

AMSER Rad/Path Case of the Month

9-year-old female with a 2-month history of
worsening headaches and vomiting

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Patient Presentation

- 9-year-old female with no significant PMH presented in the emergency department with a 2-month history of worsening headaches and vomiting. She had seen her PCP at the onset of her headaches and was diagnosed with migraines and started on magnesium without relief. Her headaches and vomiting became more severe and she began having difficulty in seeing the board at school which brought her into the ED.
- Family history is significant for migraines in her father.

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

Scenario	Scenario ID	Procedure	Adult RRL	Peds RRL	Appropriateness Category
Headache, secondary, initial exam	3081592	● MRI head without and with IV contrast	0 mSv O	0 mSv [ped] O	Usually appropriate
		● MRI head without IV contrast	0 mSv O	0 mSv [ped] O	Usually appropriate
		● MRA head without IV contrast	0 mSv O	0 mSv [ped] O	May be appropriate
		● MRV head without IV contrast	0 mSv O	0 mSv [ped] O	May be appropriate
		● CT head without IV contrast	1-10 mSv ☼☼☼	0.3-3 mSv [ped] ☼☼☼	May be appropriate
		● CTA head with IV contrast	1-10 mSv ☼☼☼	3-10 mSv [ped] ☼☼☼☼	May be appropriate
		● CTV head with IV contrast	1-10 mSv ☼☼☼	3-10 mSv [ped] ☼☼☼☼	May be appropriate
		● Radiography skull	<0.1 mSv ☼	0.03-0.3 mSv [ped] ☼☼	Usually not appropriate
		● Arteriography cerebral	1-10 mSv ☼☼☼	3-10 mSv [ped] ☼☼☼☼	Usually not appropriate
		● MRV head with IV contrast	0 mSv O	0 mSv [ped] O	Usually not appropriate
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This imaging modality was initially ordered by the ER physician

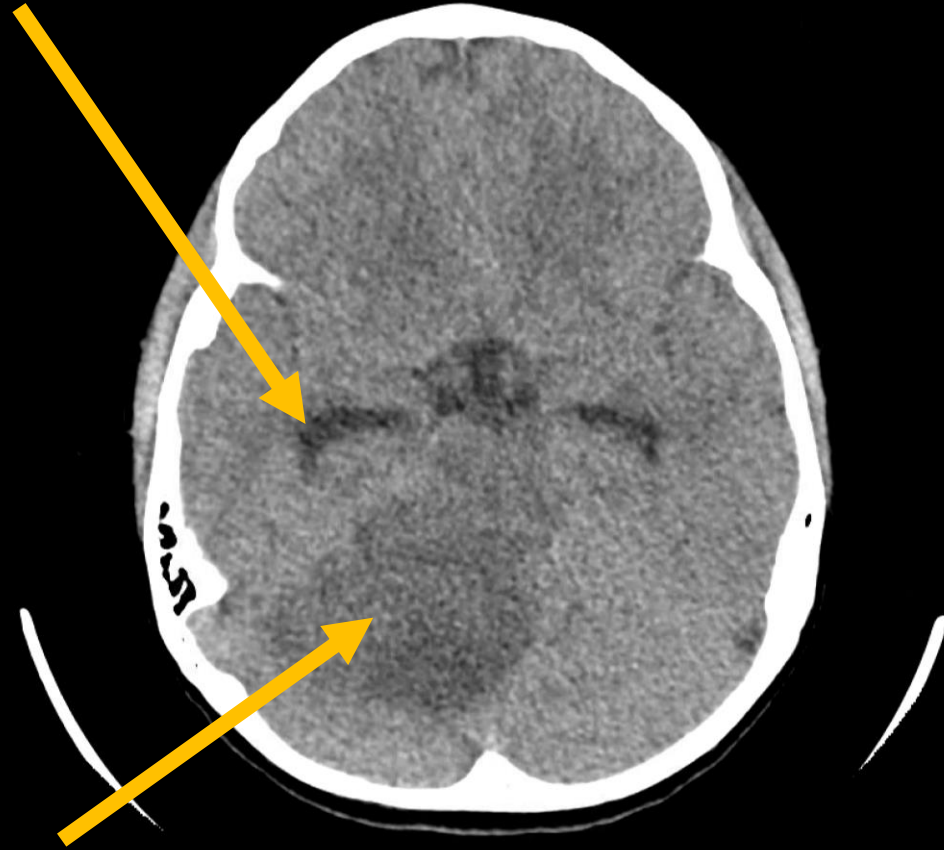
CT w/out Contrast



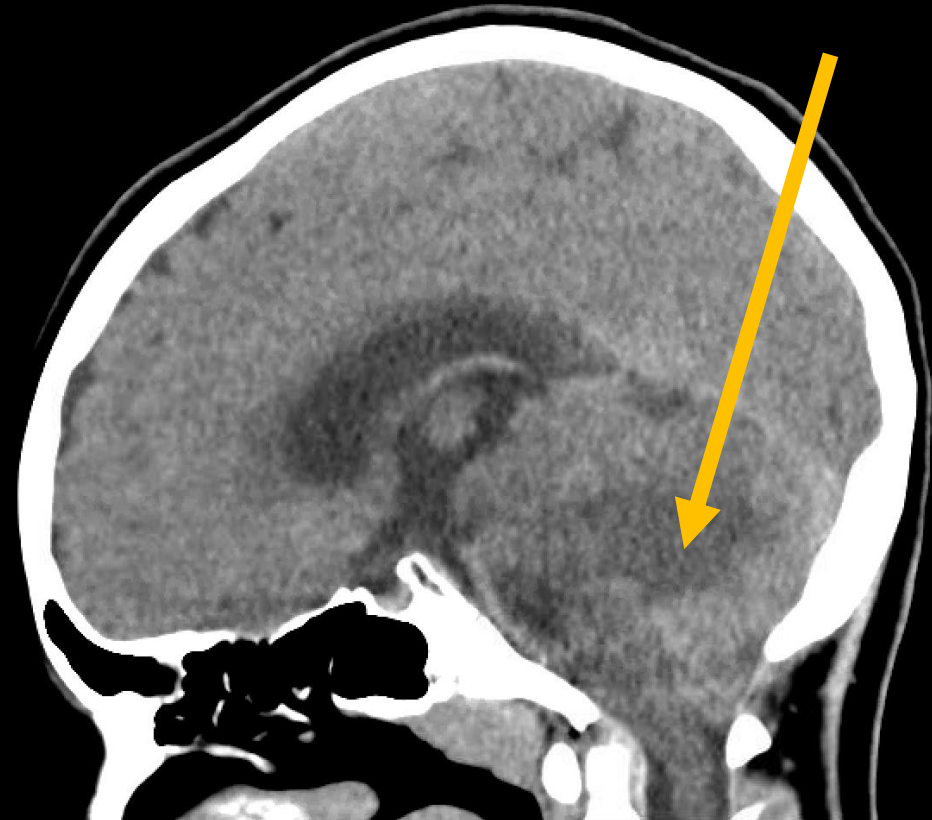
Dilation of temporal horns lateral ventricles due to obstructive hydrocephalus secondary to effacement of 4th ventricle by mass.

CT w/out Contrast

Hypodense mass extending into the pons with compression of the cerebral aqueduct affecting the 4th ventricle



5 cm hypodense mass extending into the right cerebellum.

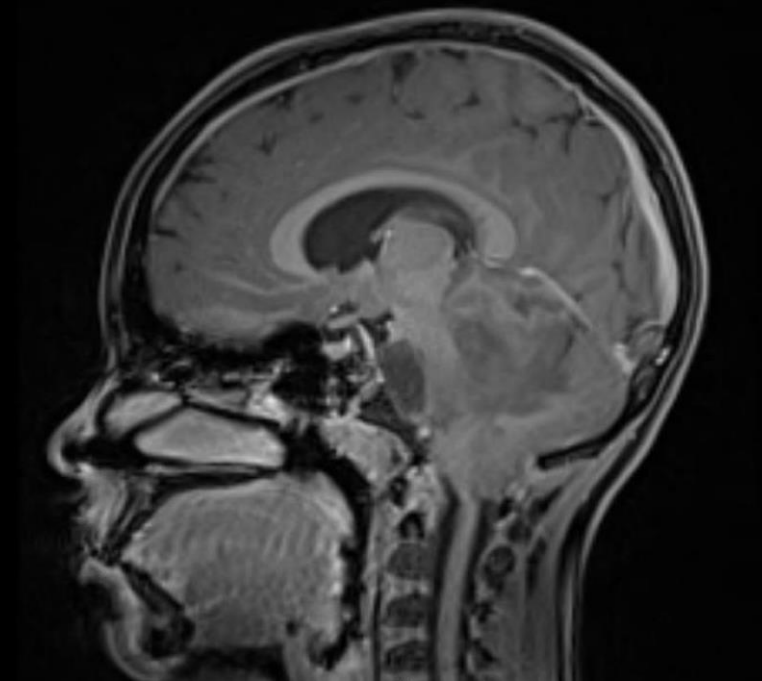
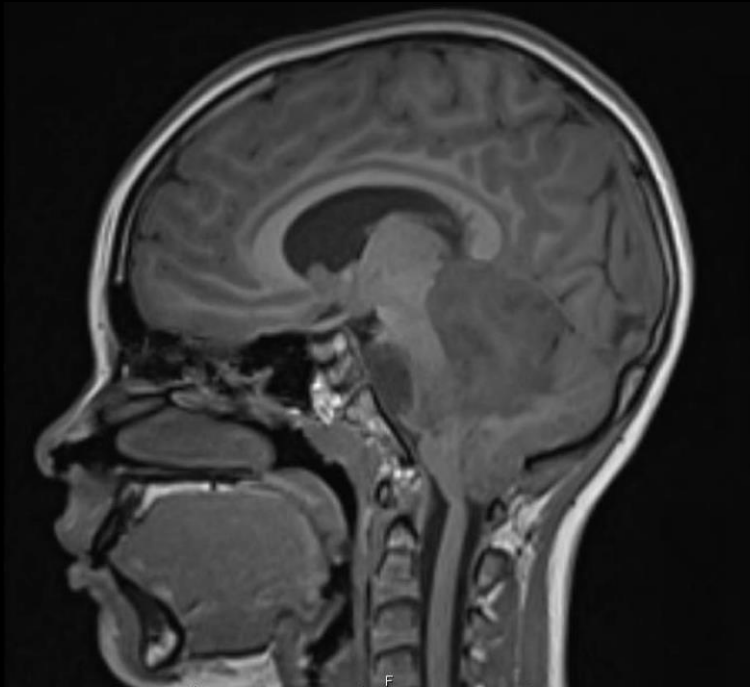


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This imaging was ordered as a follow up to the CT without contrast

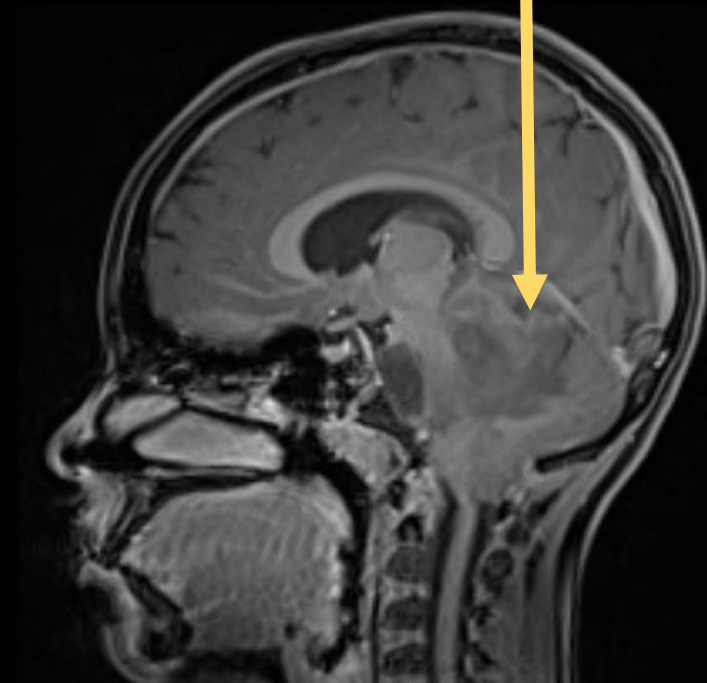
MRI Pre (Left) and Post (Right) Gadolinium



MRI Pre (Left) and Post (Right) Gadolinium

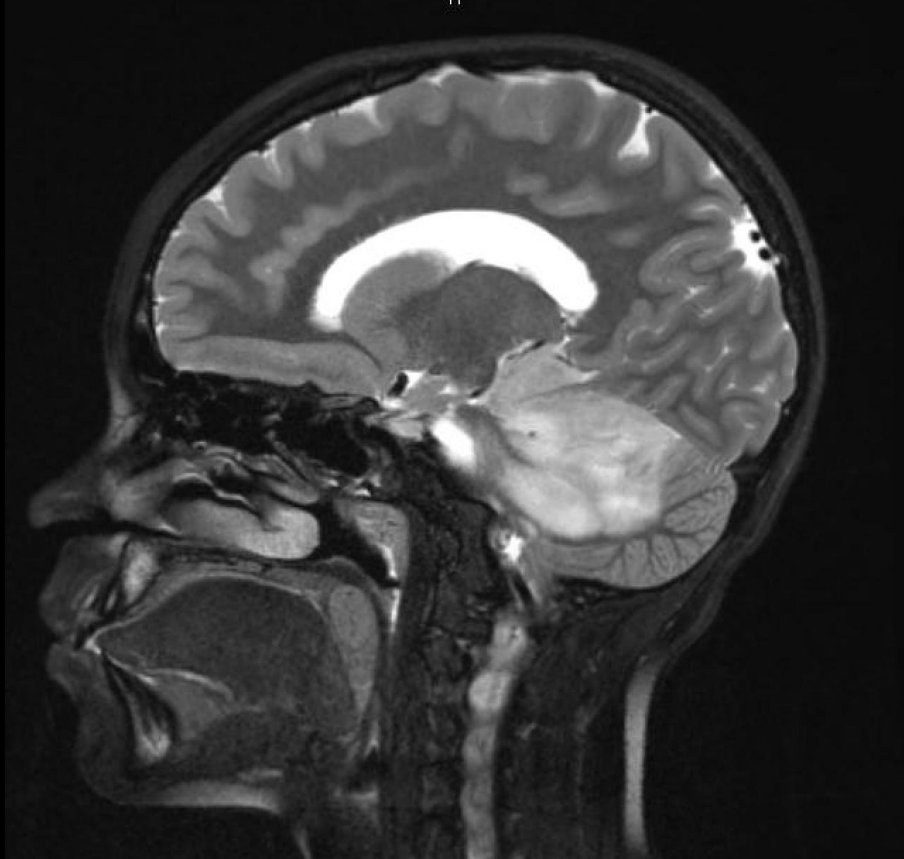


Hypointense posterior fossa lesion involving both the cerebellum and brainstem with effacement of the 4th ventricle .

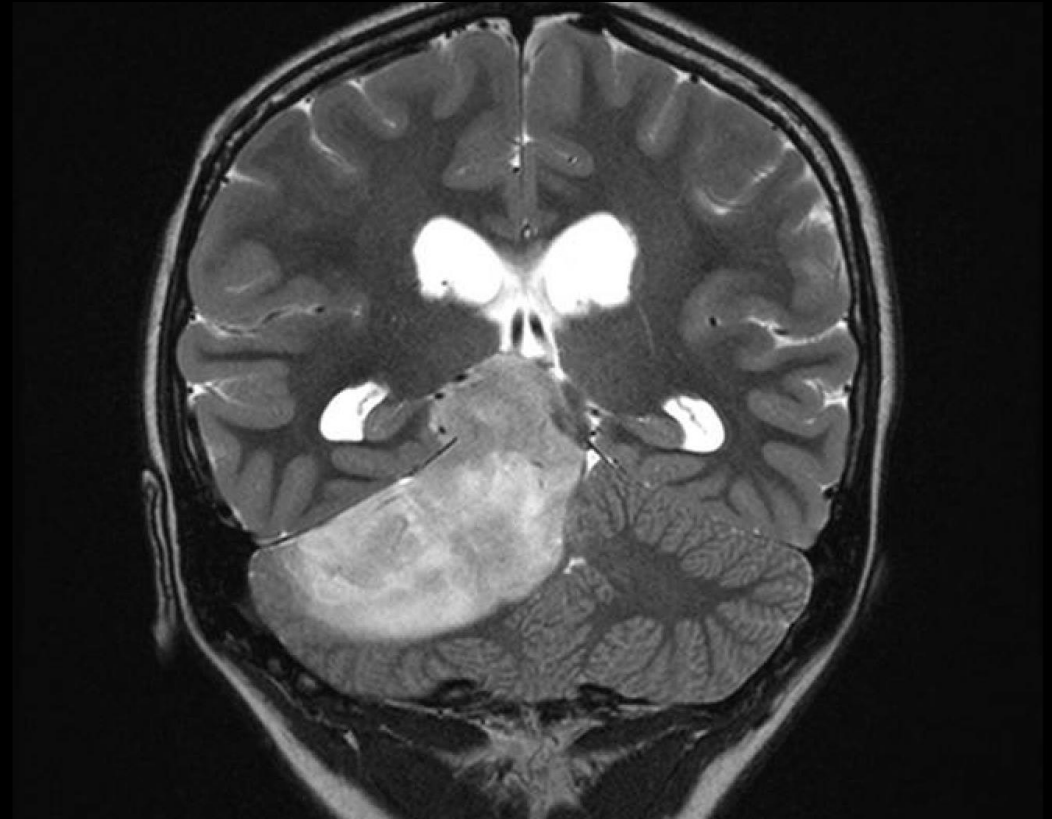


Post contrast enhancement supports neoplasm.

MR T2

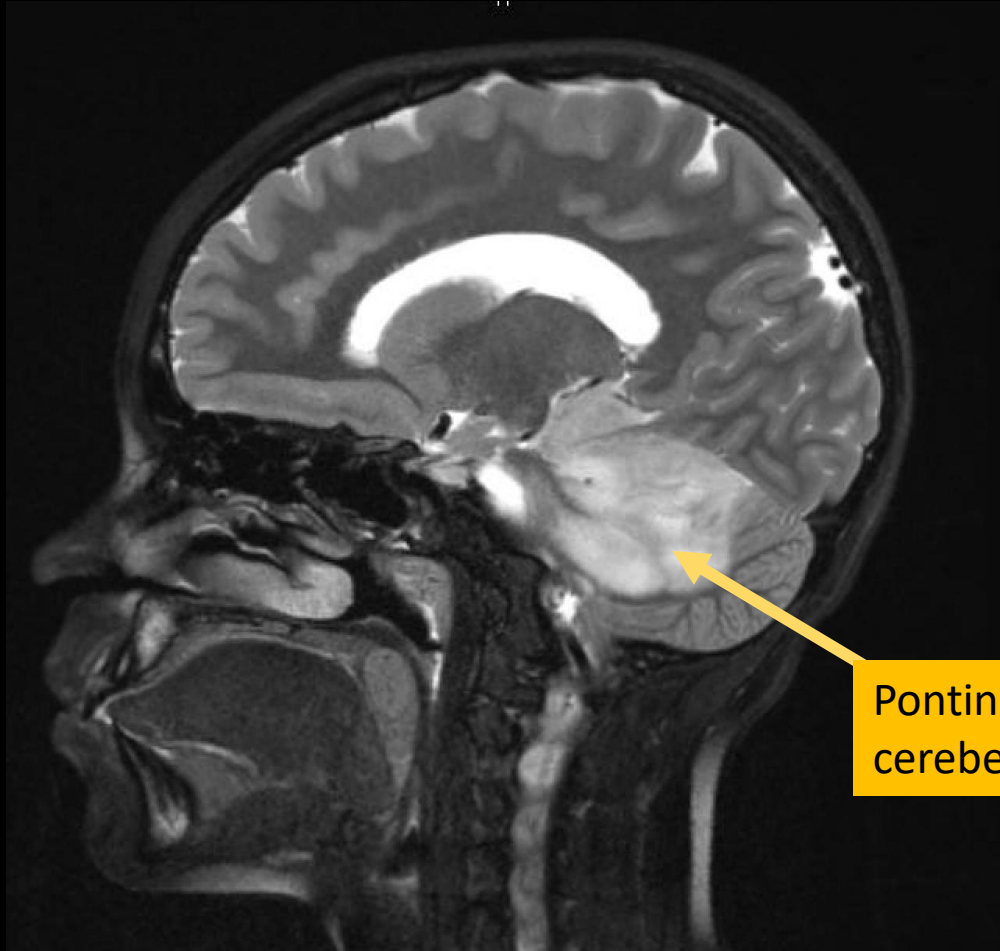


Sagittal

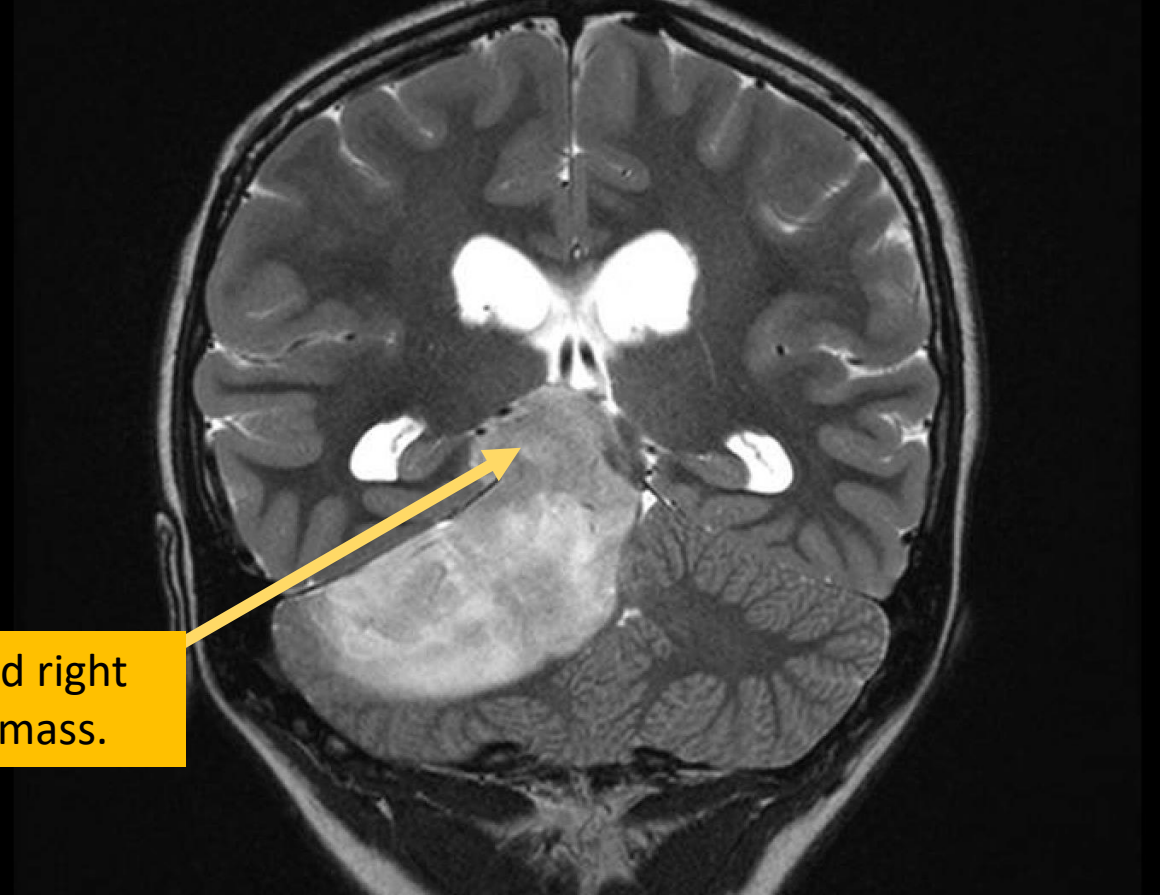


Coronal

MR T2



Sagittal



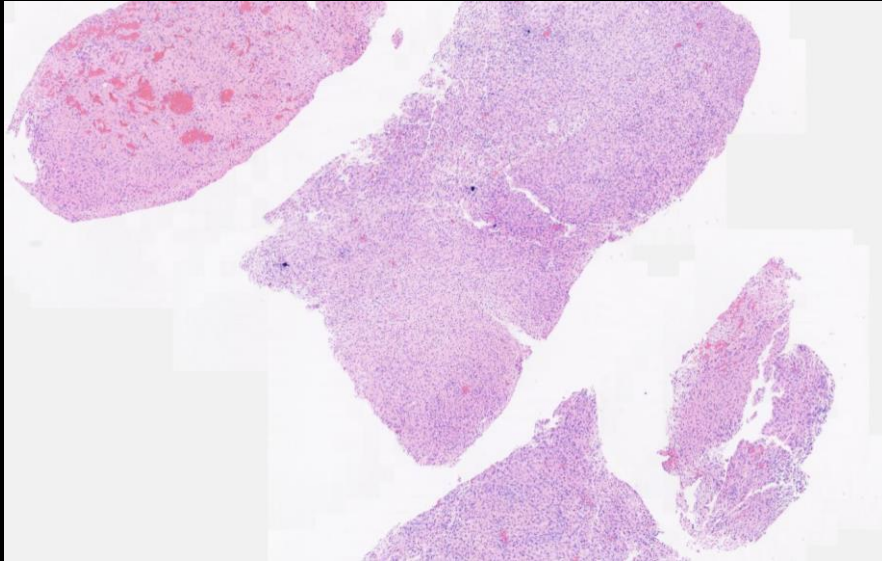
Coronal

Pontine and right cerebellar mass.

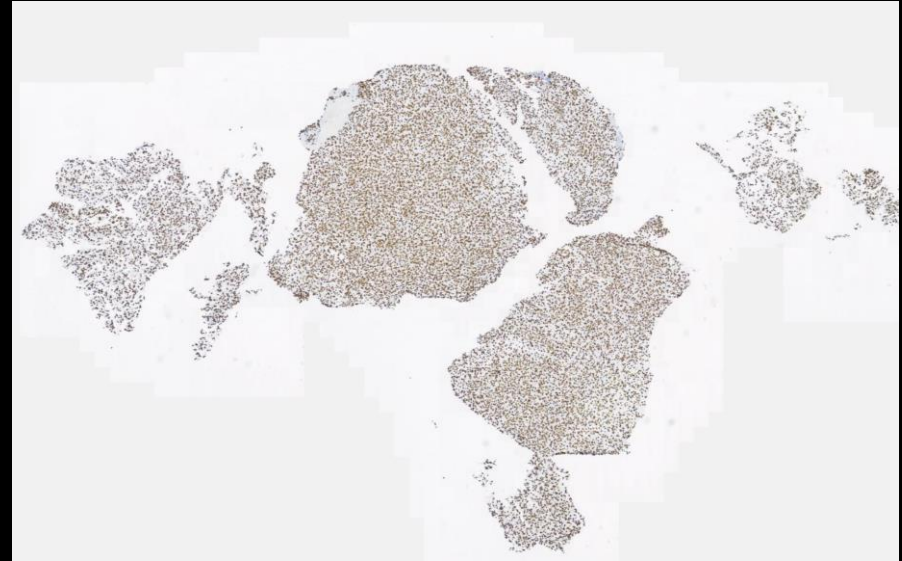
DDX

- Astrocytoma
- Midline Glioma
- Ependymoma

Pathology



H&E Stain demonstrating densely packed cells and increased mitotic activity.



H3K27 immunohistochemical stain positive in nuclei of cells and demonstrates significant proliferation. This supports diagnosis of high-grade glioma.

Final Dx:

Diffuse Midline Glioma

Diffuse Midline Glioma

- **Demographics:**

- 9% of pediatric brain tumors are high grade gliomas with the majority occurring in patients 5-9 years old

- **Risk Factors:**

- Prior radiation exposure
- Genetic predisposition (Li-Fraumeni, NF1, Mismatch repair deficiency)

Diffuse Midline Glioma

- **Presentation:**

- Location dependent, however most common initial signs include headache, vomiting, fatigue, and seizures

- **Imaging:**

T1 reveals a hypointense lesion

T2 shows hyperintense lesion with irregular border and infiltration

- **Pathology/Histology:**

Increased cellularity and mitotic activity

Most common is an H3K27 mutation

Treatment

Focal radiation and chemotherapy

Surgical resection is considered depending on various factors (location, structural involvement, infiltration)

Despite therapy median survival is 18 months

This patient specifically did not qualify for surgical resection due to depth of invasion

References:

1. Louis DN, Perry A. Classification and pathologic diagnosis of gliomas, glioneuronal tumors, and neuronal tumors. UpToDate. Waltham, MA: UpToDate Inc.
<https://www.uptodate.com/contents/classification-and-pathologic-diagnosis-of-gliomas-glioneuronal-tumors-and-neuronal-tumors>. Accessed June 24, 2025.
2. Al Sharie S, Abu Laban D, Al-Hussaini M. Decoding diffuse midline gliomas: A comprehensive review of pathogenesis, diagnosis and treatment. Cancers. October 6, 2023.
<https://pmc.ncbi.nlm.nih.gov/articles/PMC10571999/#sec6-cancers-15-04869>. Accessed August 13, 2025.
3. Packer RJ, et al. Epidemiology and classification of central nervous system tumors in children. *UpToDate*. Waltham, MA: UpToDate Inc; published approximately 1.8 years ago.
<https://www.uptodate.com/contents/epidemiology-and-classification-of-central-nervous-system-tumors-in-children>. Accessed June 24, 2025.
4. AMBOSS. Brain tumors. AMBOSS Medical Knowledge. Last updated February 4, 2025.
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