SENRS 2024

A rare intraventricular mimic

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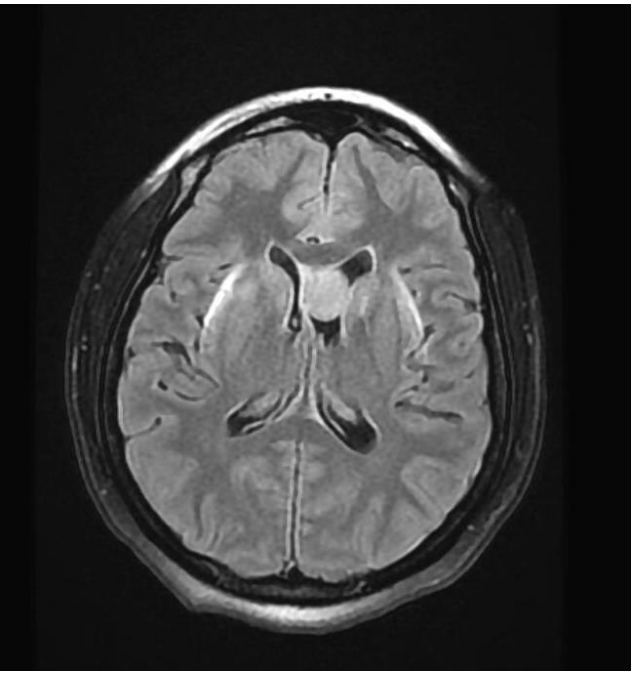
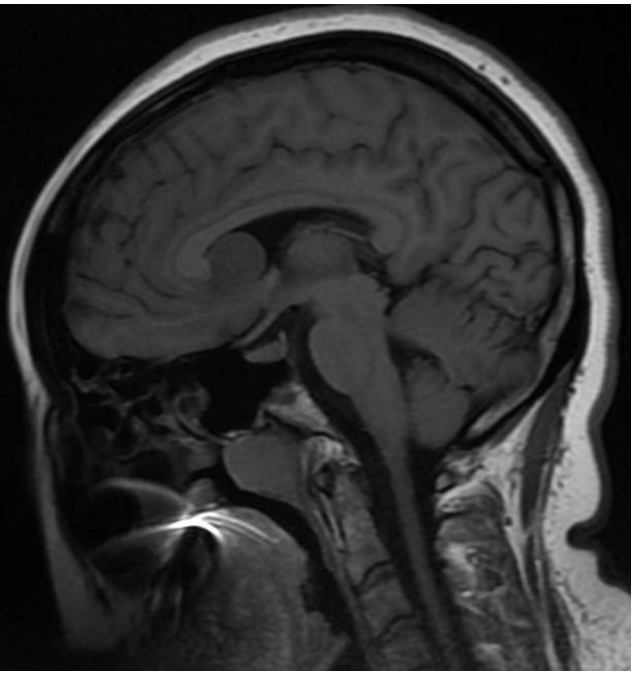
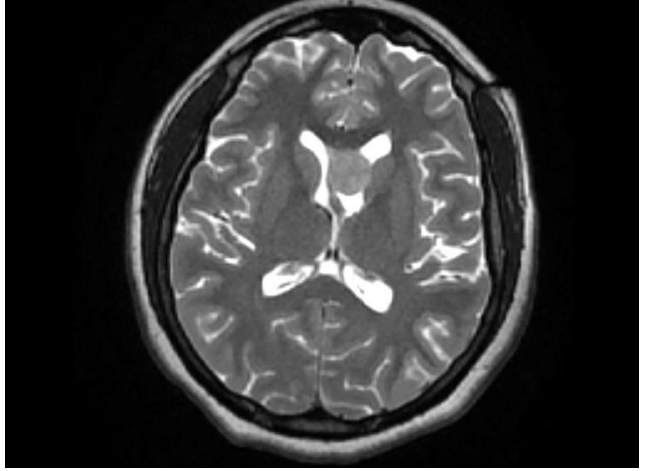
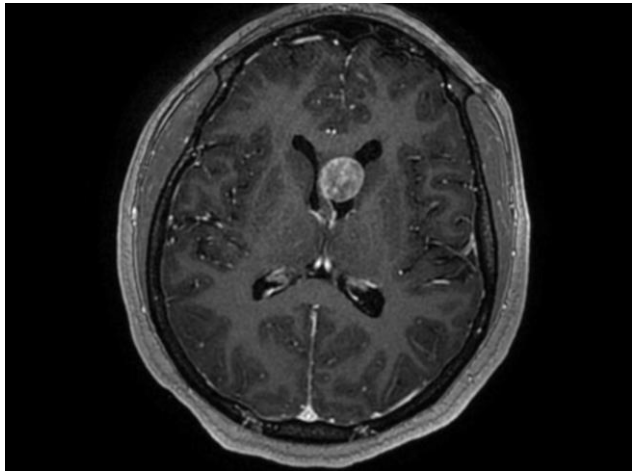
Presentation

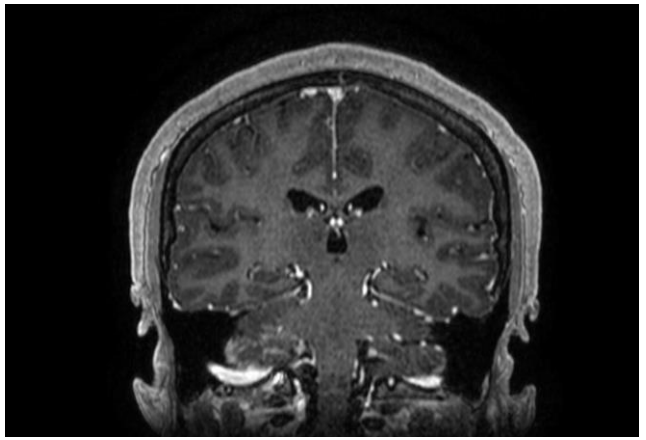
- 25-year-old female with history of hypertension, asthma, and migraines.
- No prior surgical history.
- Outside imaging (images not available):
  - 2018: Reported as normal
  - May 2024: Left intraventricular mass, surgery planned for July 2024.
- Presented in June 2024 acutely to emergency room with persistent headaches (stabbing and tension-like), nausea and vomiting, double vision, intermittent dizziness, and generalized weakness.
- Hypertension on physical exam.
- New imaging obtained.









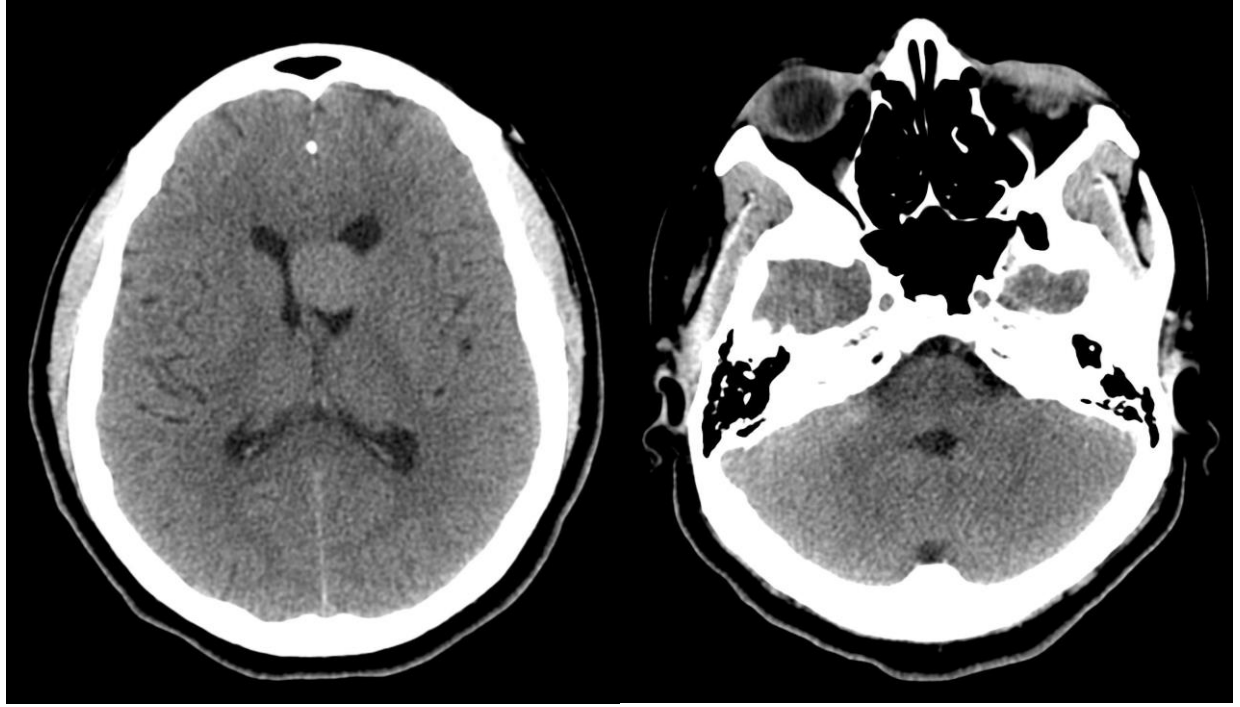


Imaging 6/7

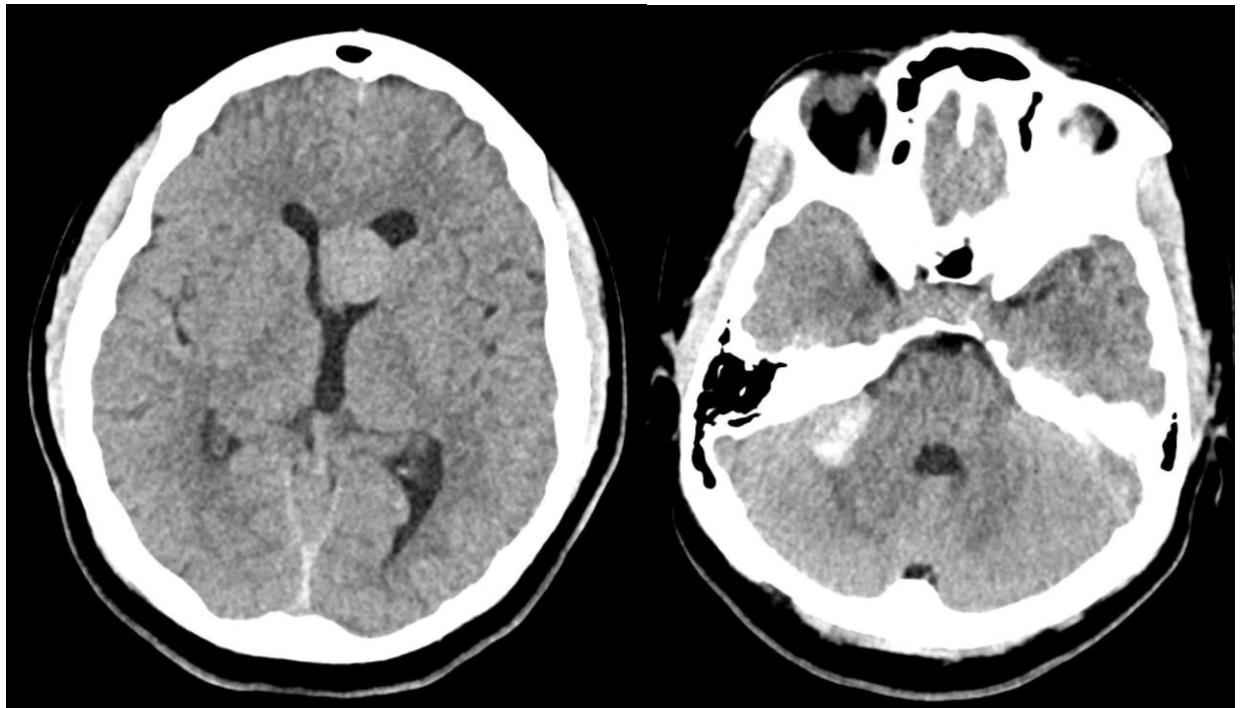
- Intraventricular mass along the left aspect of the septum
- FLAIR hyperintense to gray matter
- T2 isointense to gray matter
- T1 hypointense
- Heterogeneously enhancing
- Leptomeningeal enhancement at the right cerebellum

CT imaging

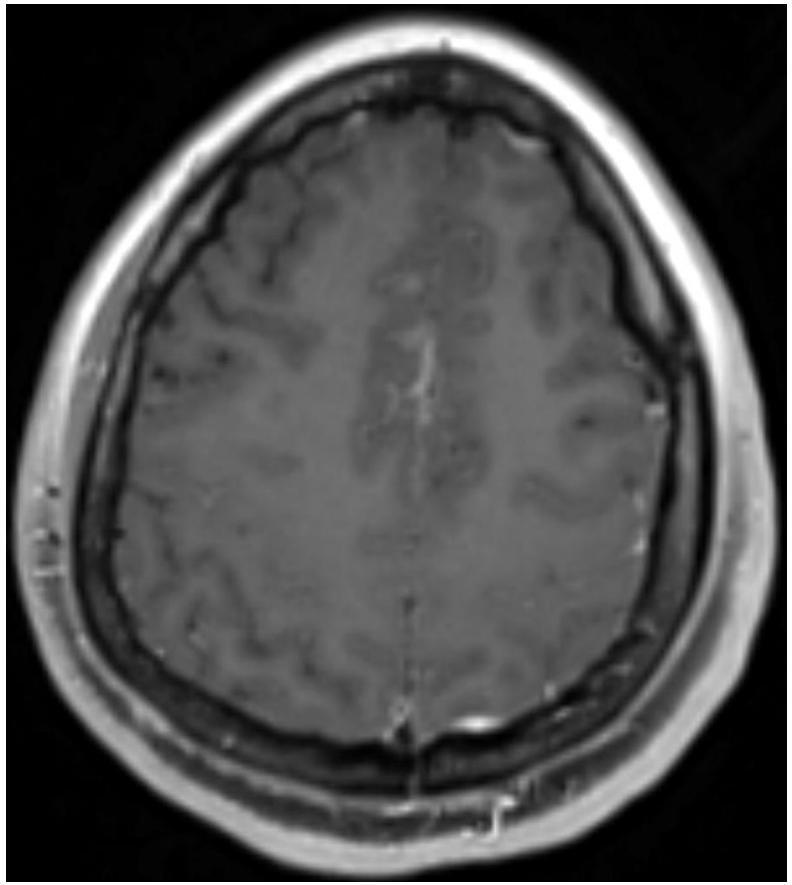
- 6/22 (top)
  - CT hyperdense
  - Right cerebellar hyperdensity

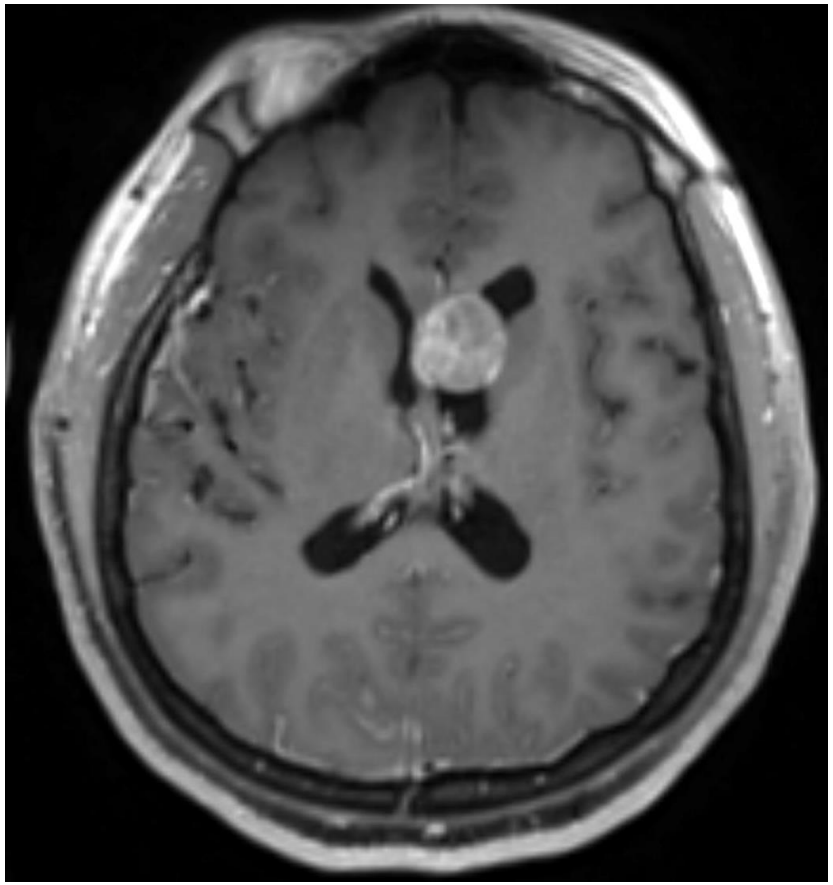


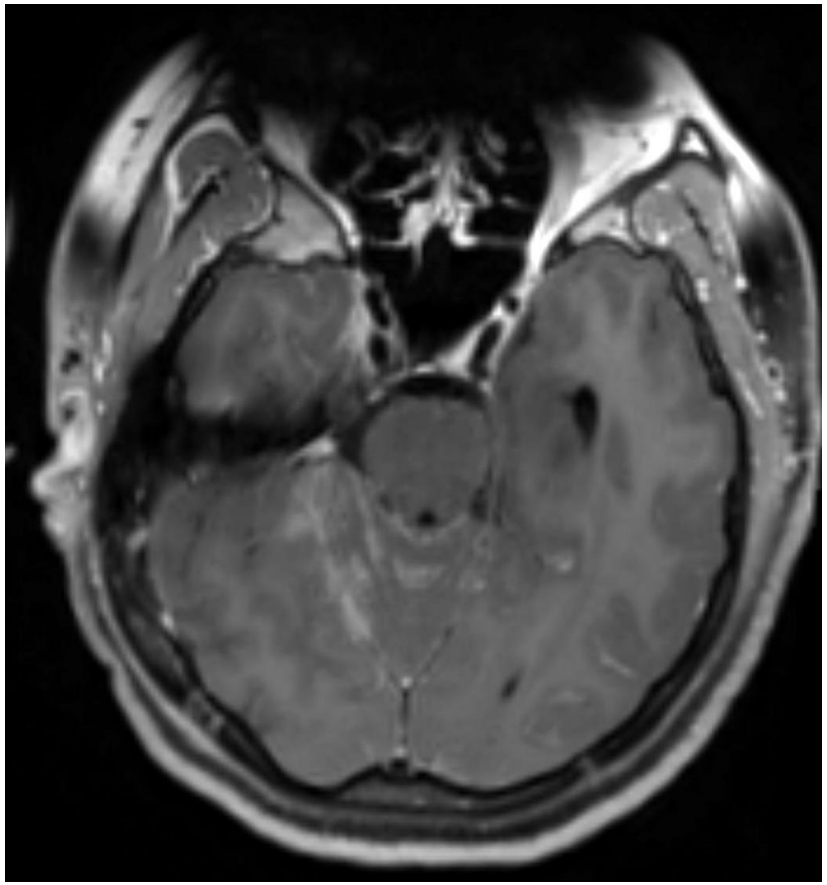
- 6/24 (bottom)
  - Right cerebellar hemorrhage



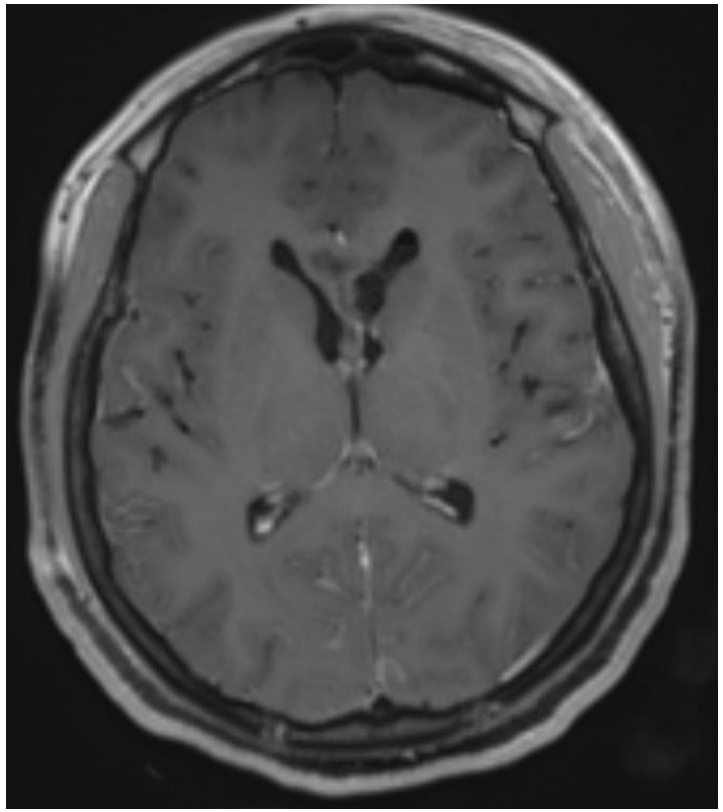
Imaging 6/25







Post-operative imaging



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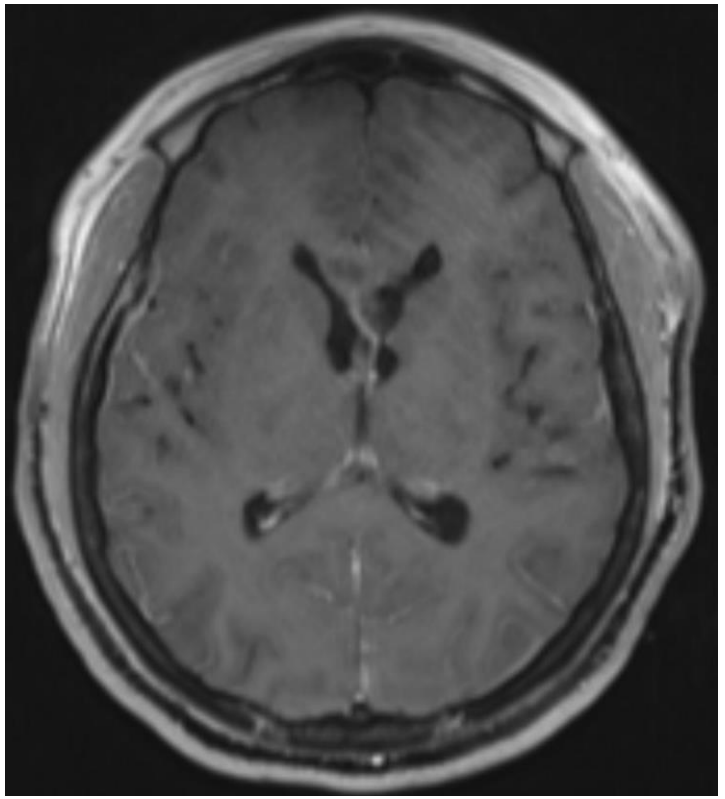

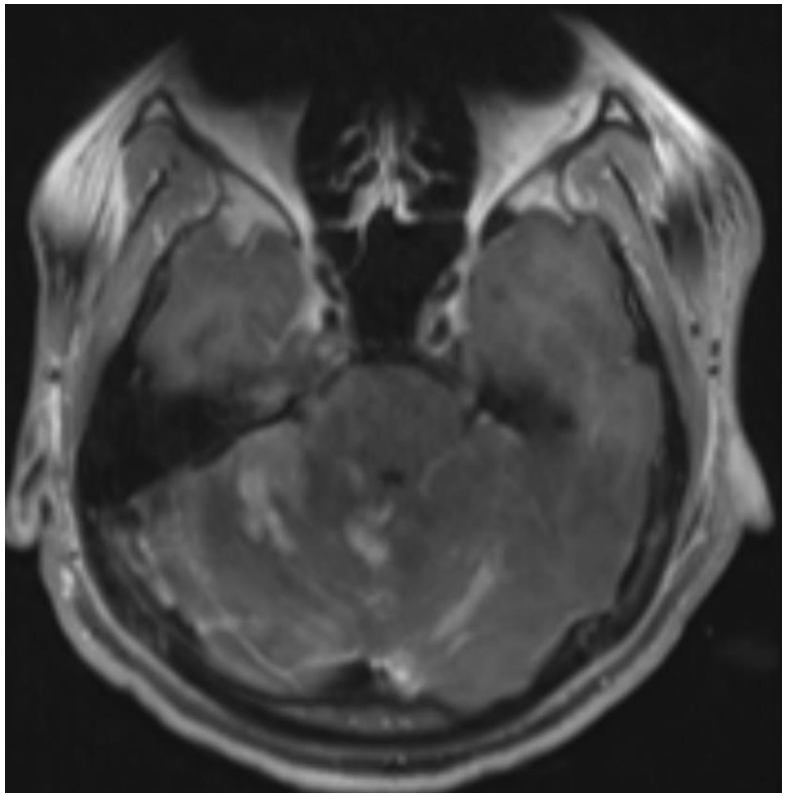

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Findings

Intraventricular tumor with initially benign features

- Meningioma
- Central neurocytoma
- Ependymoma or subependymoma

Rapid progression of features

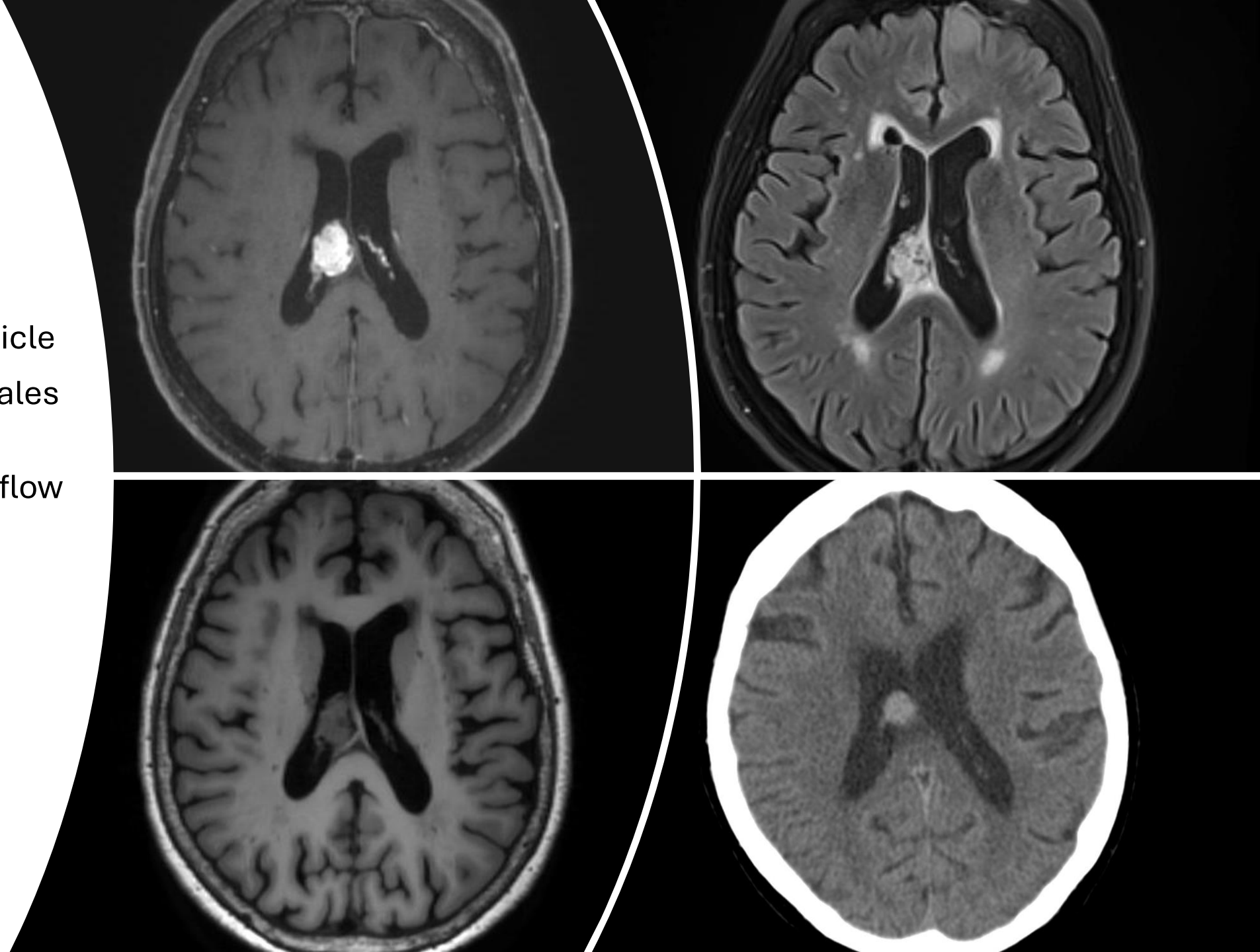
- Leptomeningeal enhancement
- Hemorrhage

New differential diagnosis

- High grade glioma

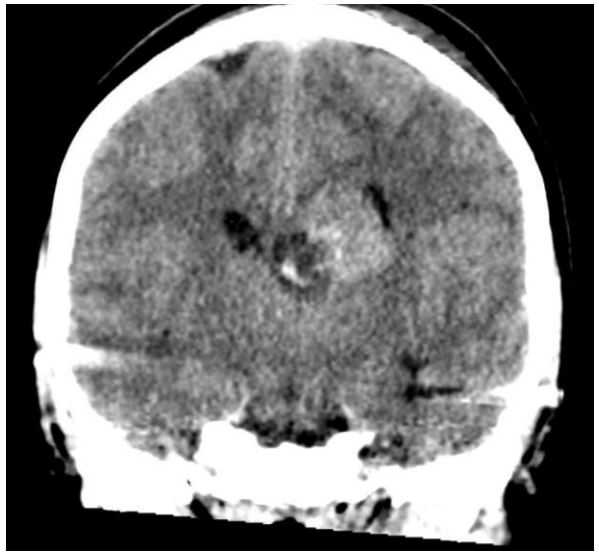
Differential: meningioma

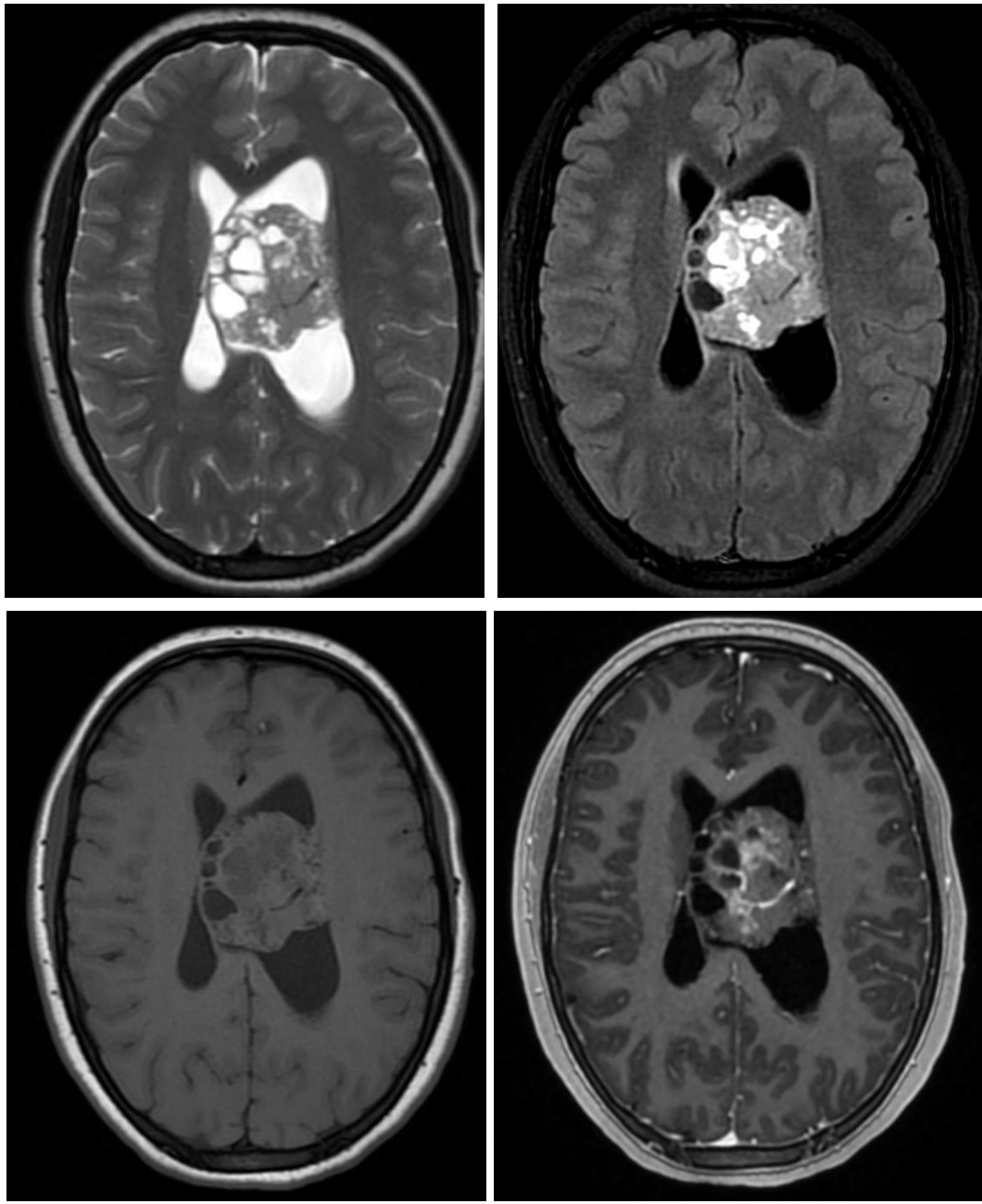
- Atrium of lateral ventricle
- More common in females
- Can calcify and have transependymal CSF flow
- Avid enhancement
- T2 iso-hyperintense
- T1 iso-hypointense
- May restrict diffusion
- CT iso-hyperdense



Differential: central neurocytoma

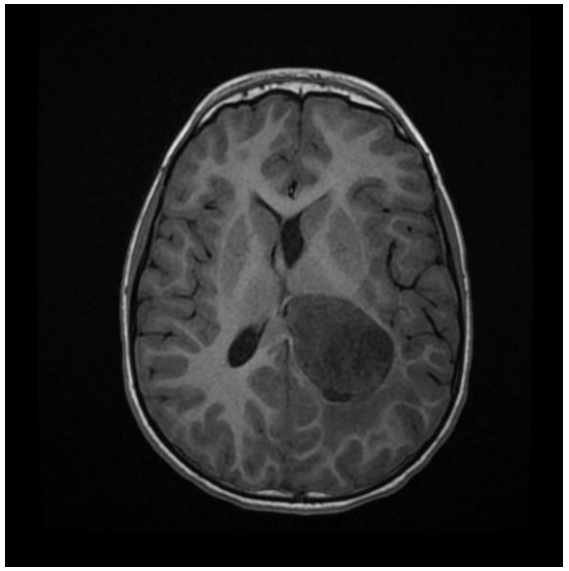
- Lateral ventricle at the septum
- Can calcify and hemorrhage
- Lobulated, cystic spaces
- Surgery is curative, rare recurrence
- CT hyperdense
- T2/FLAIR hyperintense
- T1 isointense to gray matter
- Variable enhancement
- May have flow voids

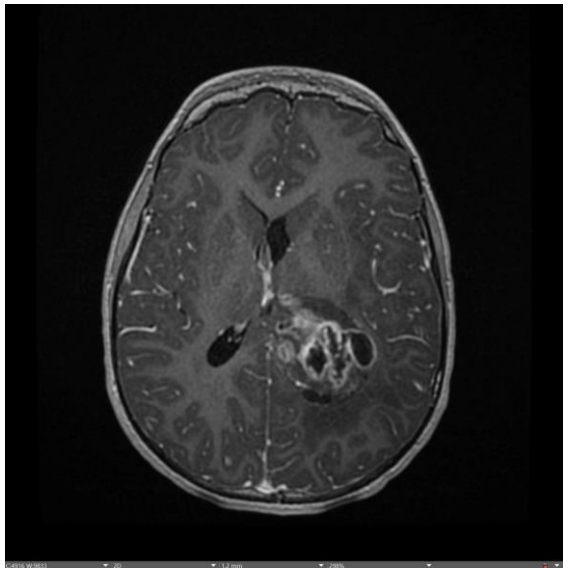


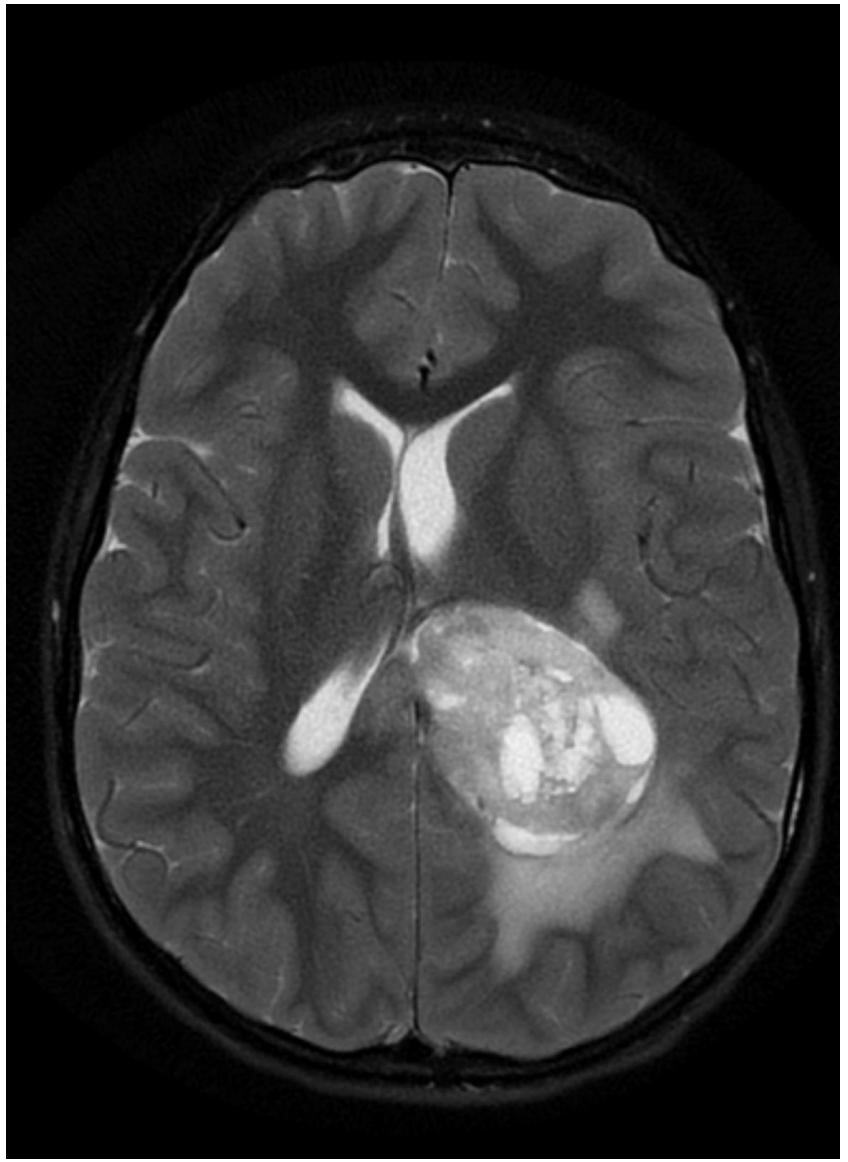


Differential: ependymoma

- 40% are supratentorial, less than half of those are intraventricular (often 4th ventricle)
- Difficult to resect and can recur
- Can have calcification and hemorrhage and cause adjacent edema
- T1 iso-hypointense
- Heterogenous enhancement
- T2 iso-hyperintense
- Variable diffusion
- CT iso-hypodense (soft tissue component)

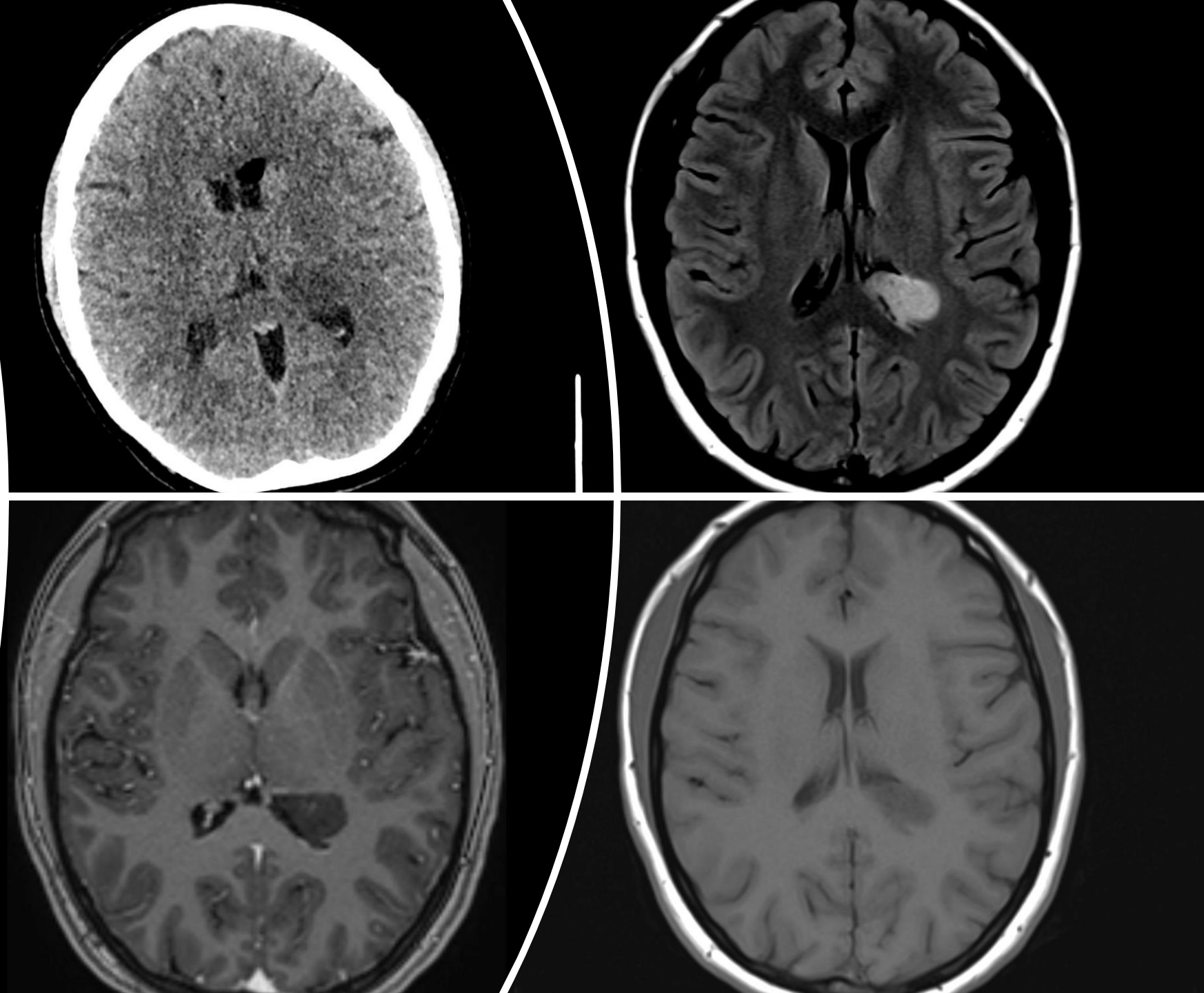






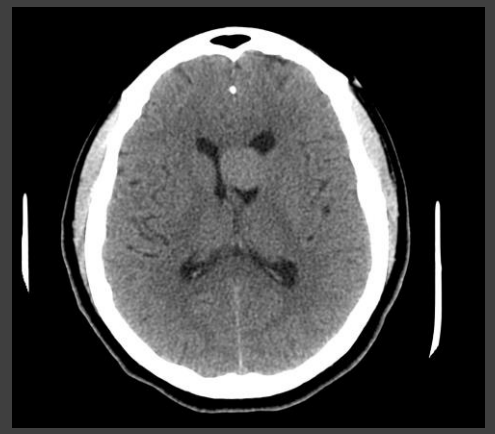
Differential: subependymoma

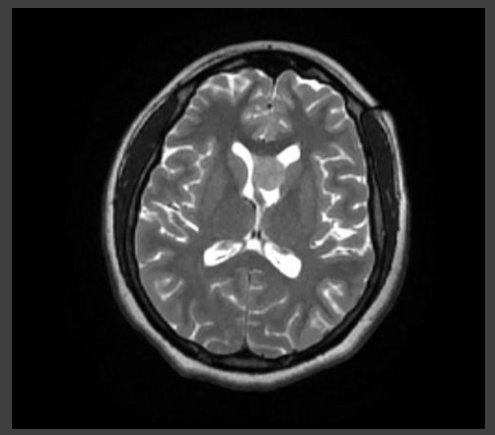
- Intraventricular (lateral ventricles in 30-40%)
- Good prognosis, rarely recurs after resection, asymptomatic unless large
- Heterogenous, blood product and calcification may occur
- CT iso-hypodense
- T2/FLAIR hyperintense
- Variable enhancement
- T1 iso-hypointense to white matter

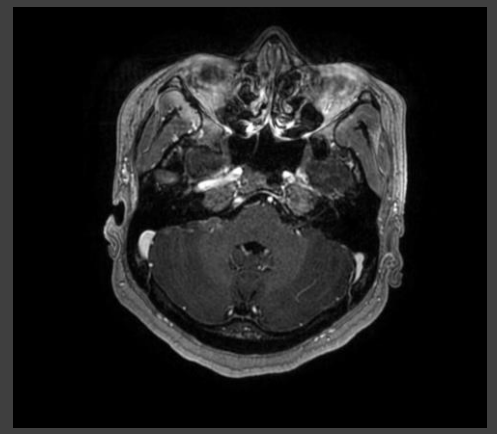


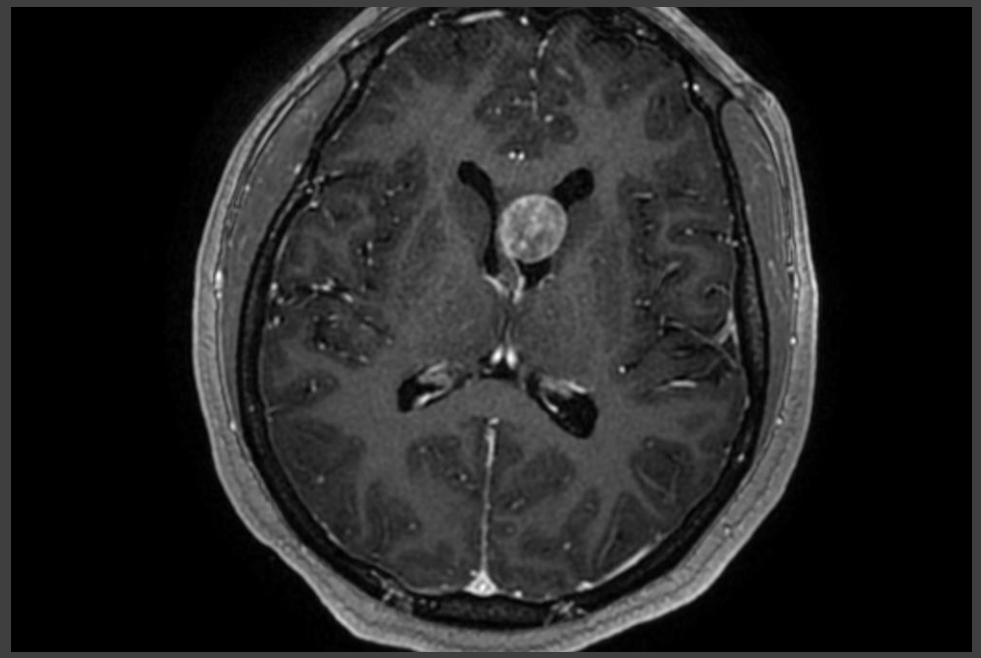
DIAGONSIS: epithelioid glioblastoma (eGBM)

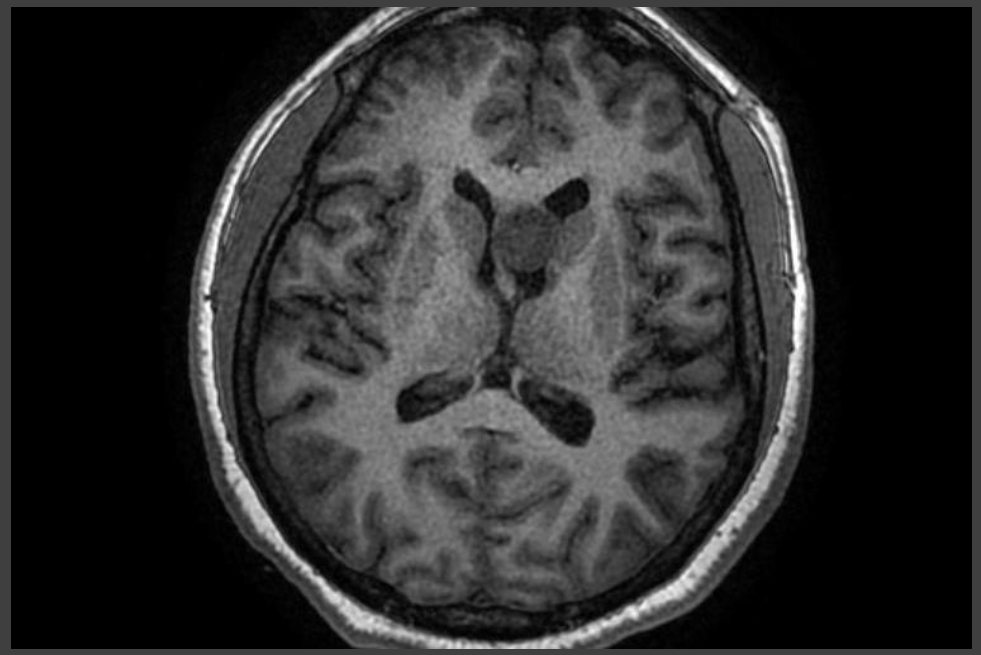
- Glioblastoma is rare in the ventricles
- Epithelioid glioblastoma is rare with previous case studies demonstrating cortical eGBM, making this case very rare
- Some reports demonstrate transformation from more benign lesions such as pilocytic astrocytoma
- CT hypodense per literature, our case demonstrates iso-hyperdensity
- T2 hyperintense
- T1 iso-hypointense
- Can see mild edema in cortical cases
- Watch for aggressive features
  - Irregular heterogenous/ring enhancement
  - Infiltrative margins
- CSF was positive for high grade neoplastic cells, MGMT negative





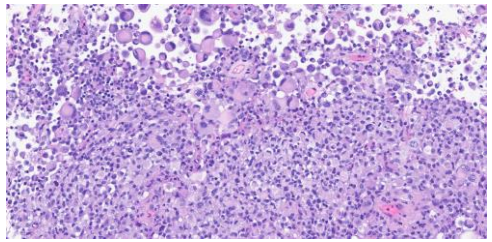
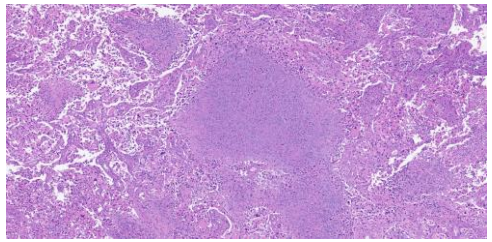
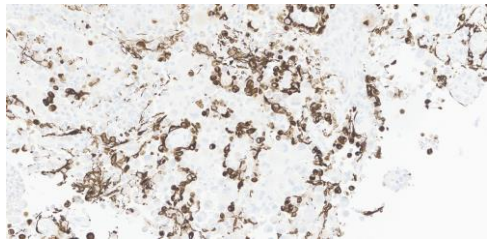
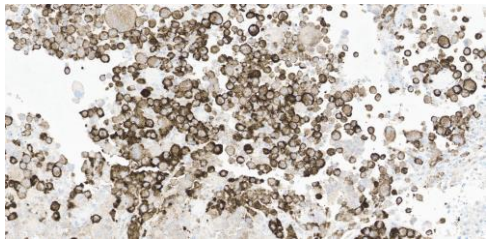






Histology

Epithelioid glioblastoma, WHO grade 4, focal desmoplasia without convincing sarcomatous element, positive BRAF (V600E), GFAP, OLIG2. p53 shows wildtype expression pattern.

H&E stain, 250x	H&E stain, 36x	Glial fibrillary acidic protein (GFAP) immunostain, 250x	BRAF V600E immunostain, 200x
Patternless sheets of malignant epithelioid cells with loss of cohesion (upper). No discernible fibrillarity as expected in a glioma.	Central necrosis, indicating high grade tumor	A subpopulation of tumor cells are positive for the marker, with most of the cells being negative, typical for epithelioid GBM	Positive
			

Management

Due to the patient age and acute presentation, surgery was most appropriate with plans for chemotherapy and radiation.

In the literature and historically, glioblastoma is best treated with surgery, chemotherapy, radiation and close follow-up with expectation for recurrence.

Because of CSF seeding, imaging of the spine is also necessary.

Outcome

- Unfortunately, due to the presentation with hemorrhage, hypertension and aggressivity of the disease, the patient declined despite surgery with increased bleeding followed by significant intracranial edema and multiple territory infarction. After much discussion, palliative care was chosen as the next step in management.

- In the literature, glioblastoma has a median survival of 2 years
- In a case series of 7 cases of cortical eGBM, average survival was around 10 months.

Take home points

- Intraventricular epithelioid glioblastoma is very rare.
  - Epithelioid glioblastoma has previously been described as a rare subtype of glioblastoma that occurs predominantly cortically, with one case report of intraventricular eGBM that we are aware of.
- Initially it can be indistinguishable from benign tumors and should therefore be considered in the differential.
- Look for aggressive or atypical features: subtle leptomeningeal enhancement, heterogeneity, irregular margins, and growth.
- Consider early CSF analysis if surgery or biopsy are going to be delayed.

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