AMSER Case of the Month October 2023

24-year-old female with dizziness and blurry vision.

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Patient Presentation

- HPI: Pt. was experiencing 8 months of daytime drowsiness, dizziness, and blurry vision. Pt. denied hearing changes, ptosis, facial asymmetry, headaches, gait imbalance. Pt. was initially diagnosed with hypersomnia with no response to treatment.

- Physical Exam: Cranial Nerves II-XII were intact, Strength in all four extremities was 5/5. No speech impairments noted.

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Pertinent Labs

- All initial Lab results were within normal limits.

What Imaging Should We Order?

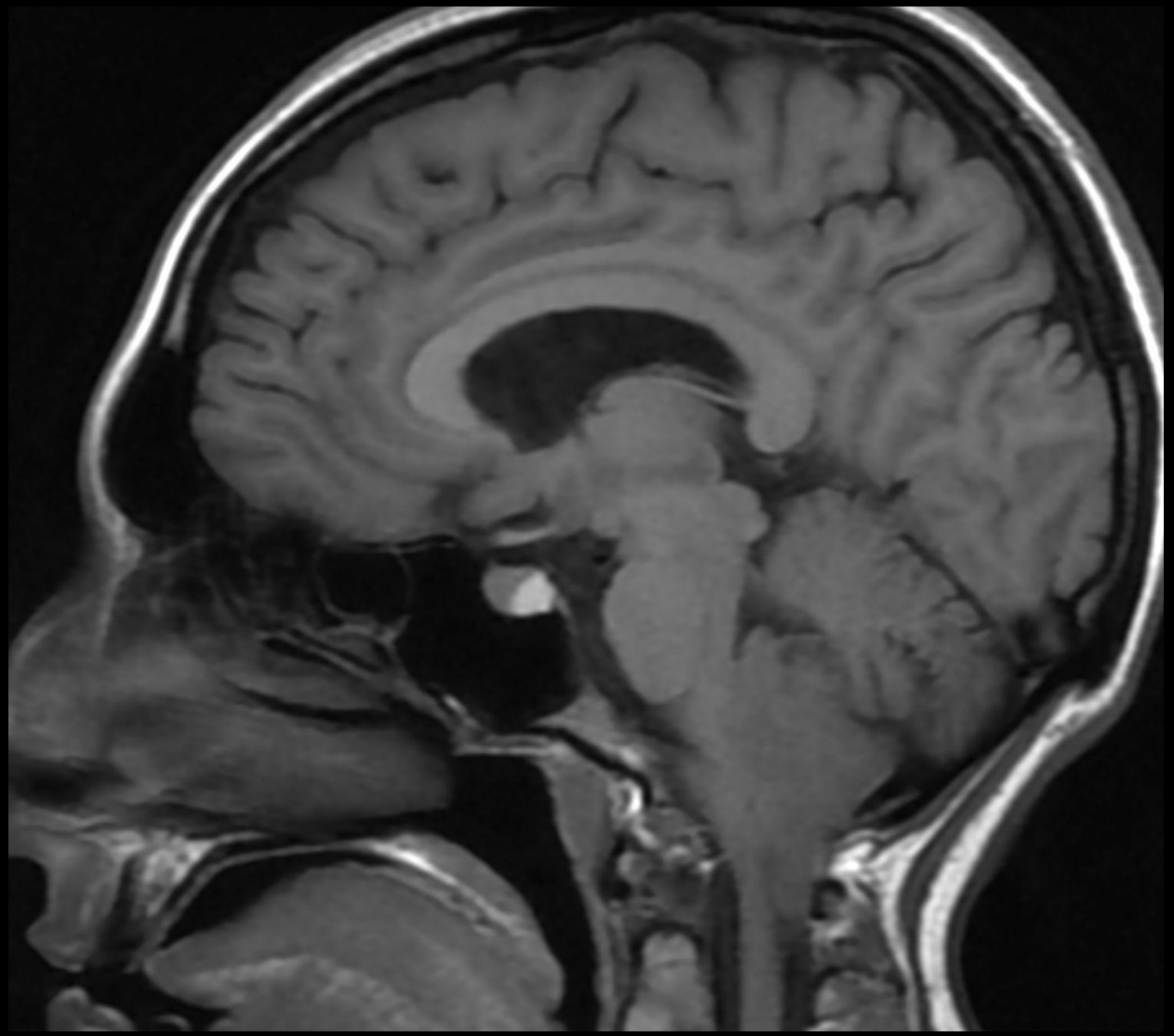
Select the applicable ACR Appropriateness Criteria

Variant 4: **Persistent or worsening mental status change despite clinical management of the suspected underlying cause (intoxication, medication-related, hypoglycemia, sepsis, etc) or acute change in mental status of unknown cause. 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	☢☢☢
CT head without and with IV contrast	May Be Appropriate	☢☢☢
CT head with IV contrast	Usually Not Appropriate	☢☢☢

This imaging modality was ordered by the Primary physician

Findings (unlabeled)



Sagittal T1

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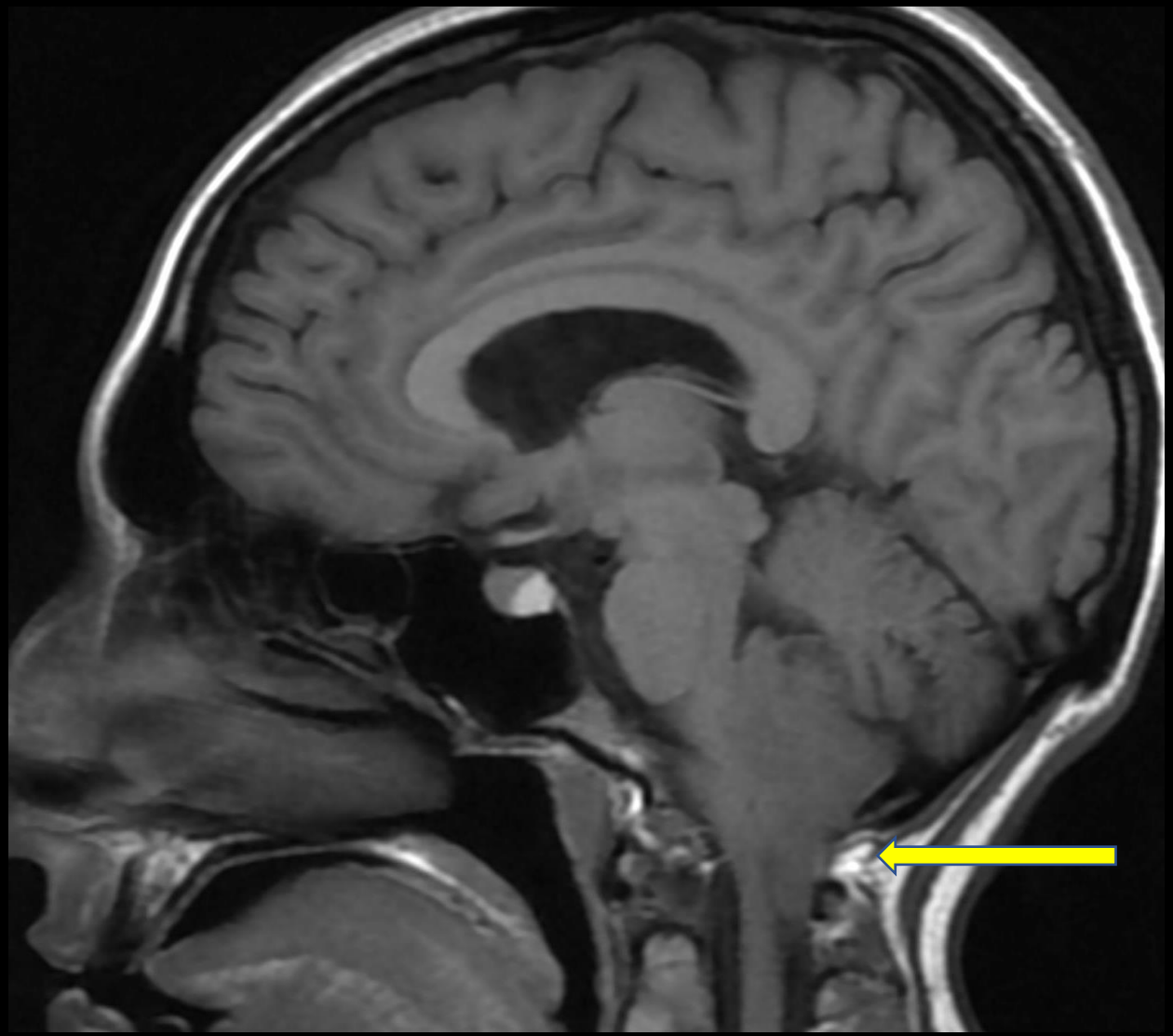
Findings (labeled)



Outside institution's diagnosis of brain MRI was Chiari I malformation due to apparent cerebellar tonsillar ectopia of > 5 mm, pointed morphology of the cerebellar tonsils, and crowded foramen magnum

The patient was then referred to a neurosurgeon for further evaluation

Neurosurgeon's review of images were concerning for a mass lesion, not Chiari I

Sagittal T1

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Select the applicable ACR Appropriateness Criteria

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This imaging modality was ordered by the Primary physician

Variant 2: **Acute or progressively worsening mental status change in patient with a known intracranial process (mass, recent hemorrhage, recent infarct, central nervous system infection, etc). Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
CT head without IV contrast	Usually Appropriate	☢☢☢
MRI head without and with IV contrast	Usually Appropriate	O
MRI head without IV contrast	Usually Appropriate	O
CT head without and with IV contrast	May Be Appropriate	☢☢☢
CT head with IV contrast	May Be Appropriate	☢☢☢

This imaging modality was ordered by the Neurosurgeon

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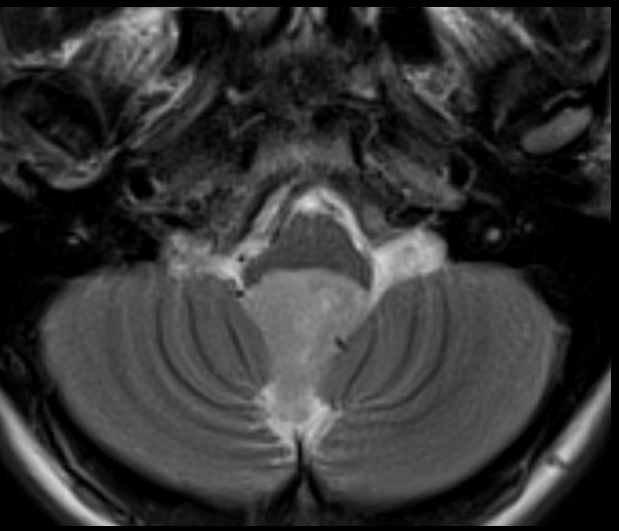
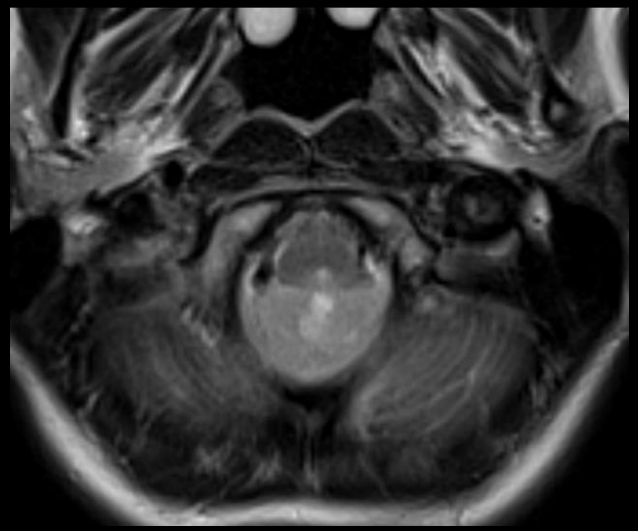
Findings (unlabeled)

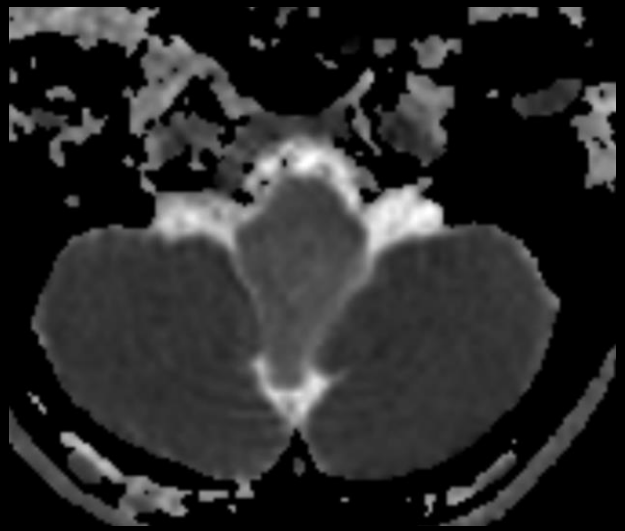
Axial T2



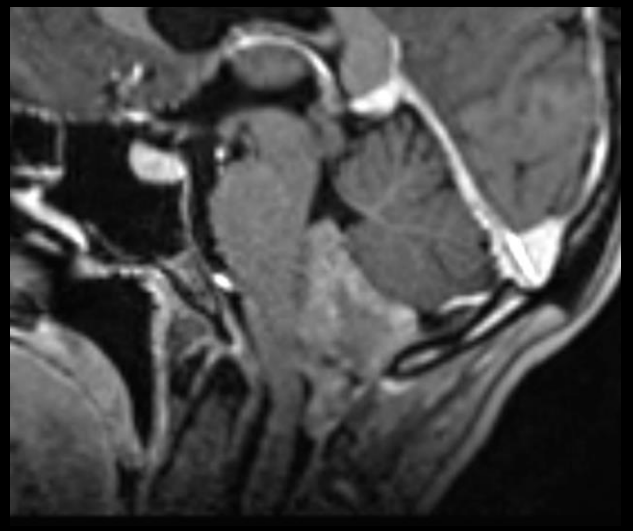
Axial T2



Axial ADC

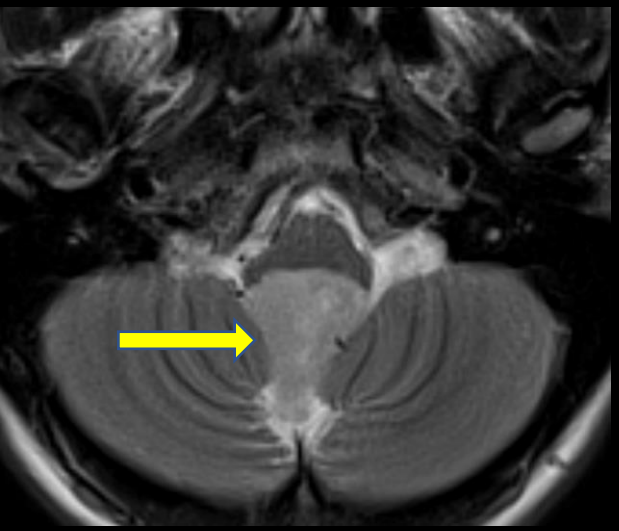


Sag T1 Post Contrast



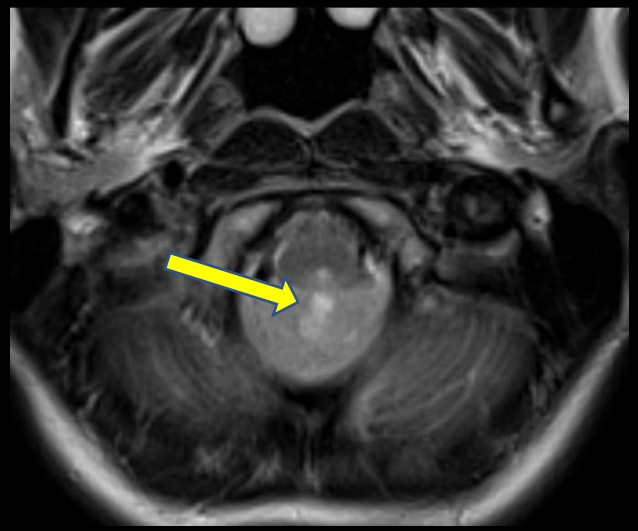
Findings (labeled)

Axial T2



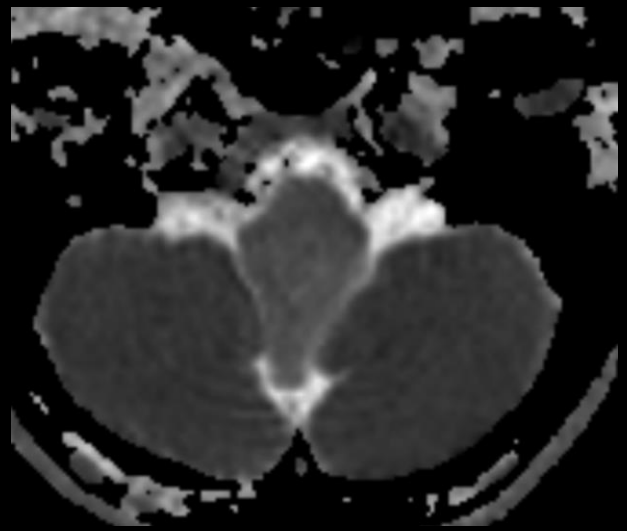
4th ventricular mass that is slightly hyperintense to brain parenchyma

Axial T2



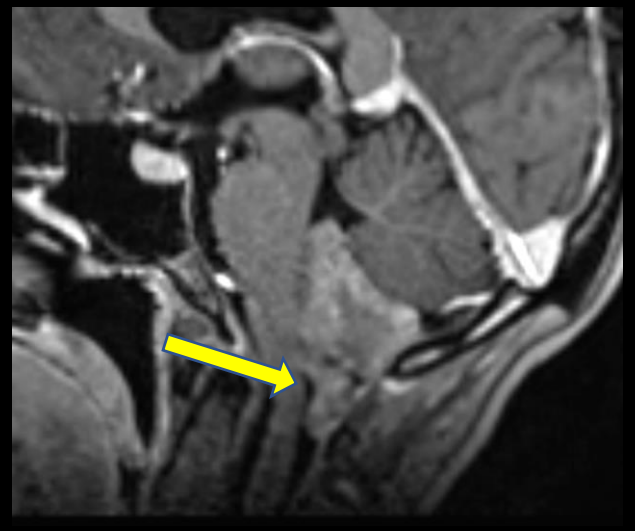
Mass is predominantly solid with small cystic components

Axial ADC



No restricted diffusion, indicating that the mass is not hypercellular

Sag T1 Post Contrast



Heterogeneously enhancing solid tumor. Mass “squeezes” through foramen of Magendie and cisterna magna

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Final Dx:

Fourth Ventricle Ependymoma
Posterior Fossa Group B

Case Discussion

- Ependymomas are primary CNS tumors that arise from ependymal cells that occur in 3 distinct places in the CNS:
  - Infratentorial ependymoma:
    - Most common location in brain (70%)
    - Arise from ependymal cells lining the 4^{th} ventricle
  - Supratentorial ependymoma:
    - Less common location in brain (30%)
    - Usually ***extraventricular*** in location
    - Arise from ependymal rests within the brain parenchyma
  - Spine (not further discussed)
    - Arise from ependymal lining of the central canal
    - Usually intramedullary in location

Case Discussion - Epidemiology

- Infratentorial ependymomas depend on molecular profile:
  - Posterior Fossa Group A
    - Much more common in young children (< 5 years of age) >> adolescents >> adults
    - Worse prognosis
  - Posterior Fossa Group B
    - Much more common in adults >> adolescents >> young children (< 5 years of age)
    - Better prognosis
- Supratentorial ependymomas:
  - Bimodal age distribution: young children (<5 years old) >> young adult
- Up to 10% can have leptomeningeal dissemination, thus imaging of the entire neuroaxis is essential

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Case Discussion - Presentation

- Infratentorial Ependymomas: Related to increased intracranial pressure
  - Nausea/Vomiting
  - Headaches
  - Blurry vision/papilloedema
  - Gait ataxia
  - Dizziness
- Supratentorial Ependymomas: Based on location in cerebrum
  - Seizures (most common)
  - Headaches
  - Focal neurologic deficit

Case Discussion – Imaging Findings

- Infratentorial Ependymomas:
  - Soft tumor known to “squeeze” through the 4th ventricle foramina, unlike 4th ventricular medulloblastomas (main ddx), which grown like “balls”
  - Heterogeneous tumors
    - Often both cystic and solid
    - Commonly have small calcifications and areas of hemorrhage
    - Minimal to no restricted diffusion
    - Heterogeneous enhancement
- Supratentorial Ependymomas:
  - Cystic and solid masses, with varying degrees of cystic and solid components
  - Tend to be larger and associated with surrounding vasogenic edema
  - Calcifications and hemorrhage are common
  - Solid components may show restricted diffusion
  - Heterogeneous enhancement

Case Discussion – Management

- Imaging of the entire neuroaxis to assess for leptomeningeal dissemination
- Treatment: Surgical resection and radiation therapy

References:

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- Koeller, K; Sandberg, G. D. (2002). Cerebral Intraventricular Neoplasms: Radiologic-Pathologic Correlation. RadioGraphics, 22(6), 1473–1505. https://doi.org/10.1148/rg.226025118

- Mu W, Dahmoush, H. (2023). Classification and neuroimaging of ependymal tumors. Frontiers in Pediatrics, 11. https://doi.org/10.3389/fped.2023.1181211

- Routman DM, Raghunathan A, Giannini C, Mahajan A, Beltran C, Nagib MG, Nageswara Rao AA, Skrypek MM, Laack NNI. Anaplastic Ependymoma and Posterior Fossa Grouping in a Patient With H3K27ME3 Loss of Expression but Chromosomal Imbalance. Adv Radiat Oncol. 2019 Mar 14;4(3):466-472. doi: 10.1016/j.adro.2019.03.003.

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