AMSER Rad Path Case of the Month:

67yo F with difficulty ambulating, hearing loss, and intermittent facial weakness

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Patient Presentation

HPI: 67yo F with complaints of several years of progressive difficulty ambulating, weakness in lower extremities and hearing loss in her R ear along with intermittent right-sided facial weakness and hand tremor.

Previous partial workup for mass of the R cerebellopontine angle, lost to follow up in 2021

PMHx: stage III CKD, prior CVA (on Coumadin), T2DM, HTN, seizure (on Lamictal and Dilantin)

Physical Exam: Oriented to person, place, and time. Normal speech, concentration and attention. Cranial nerve function intact. Motor and sensory function intact. Coordination assessment unremarkable. Gait exam limited as patient was wheelchair bound

What imaging should we order?

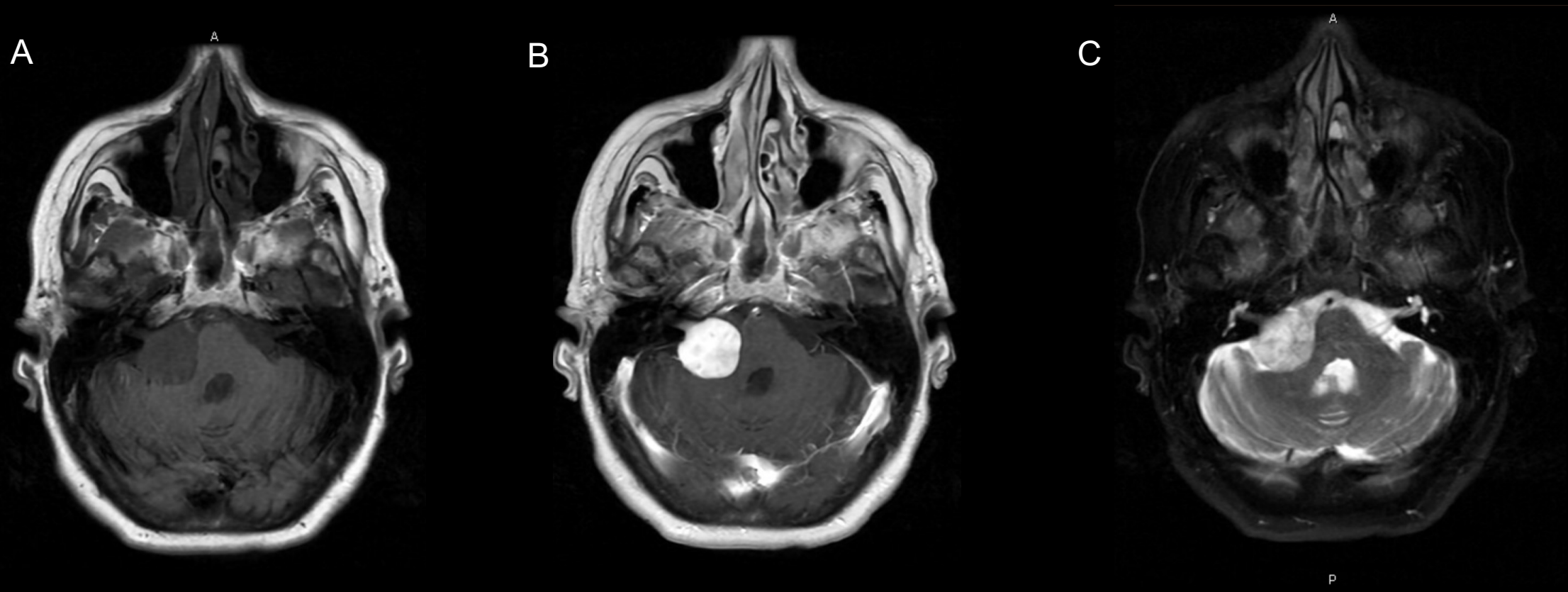
Select the applicable ACR Appropriateness criteria

Variant 6: **Adult. Chronic disequilibrium with signs of cerebellar ataxia. Initial imaging.**

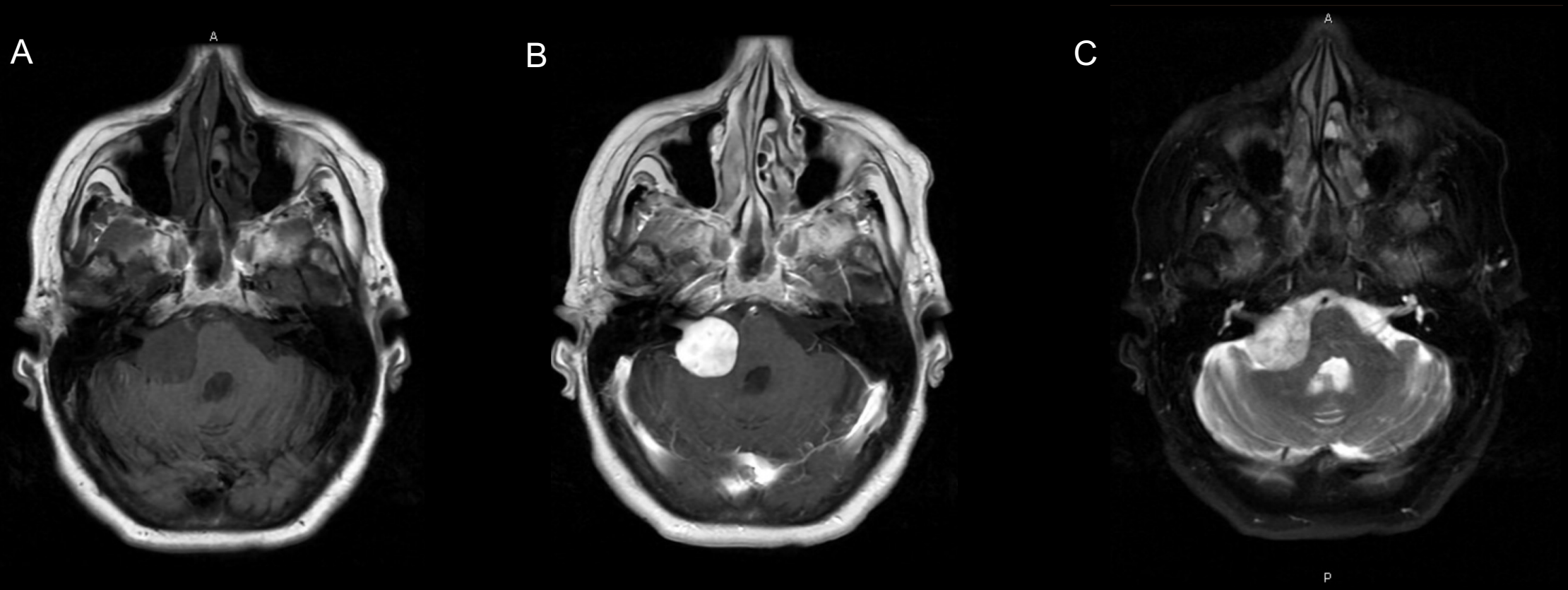
Procedure	Appropriateness Category	Relative Radiation Level
MRI head without and with IV contrast	Usually Appropriate	O
MRI head without IV contrast	Usually Appropriate	O
MRI cervical and thoracic spine without IV contrast	May Be Appropriate	O
MRA head and neck with IV contrast	Usually Not Appropriate	O
MRA head and neck without and with IV contrast	Usually Not Appropriate	O
MRA head and neck without IV contrast	Usually Not Appropriate	O
MRI cervical and thoracic spine with IV contrast	Usually Not Appropriate	O
MRI cervical and thoracic spine without and with IV contrast	Usually Not Appropriate	O
MRI head and internal auditory canal with IV contrast	Usually Not Appropriate	O
MRI head and internal auditory canal without and with IV contrast	Usually Not Appropriate	O
MRI head and internal auditory canal without IV contrast	Usually Not Appropriate	O
MRI head with IV contrast	Usually Not Appropriate	O
CT head with IV contrast	Usually Not Appropriate	☢☢☢
CT head without and with IV contrast	Usually Not Appropriate	☢☢☢
CT head without IV contrast	Usually Not Appropriate	☢☢☢
CT temporal bone with IV contrast	Usually Not Appropriate	☢☢☢
CT temporal bone without and with IV contrast	Usually Not Appropriate	☢☢☢
CT temporal bone without IV contrast	Usually Not Appropriate	☢☢☢
CTA head and neck with IV contrast	Usually Not Appropriate	☢☢☢

This imaging modality was ordered

MRI Brain With and Without Contrast (unlabeled)



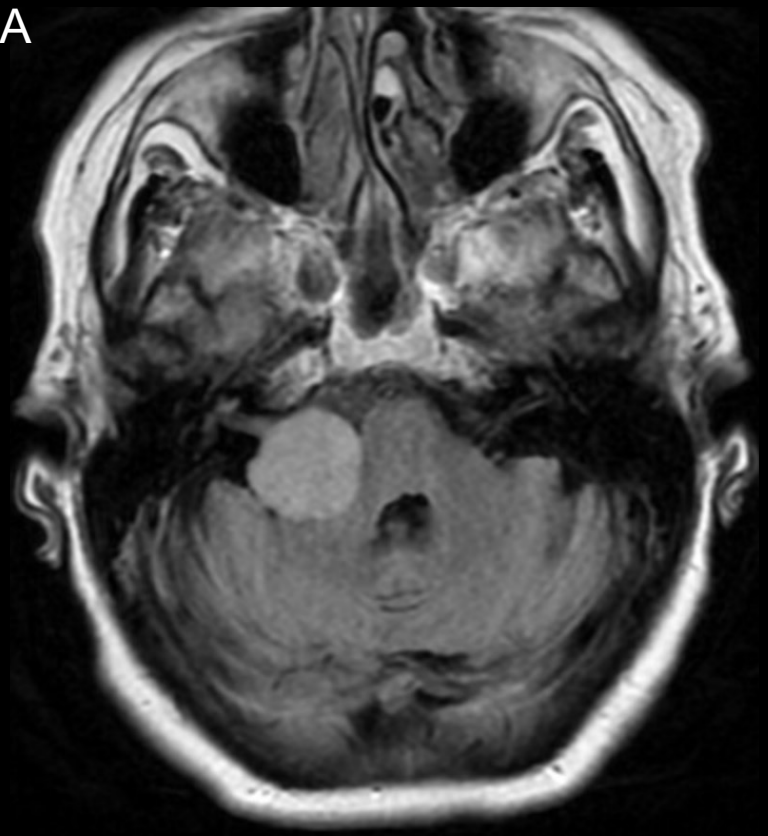
MRI Brain With and Without Contrast (labeled)



(A) Axial T1-weighted spin-echo pre-contrast and (B) post-contrast MR images localized a 2.3cm enhancing lesion at the right cerebellopontine angle. Lesion was heterogeneously hyperintense on (C) Axial T2-weighted fat-suppressed MR imaging.

MRI Brain With and Without Contrast (unlabeled)

A



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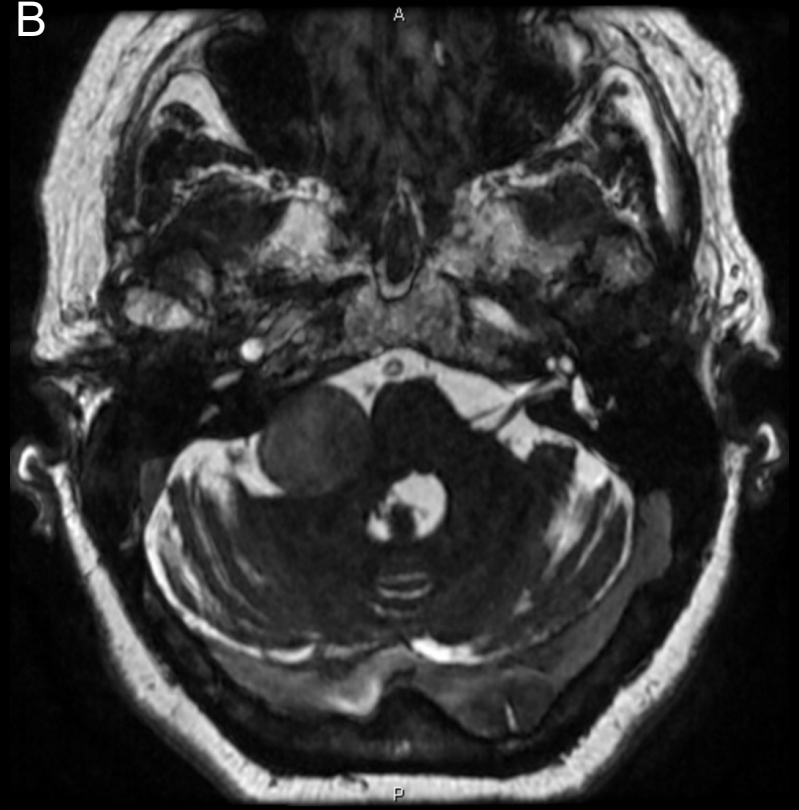
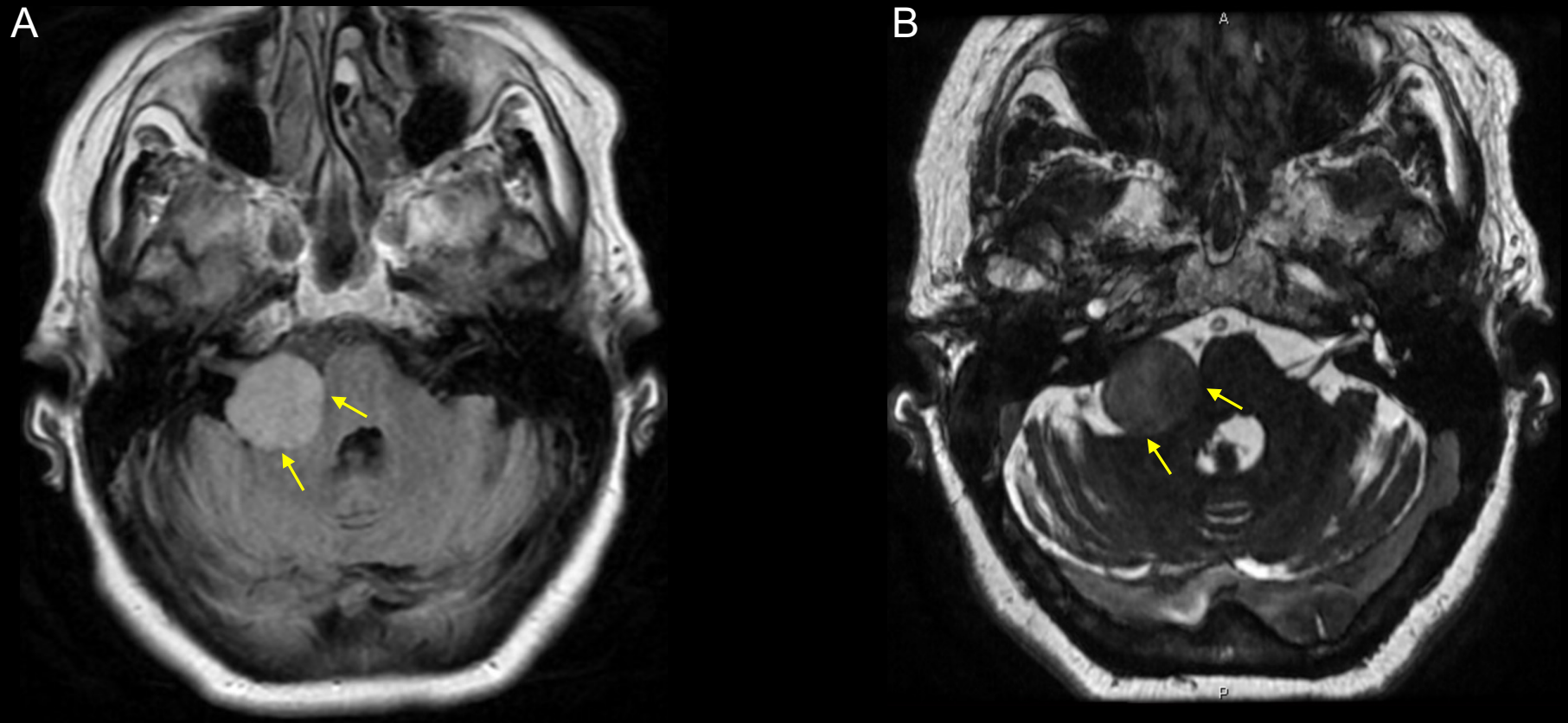
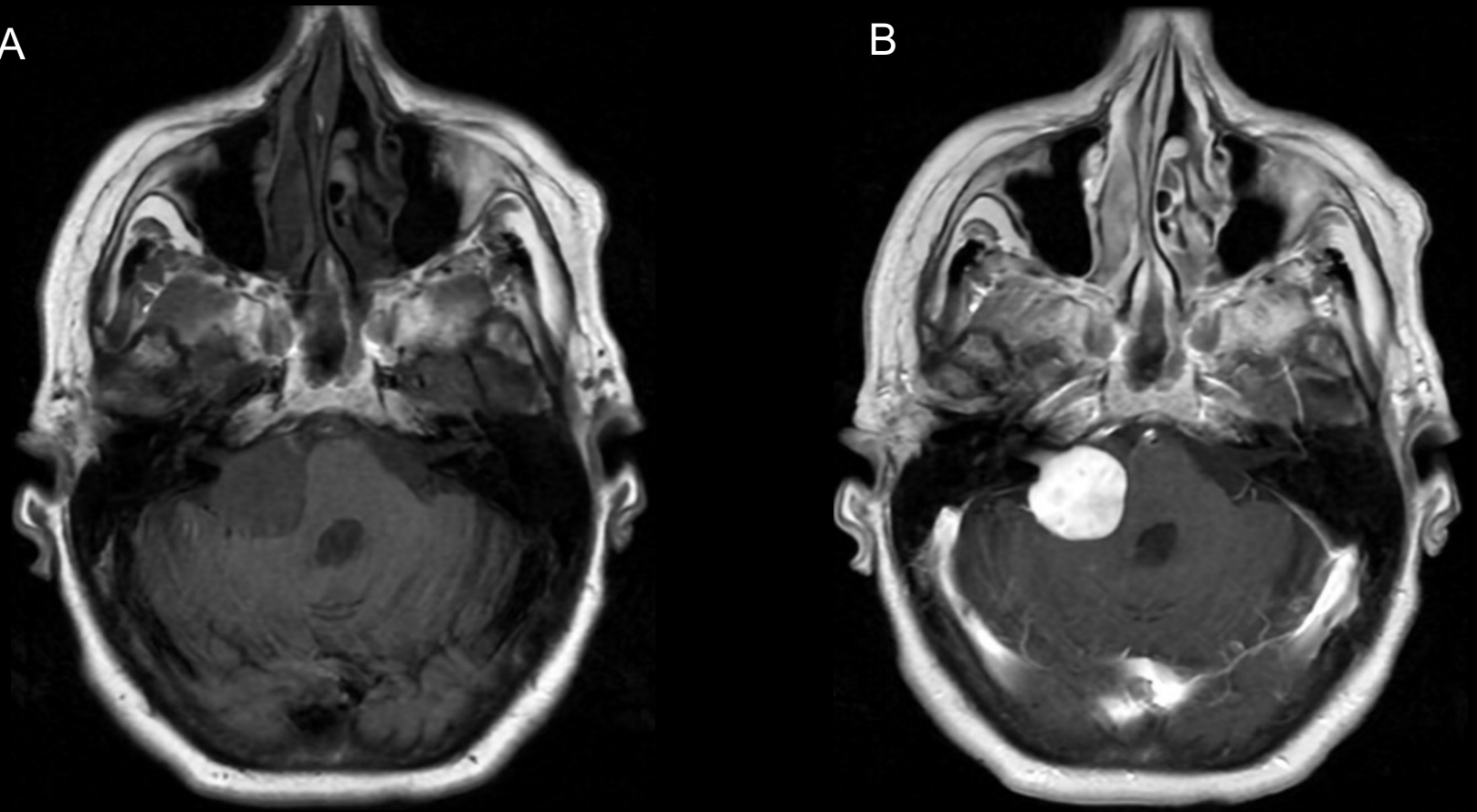


MRI Brain With and Without Contrast (unlabeled)

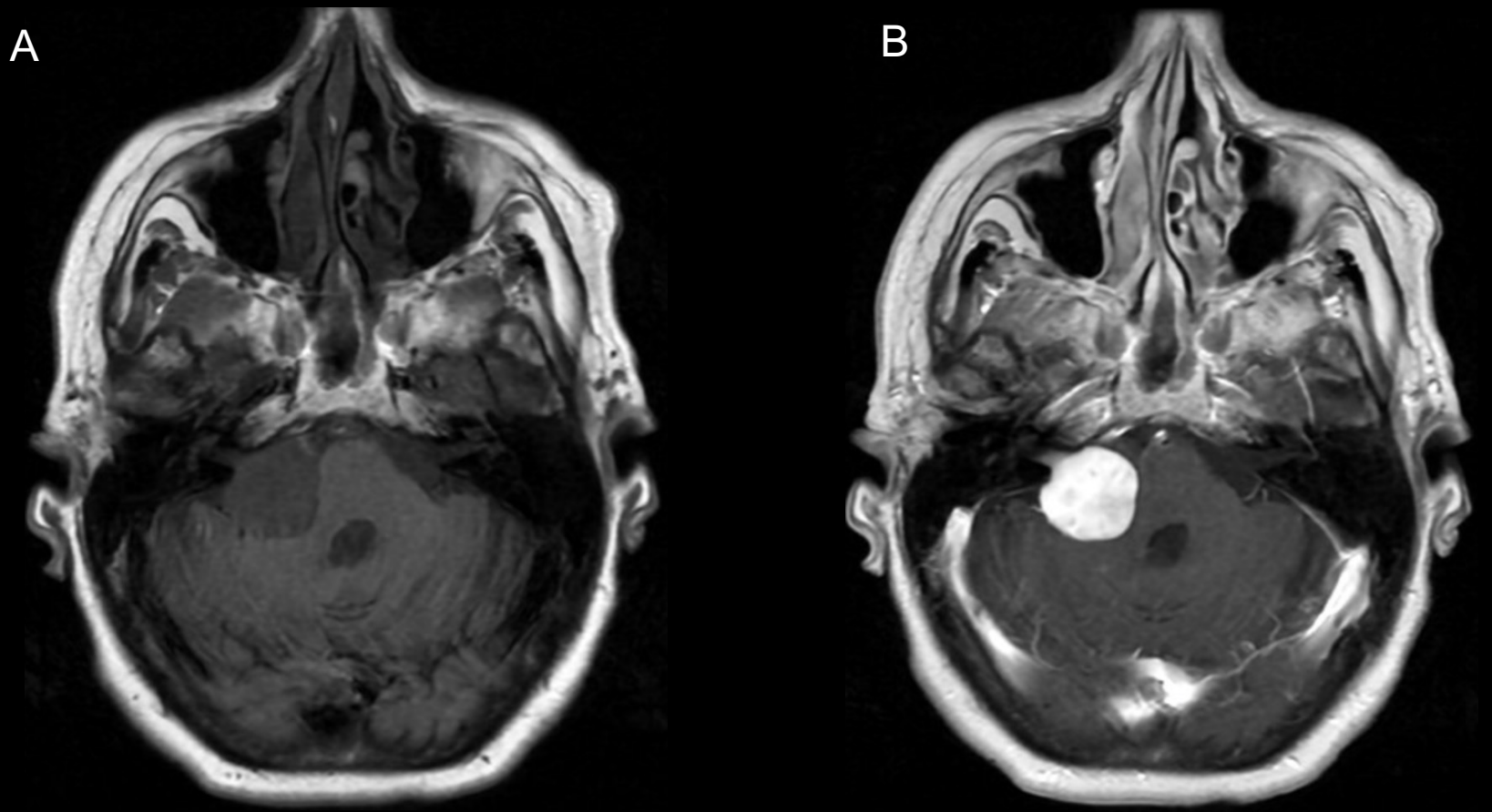


(A) Axial FLAIR and (B) Axial 3D FIESTA-C MR images characterized lesion as extra-axial (arrows)

MRI Brain With and Without Contrast (unlabeled)



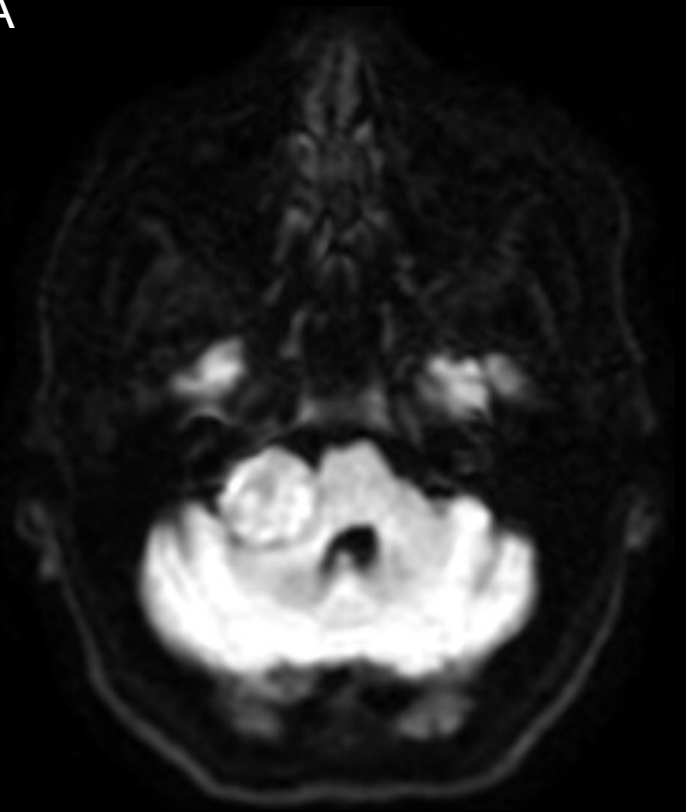
MRI Brain With and Without Contrast (labeled)



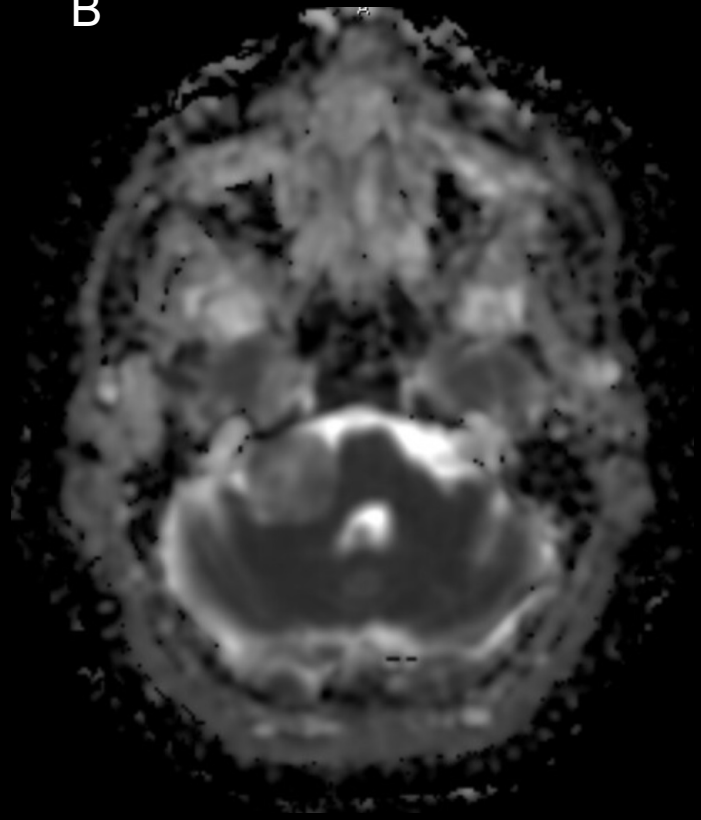
(A) Axial T1-weighted spin-echo pre-contrast and (B) post-contrast MR images demonstrated a lesion with intense heterogenous enhancement

MRI Brain With and Without Contrast (unlabeled)

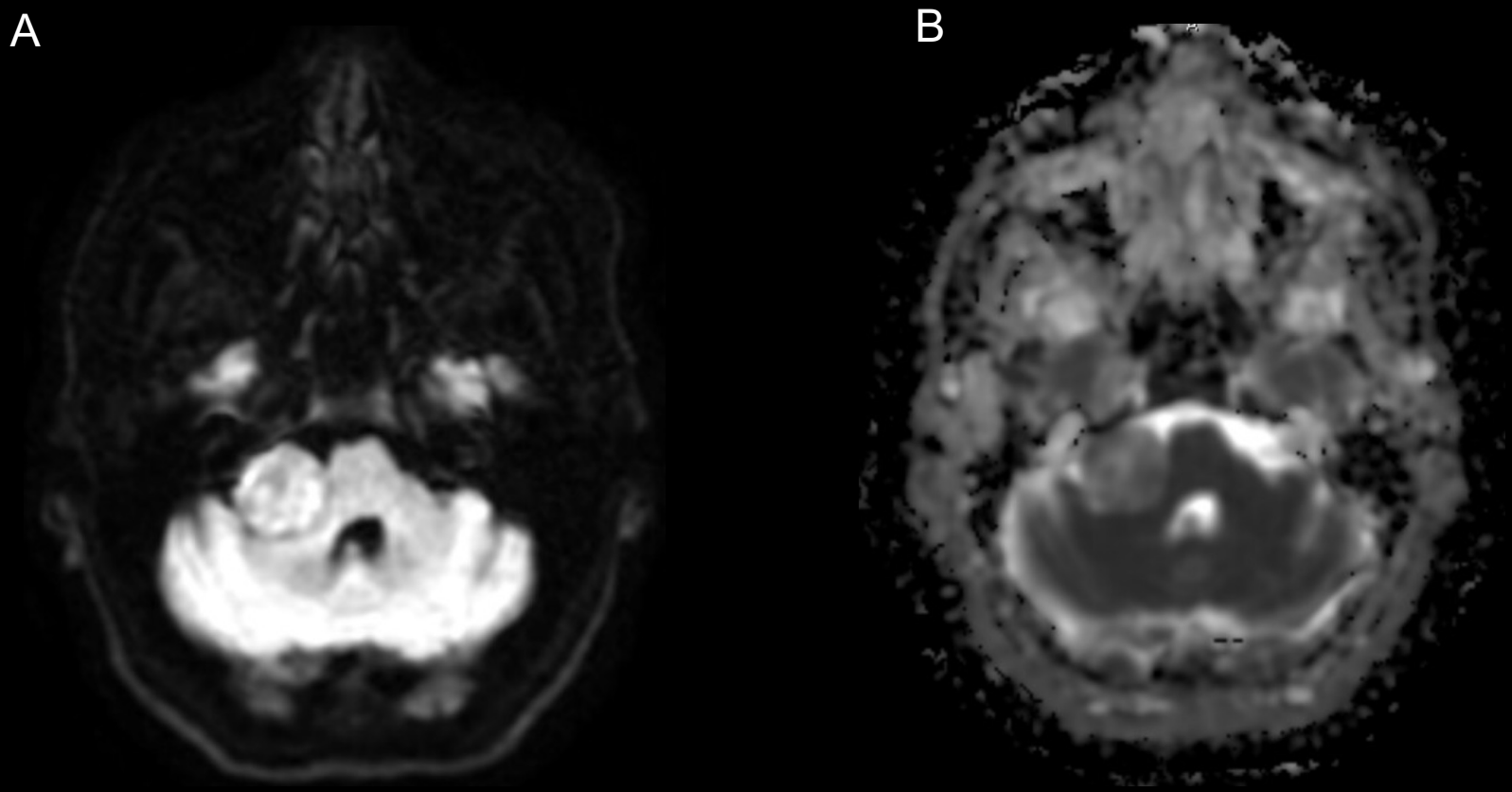
A



B



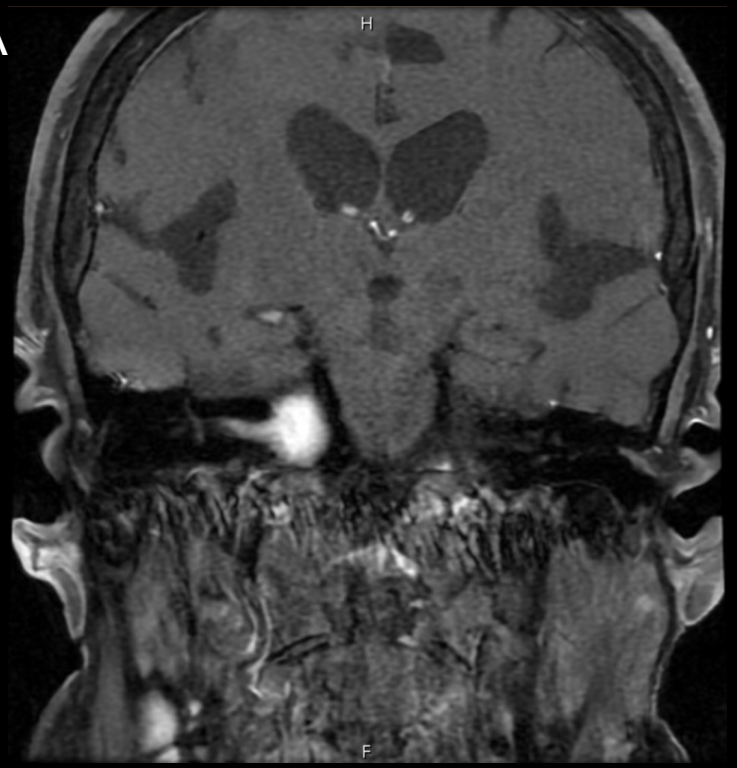
MRI Brain With and Without Contrast (labeled)



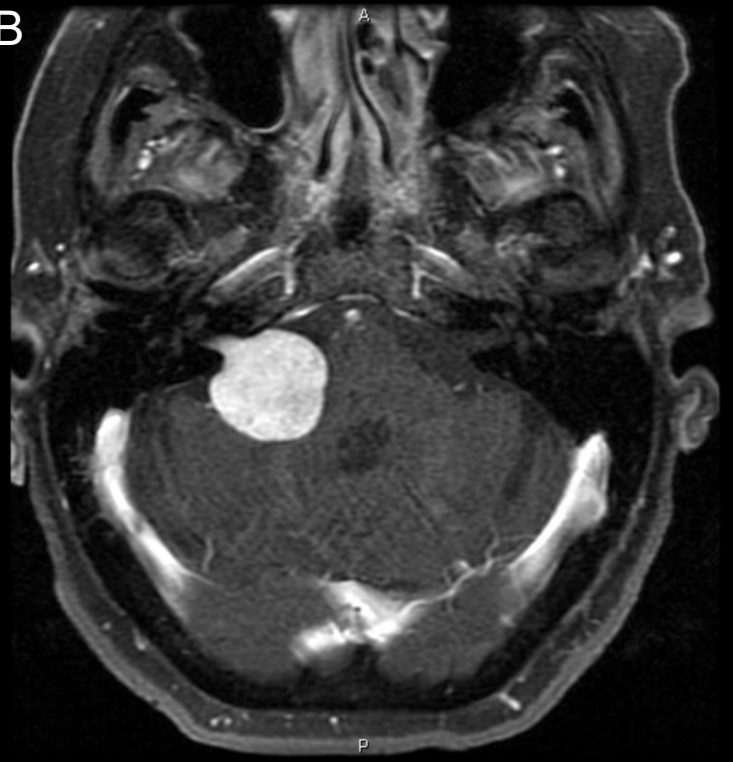
(A) Axial diffusion-weighted MR image and (B) Axial ADC map demonstrated lesion with some subtle diffusion restriction

MRI Brain With and Without Contrast (unlabeled)

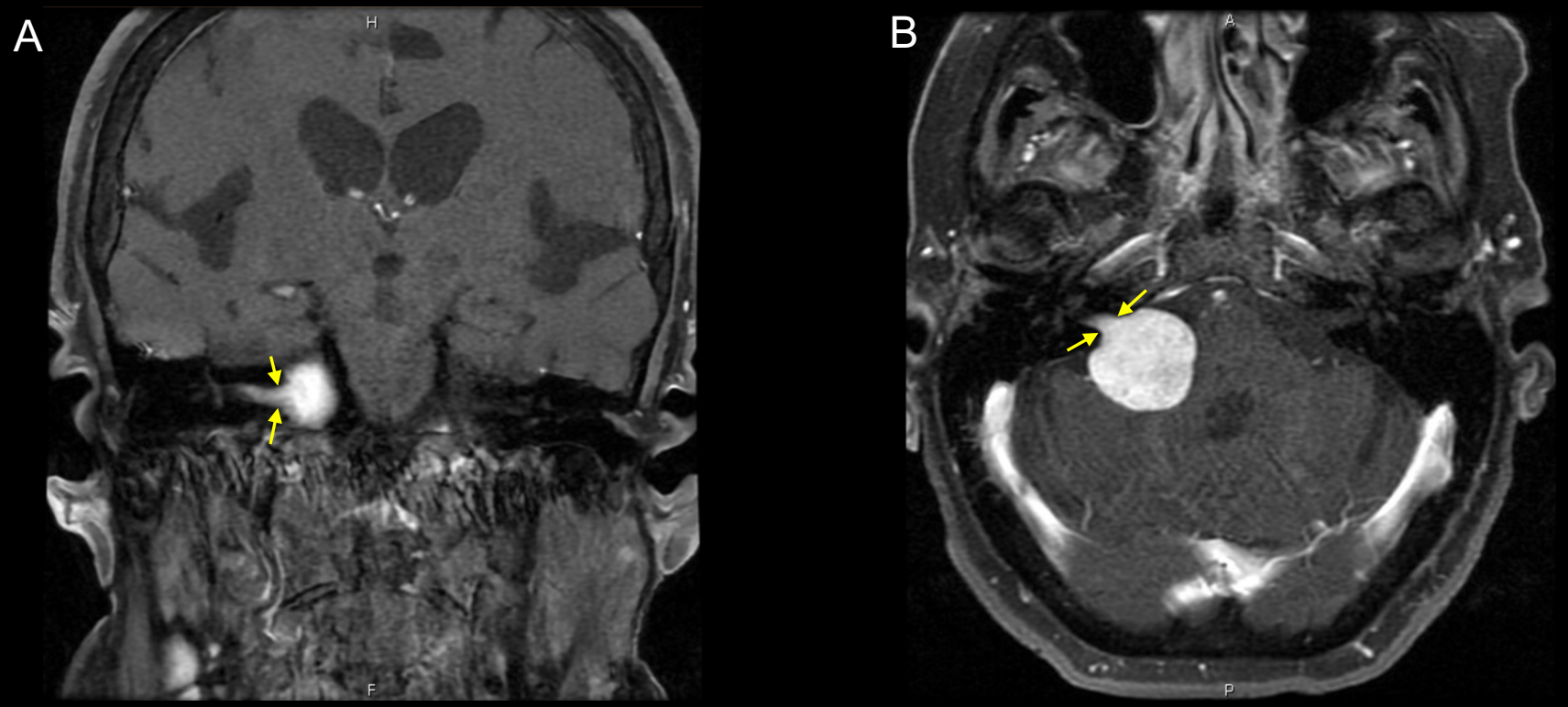
A



B



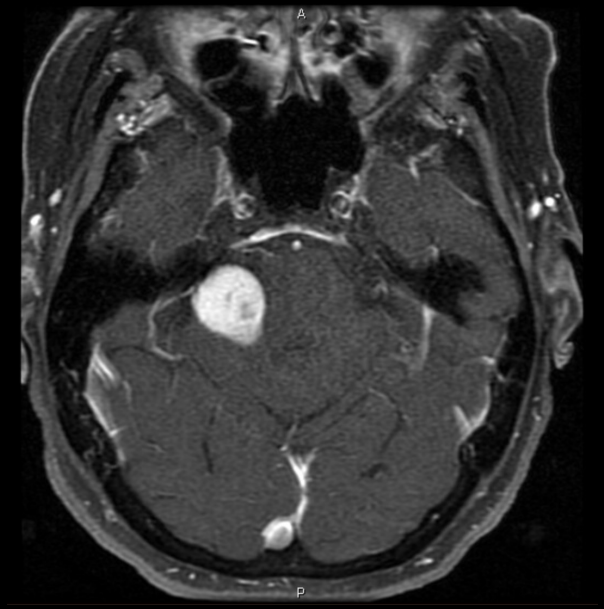
MRI Brain With and Without Contrast (labeled)



(A) Coronal T1-weighted fat-saturated post-contrast and (B) Axial T1-weight fat-saturated post-contrast MR images demonstrated extension of the lesion into the internal acoustic canal and widening of the porus acusticus (arrows)

MRI Brain With and Without Contrast (unlabeled)

A



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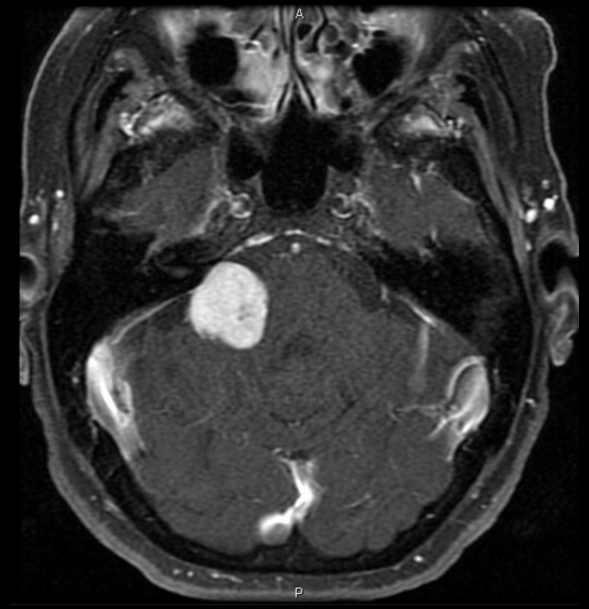
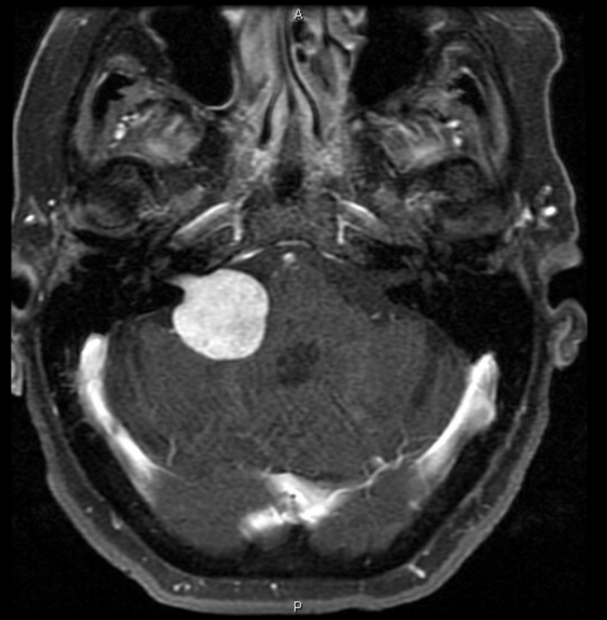
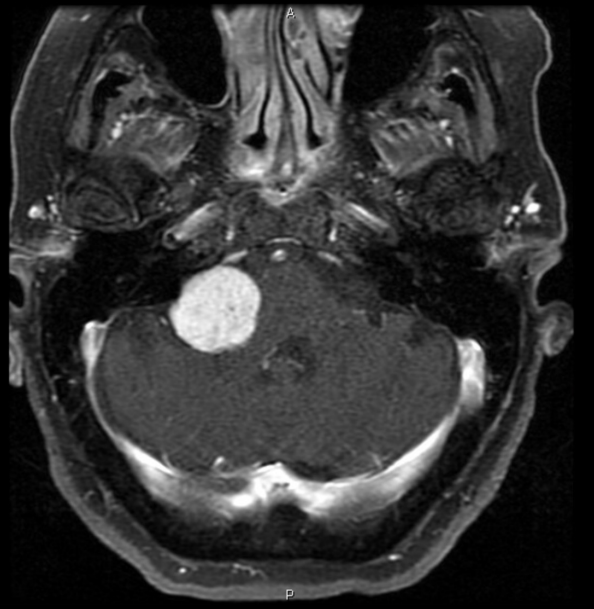


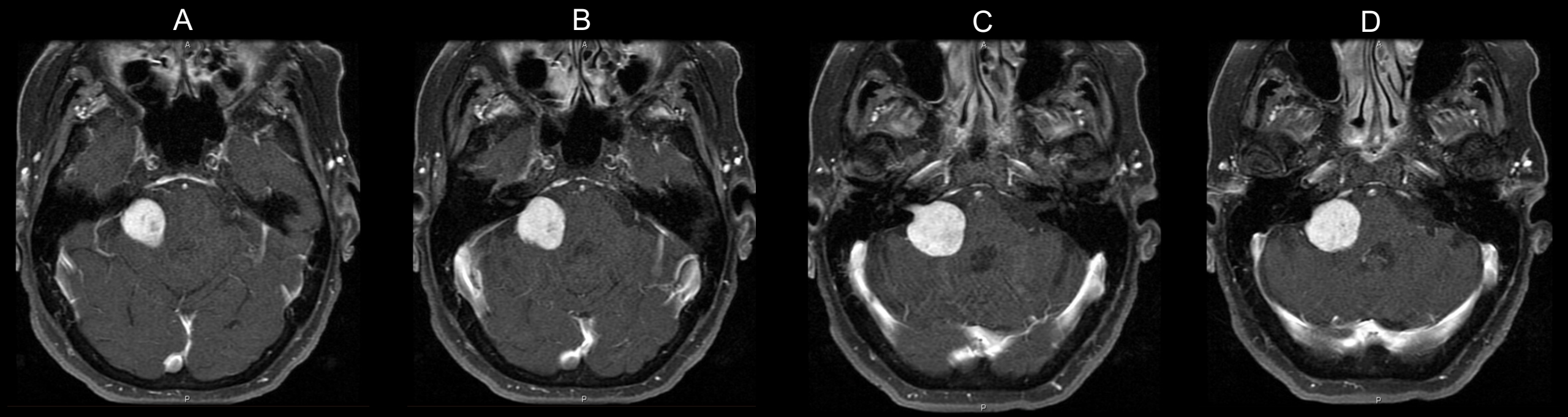
C



D



MRI Brain With and Without Contrast (labeled)



Axial T1-weighted fat-saturated post-contrast MR images from cranial to caudal (A-D) demonstrated no evidence of an enhancing dural tail

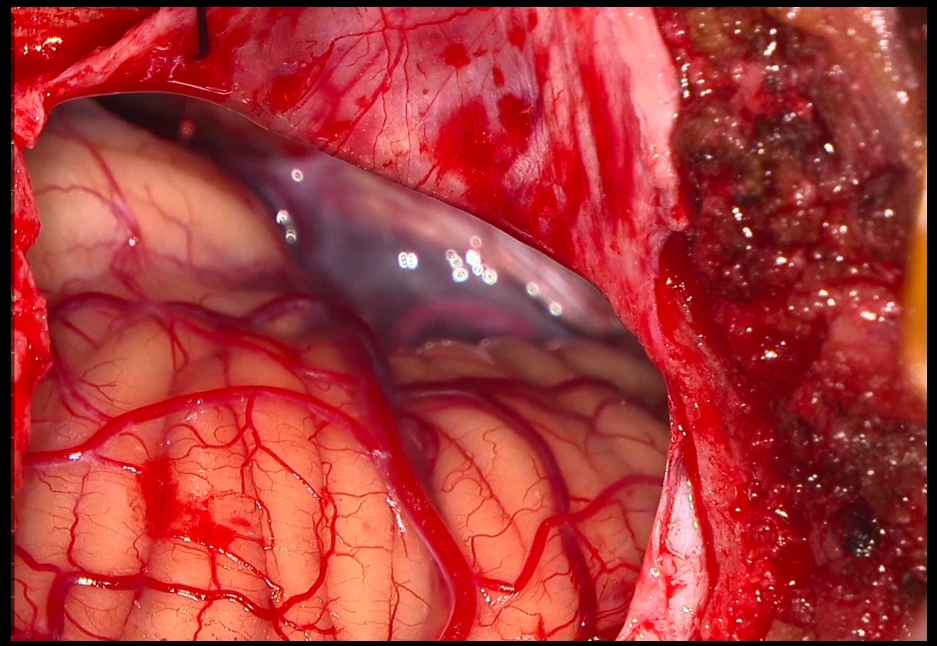
Differential Diagnosis (based on imaging)

- Vestibular Schwannoma
- Meningioma
- Ependymoma

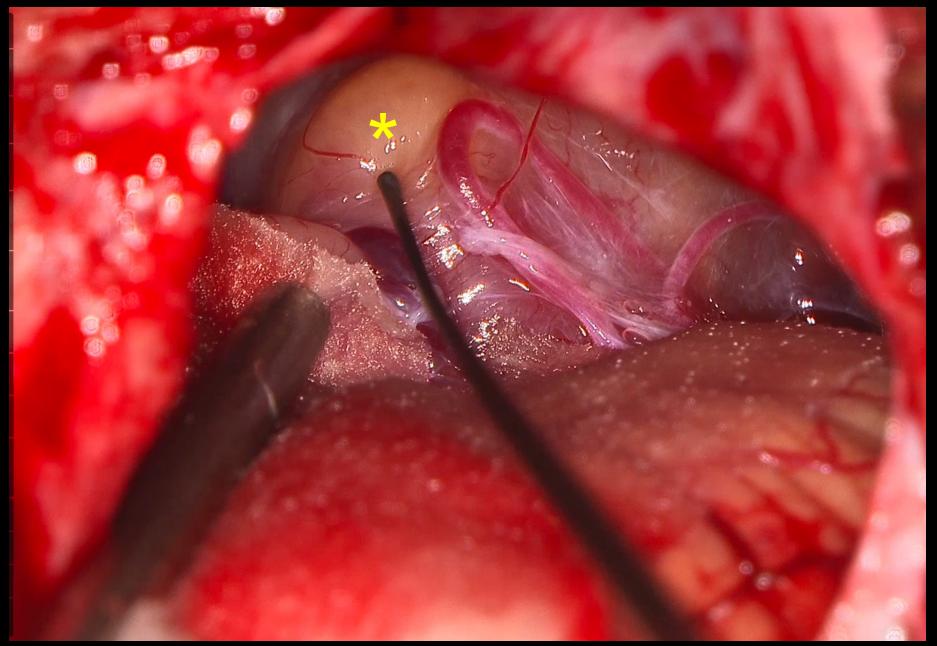
Case Progression

Given progressive symptoms and differential diagnosis patient elected to undergo retrosigmoid craniotomy for tumor debulking

Gross Path (labeled)



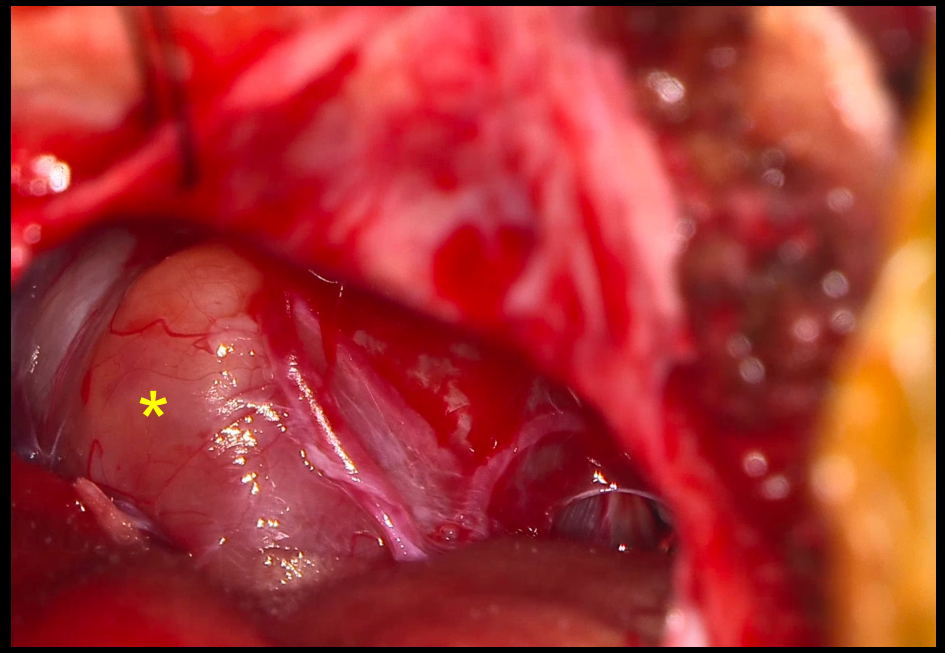
View of right lateral cerebellar hemisphere with the mass located deep behind connective tissue



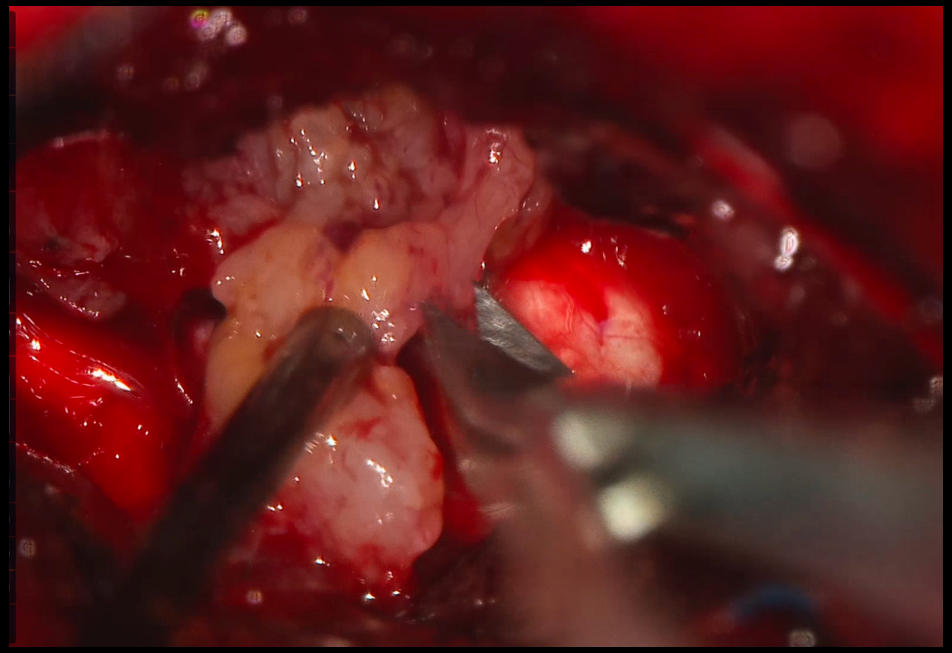
Partial view of the mass(*) behind vasculature after displacement of the right lateral cerebellar hemisphere

Images provided via Synpative Modus X robotic exoscope

Gross Path (labeled)



Partial view of mass(*) after dissection of connective tissue in the direct field of view



Final dissections of the mass demonstrate its internal color and ‘fat-like’ consistency

Images provided via Synpative Modus X robotic exoscope

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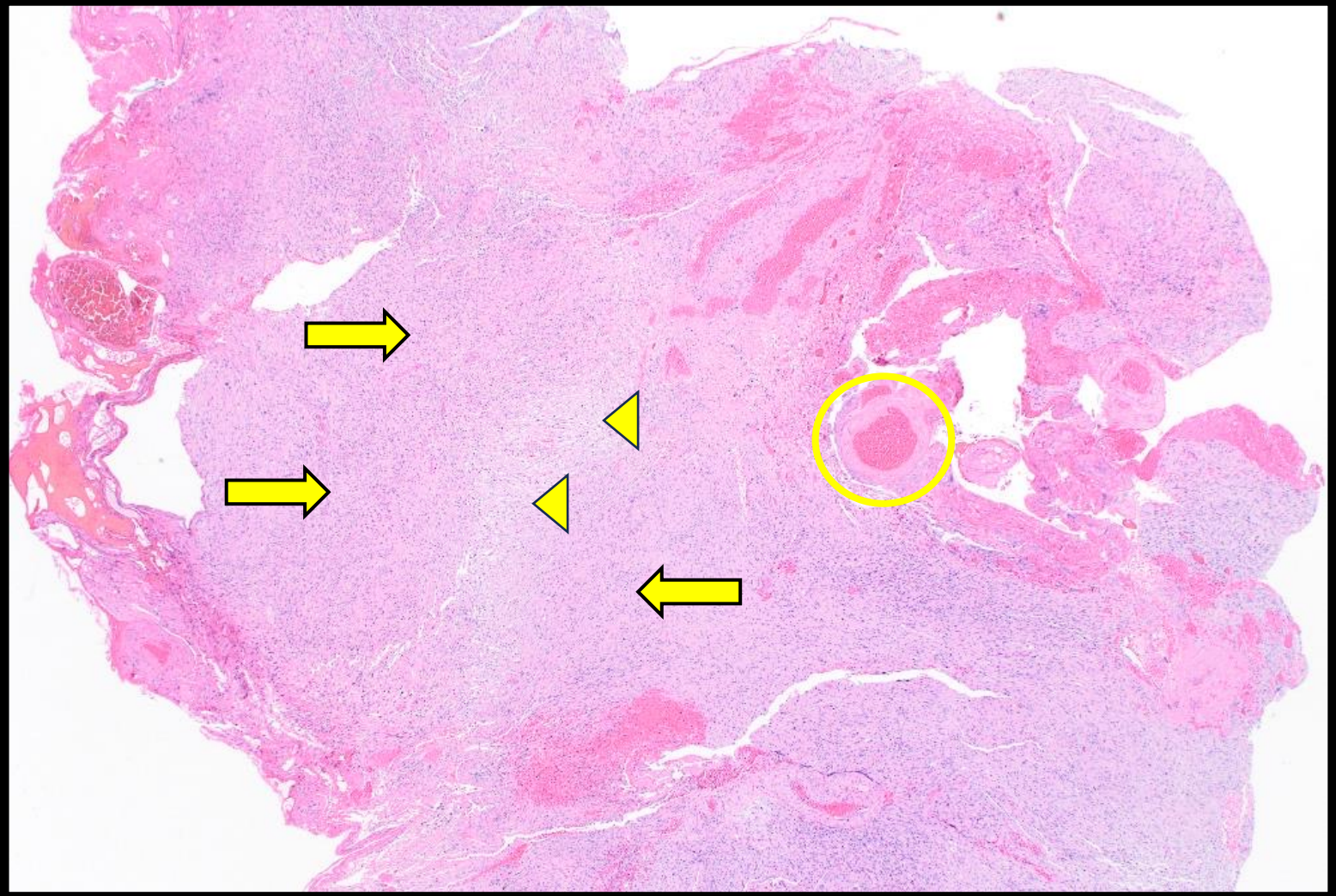
Pathology (labeled)



H&E (20x) section demonstrating biphasic morphology with compact hypercellular areas (Antoni A, arrows) and loose microcystic areas (Antoni B, arrowhead) and thick walled hyalinized blood vessels (circle).

Pathology (labeled)

H&E section at 40x (image 1) and 100x (image 2) demonstrating nuclear palisading, Verocay bodies

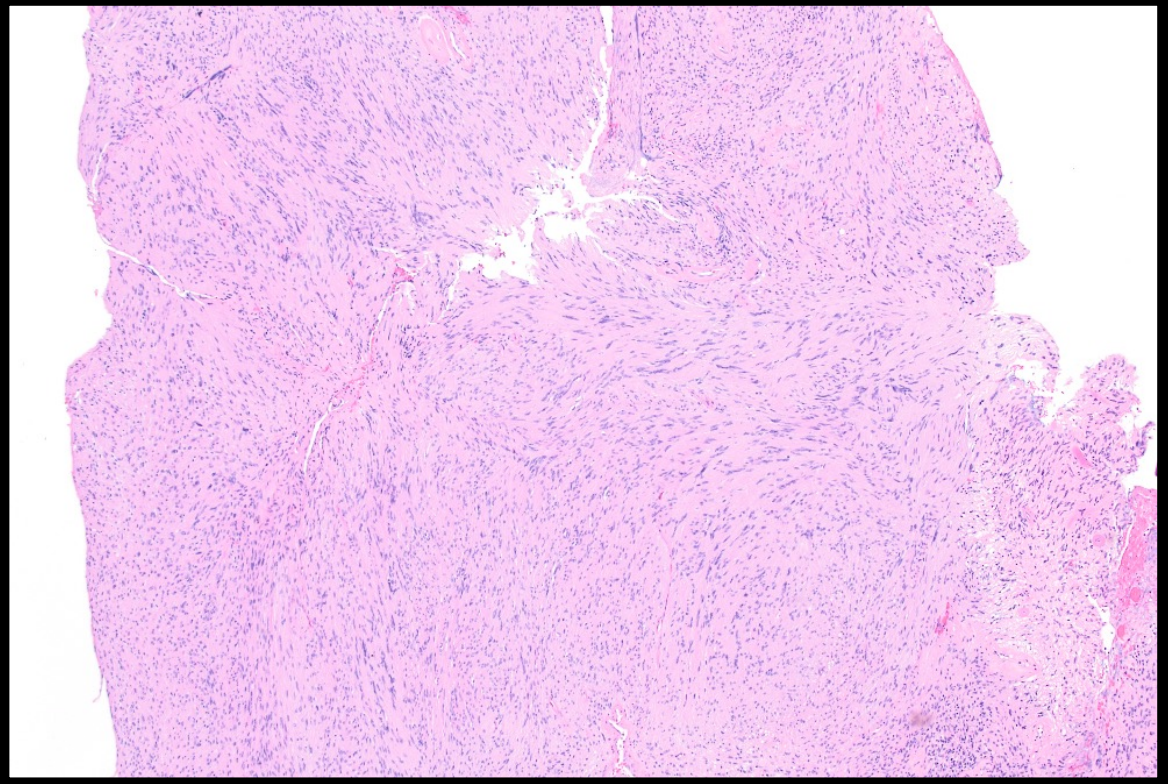


Image 1

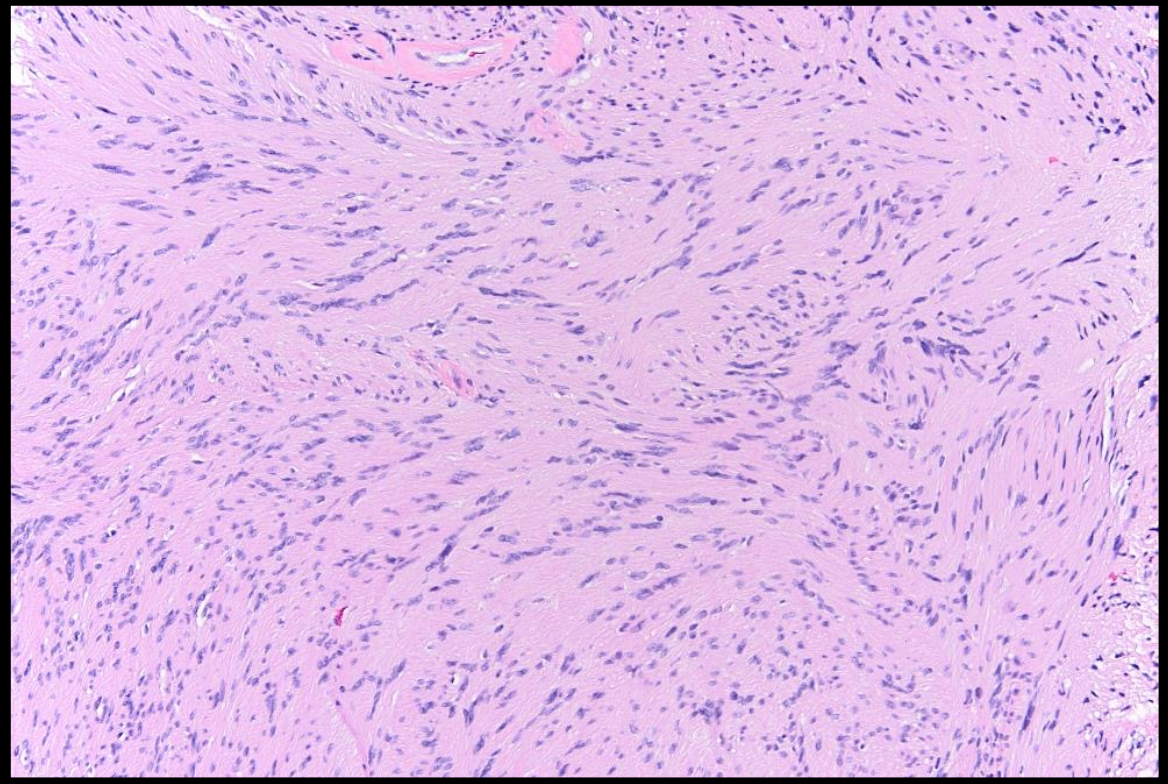


Image 2

Final Dx:

Vestibular Schwannoma

Case Discussion

- Differential diagnosis and relative frequency for cerebellopontine angle masses can be recalled from the acronym ‘A M E N’
  - Acoustic neuroma (aka vestibular schwannoma) – 80%
  - Meningioma – 10%
  - Ependymoma – 5%
  - Neuroepithelial cysts (epidermoid/arachnoid) – 5%
- Imaging was most consistent with vestibular schwannoma given:
  - Intense heterogenous contrast enhancement
  - Mass extension into medial internal auditory canal
  - No evidence of a dural tail
- Pathology was most consistent with vestibular schwannoma given:
  - Presence of Antoni A and Antoni B cells, and Verocay bodies
  - Meningiomas lack the features above and are more cellularly compact

Case Discussion

Vestibular Schwannoma aka acoustic neuroma

- Benign tumors of CN VIII
  - 85% involve the cochlear nerve
  - 61% involving vestibular nerve
  - Trigeminal nerve 17% and Facial nerve 6%
- Epidemiology
  - Incidence of 3-5/100,000 person-years with increasing incidence with age
  - Median age of diagnosis, 50yrs
  - Represent 8% of all primary intracranial tumors
- Common symptoms: unilateral hearing loss (often unrecognized by patient), dizziness, vestibular dysfunction, and facial weakness or sensory change
- Bilateral schwannomas associated neurofibromatosis type 2 (NF2)
  - Present earlier than unilateral, commonly in the third decade
  - Represent ~5% of vestibular schwannoma cases

Case Discussion

- Management usually based on the Koos grading scale

Koos Grading Scale		
Grade 1	Small intracanalicular	Observation
Grade 2	Small tumor with protrusion into CP cistern, no brainstem contact	Observation; SRS/SRT if symptomatic
Grade 3	Tumor occupying the CP cistern with no brainstem displacement	SRS/SRT vs surgery
Grade 4	Large tumor with brainstem and cranial nerve displacement	Surgery

SRS – stereotactic radiosurgery; SRT – stereotactic radiotherapy

- Surgical approaches include retrosigmoid, middle fossa, and translabyrinthine depending on preservation of hearing function
- Our patient had a Grade 4 vestibular schwannoma and completed successful debulking via a retrosigmoid approach

References:

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