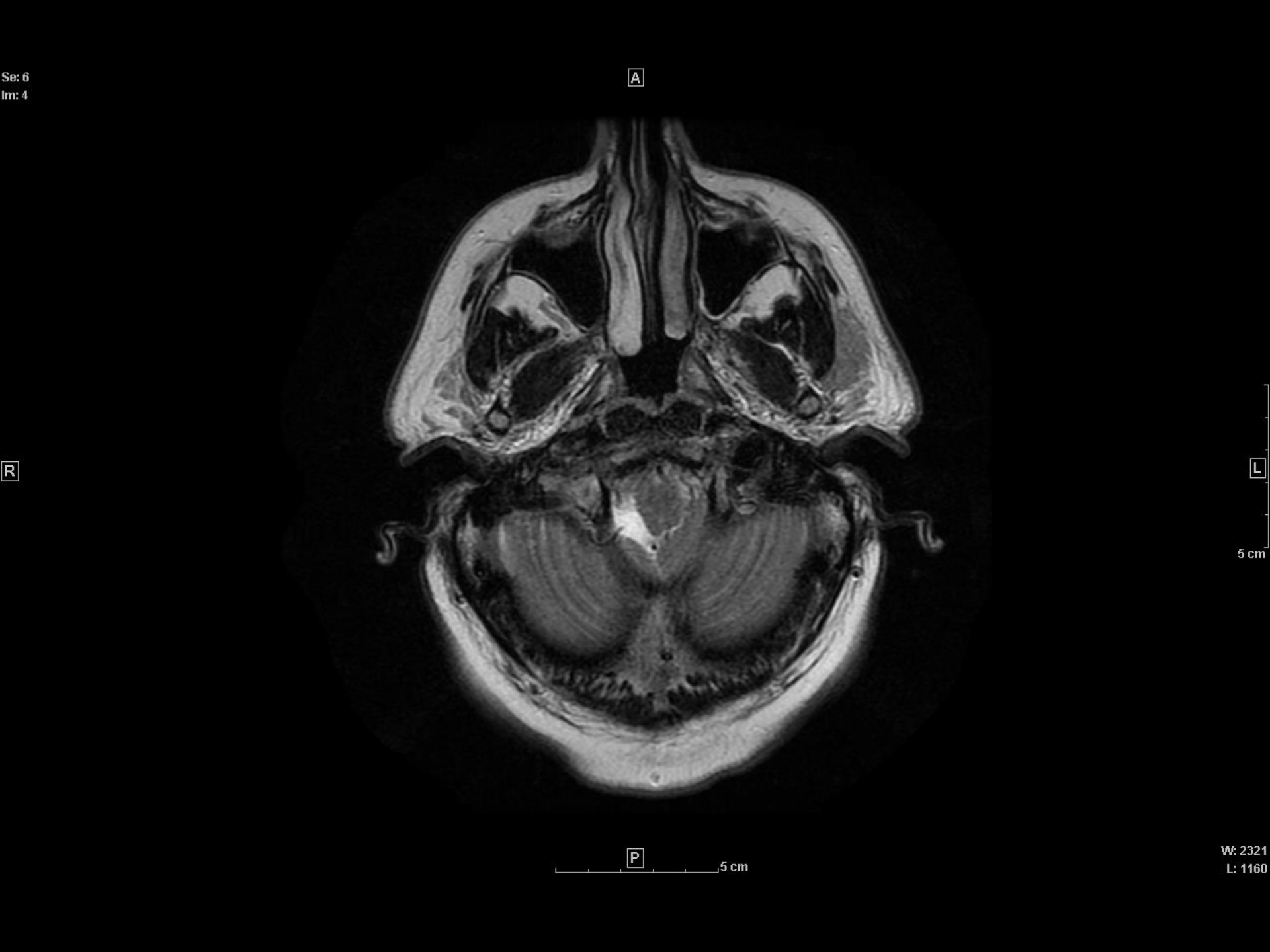
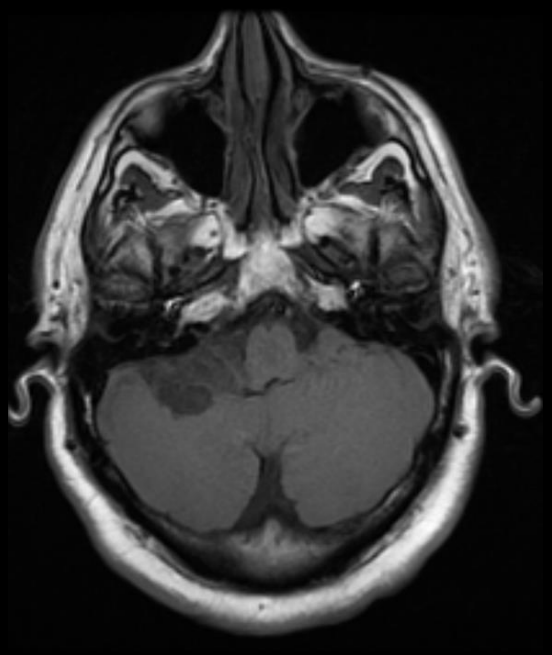
Im: 5
A
R
L
5 cm
P
5 cm
W: 2215
L: 1107


Se: 6
Im: 4
A
R
L
5 cm
P
5 cm
W: 2321
L: 1160



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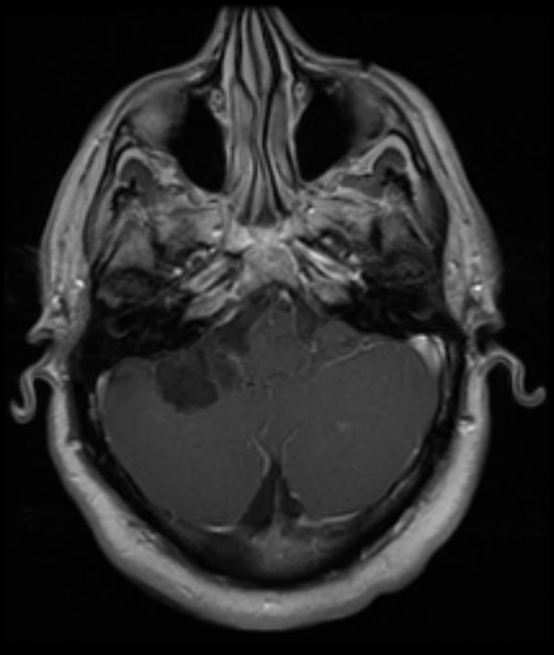

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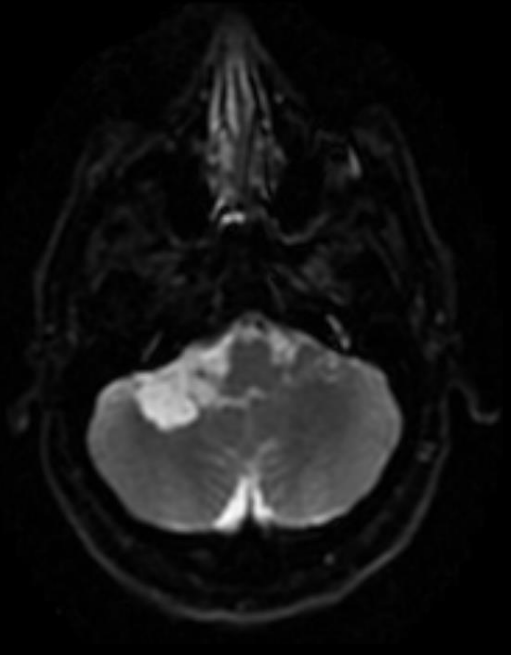
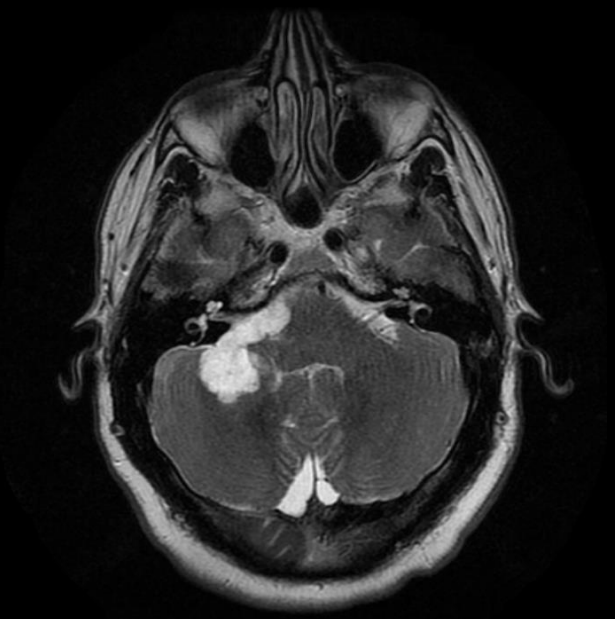


DWI



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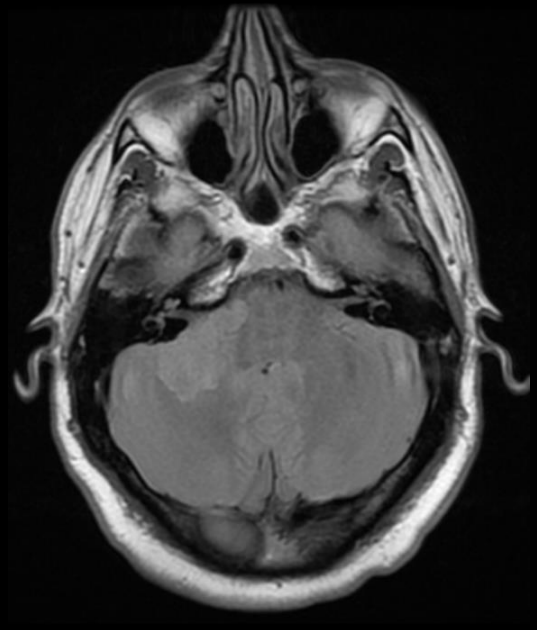
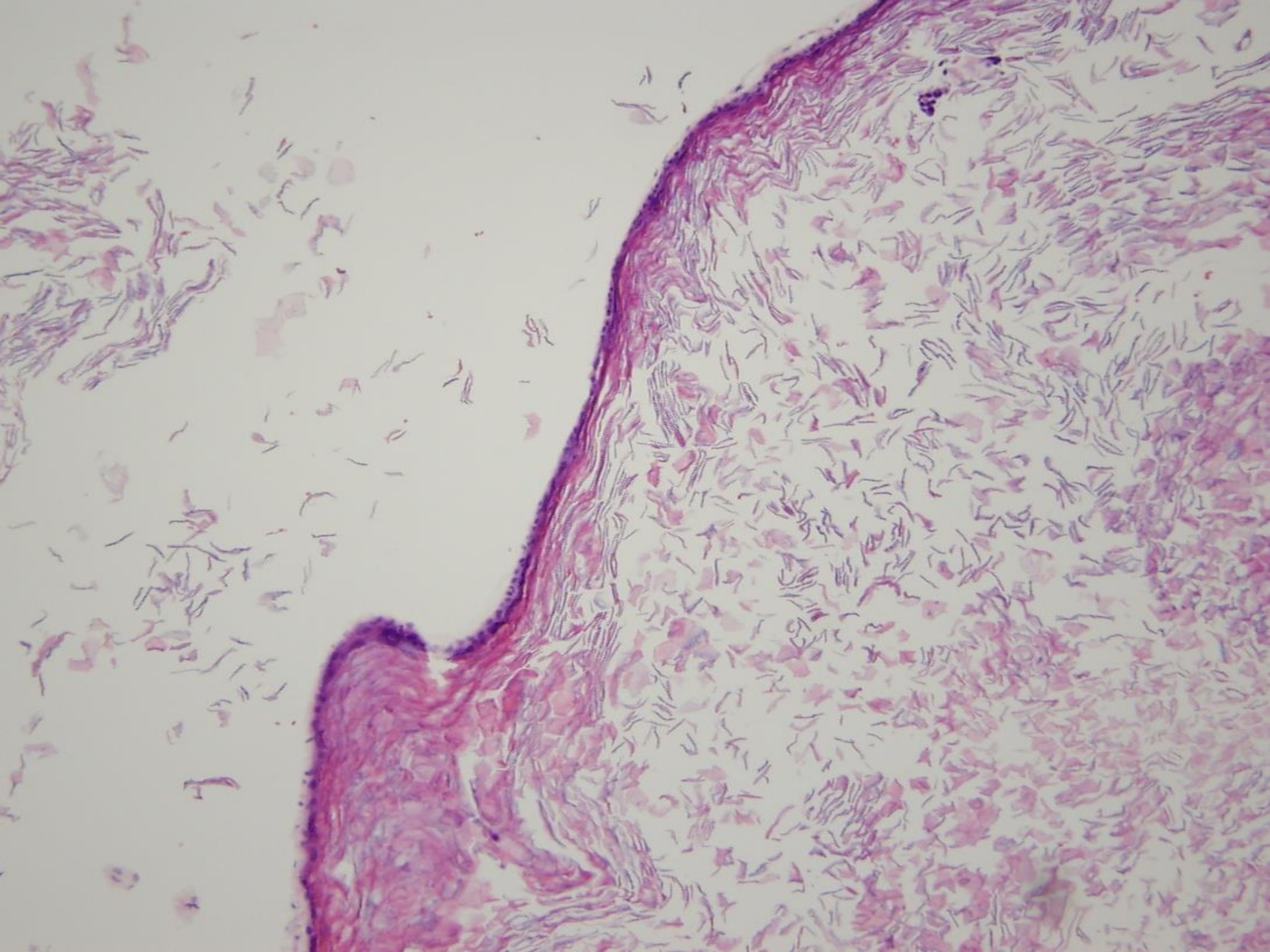
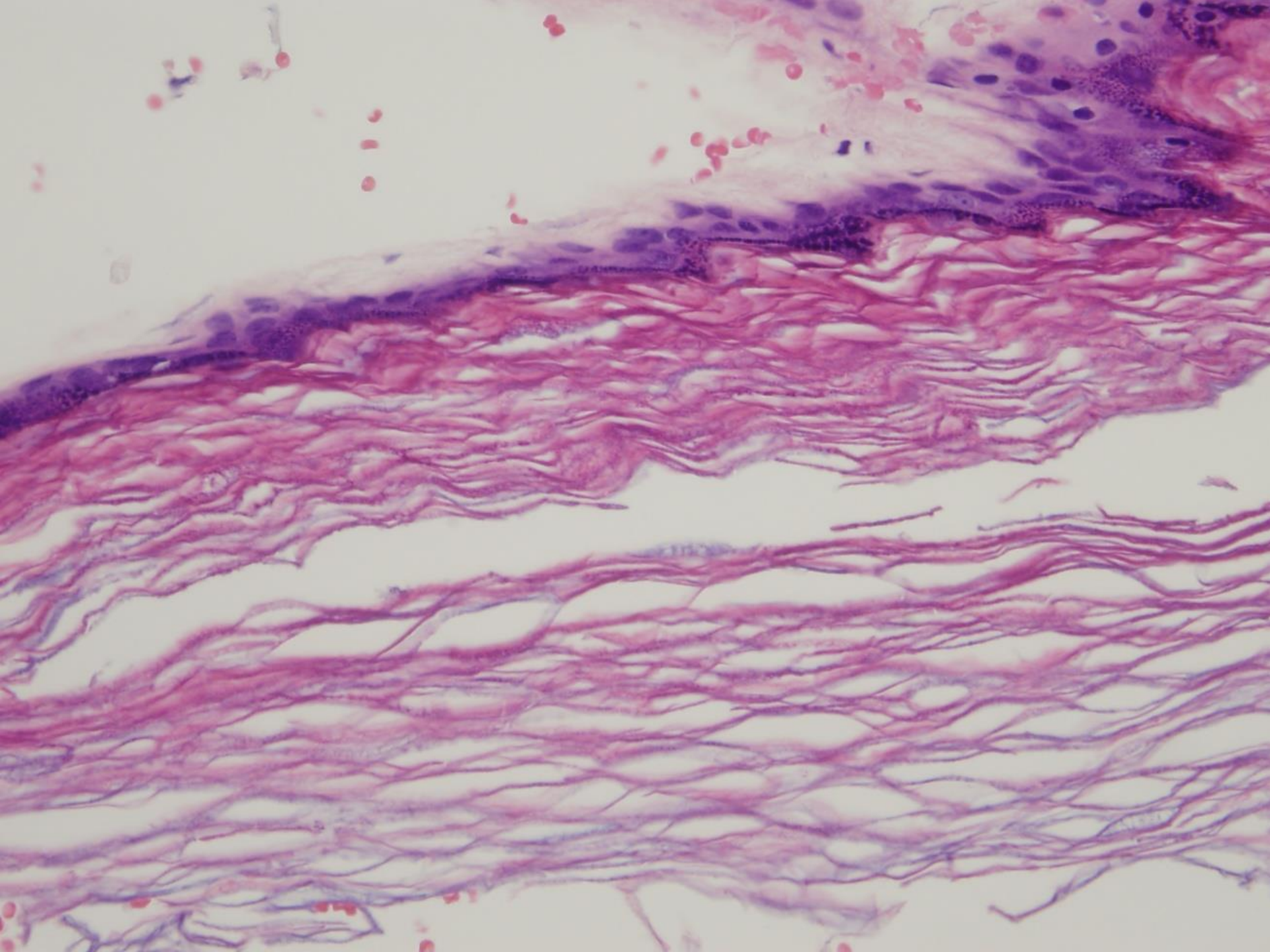


PD

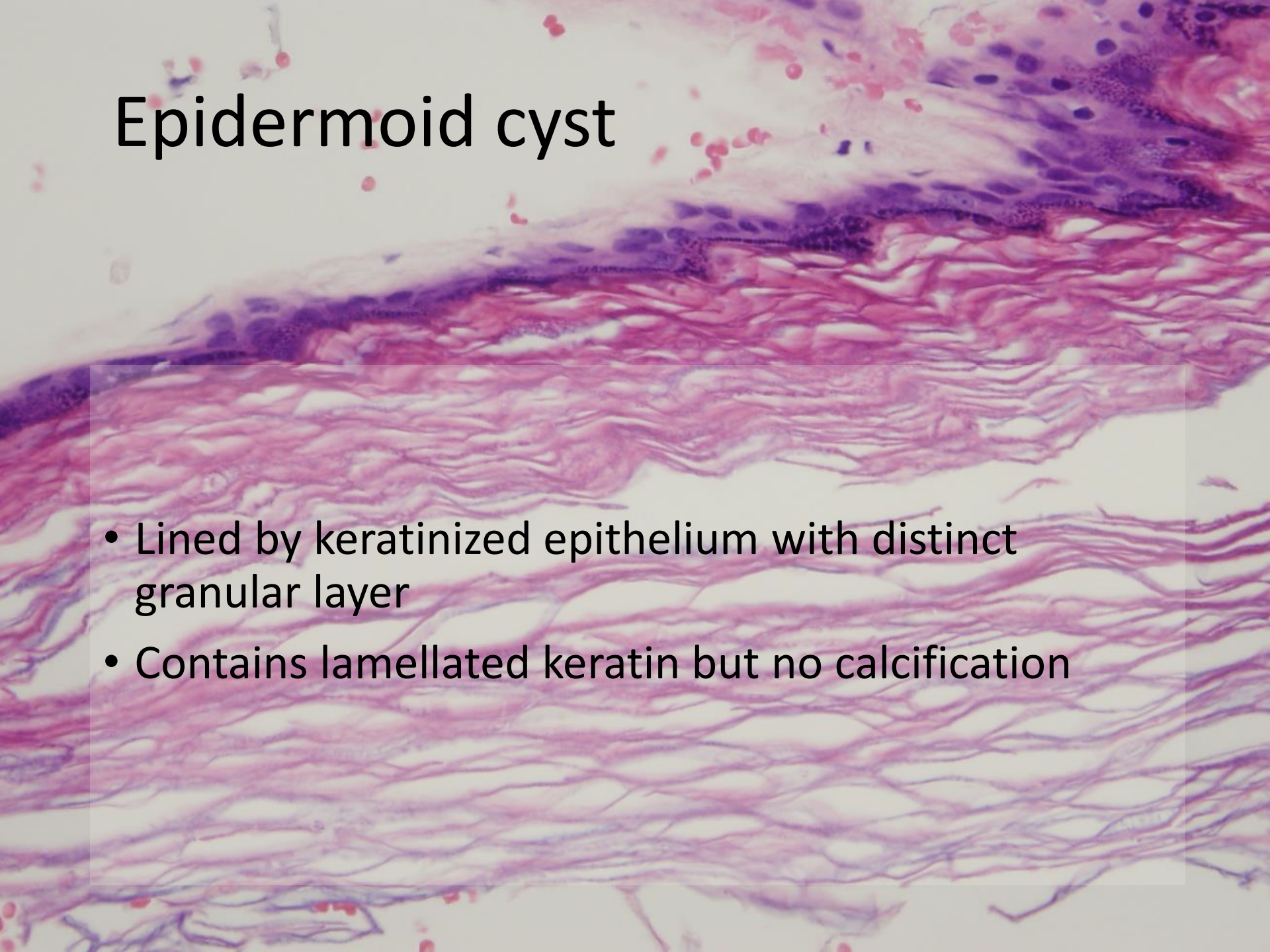




SPECIMEN DESIGNATED "RIGHT CEREBELLAR MASS":

- EPIDERMOID CYST.

Epidermoid cyst



- Lined by keratinized epithelium with distinct granular layer
- Contains lamellated keratin but no calcification

Epidermoid

- T1 usually iso to CSF, may be high signal
- T2 usually iso to CSF or slightly hyperintense
- FLAIR often heterogeneous
- DWI increased restriction
- T1 C+ non-enhancing, sometimes thin rim

- Benign inclusion of ectodermal elements during neural tube closure (desquamated epithelial cells)
- Identical to cholesteatoma

Epidermoid

- Slow-growing congenital lesions that remain clinically silent for many years
- Smaller lesions are readily cured with surgery
- Larger lesions with upward supratentorial herniation are more difficult to completely remove, but recurrence is slow growing

Case 4

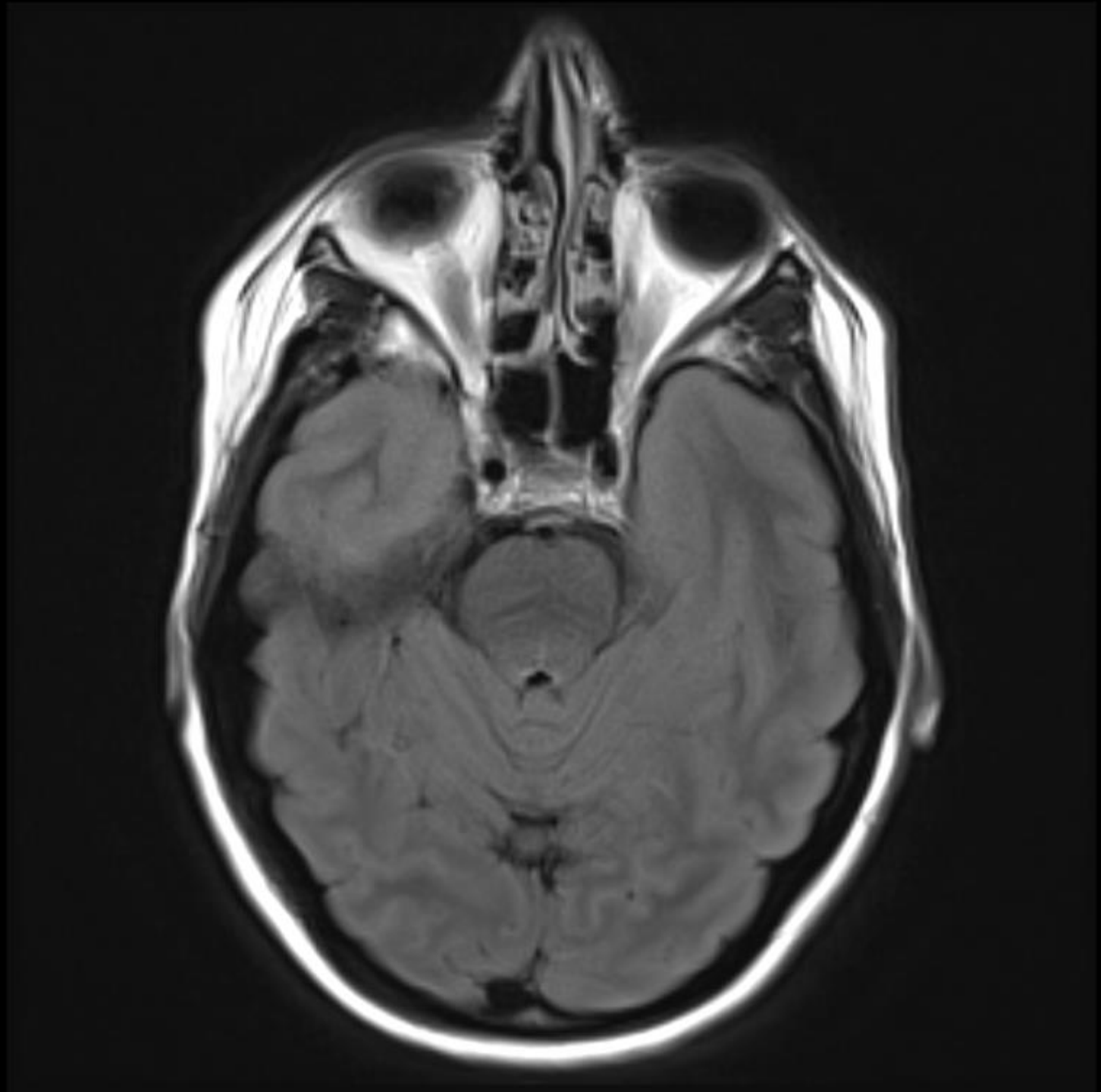
Clinical Condition: **Orbits, Vision and Visual Loss**

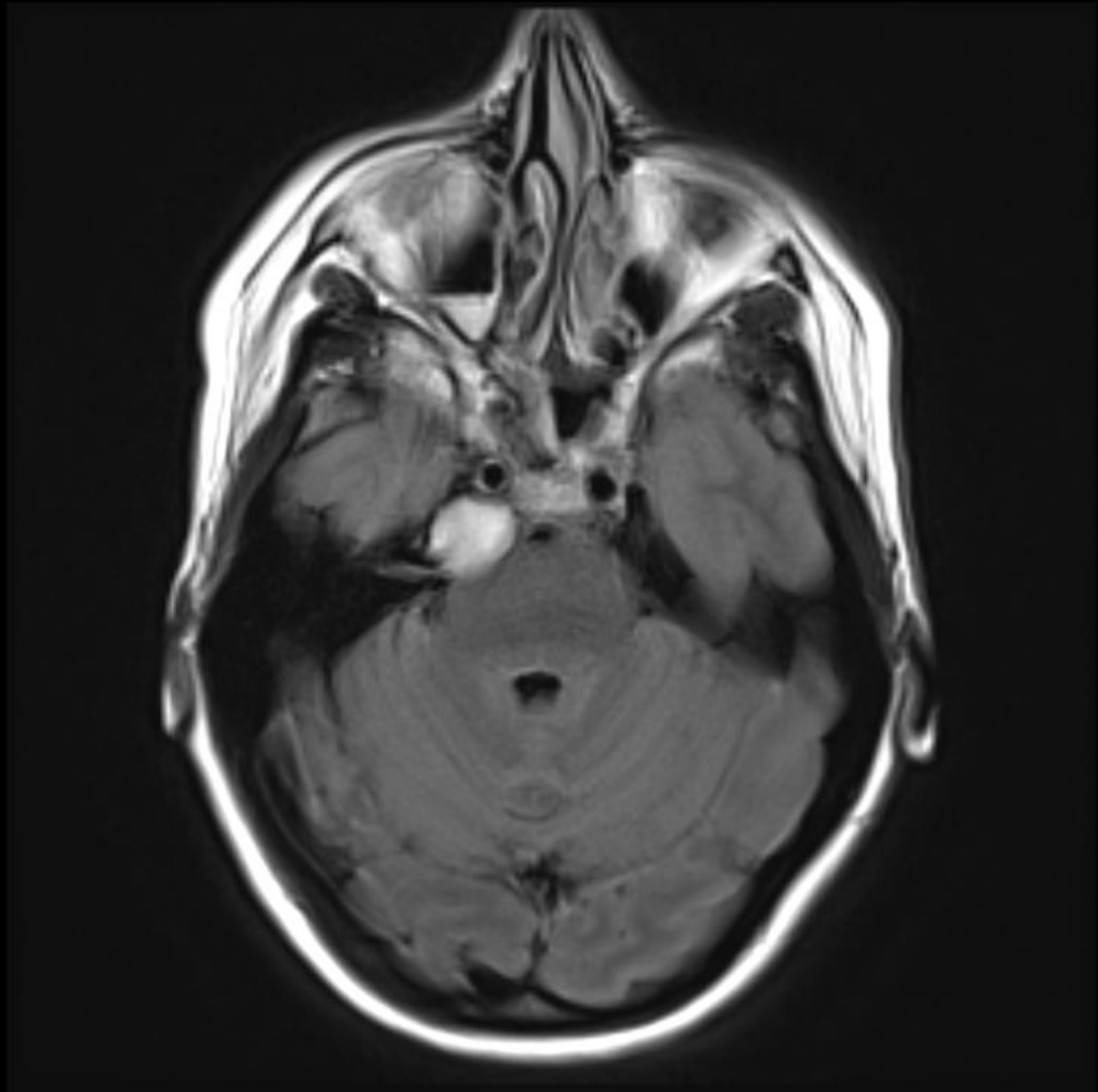
Variant 6: **Adult patient with ophthalmoplegia.**

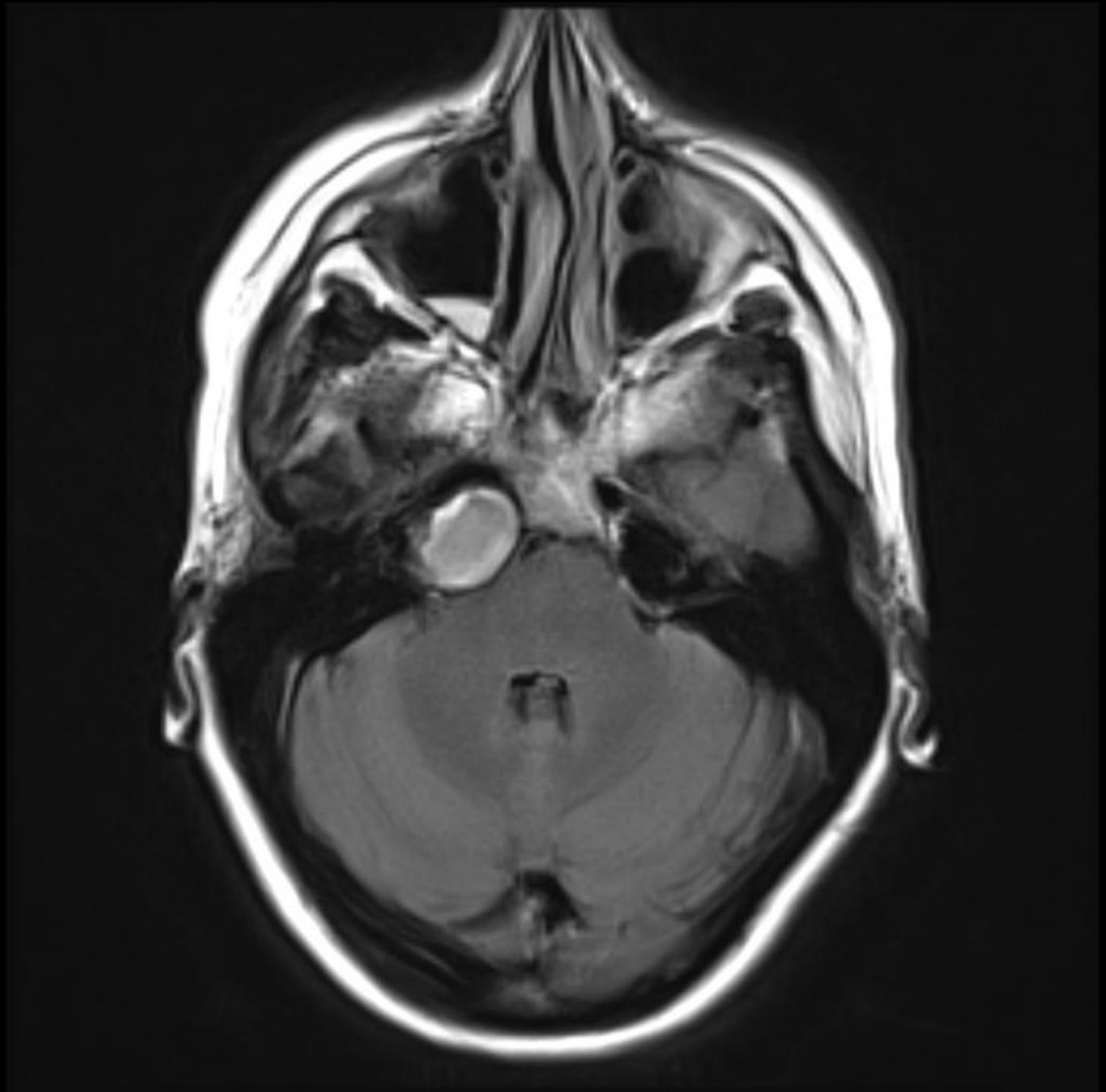
Radiologic Procedure	Rating	Comments	RRL*
MRI head and orbits without and with contrast	9	See statement regarding contrast in text under "Anticipated Exceptions."	O
MRI head and orbits without contrast	6		O
MRA head and neck without contrast	6		O
MRA head and neck without and with contrast	6	See statement regarding contrast in text under "Anticipated Exceptions."	O
CT head with contrast	6	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢☢☢
CT head without and with contrast	6	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢☢☢
CTA head and neck with contrast	6	If vascular disease is suspected.	☢☢☢
CT head without contrast	5	Thin slices dedicated to the orbits are useful for orbit disease and may be substituted for the complete head examination in selected patients.	☢☢☢
X-ray orbit	1		☢
Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			*Relative Radiation Level

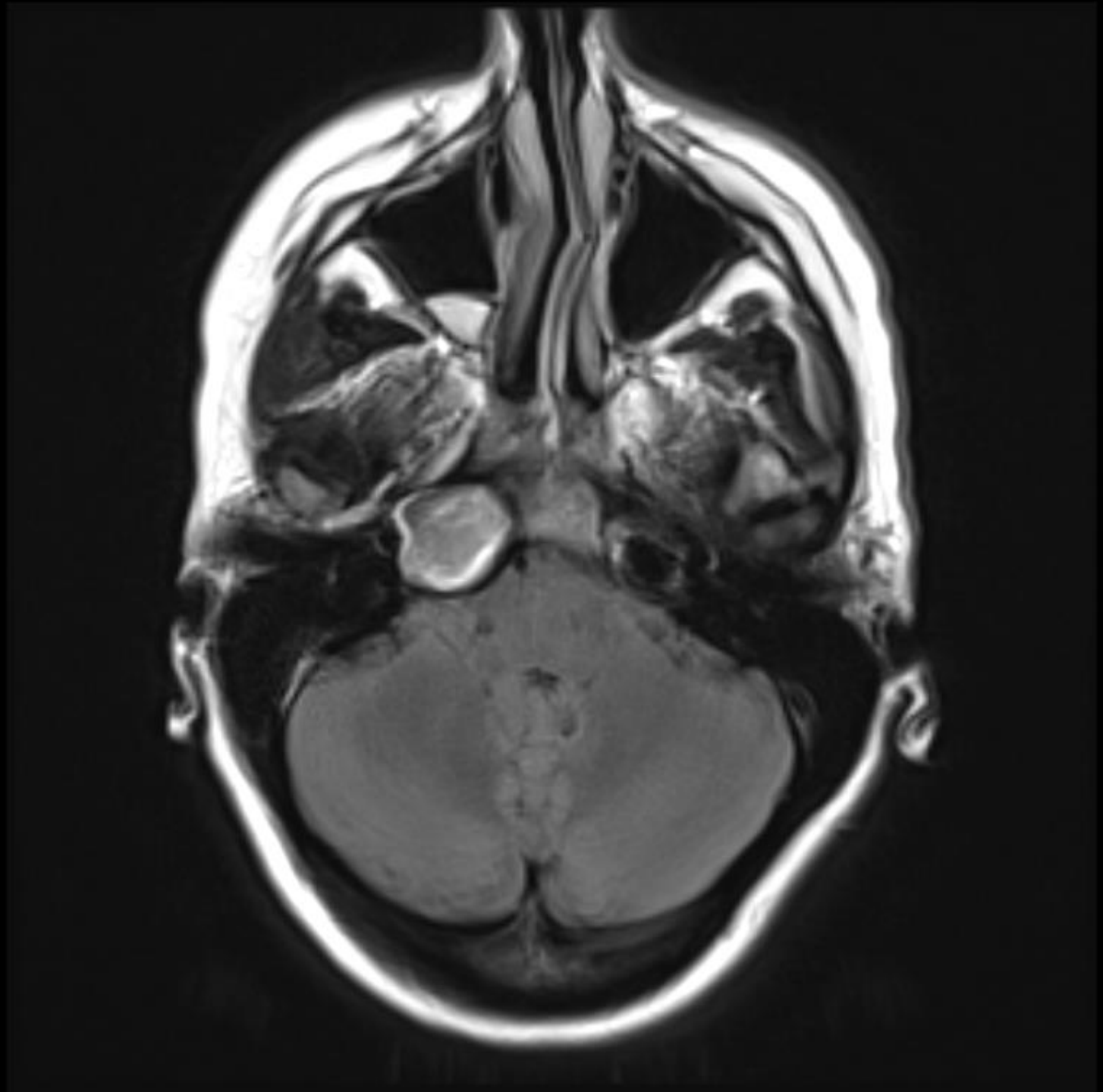
38 Female

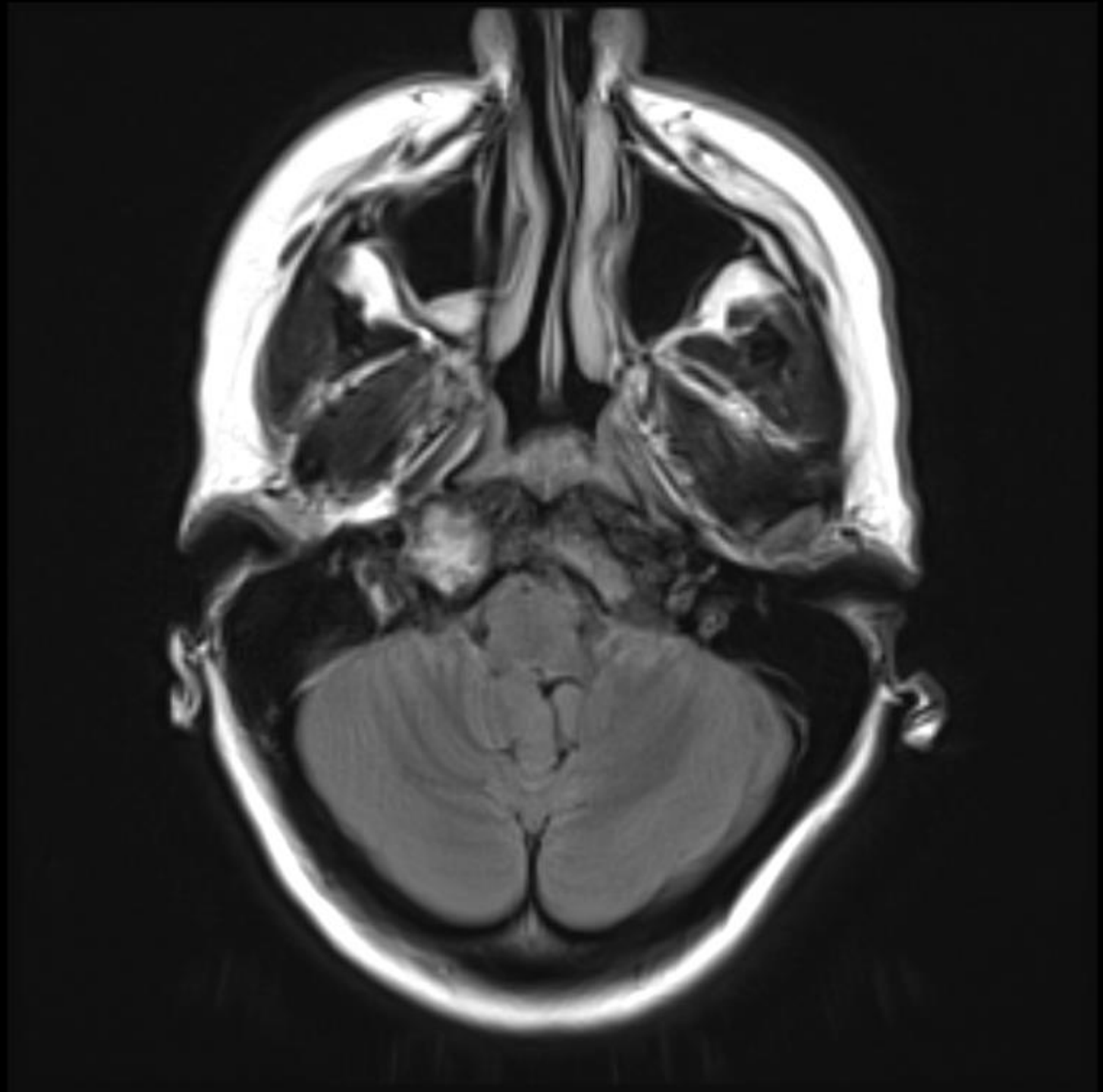
- Difficulty focusing x 8 days
- Double vision w/ lateral gaze x 7 days
- Right-sided weakness x few days
- No hearing loss
- Right aural fullness
- Intermittent dizziness x 1 week
- Reduced right facial sensation on exam

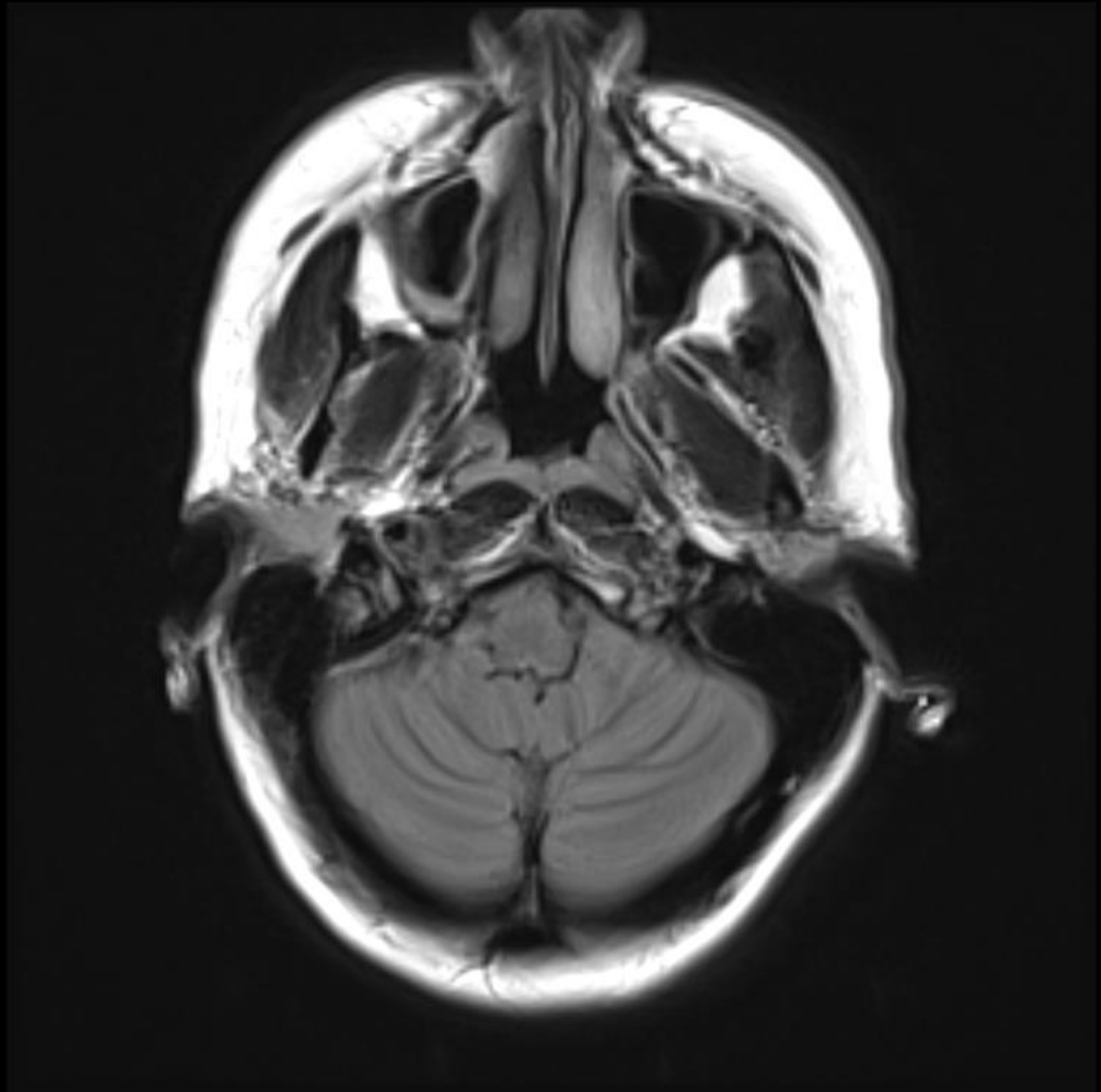


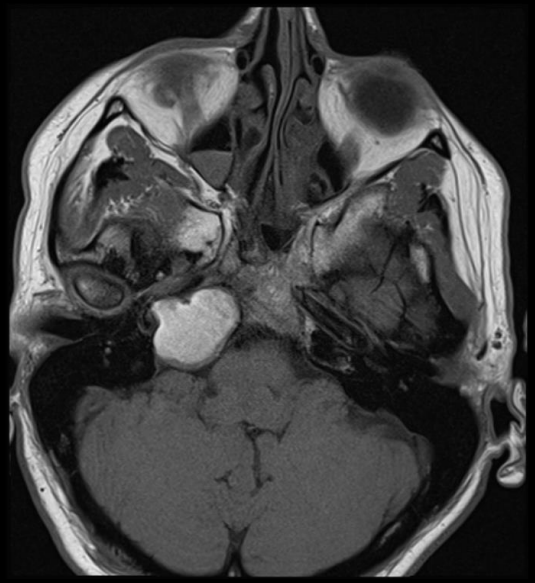












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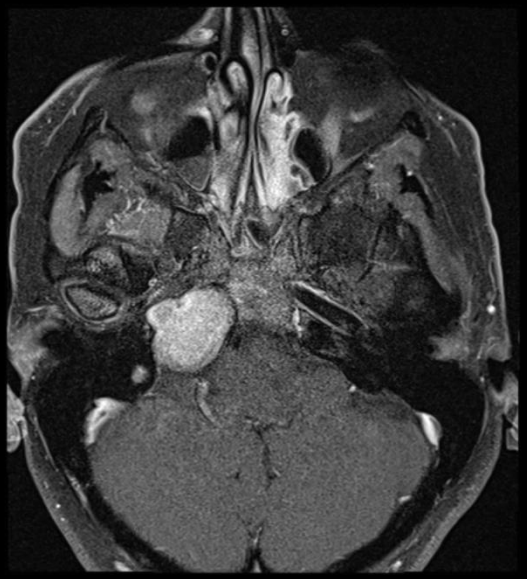

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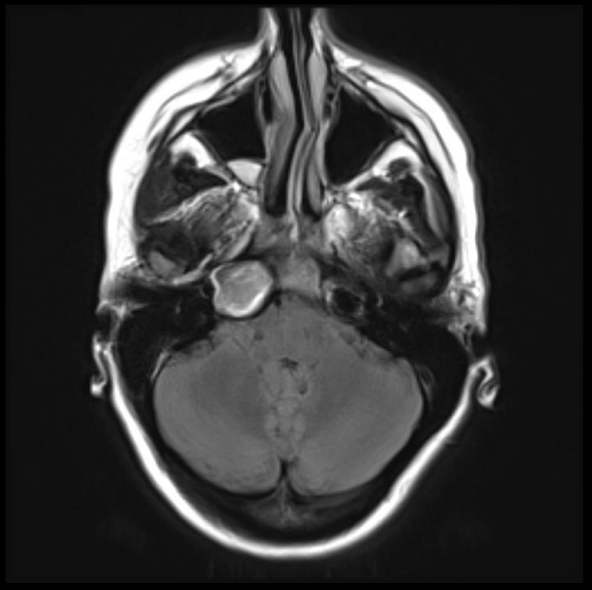
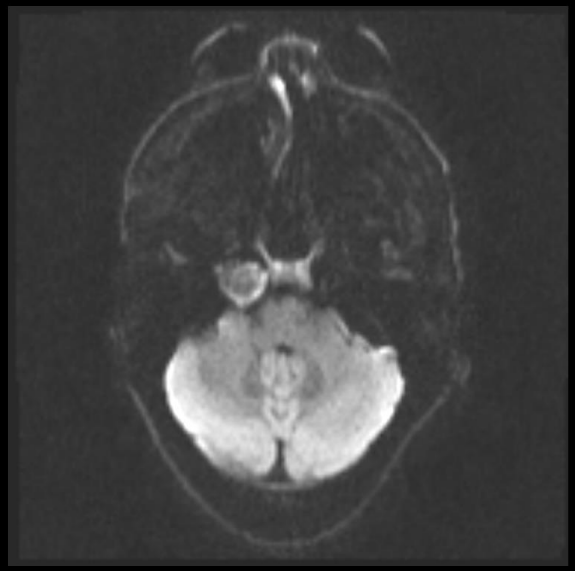
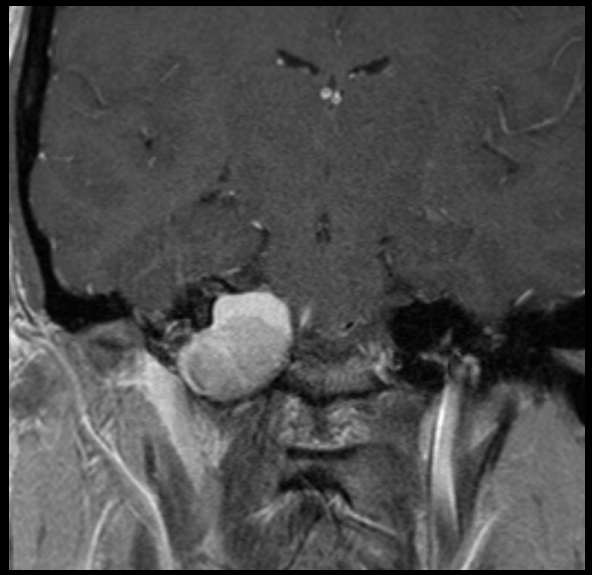


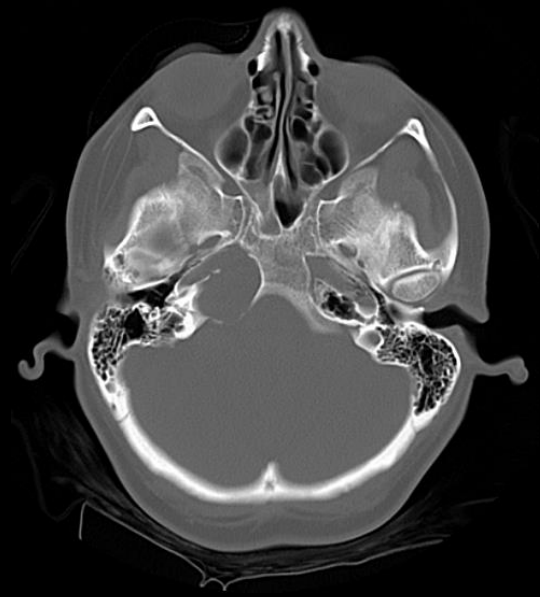
T2 FLAIR



DWI



T1 VIBE



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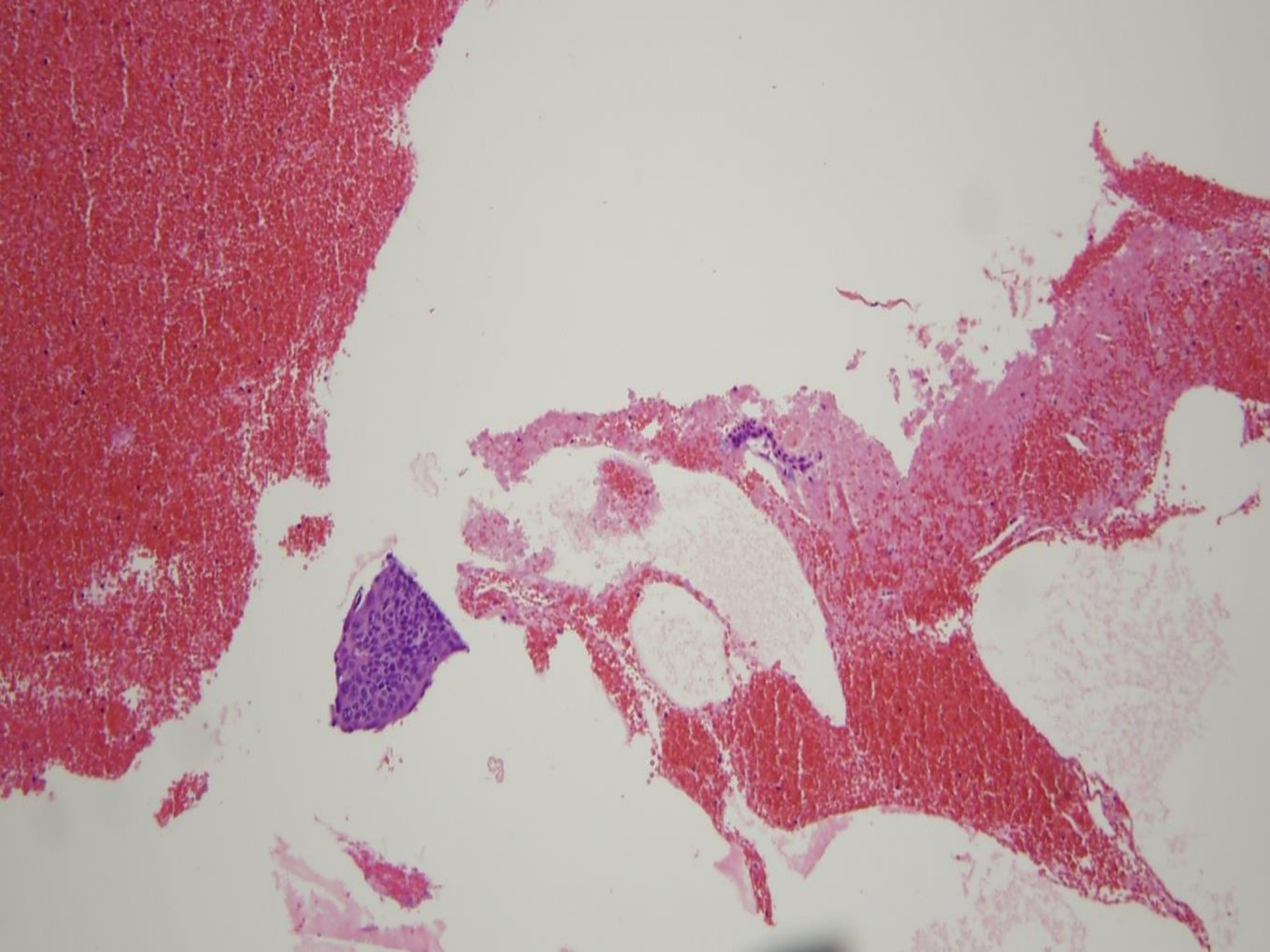
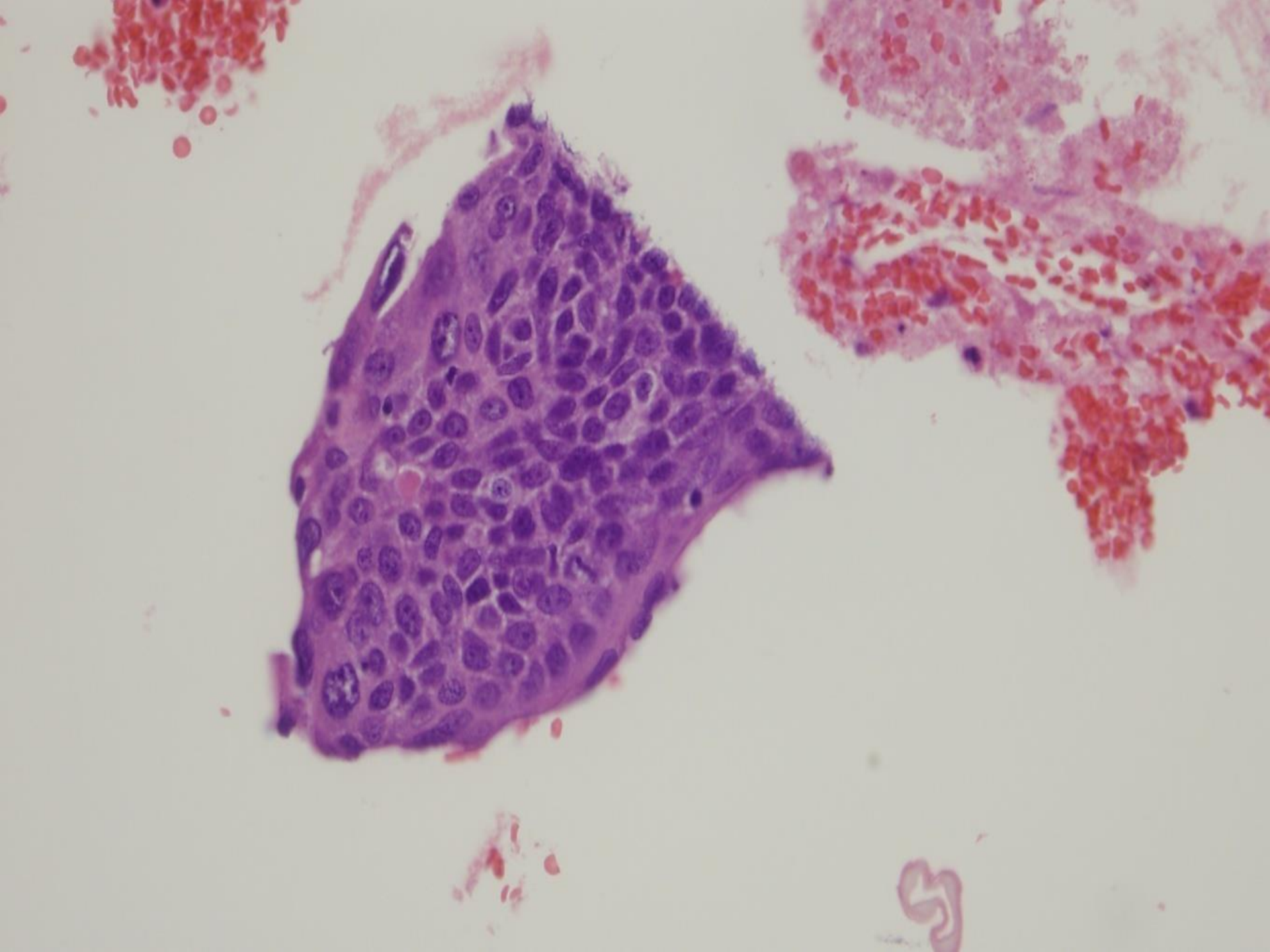
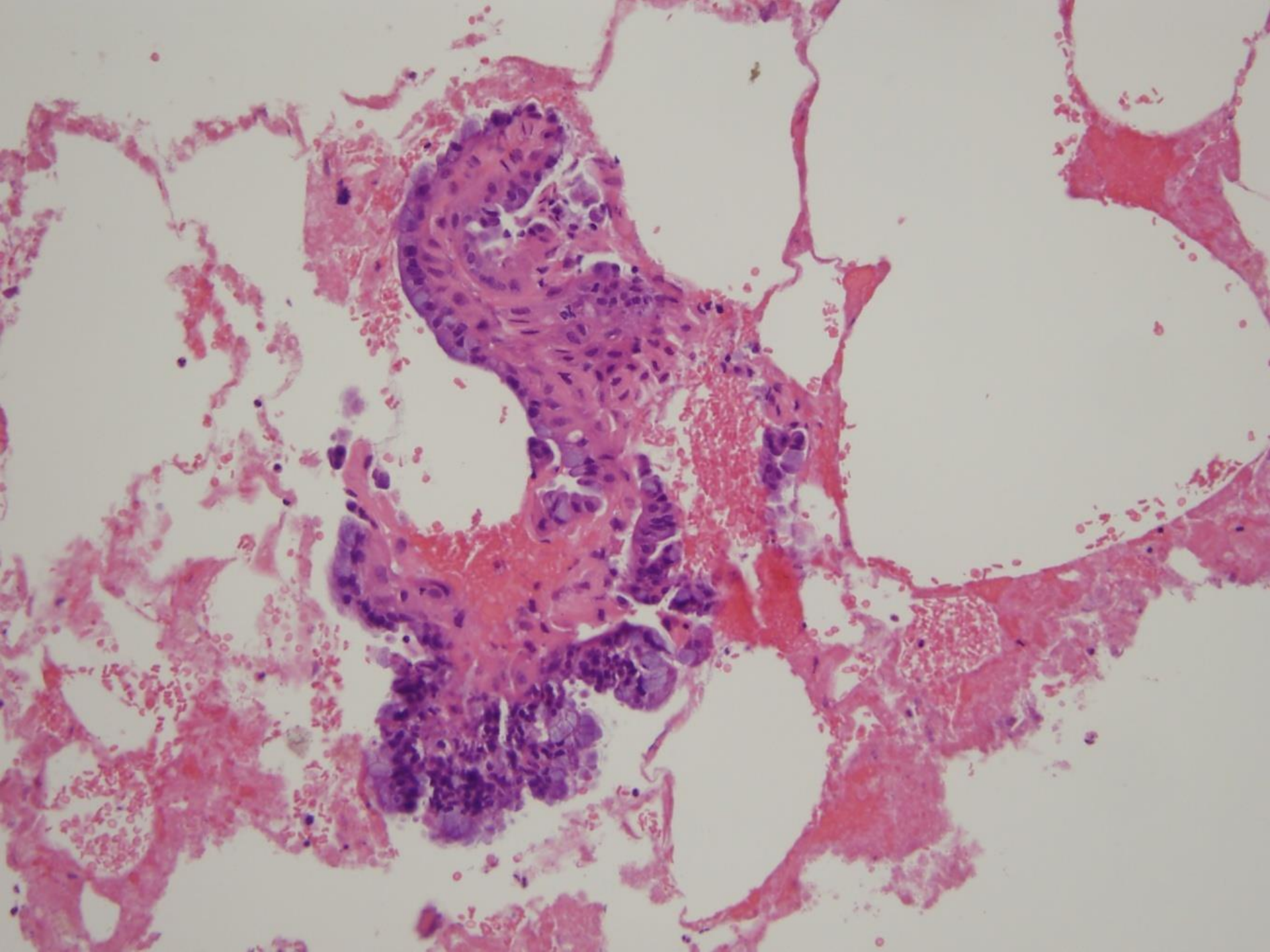
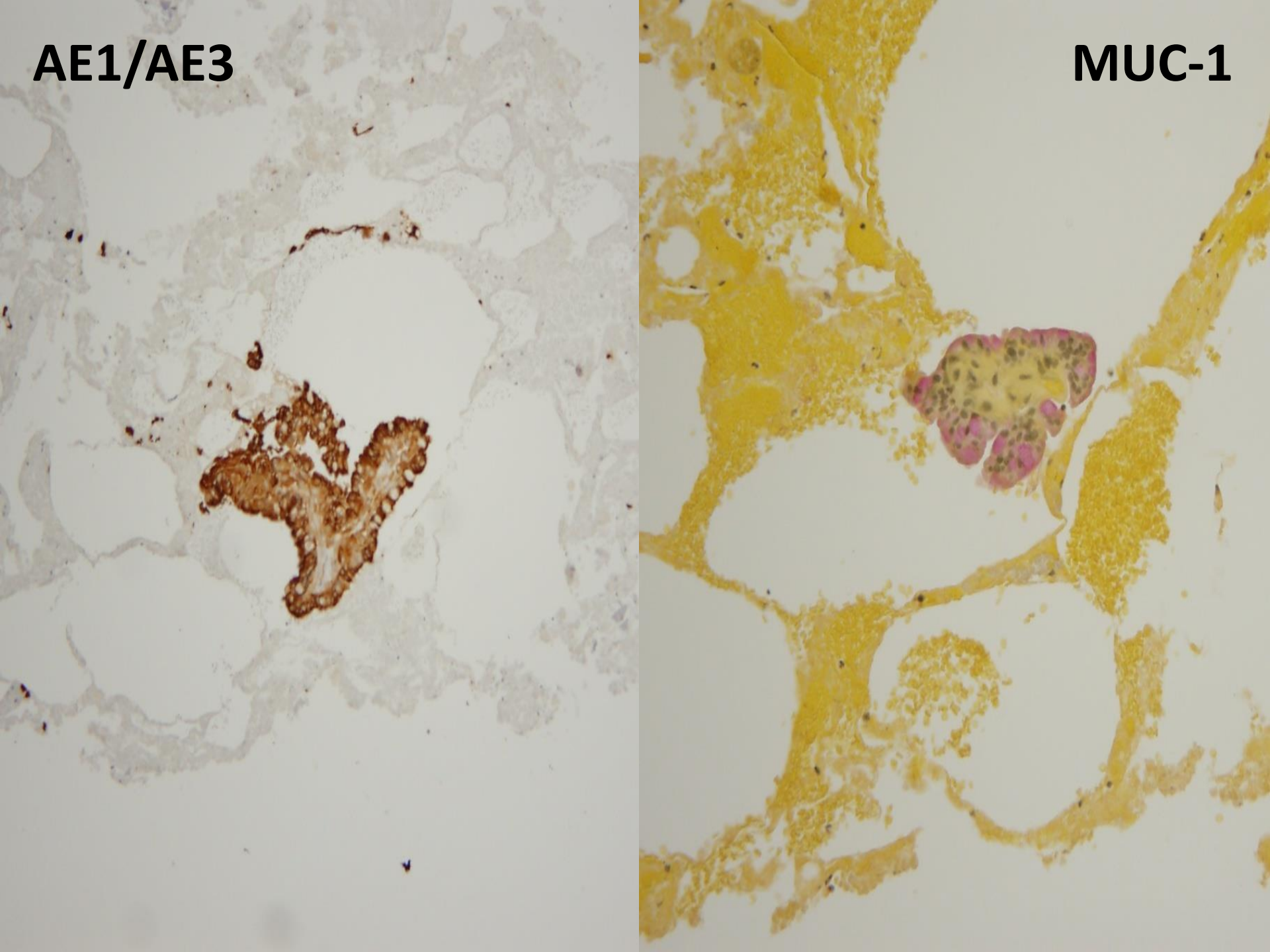







AE1/AE3
MUC-1

SPECIMEN DESIGNATED "WALL OF CHOLESTEROL GRANULOMA":

UNCLASSIFIED CYST LINING consisting of strands of dense fibrous connective tissue lined by epithelium of variable histologic characteristics (See NOTE).

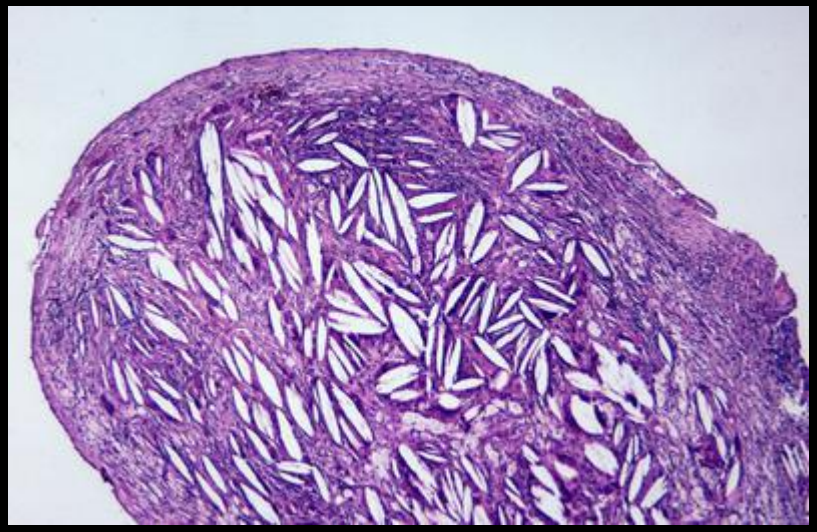
NOTE: The tissue consists of large pools of acute hemorrhage. In addition, there are several strands of fibroconnective tissue with variable numbers of chronic inflammatory cells. In some areas, the connective tissue is lined by epithelium that also contains mucin-secreting cells (demonstrated by AE1/3 keratin immunostain and mucicarmine stain).

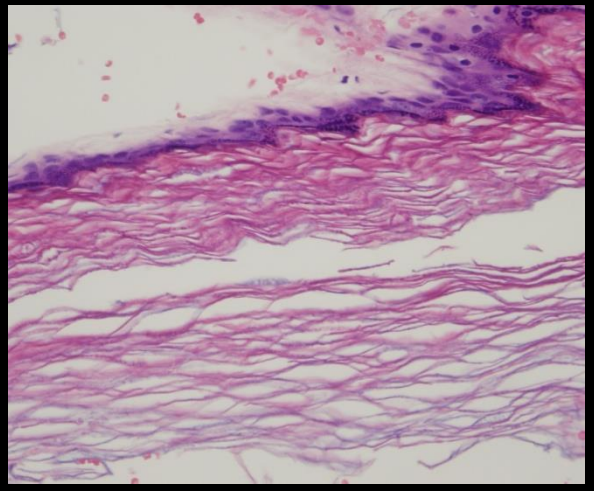
These strands likely represent the lining of a cyst, the nature of which is not entirely clear; its histological properties are not especially in keeping with the clinical diagnosis of a “cholesterol granuloma” inasmuch as there are no granulomas or cholesterol clefts (or desquamating keratin deposits) in the material received.

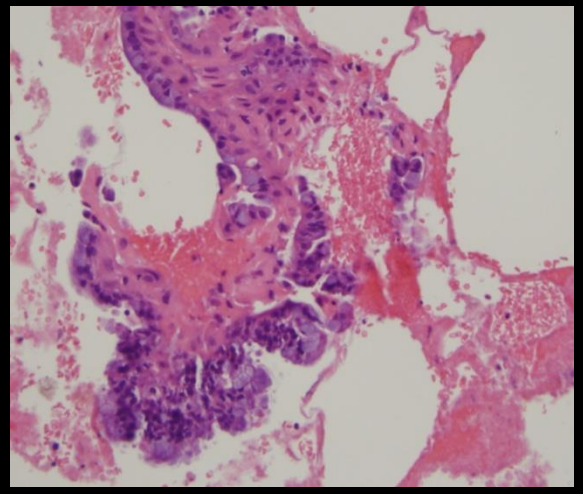
The case was reviewed by Dr. Harry Kozakewich (Pathology, Boston Children Hospital) who raised the possibility of a petrous apex “mucocele.”

Common cystic lesions of the petrous apex:

- Cholesterol granuloma
- Epidermoid cyst
- Mucocele







Cholesterol Granuloma vs Mucocele

Cholesterol Granuloma

- T1 high w/ maybe low signal rim and thinned adjacent bone
- T2/FLAIR central high peripheral low
- T1 C+ possible faint peripheral enhancement

- Middle ear granulation tissue – eustachian tube dysfunction vs exposed marrow -> bleeding -> inflammation
- Cholesterol crystals, multi-nucleated giant cells, hemosiderin, RBC breakdown

Cholesterol Granuloma

- Growth rate highly variable (depends on frequency & severity of microhemorrhages)
- Asymptomatic patients can be safely followed with imaging
- Extended middle cranial fossa approach with extradural removal of PA-CG & obliteration of its cavity

Mucocele

- T1 low (CG T1 high)
- T2/FLAIR high
- T1 C+ nonenhancing
- DWI increased diffusion

- Mucus-containing, expanded petrous apex (PA) air cells(s) lined by secretory epithelium resulting from chronic ostial obstruction

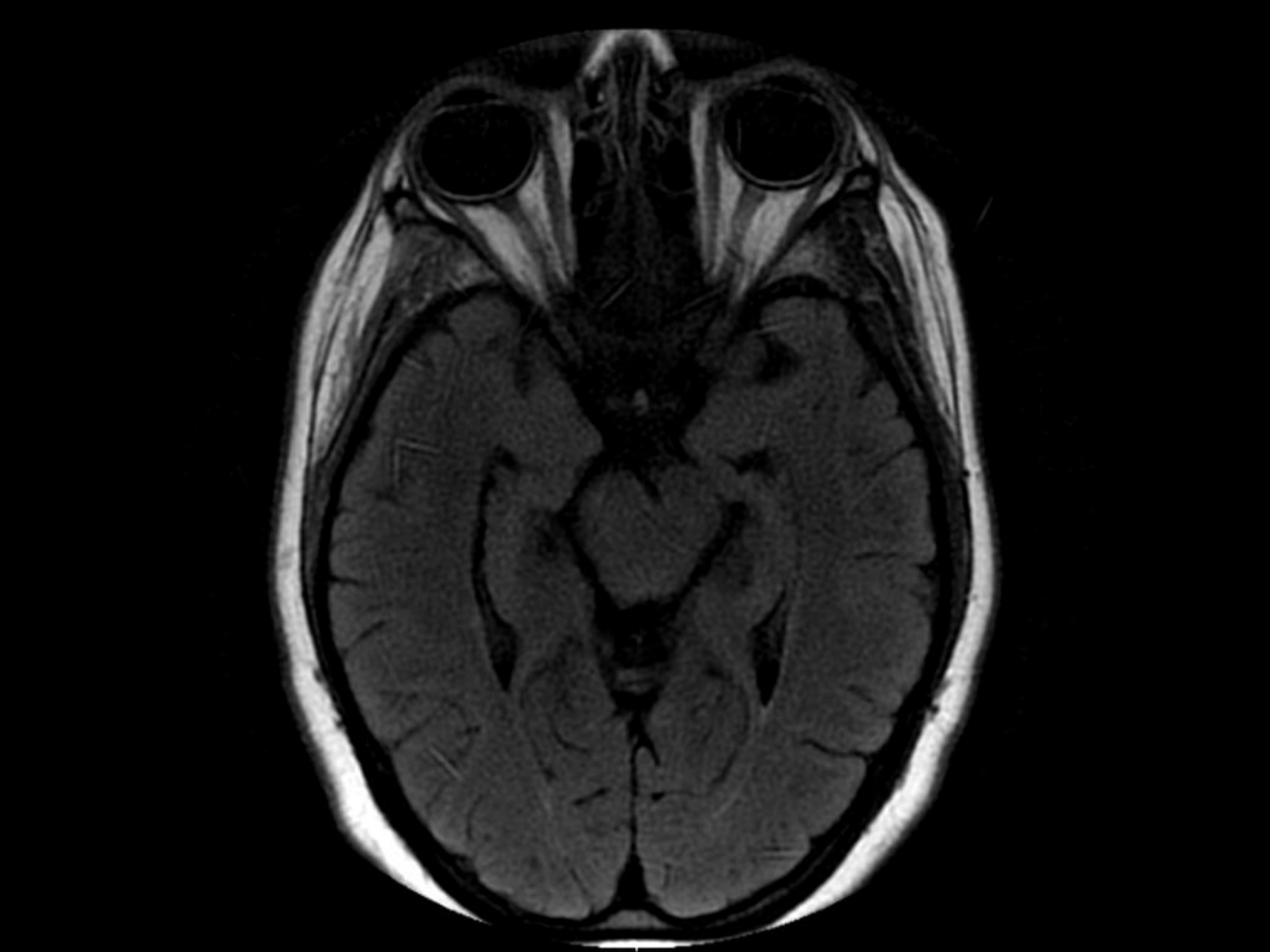
Mucocele

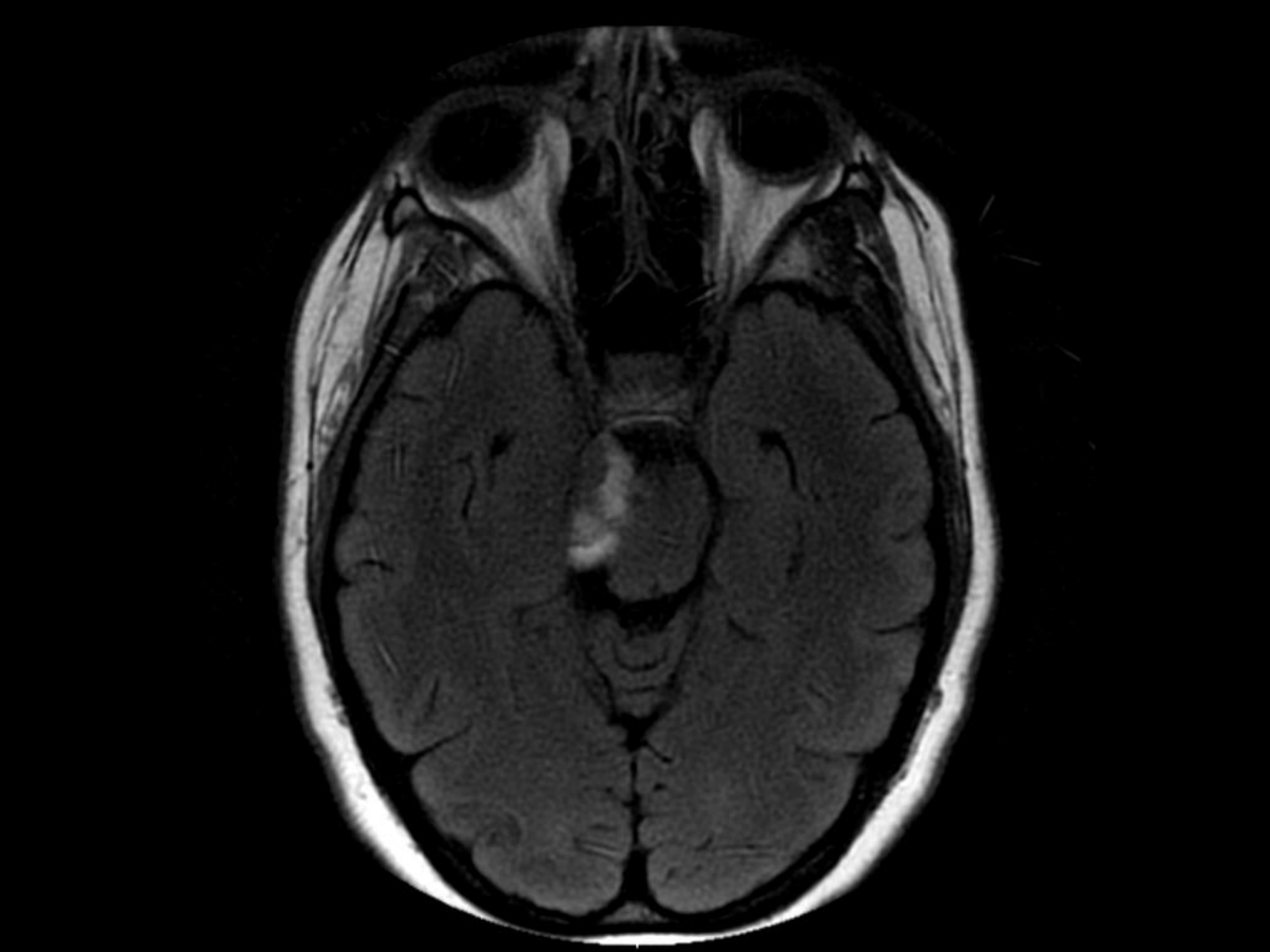
- Usually incidental finding
- Treatment controversial

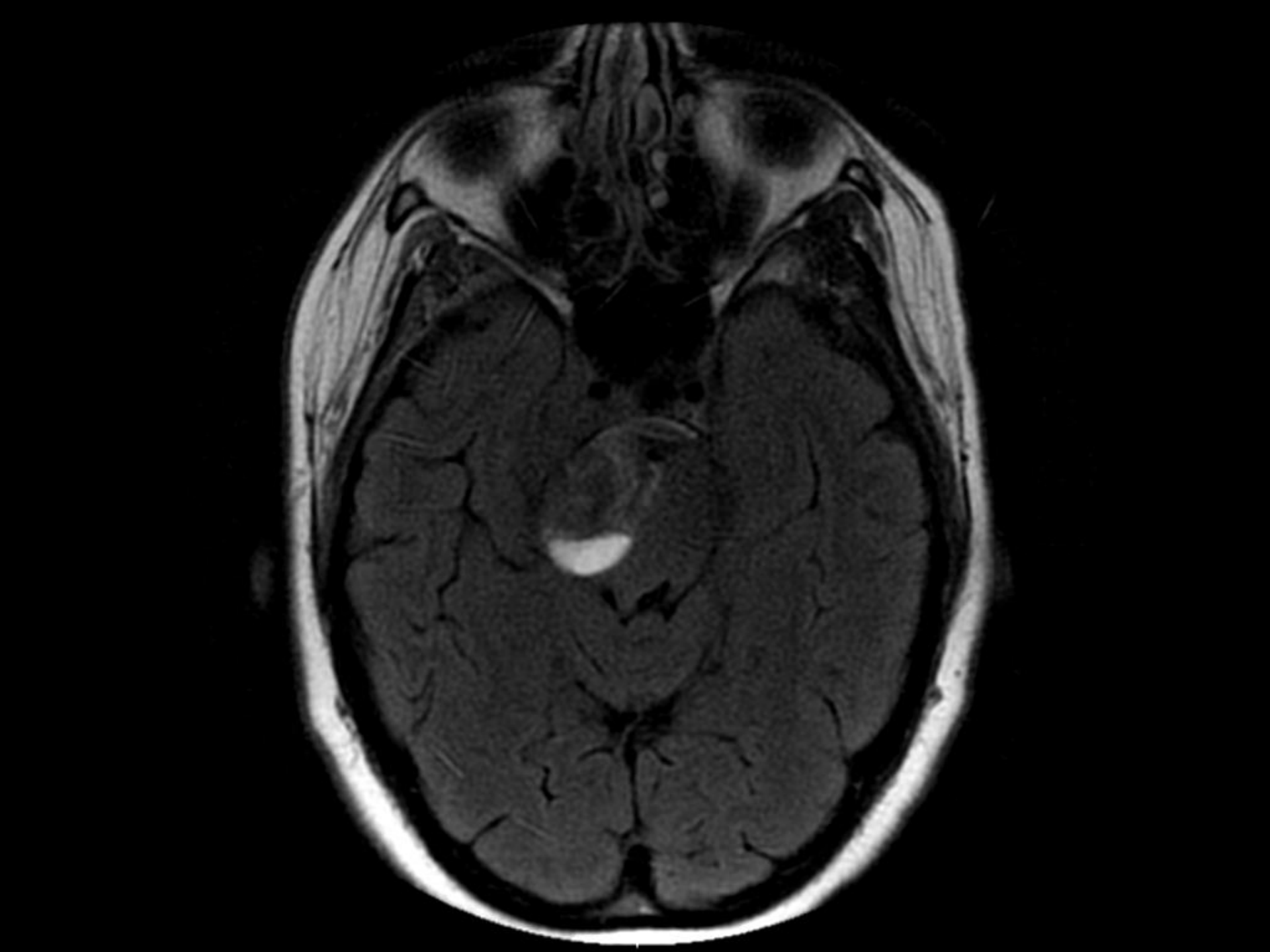
Case 5

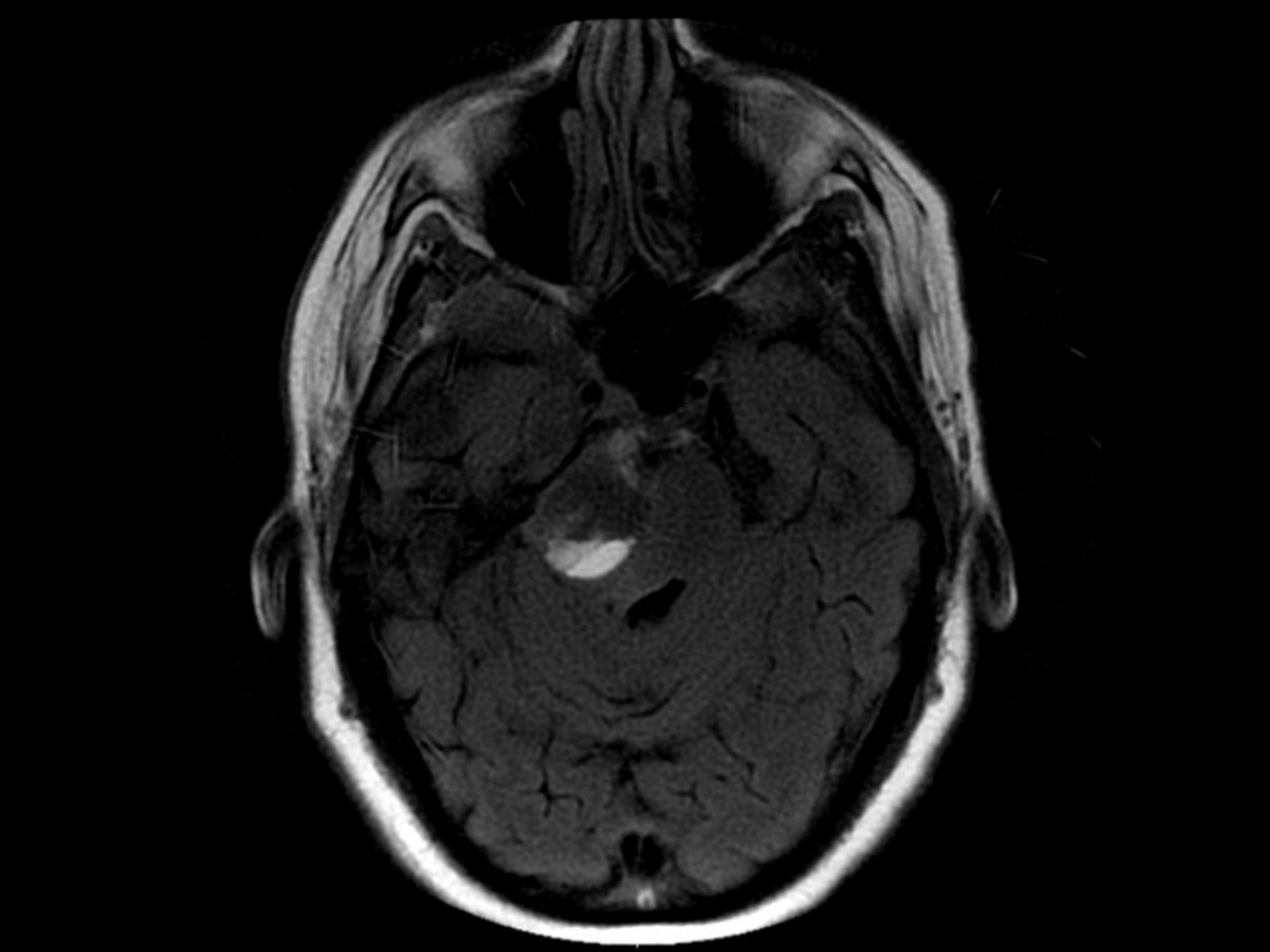
25 Female

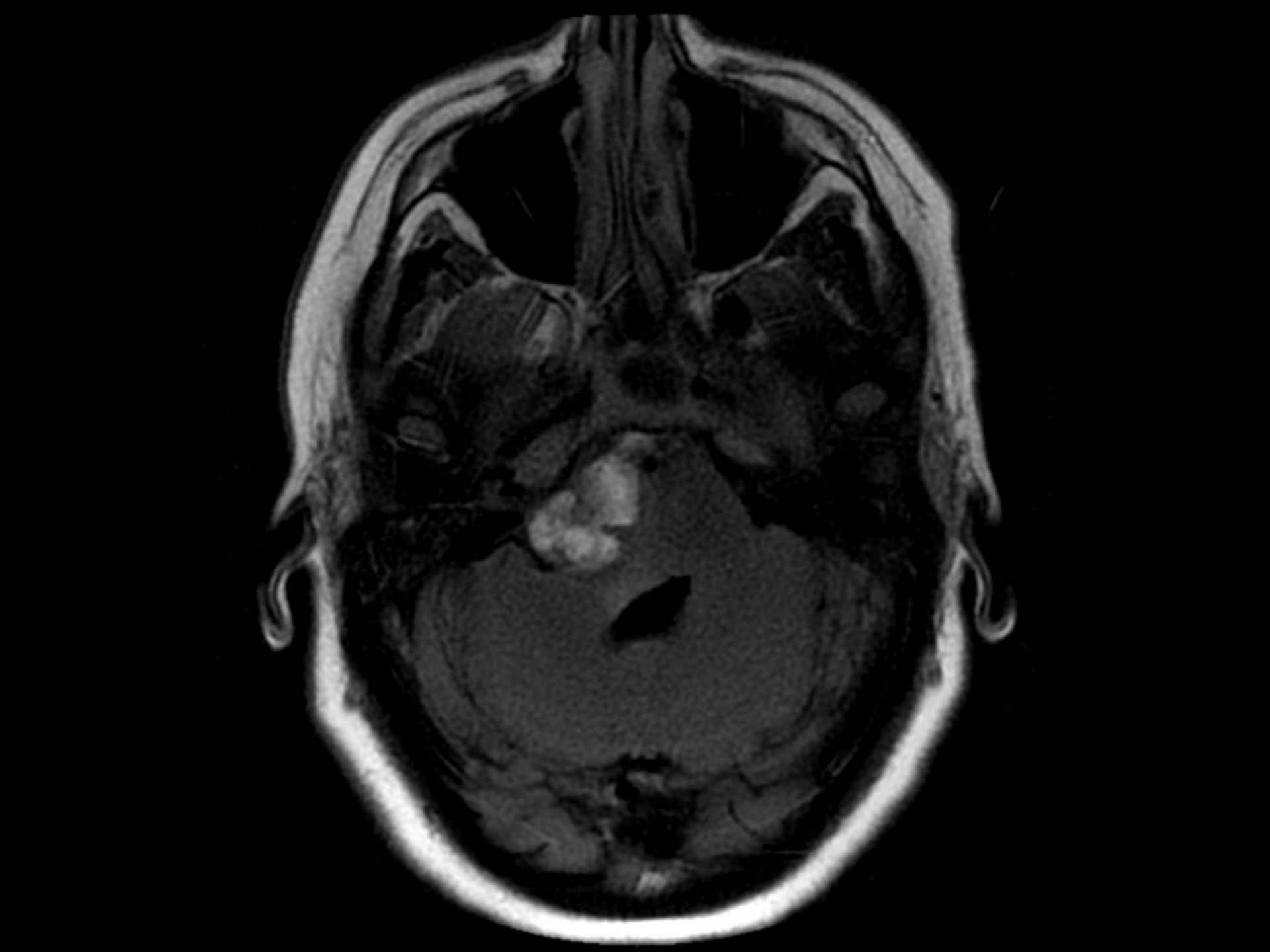
- One day s/p c-section
- Right V1-3 facial numbness/tingling x 1 month
- Occasional right facial pain
- Progressive hearing loss x 1 month
- Diplopia in all directions x 1 week
- Nausea x 1 week
- Brother with congenital hydrocephalus

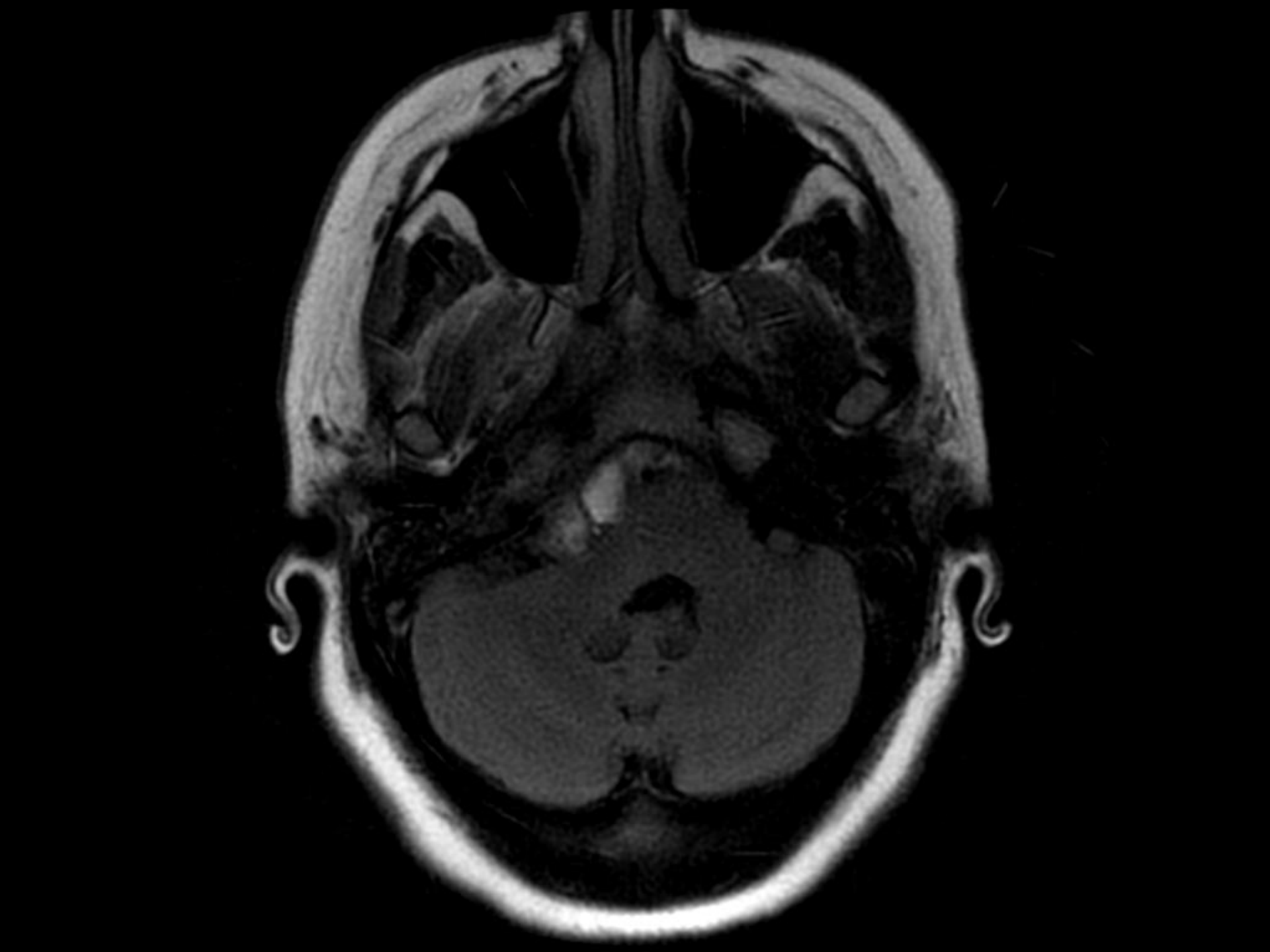


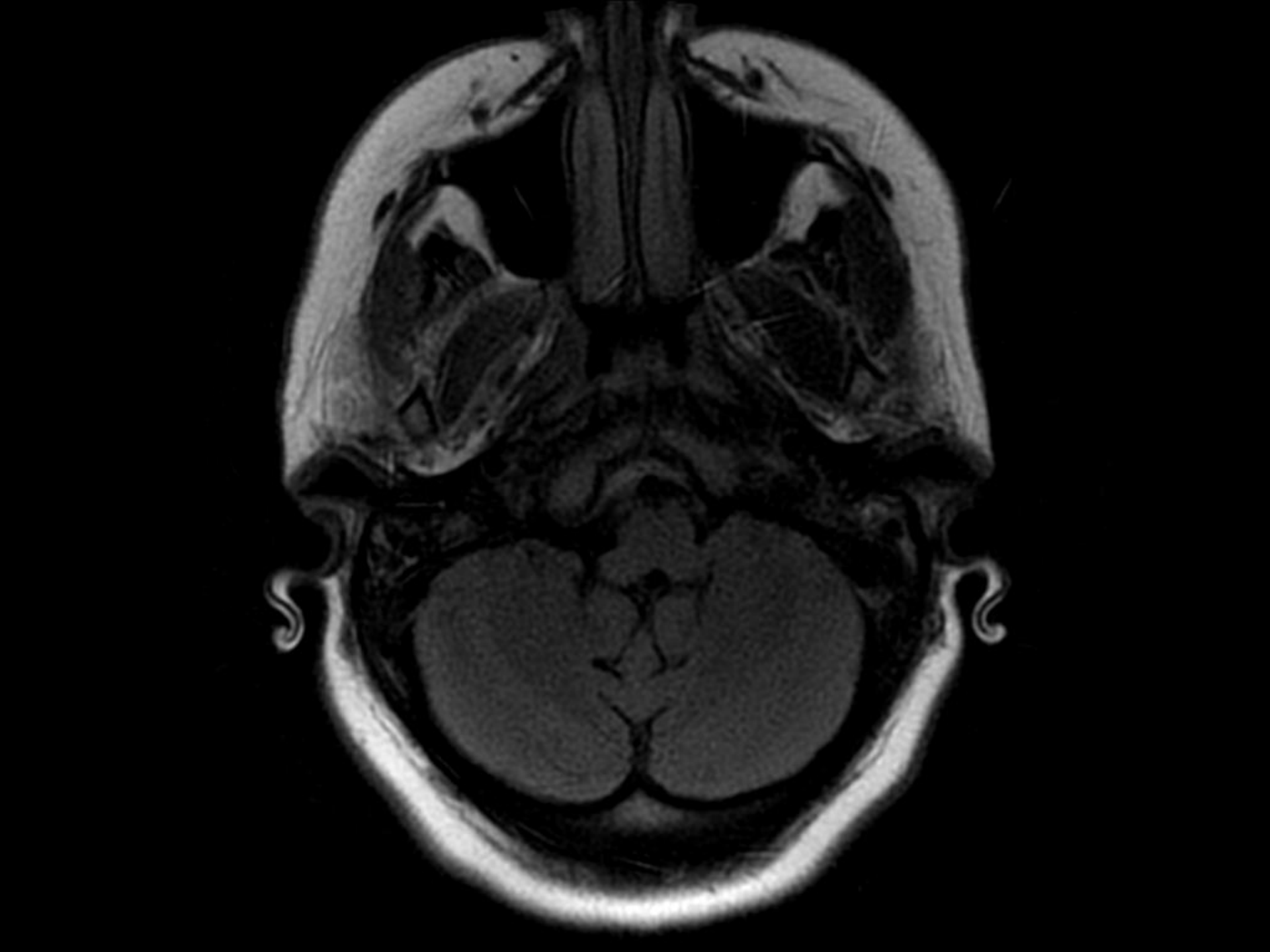


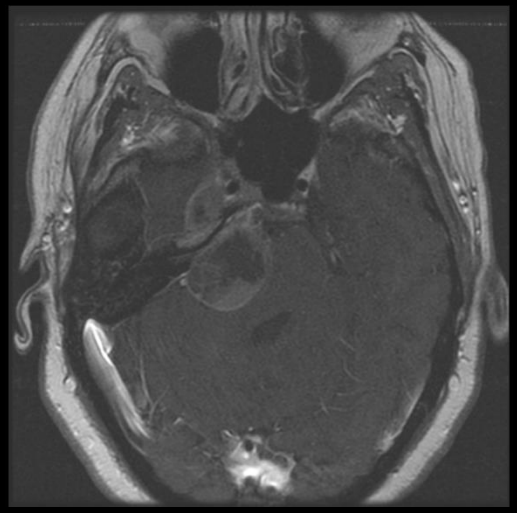












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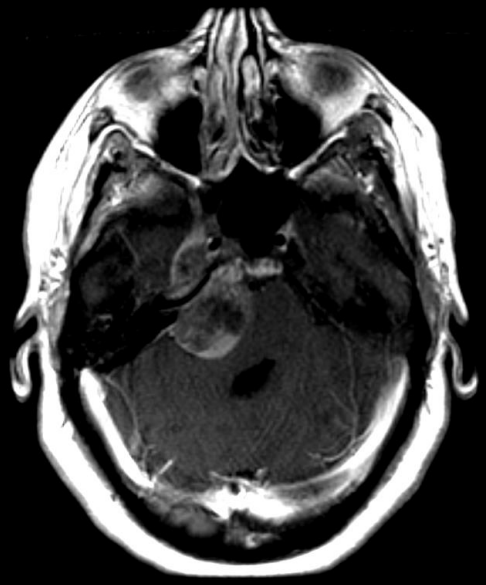
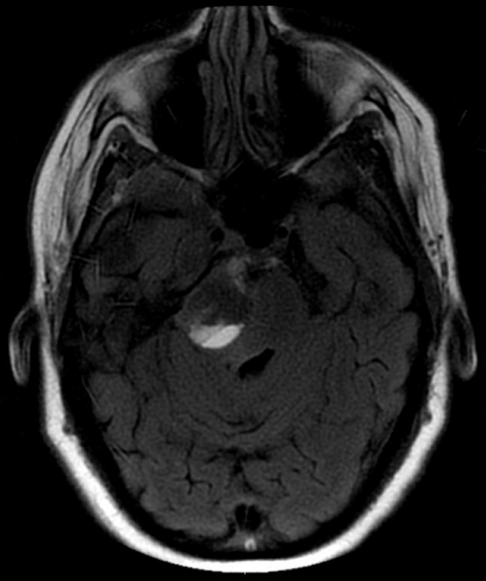
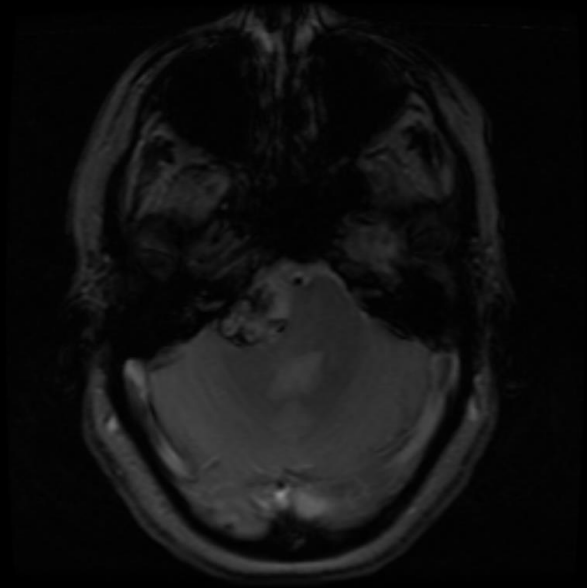


T1 Post



T2 FLAIR



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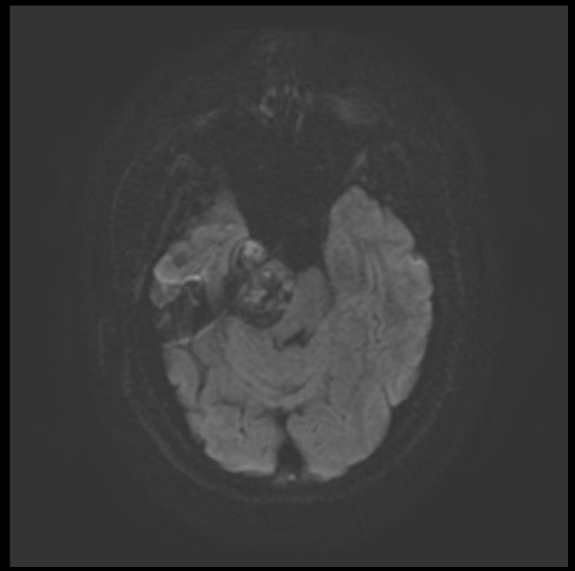

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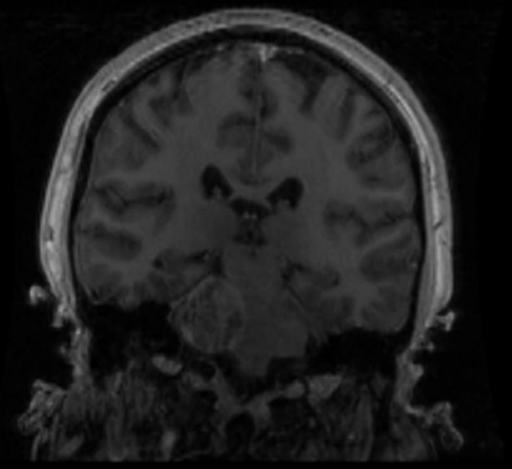
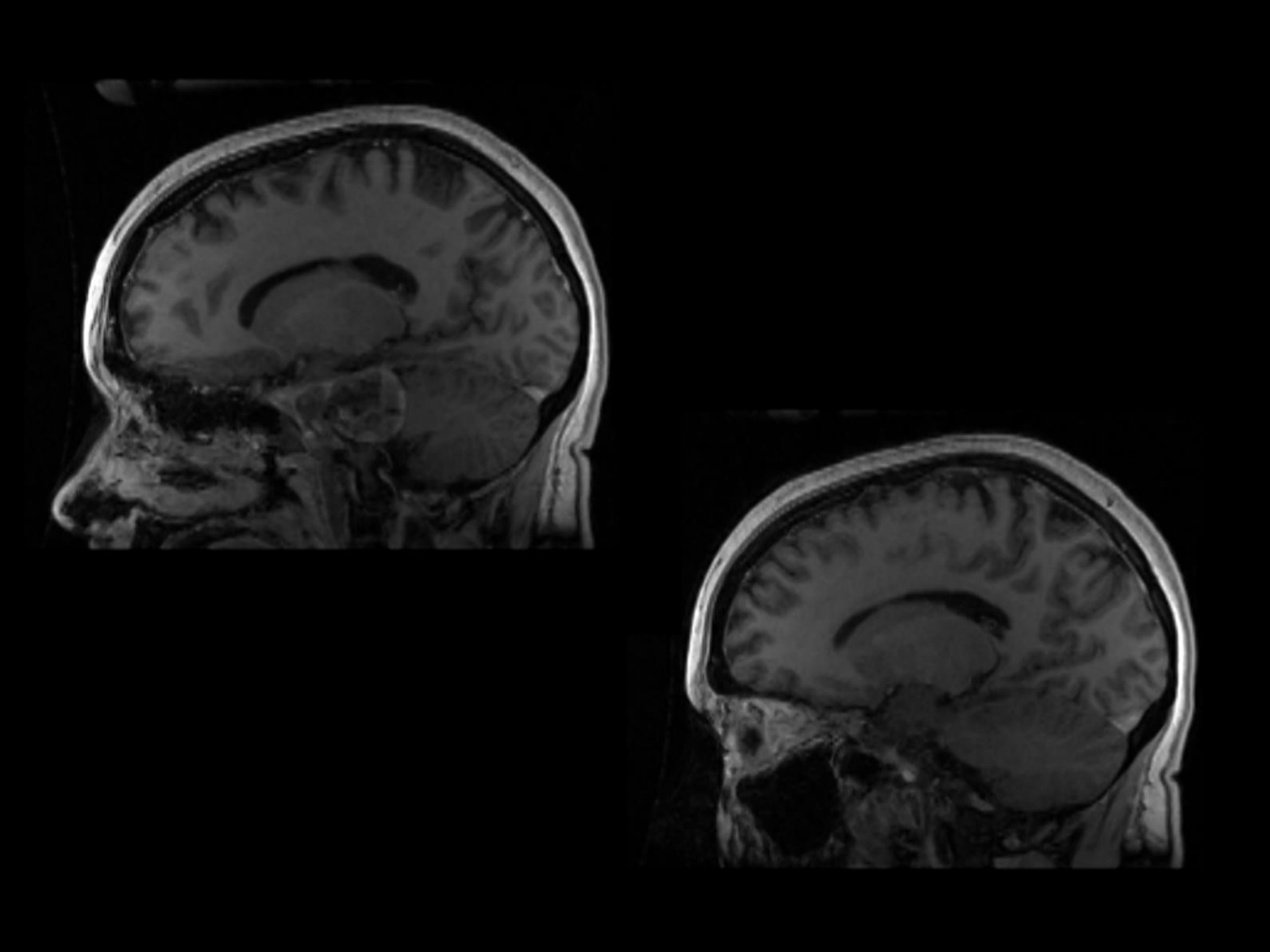
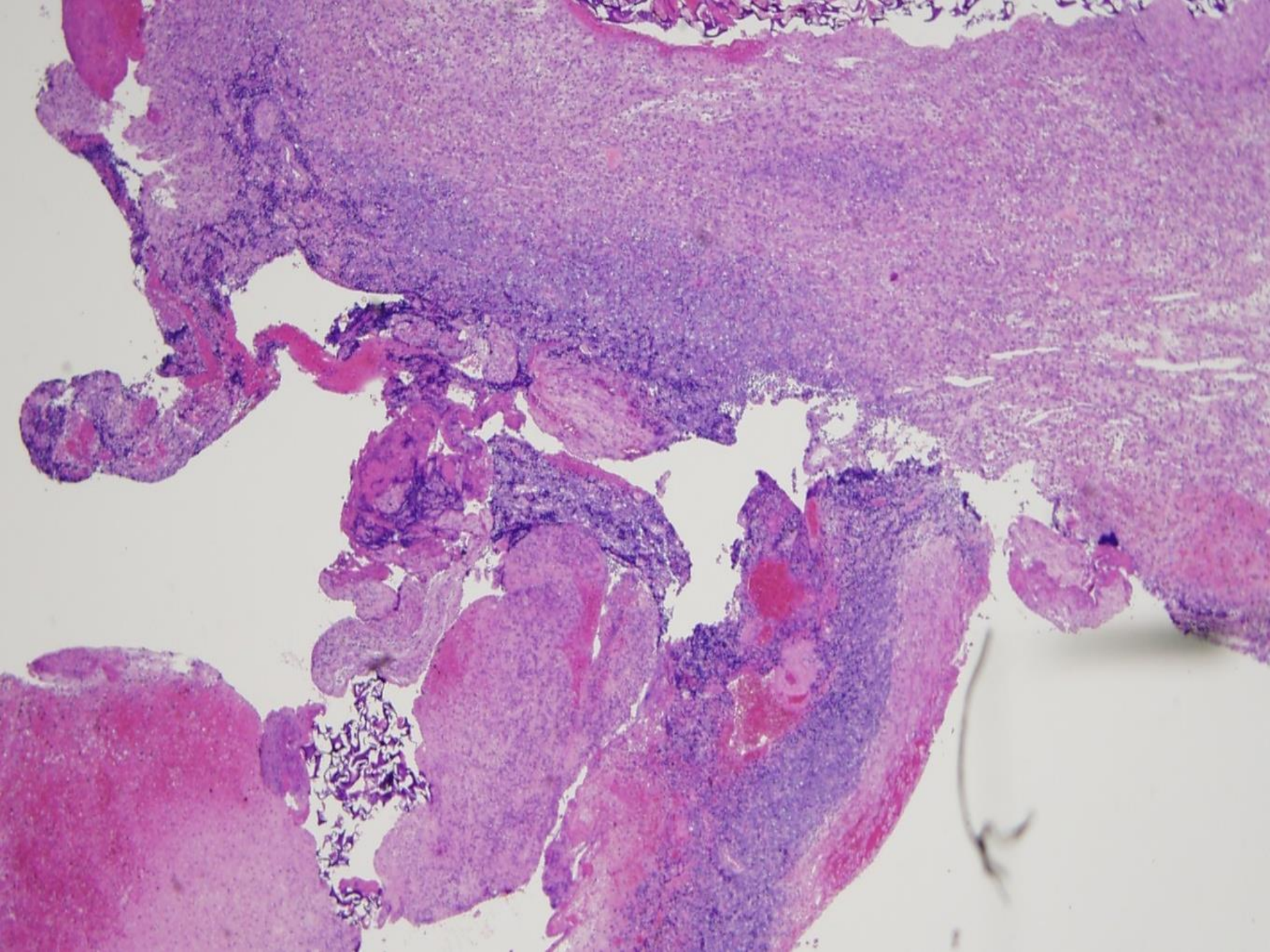
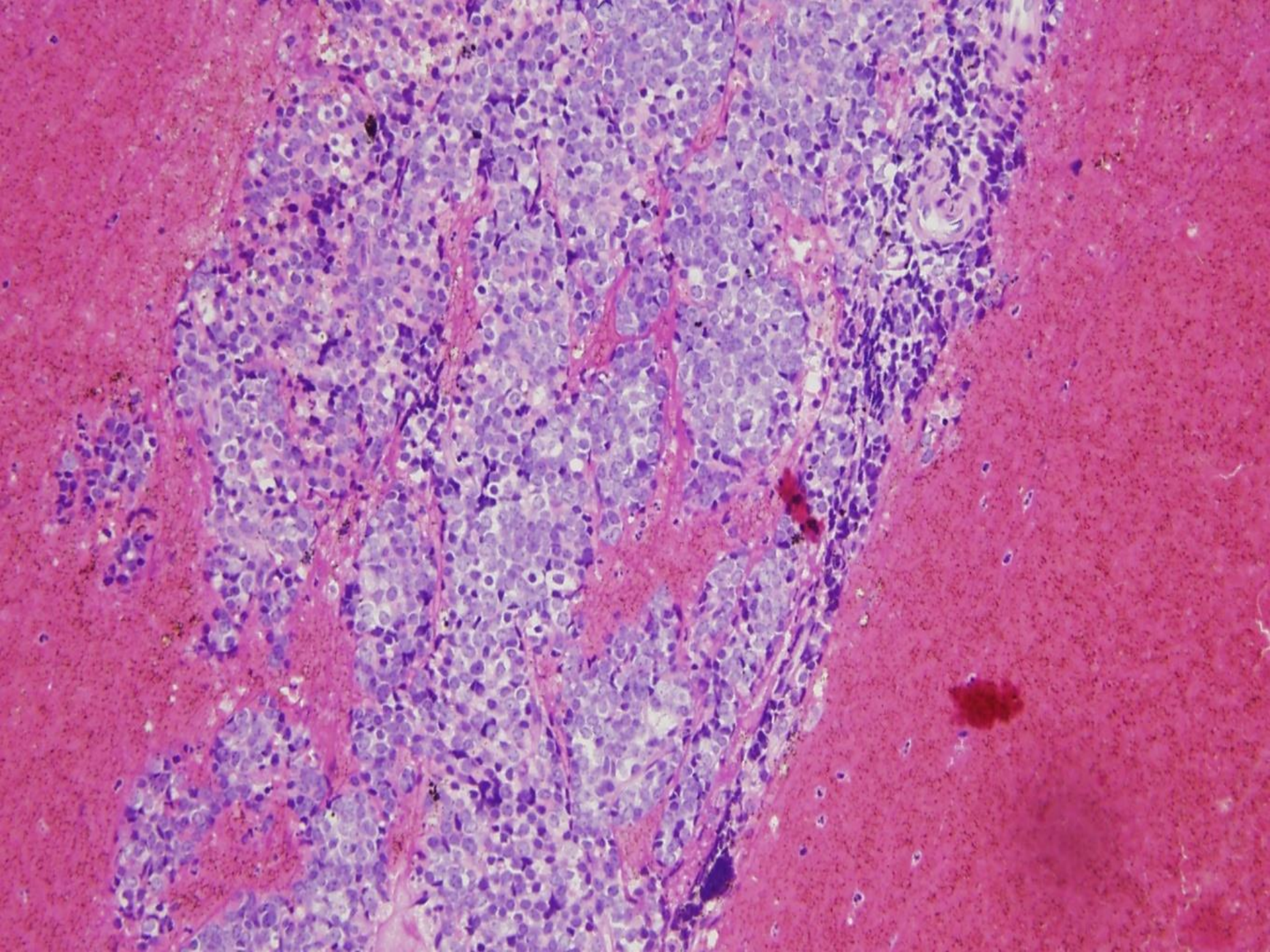
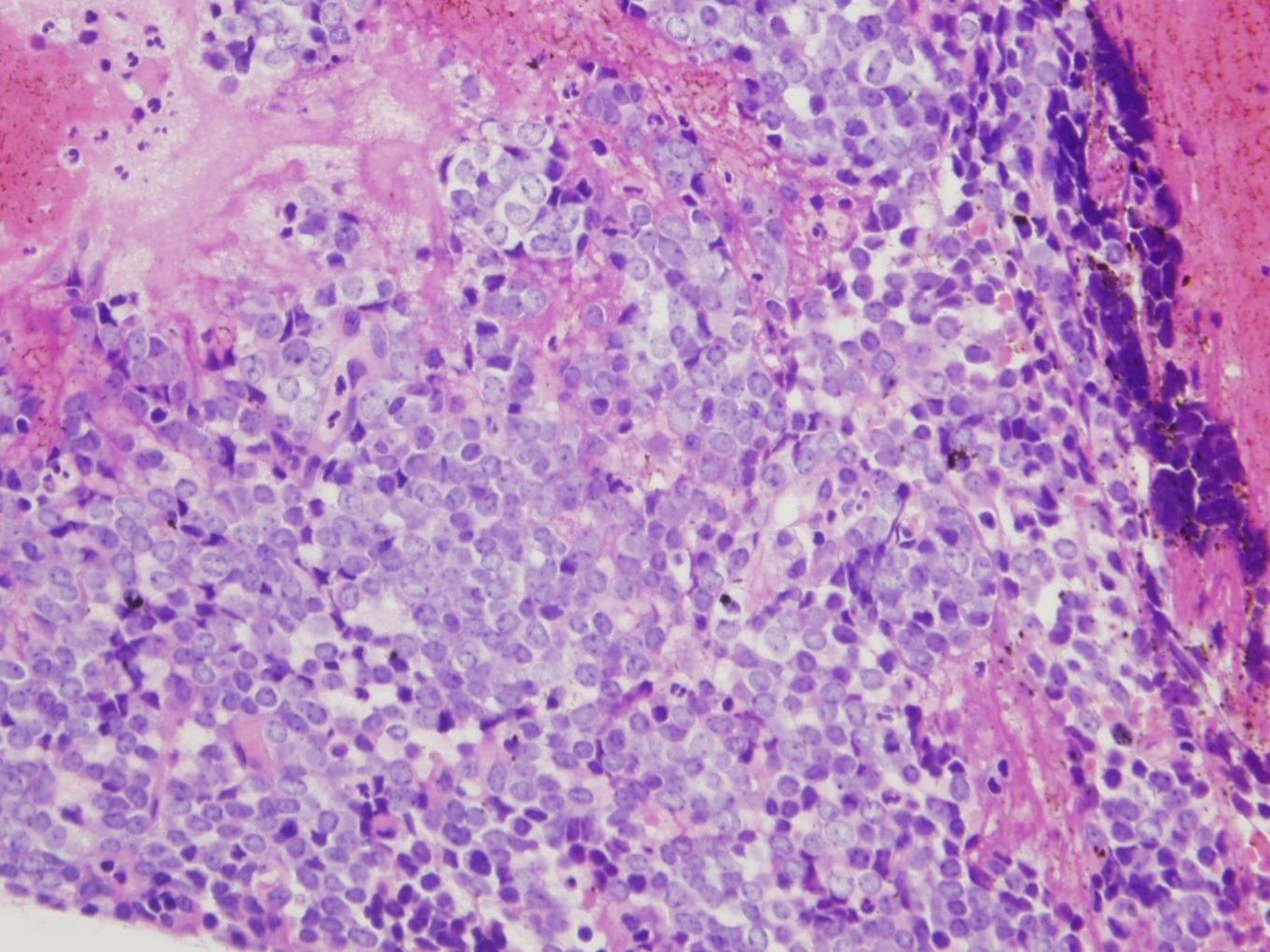


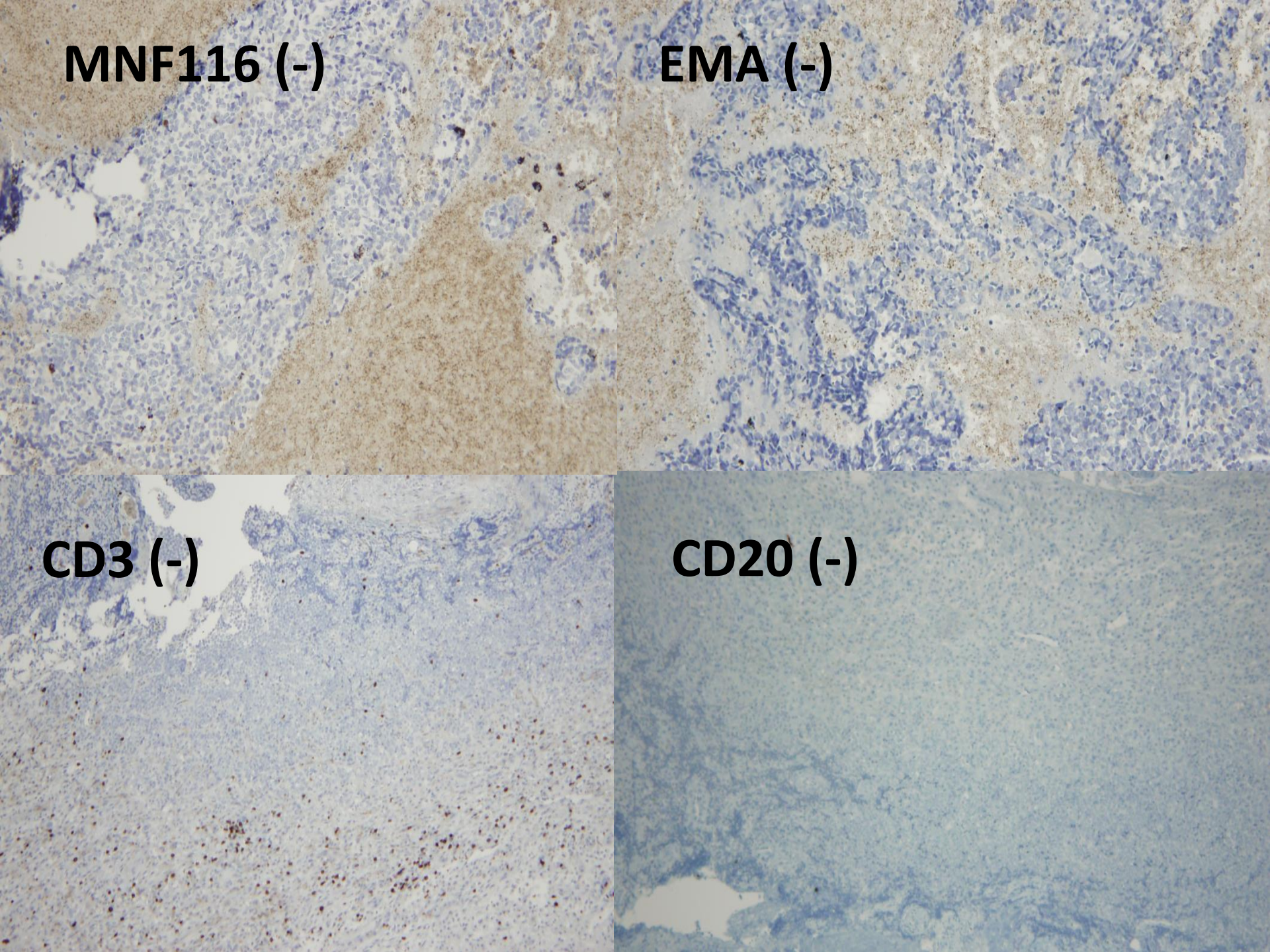
T1 MPRAGE

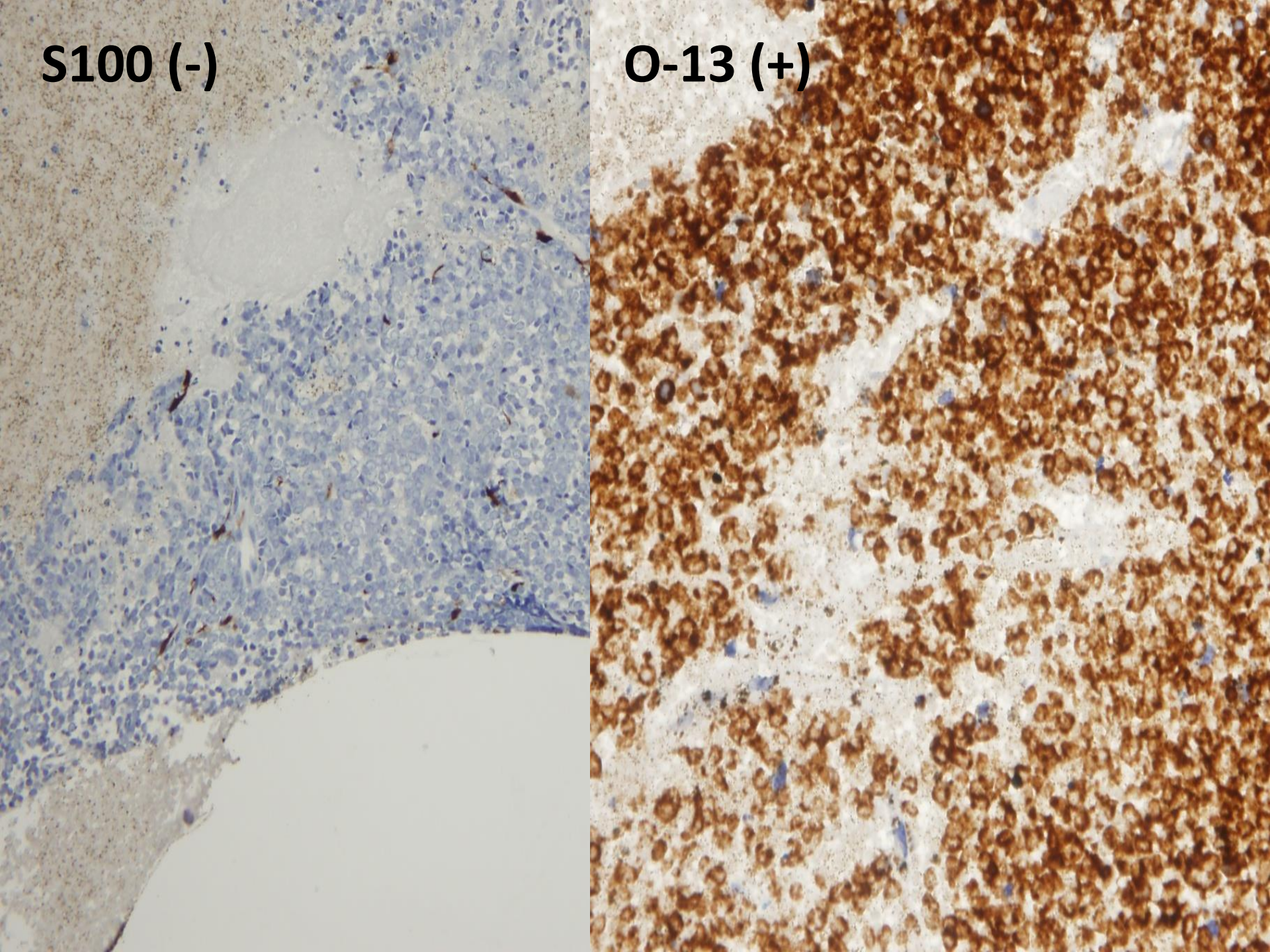









MNF116 (-)
EMA (-)
CD3 (-)
CD20 (-)


S100 (-)
O-13 (+)

RIGHT CEREBELLO-PONTINE ANGLE BIOPSY:

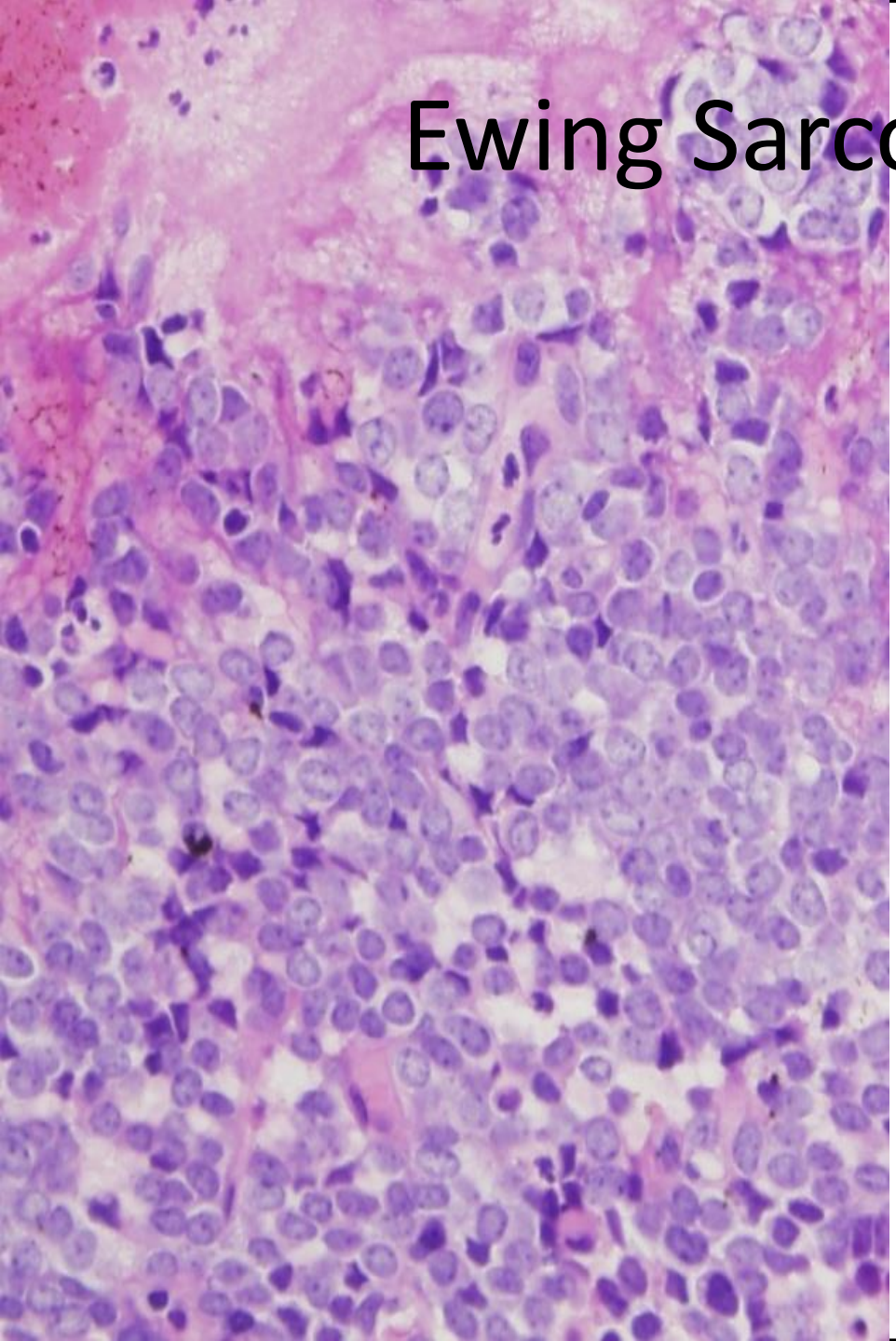
-EWING SARCOMA.

Immunohistochemistry performed at BWH demonstrates the following staining profile in lesional cells:

Positive - O13 (strong, diffuse, membranous)

Negative - CD3, CD20, S100, EMA, MNF-116

Cytogenetic analysis (CG09-4966) showed a translocation of chromosomes 11 and 22 in all metaphases analyzed. This translocation is associated with EWSR1 rearrangement and is a characteristic finding in Ewing sarcoma.



Ewing Sarcoma/PNET

- Sheets of small, round, uniform cells divided into irregular lobules by fibrous strands
- Round nuclei with small nucleoli
- Scant clear cytoplasm with indistinct cell membranes
- t(11,22)(q24;q12); *EWS-FL1*
- CD99/O-13 positive (membranous)
- S100 and EMA negative

Ewing's Sarcoma

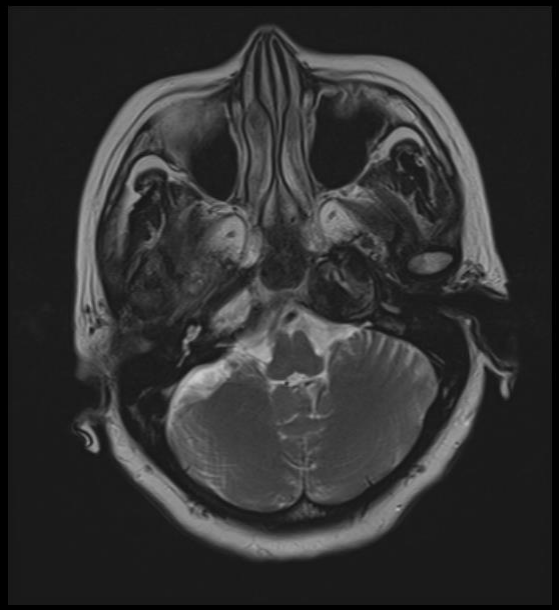
- T1 low to intermediate
- T2 heterogeneously high, maybe low hair on end
- T1 C+ heterogeneous, prominent enhancement

- Moth eaten, destructive, permeative, with large soft tissue component without osteoid matrix, also occurs in flat bones, 2% skull/face
- 95% between 4 and 25 years of age
- Small round blue cell tumor

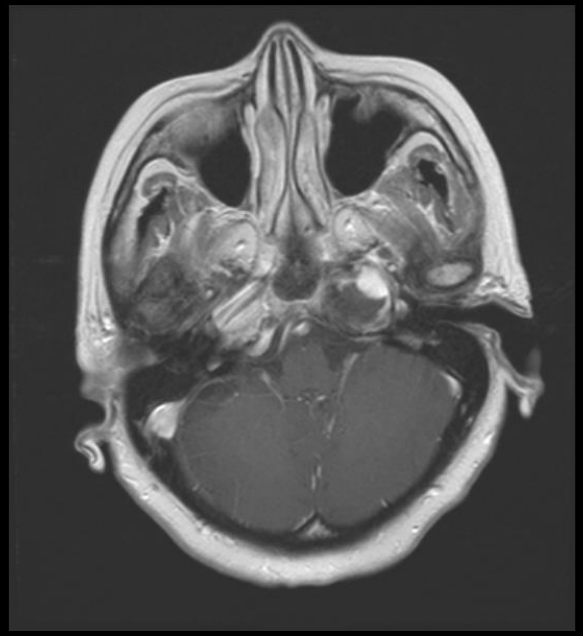
Bonus Case

71 Female

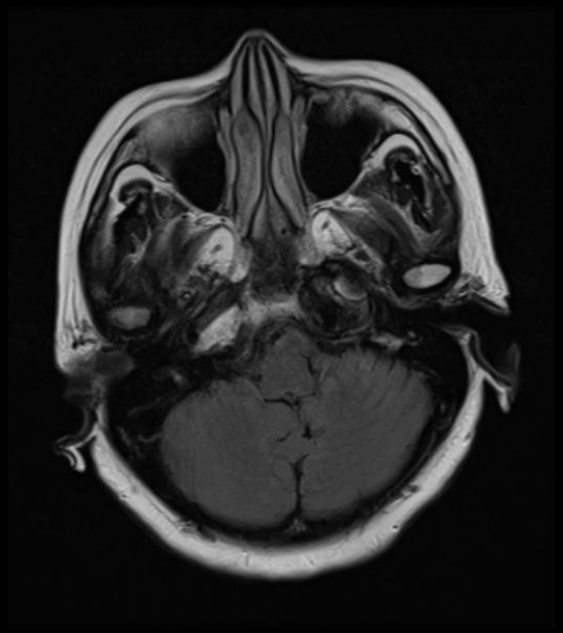
- Agitation
- Worsened Memory
- Auditory Hallucinations



T1 Pre

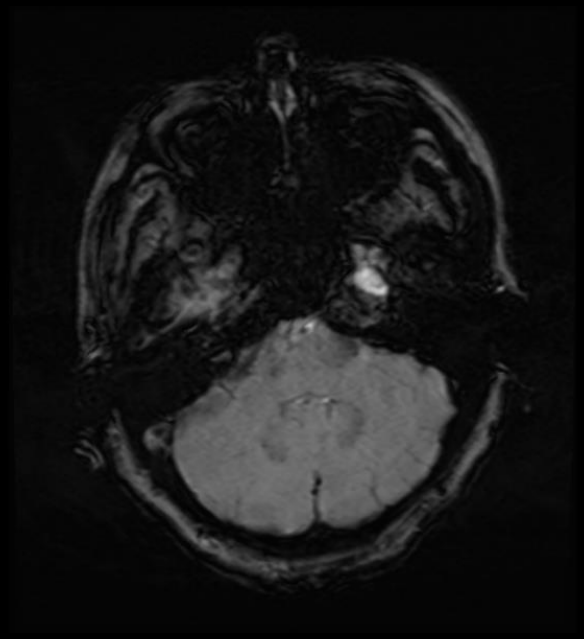


T1 Post

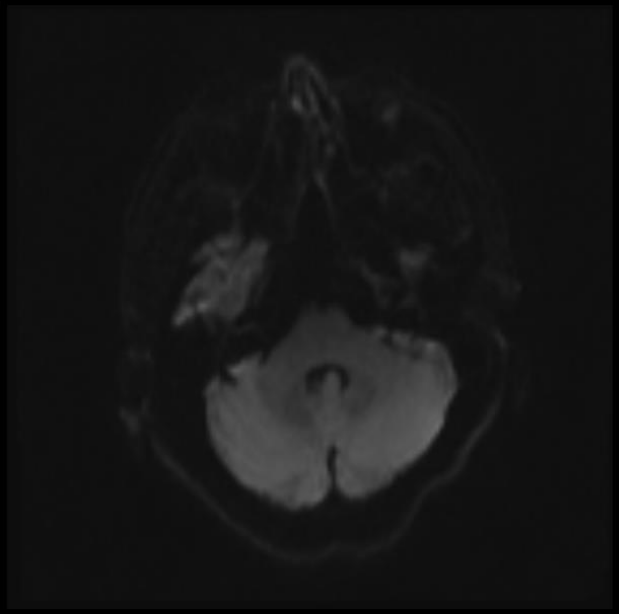


T2 FLAIR

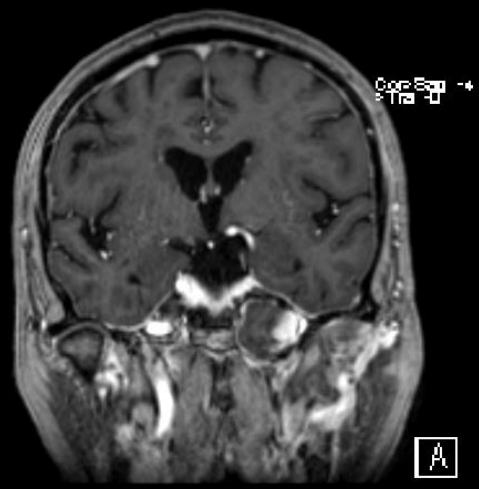
MEDCOM RESAMPLED



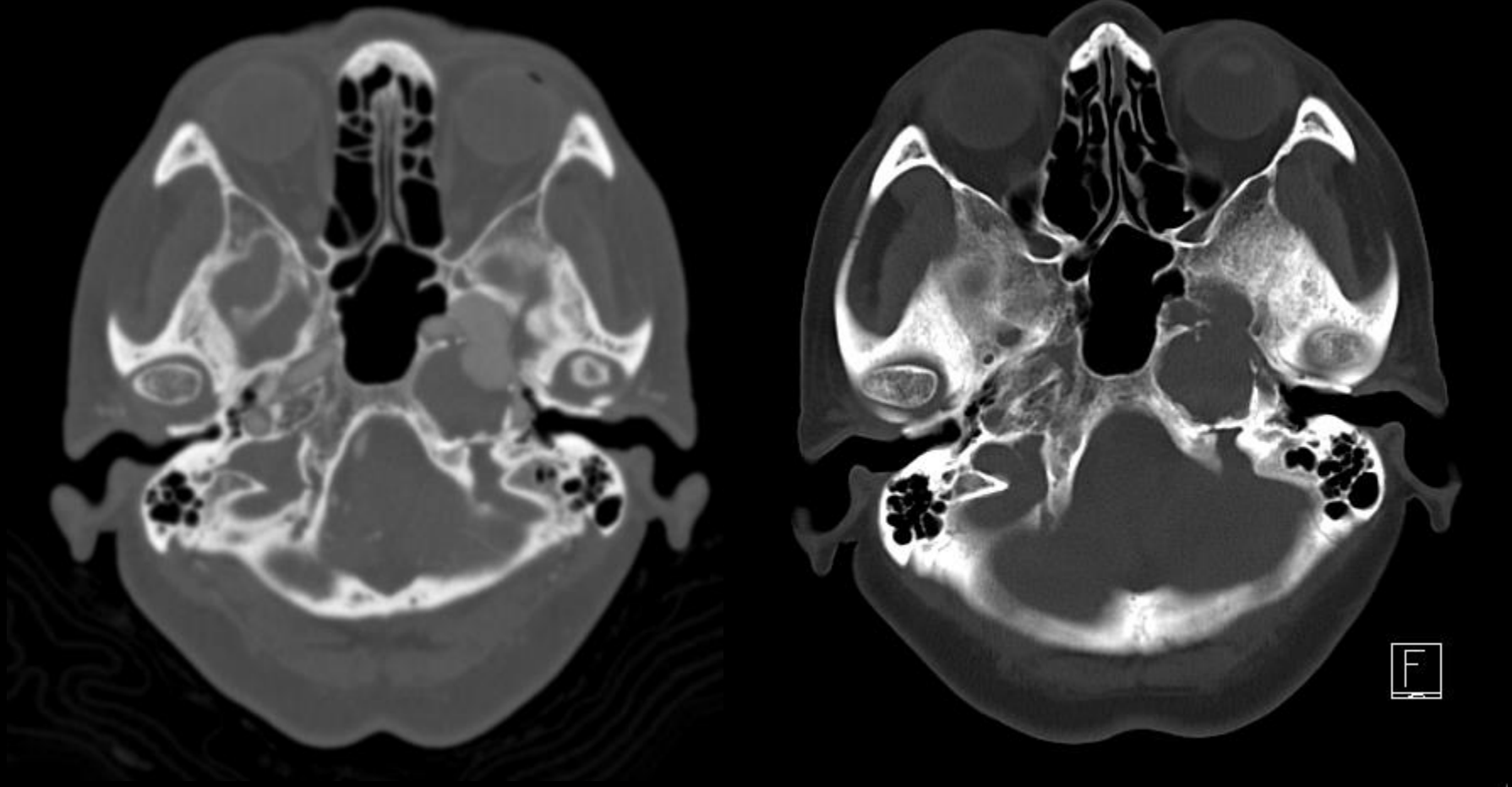
SWI



DWI



T1 MPRAGE



Petrous Carotid Aneurysm

Thank You

- Asha Sarma
- Paul Bunch
- Liangge Hsu
- Srini Mukundan
- Waihay Wong