

AMSER Rad Path

Case of the Month:

49-year-old female with headaches

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Patient Presentation

- **History of Present Illness**: 49F with PMHx of WPW and MDD presents with a headache. She presents from home after receiving a call from her PCP about the results of an outpatient non-contrast CT head and was told to go to the ED. Her headaches have been occurring daily for the past 5 months. It has been growing in intensity and worsens with rapid positional changes and sneezing. It is associated with a sense of imbalance. Notably she has also had 2 episodes of syncope where vision “goes dark” and she passes out.
- Denies numbness, weakness, and tingling in her extremities. Also denies nausea, vomiting, blurry or double vision.
- **Social History**: At baseline, she is very active, runs marathons and goes out dancing with friends regularly
- **Physical Exam**: PERRLA, EOM intact; 5/5 strength throughout, equal and bilateral sensation throughout. Reflexes present and symmetric throughout. CN II-XII grossly intact. Finger-nose-finger performed bilaterally without difficulty. No pronator drift. Gait steady and regular.

Pertinent Labs

- Pre-operative
 - Hgb 15.6 borderline high
 - Hct 46.7
- Baseline (2 years ago)
 - Hgb 13

What Imaging to Order?

Variant 5: Headache with features of intracranial hypotension (eg, positional, worse when upright, better when lying down). Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
MRI head without and with IV contrast	Usually Appropriate	0
MRI thoracic spine with IV contrast	May Be Appropriate (Disagreement)	0
MRI thoracic spine without and with IV contrast	May Be Appropriate	0
MRI thoracic spine without IV contrast	May Be Appropriate	0
Arteriography cervicocerebral	Usually Not Appropriate	⊕⊕⊕
MRA head with IV contrast	Usually Not Appropriate	0
MRA head without and with IV contrast	Usually Not Appropriate	0
MRA head without IV contrast	Usually Not Appropriate	0
MRI head with IV contrast	Usually Not Appropriate	0
MRI head without IV contrast	Usually Not Appropriate	0
MRV head with IV contrast	Usually Not Appropriate	0
MRV head without and with IV contrast	Usually Not Appropriate	0
MRV head without IV contrast	Usually Not Appropriate	0
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	⊕⊕⊕
CTA head with IV contrast	Usually Not Appropriate	⊕⊕⊕
CTV head with IV contrast	Usually Not Appropriate	⊕⊕⊕

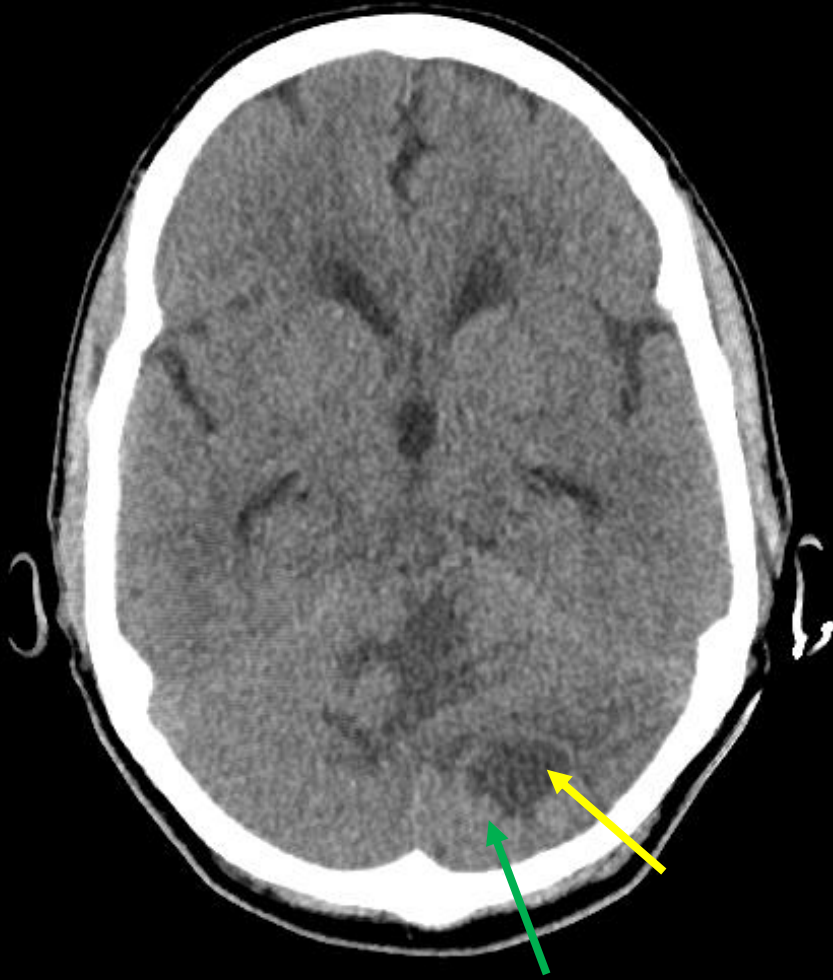
Ordered 2nd as follow-up imaging in ED

Ordered 1st in Out-Patient setting by PCP

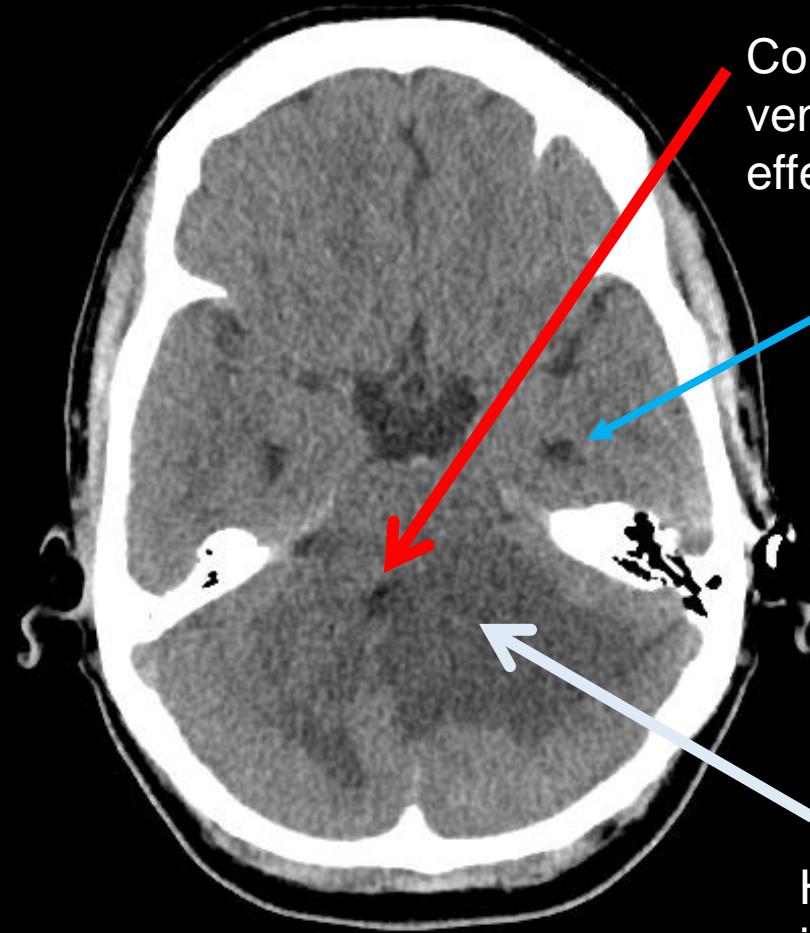
CT Head Non-contrast (not labeled)



CT Head Non-contrast (labeled)



Tumor with solid (green arrow) and cystic (yellow arrow) components.

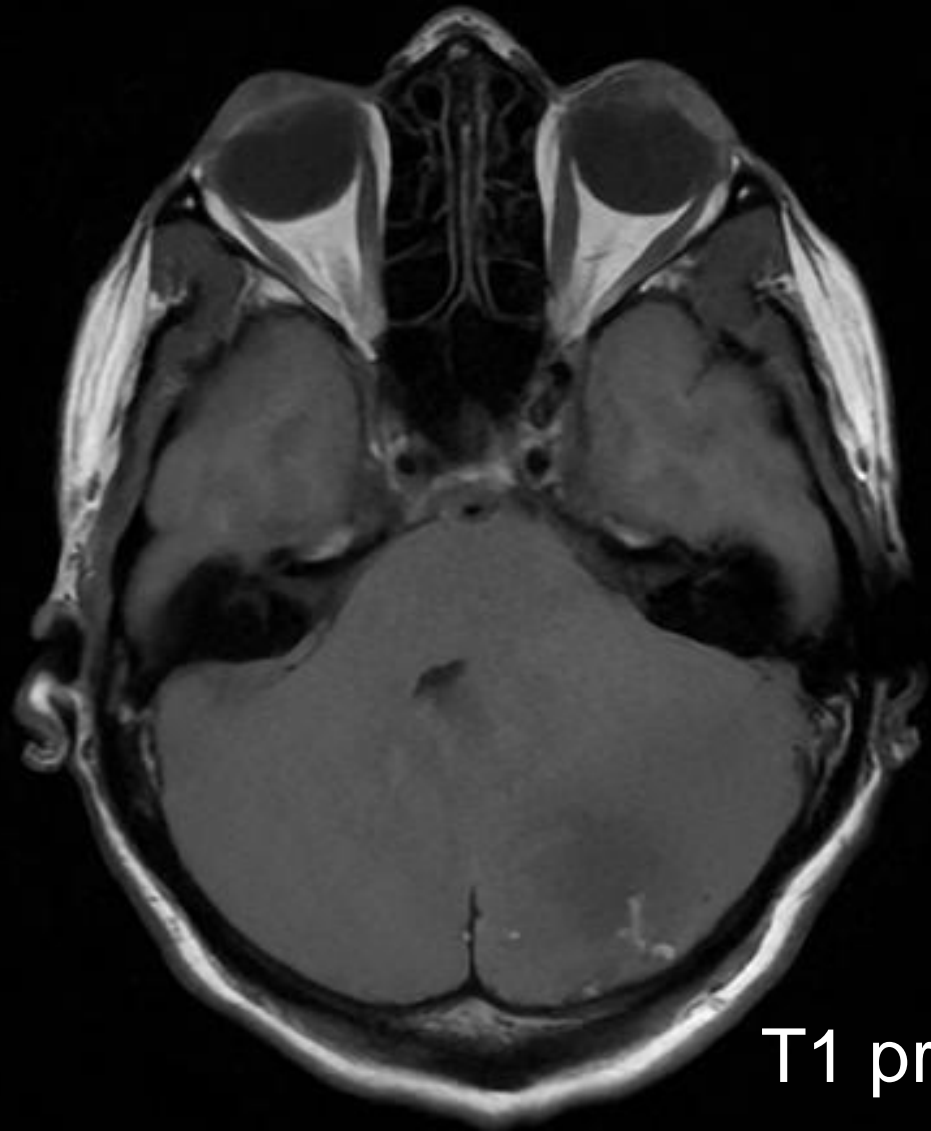


Compression of the 4th ventricle due to mass effect

Slight dilation of temporal horns of lateral ventricles

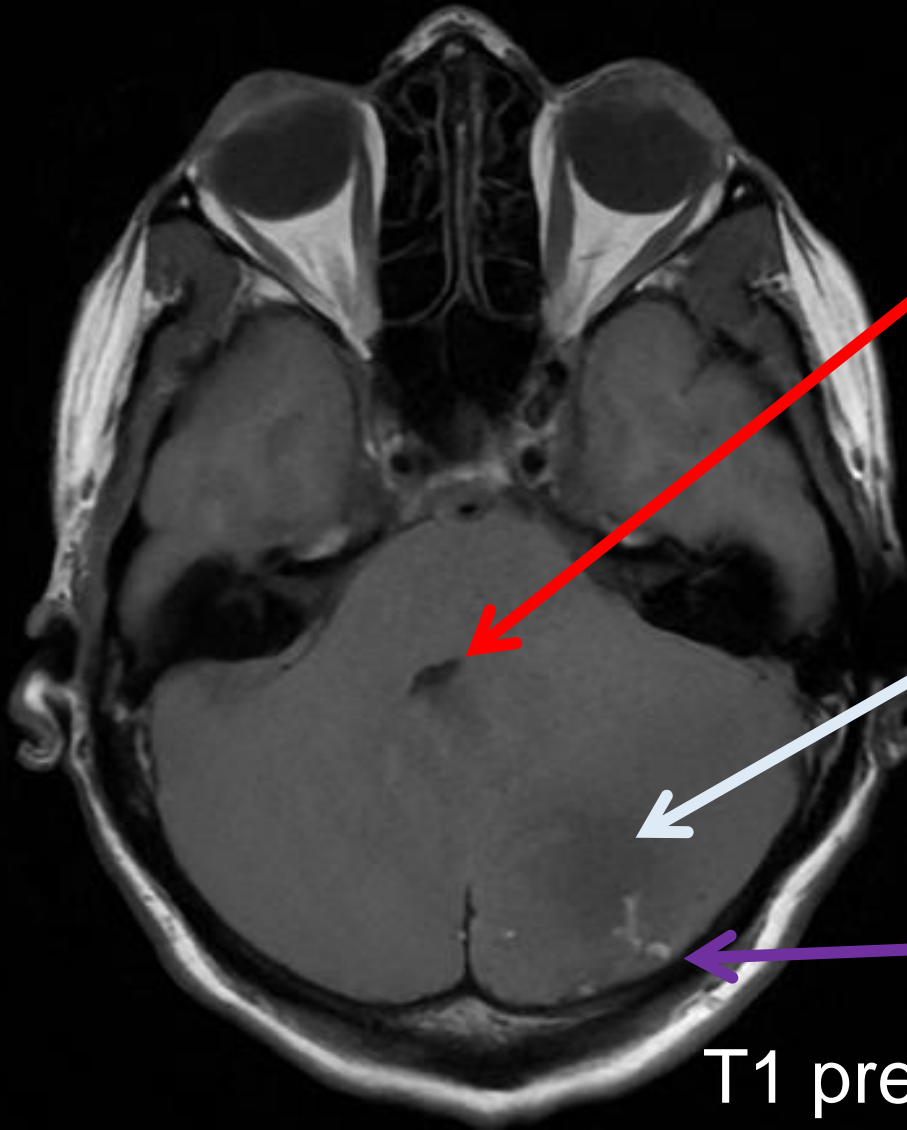
Hypodense area indicating surrounding edema

MRI (T1 Pre-Contrast - not labeled)



T1 pre-contrast

MRI (T1 Pre-Contrast - labeled)



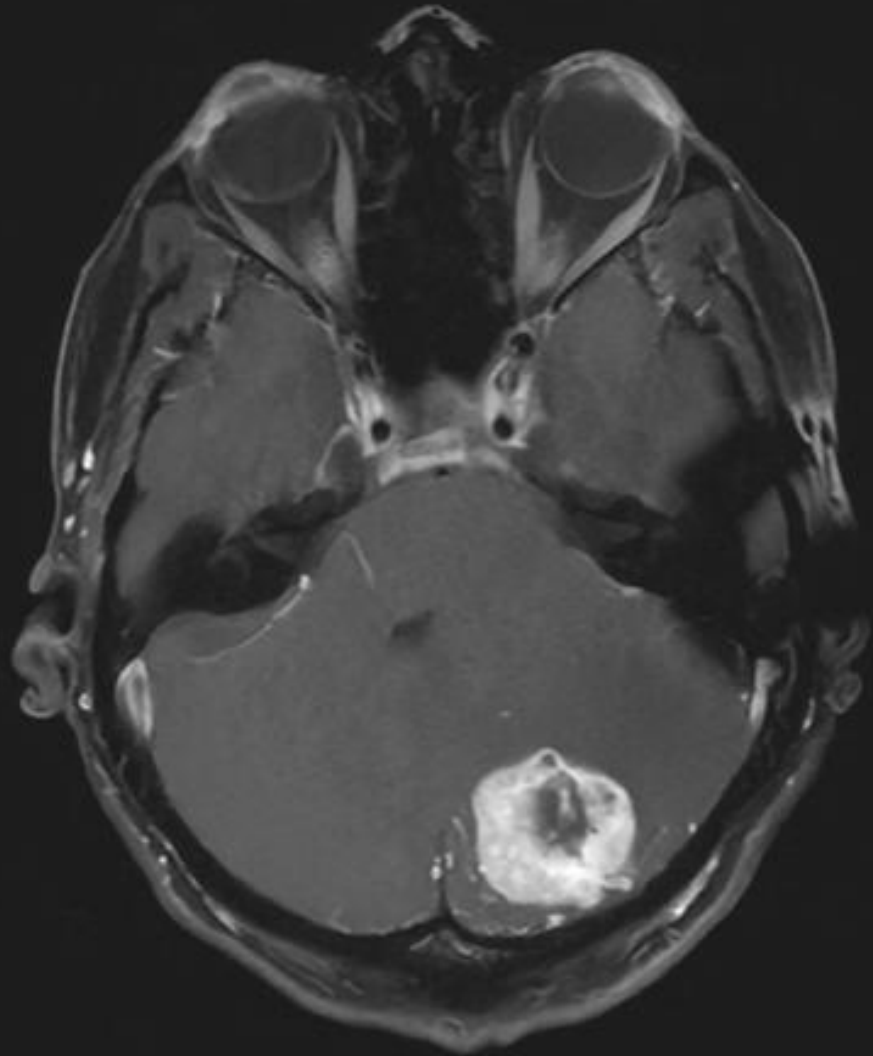
Compression of the 4th Ventricle due to mass effect

Hypointense area indicating surrounding edema

Slow flow in a feeding vessel

T1 pre-contrast

MRI (T1 Post-Contrast - not labeled)



T1 post-contrast

MRI (T1 Post-Contrast - labeled)

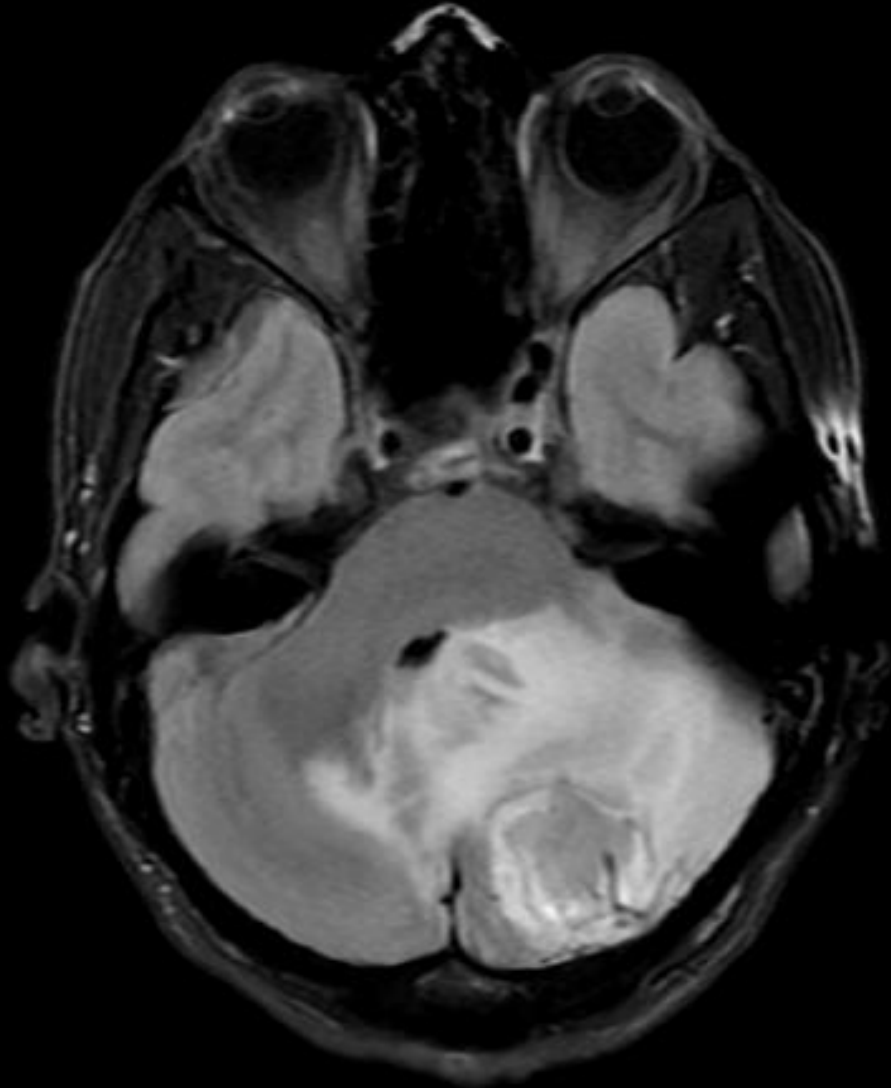


Enhancing solid component

Non-enhancing cystic component centrally

T1 post-contrast

MRI (T2 FLAIR and T2 Series - not labeled)



T2 FLAIR

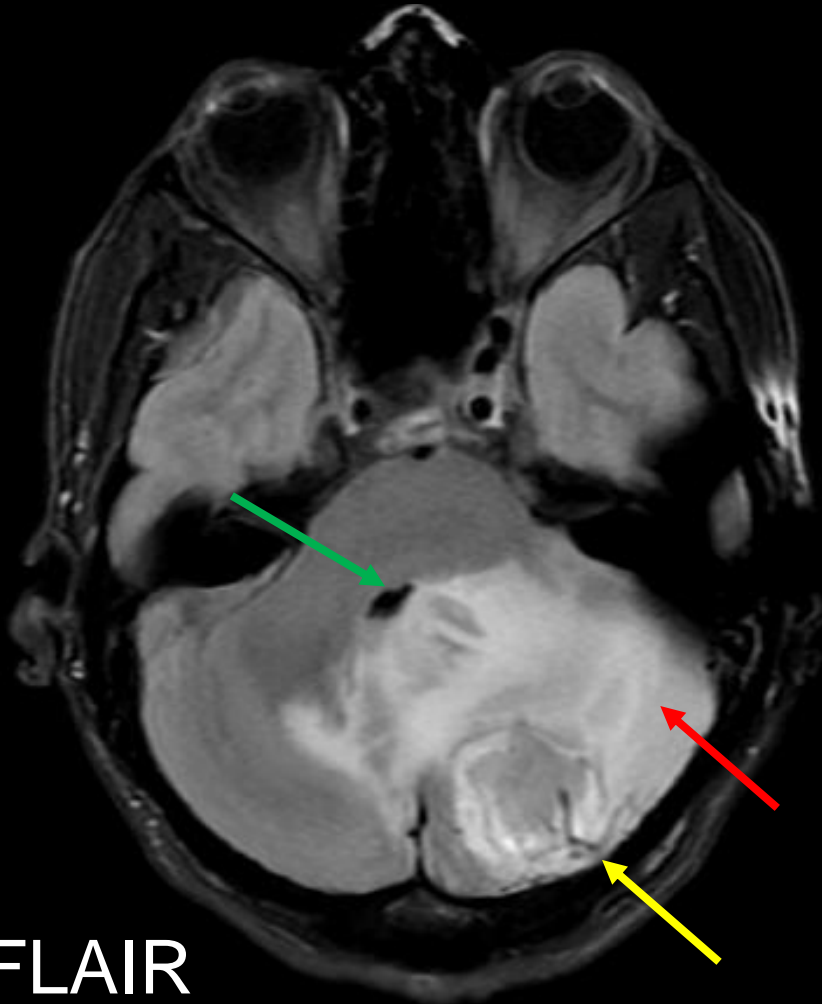


T2

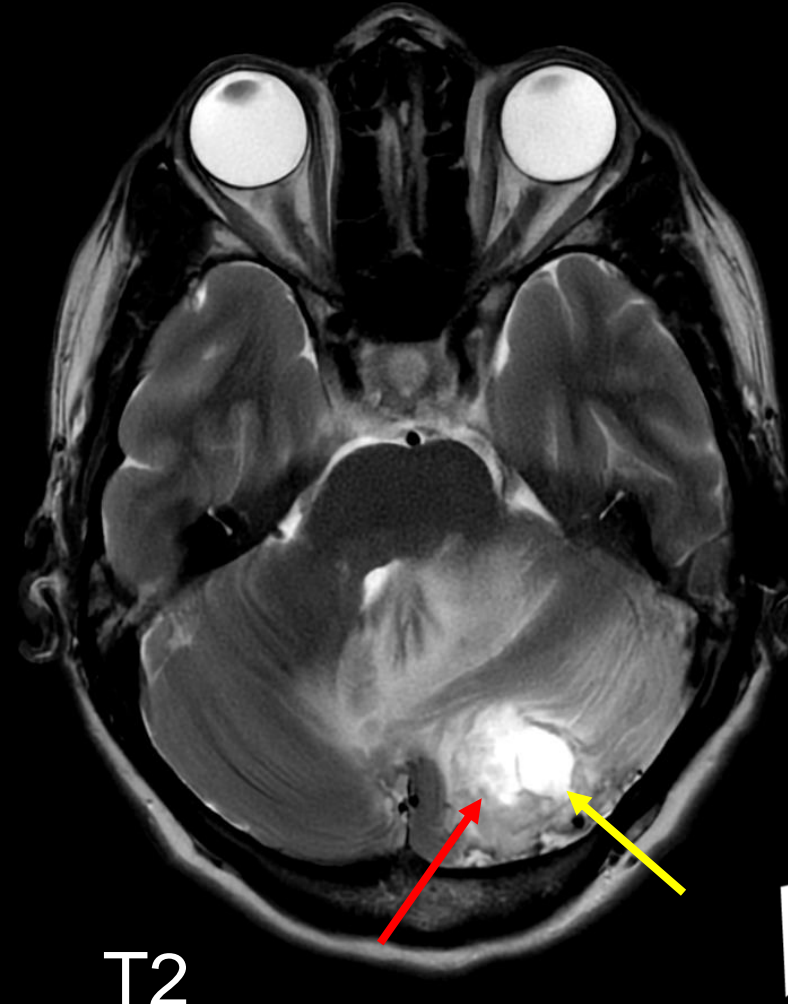
MRI (T2 FLAIR and T2 Series- labeled)

T2 FLAIR: surrounding edema (red arrow) and flow void in feeding vessel (yellow arrow). Mass effect upon 4th ventricle (green arrow).

T2 Series: Tumor with solid (red arrow) and cystic (yellow arrow) components.



T2 FLAIR



T2

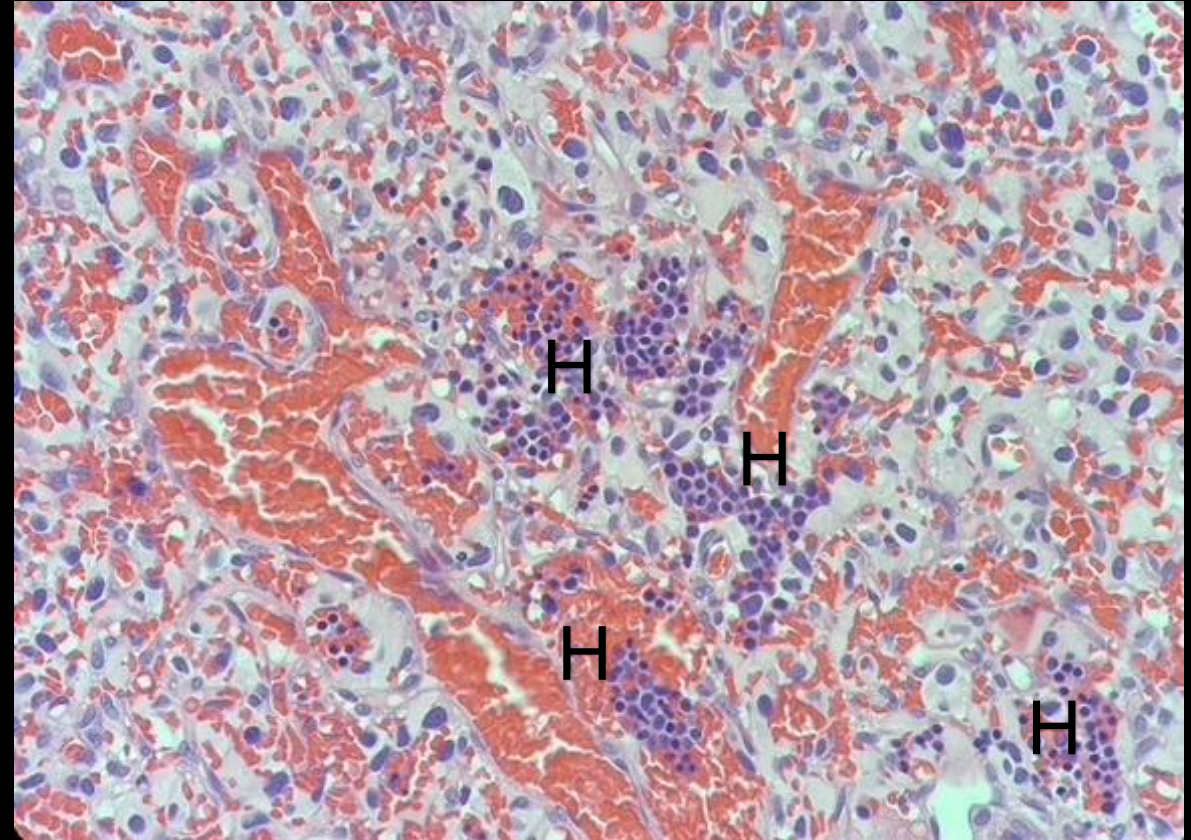
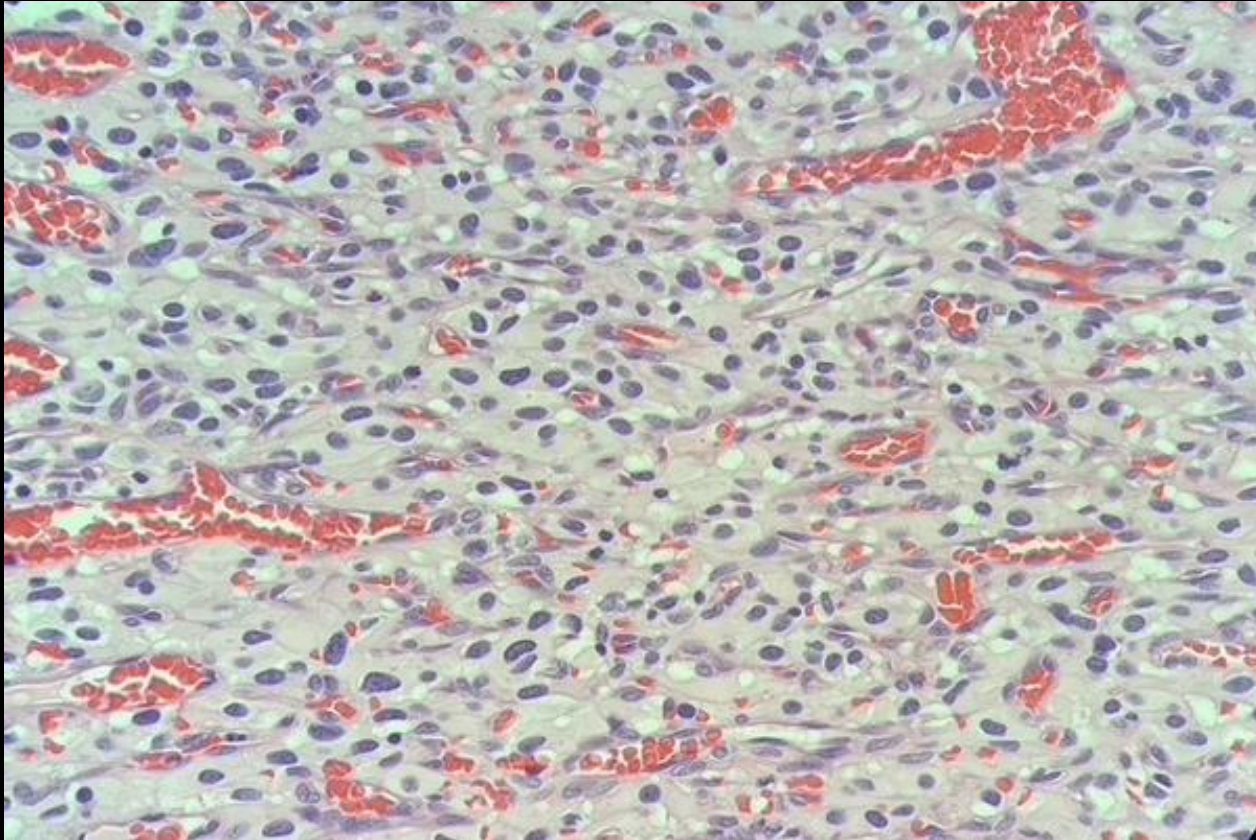
DDX (based on imaging)

- Brain metastases
 - Ependymoma
 - Vascular lesions
 - Pilocytic Astrocytoma
 - Medulloblastoma
-
- Brain metastases are more common - Diagnosis is difficult and it can be challenging to distinguish the above based on imaging.
 - Initial read pushed DDx towards malignancy, which led to prioritizing surgical intervention over further evaluation

Gross Path Findings

- Large, highly vascular cerebellar mass observed during operation in OR

Micro Path (labeled)



H&E Microscopic Pathology:

Left: Various thin-walled vessels of different calibers with surrounding polygonal stromal cells laden with lipids, with a “clear cell” appearance.

Right: Hypercellular tumor with areas of extramedullary hematopoiesis (H).

Final Dx:

Cerebellar Hemangioblastoma

Hemangioblastoma Overview

- Hemangioblastomas are rare, **benign**, highly vascularized neoplasms with a peak incidence at 20-50 years; most commonly appearing in the cerebellum, though can also be in the kidneys, liver, and pancreas.
- Symptoms relate to compression, edema, hemorrhage or less commonly, paraneoplastic erythrocytosis
- Sporadic cases account for ~75-80%, remainder found in patients with von Hippel-Lindau (vHL)
- When associated with vHL, ~60-80% of patients are found to have CNS hemangioblastomas, and incidence tends to be earlier for these patients as well.

Case Discussion

- Stromal cells are mutated -> VEGF, EPO upregulated
- Differential based on imaging include brain metastases, ependymoma, vascular lesions (in adults), medulloblastomas, and astrocytomas, with brain metastases being most common. Can be difficult to distinguish on imaging alone, and in this case was prospectively interpreted as a likely metastasis.
- Two key features that help distinguish hemangioblastomas from other similar lesions are flow void along periphery of lesion and enhancing component contacting the pial surface.
- Histologically can appear as renal clear cell carcinoma
- At risk for increased growth during pregnancy
- Consider testing for vHL if more than one lesion, or if other features, present
- **Treatments:** surgical resection with **pre-op angiography** to identify feeding arteries, embolize if needed (not done in this case) vs. radiation vs. angiogenic inhibitors
- Caution with procedures due to high bleeding risk

Conclusion

- Hemangioblastomas can appear due to sporadic mutations or with vHL syndrome due to mutations in stromal cells causing hypervascularity
- Symptoms arise from direct compression, edema or hemorrhage or paraneoplastic erythrocytosis
- It can appear like brain metastases on neuroimaging, which is more common
- Pathologically distinguished as hypervascular mass distinct from nearby tissue
- Histologically distinguished by hypervascularity with distinct stromal cells that look like renal cell carcinoma
- If operation is deemed necessary, important to decrease hemorrhage risk by identifying targets for embolization
- Resection is generally curative; incidence of recurrence is about 25%, and is associated with vHL, male sex, younger presentation (~22 y/o), and with incomplete resection.

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