AMSER Case of the Month July 2025

50-year-old female with a 6-month history of severe headaches

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Patient Presentation

HPI: 50-year-old female presented to her primary care physician with recurrent severe headaches and a tender spot on the top of her head for 6 months. These occurred nearly daily and ranged in severity from a 1-10/10. She also endorsed dizziness, vision changes, depressive symptoms, and occasional anterior neck pain.

Past Medical History: Chronic Kidney Disease, Hypothyroidism, Hypertension, Fibromyalgia

Past Surgical History: Appendectomy, Cholecystectomy, B/L Carpal Tunnel Release, Thyroidectomy

Family History: Cancer in mother, father, maternal grandmother, and maternal grandfather

Social History: Current smoker, 3 packs/day for 30 years

Patient Presentation

Physical Exam:

Mental Status: Alert and oriented x3. Normal speech without aphasia/dysarthria. Normal attention and memory.

Cranial Nerves: Pupils equal, round, and reactive to light. No nystagmus. Visual fields full and equal. Facial sensation intact. Face symmetric. Hearing intact and equal bilaterally. Palate elevates symmetrically. Tongue midline. Shoulder shrug equal bilaterally.

Motor: 5/5 strength in bilateral upper and lower extremities.

Sensory: Intact to light touch in all dermatomes.

Reflexes: Normal.

Gait: Steady.

Coordination: Finger-to-nose intact bilaterally. No pronator drift.

Pertinent Labs

ANA: Positive at 1:320

Anti-dsDNA: Indeterminate

BMP: BUN 22, Creatinine 1.54

Thyroid: TSH 93.0, Free T4 0.41

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

Variant 7: **Headache with one or more of the following "red flags": increasing frequency or severity, fever or neurologic deficit, history of cancer or immunocompromise, older age (>50 years) of onset, or posttraumatic onset. 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
CT head without IV contrast	Usually Appropriate	☢☢☢
Arteriography cervicocerebral	Usually Not Appropriate	☢☢☢
MRA head with IV contrast	Usually Not Appropriate	O
MRA head without and with IV contrast	Usually Not Appropriate	O
MRA head without IV contrast	Usually Not Appropriate	O
MRI head with IV contrast	Usually Not Appropriate	O
MRV head with IV contrast	Usually Not Appropriate	O
MRV head without and with IV contrast	Usually Not Appropriate	O
MRV head without IV contrast	Usually Not Appropriate	O
CT head with IV contrast	Usually Not Appropriate	☢☢☢
CT head without and with IV contrast	Usually Not Appropriate	☢☢☢
CTA head with IV contrast	Usually Not Appropriate	☢☢☢
CTV head with IV contrast	Usually Not Appropriate	☢☢☢

This imaging modality was ordered by the primary care physician

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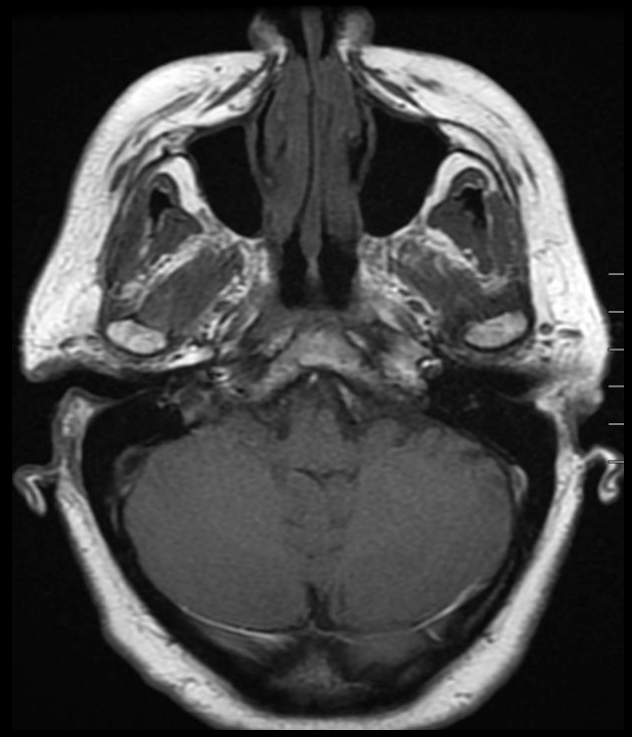
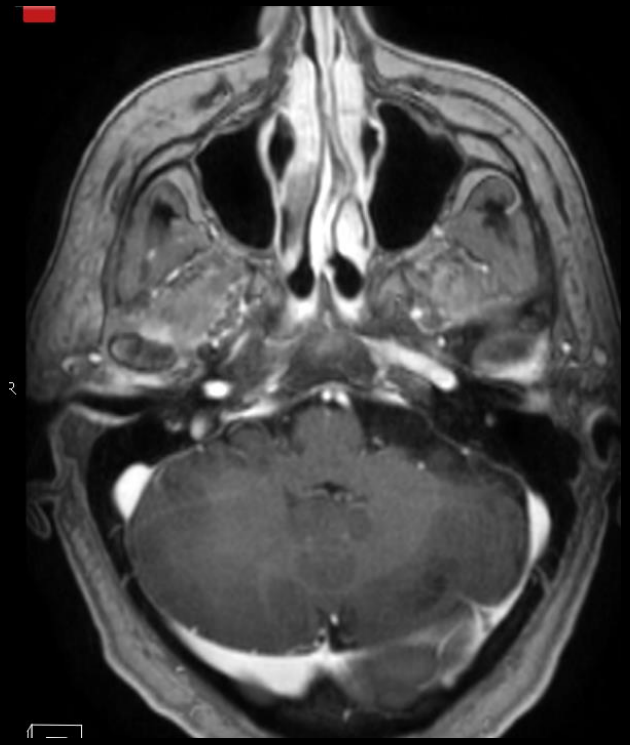
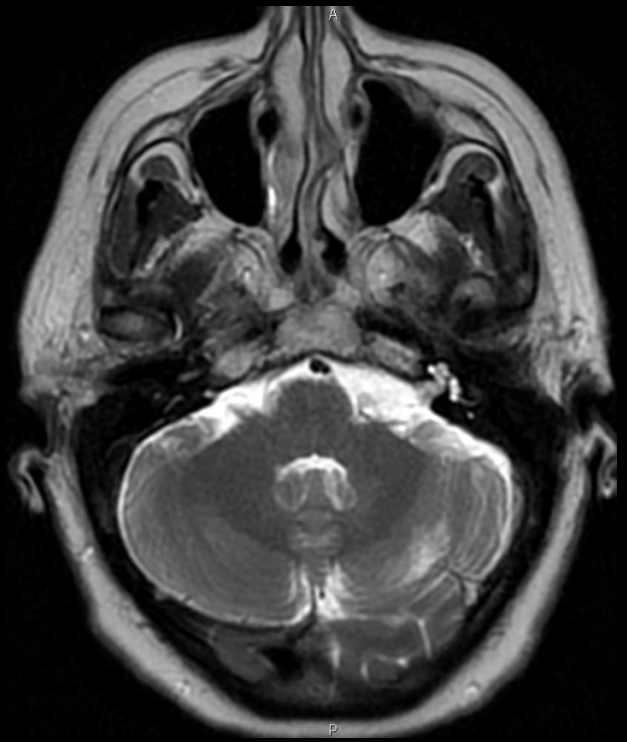
Findings (unlabeled)



T1 Pre-Contrast

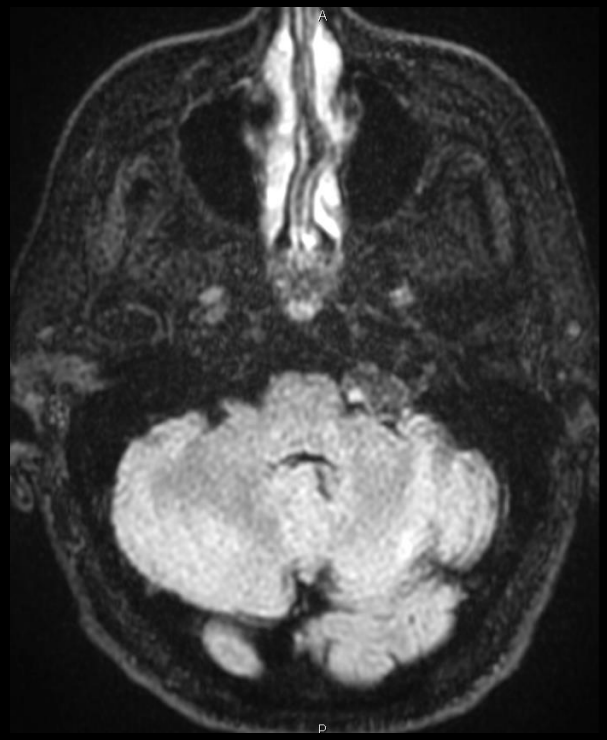


T1 Post-Contrast

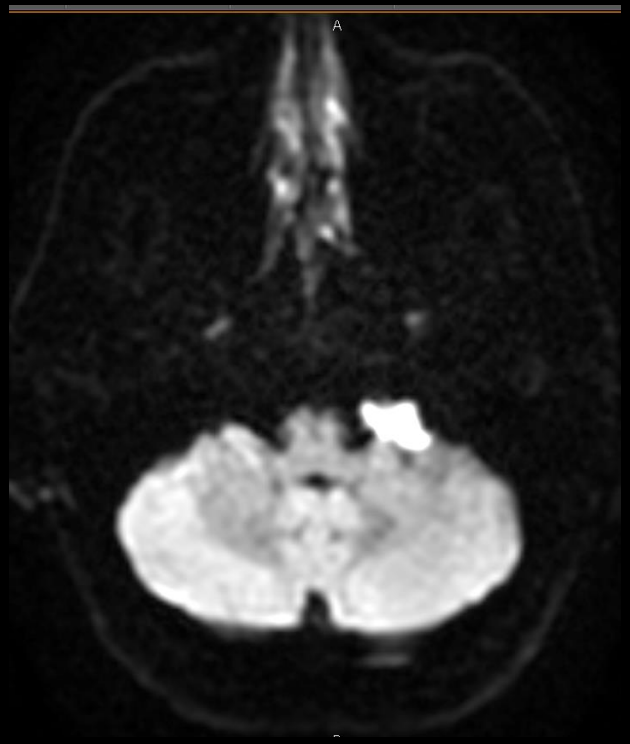


T2

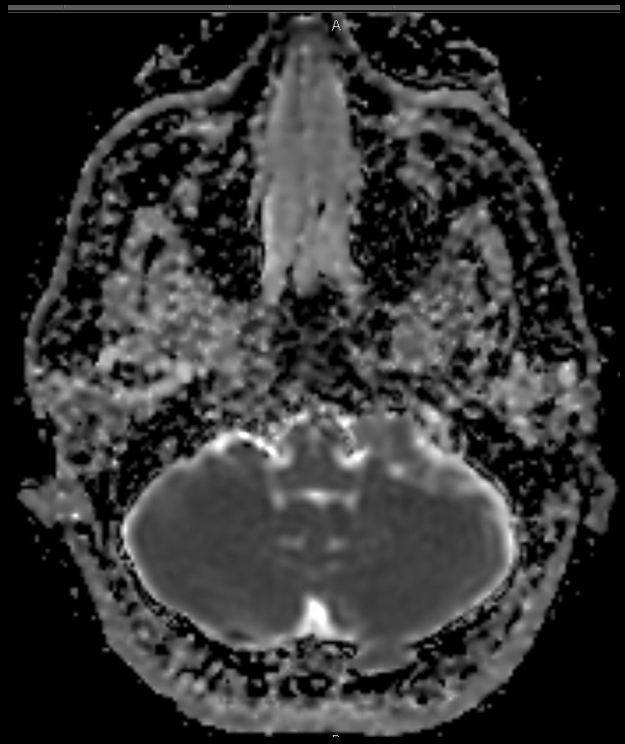
Findings (unlabeled)



FLAIR

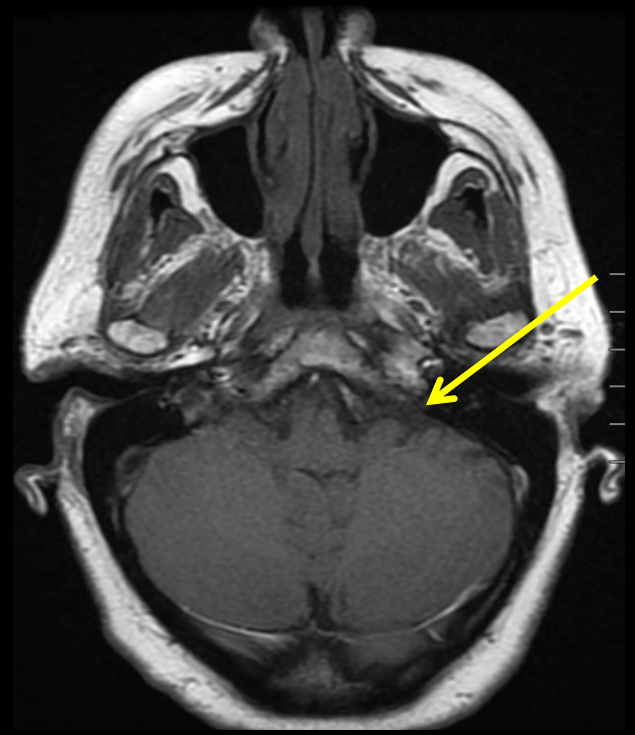


DWI

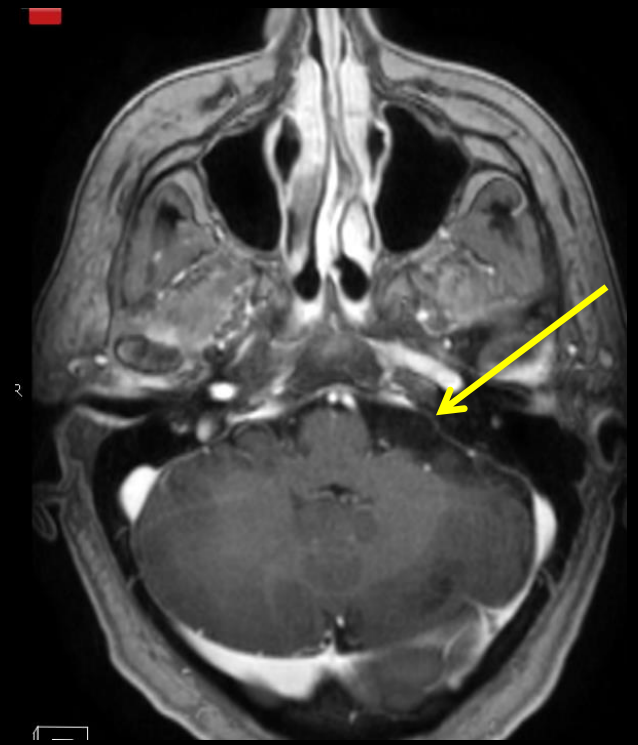


ADC

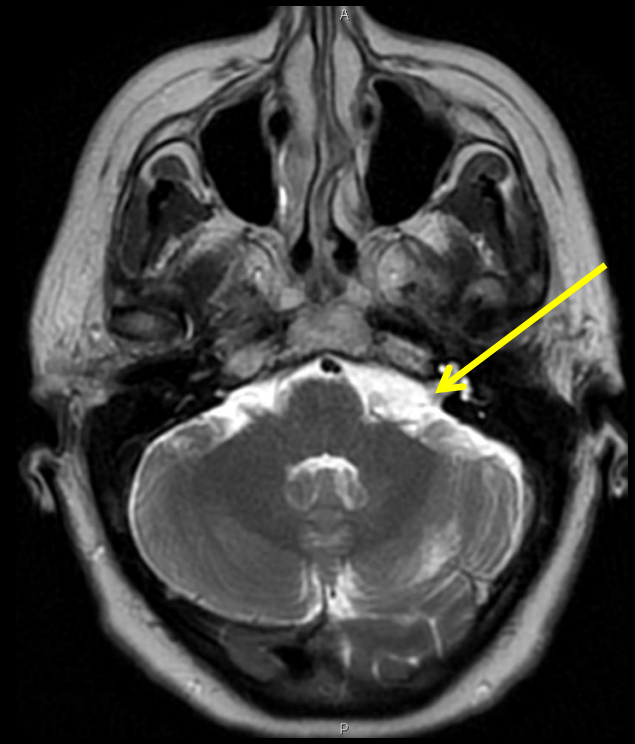
Findings (labeled)



T1 Pre-Contrast



T1 Post-Contrast



T2

- There appears to be a lesion in the left cerebellopontine angle
- It does not enhance with gadolinium contrast administration
- The lesion is T1 hypointense and T2 hyperintense

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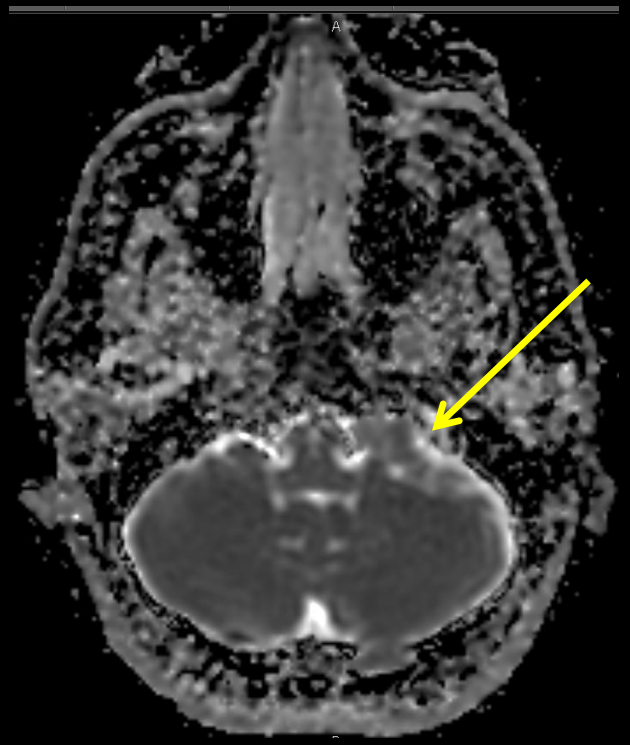
Findings (labeled)

FLAIR

DWI



ADC

- FLAIR demonstrates heterogenous intensity (“Dirty CSF” appearance)
- DWI indicates restricted diffusion within the lesion

Final Dx:

Cerebellopontine Angle (CPA) Epidermoid Cyst

Management

This patient was then referred to neurosurgery for further evaluation. On repeat examination, the patient noted worsening facial pains in addition to her previous symptoms. However, in the setting of negative focal neurologic or definite cranial nerve signs, the neurosurgical team and patient opted for conservative management with yearly MRI monitoring of the lesion.

Case Discussion: Etiology and Pathology

- Epidermoid cysts comprise about 0.2-1.8% of all intracranial tumors[1]
- These are benign, non-neoplastic lesions that are slow growing and typically present at 20-40 years of age[1]
- They are thought to be remnants of ectodermal tissue that have been encased within the neural tube and are therefore a congenital lesions[2]
  - Can also be post-surgical or post-traumatic
- They consist of a stratified squamous epithelial external lining with cystic interior contents that include desquamated keratin, epithelial cells, and cholesterol debris[2]
- These lesions are closely related to dermoid cysts, which appear largely similar however dermoid cysts contain tissue from at least two somatic tissue lines (ectoderm, mesoderm, endoderm), while epidermoid cysts only include ectodermal tissue[2]
  - Fat density is commonly visible in dermoid cysts

Case Discussion: Etiology and Pathology

CPA Lesion Differential Diagnoses (AMEN Mnemonic):

- **Acoustic Neuroma** – will enhance vividly with no diffusion restriction
- **Meningioma** – will enhance vividly with no diffusion restriction
- **Ependymoma** – tends to enhance with absent or reduced diffusion restriction
- **Neuroepithelial Cyst** (arachnoid cyst or epidermoid cyst) – arachnoid cysts have complete signal suppression on FLAIR with no diffusion restriction[3]

Intracranial Epidermoid Cyst Locations:

- Intra-axial: CPA (40-50%), Suprasellar Cistern (10-15%), Fourth Ventricle (15%), Midline Supratentorial (<5%), Spine (<5%)[4]
- Extra-axial: Within cranial bones (10%)[2]

Case Discussion: Patient Presentation and Diagnosis

Presentation:

- Symptoms of these lesions are most commonly due to gradual mass effect caused by a slowly enlarging lesion
- Headaches are the most common symptom
- May also cause cranial nerve deficits (hearing loss, vestibular symptoms, facial nerve dysfunction, etc.), cerebellar dysfunction, dizziness, elevated intracranial pressure, trigeminal neuralgia, and/or hydrocephalus
- Rarely, aseptic meningitis can occur due to cyst rupture with spillage of internal contents[5]

Imaging:

- MRI with and without contrast is most helpful for diagnosis
- **T1 MRI:** Hypointense to isointense relative to CSF, though may be slightly more heterogeneous
- **T2 MRI:** Hyperintense, often similar to or slightly brighter than CSF. May show internal heterogeneity or irregular margins
- **DWI:** Hyperintense, which is an important feature distinguishing them from arachnoid cysts (which do not restrict diffusion)
- **FLAIR:** Heterogenous intensity. “Dirty CSF” appearance due to presence of internal keratin/epithelial/cholesterol debris
- **Gadolinium Contrast Enhancement:** Can have thin peripheral enhancement, otherwise it is typically non-enhancing
- **CT:** Hypodense and non-calcified (although rare calcifications may occur). Smooth or lobulated borders

Case Discussion: Treatment and Prognosis

Treatment:

- **Conservative:** Reserved for patients with mild and/or stable symptoms or those who are poor surgical candidates
  - Management of headaches; Vestibular medications for dizziness; Carbamazepine for trigeminal neuralgia
- **Surgery:** For symptomatic lesions
  - May be hindered by envelopment of nearby structures[6]
  - Recurrence is common, especially with sub-total or partial resection (as high as 65%)[4]

Prognosis:

- Due to slow growth and benign nature, long-term prognosis is generally excellent[7]
- However, this is influenced by lesion location, size, and degree of involvement with adjacent structures

References:

1. Fu JB, Nelson MB, Bruera E. Inpatient Rehabilitation of Cerebellopontine Angle Epidermoid Cysts: Report of 3 Cases. *Neurosurg Q*. 2014;24(3):197-202. doi:10.1097/WNQ.0b013e3182a2fac7
2. Osborn AG, Preece MT. Intracranial cysts: radiologic-pathologic correlation and imaging approach. *Radiology*. 2006;239(3):650-664. doi:10.1148/radiol.2393050823
3. Bergen D, Sattouf M, Deng F, et al. Cerebellopontine angle mass (mnemonic). Reference article, Radiopaedia.org (Accessed on 21 May 2025) https://doi.org/10.53347/rID-4618
4. Hasegawa H, Vakharia K, Carlstrom LP, et al. Long-term surgical outcomes of intracranial epidermoid tumors: impact of extent of resection on recurrence and functional outcomes in 63 patients. *J Neurosurg*. 2021;136(6):1592-1600. Published 2021 Oct 15. doi:10.3171/2021.5.JNS21650
5. Fernández-de Thomas RJ, Vicenty-Padilla JC, Sánchez-Jiménez JG, et al. Obstructive Hydrocephalus and Chemical Meningitis Secondary to a Ruptured Spinal Epidermoid Cyst. *World Neurosurg*. 2019;132:173-176. doi:10.1016/j.wneu.2019.08.187
6. Pop MM, Bouros D, Klimko A, Florian IA, Florian IS. Intracranial epidermoid cysts: benign entities with malignant behavior: experience with 36 cases. *Sci Rep*. 2023;13(1):6474. Published 2023 Apr 20. doi:10.1038/s41598-023-33617-x
7. Revuelta-Gutiérrez R, Díaz-Romero Paz RF, Vales-Hidalgo LO, Hinojosa-González R, Barges-Coll J. Cerebellopontine angle epidermoid cysts. Experience of 43 cases with long-term follow-up. *Cir Cir*. 2009;77(4):257-248.

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