

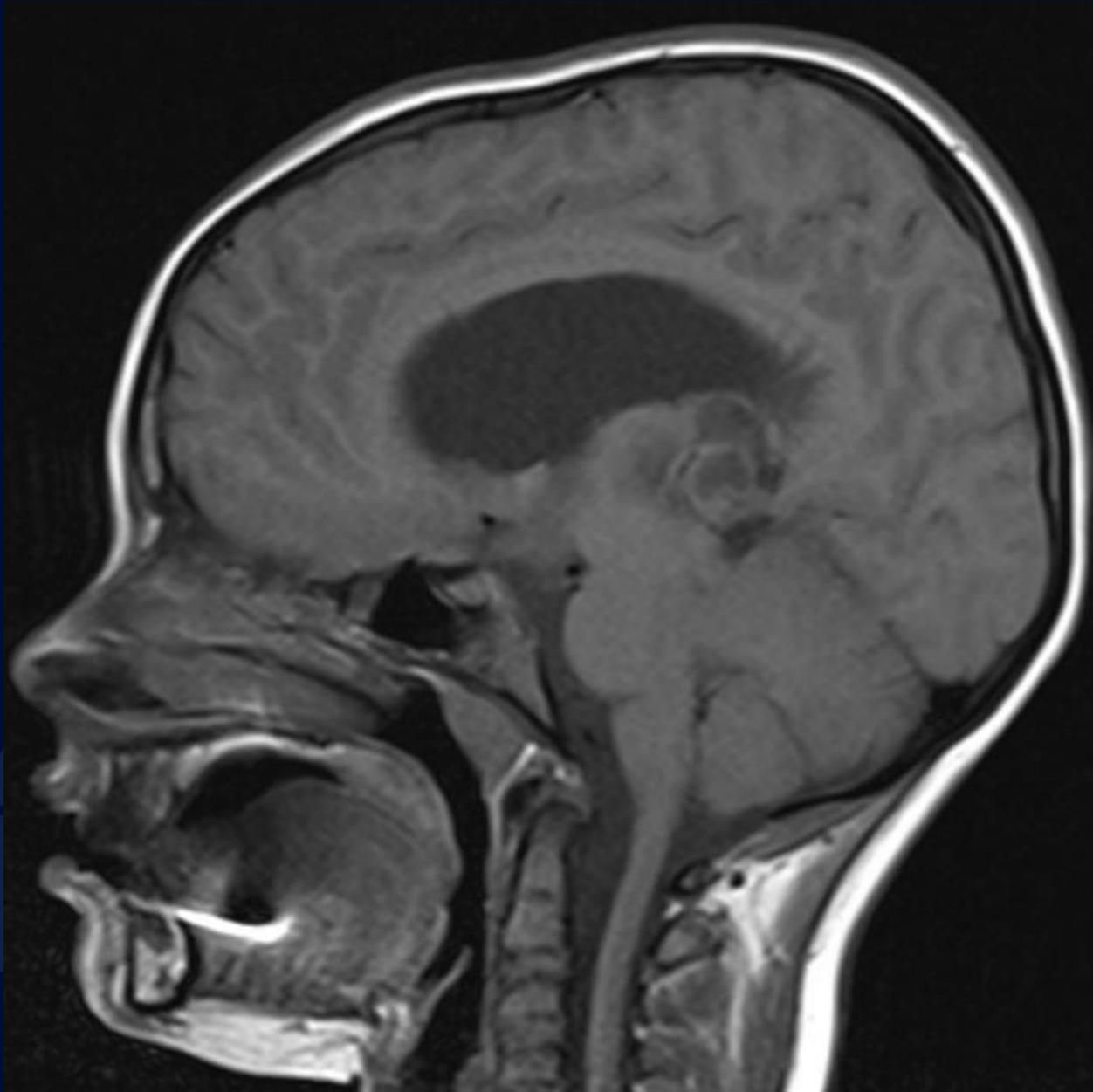
A large, stylized leaf graphic in a dark blue color, positioned on the left side of the slide, partially overlapping the text.

# 9-year-old male presenting with worsening headache and vomiting

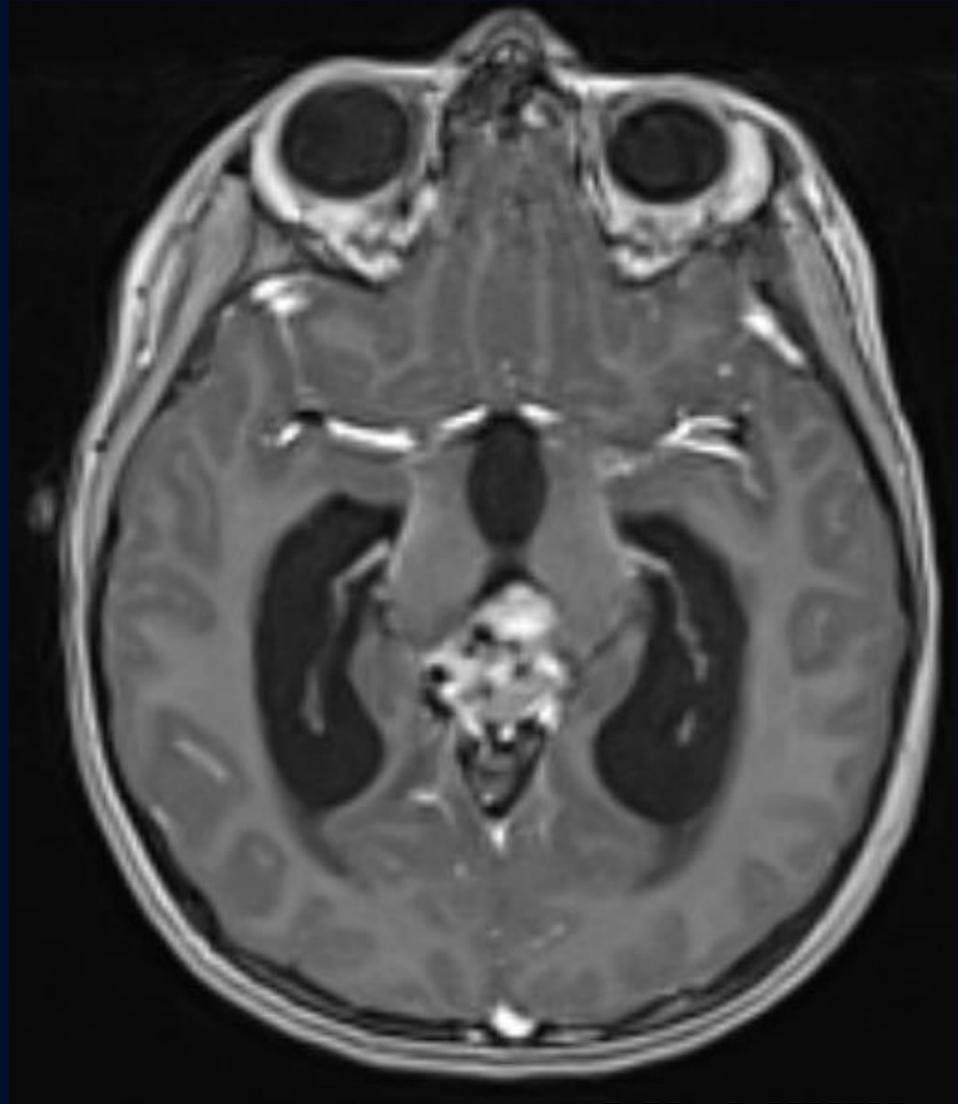
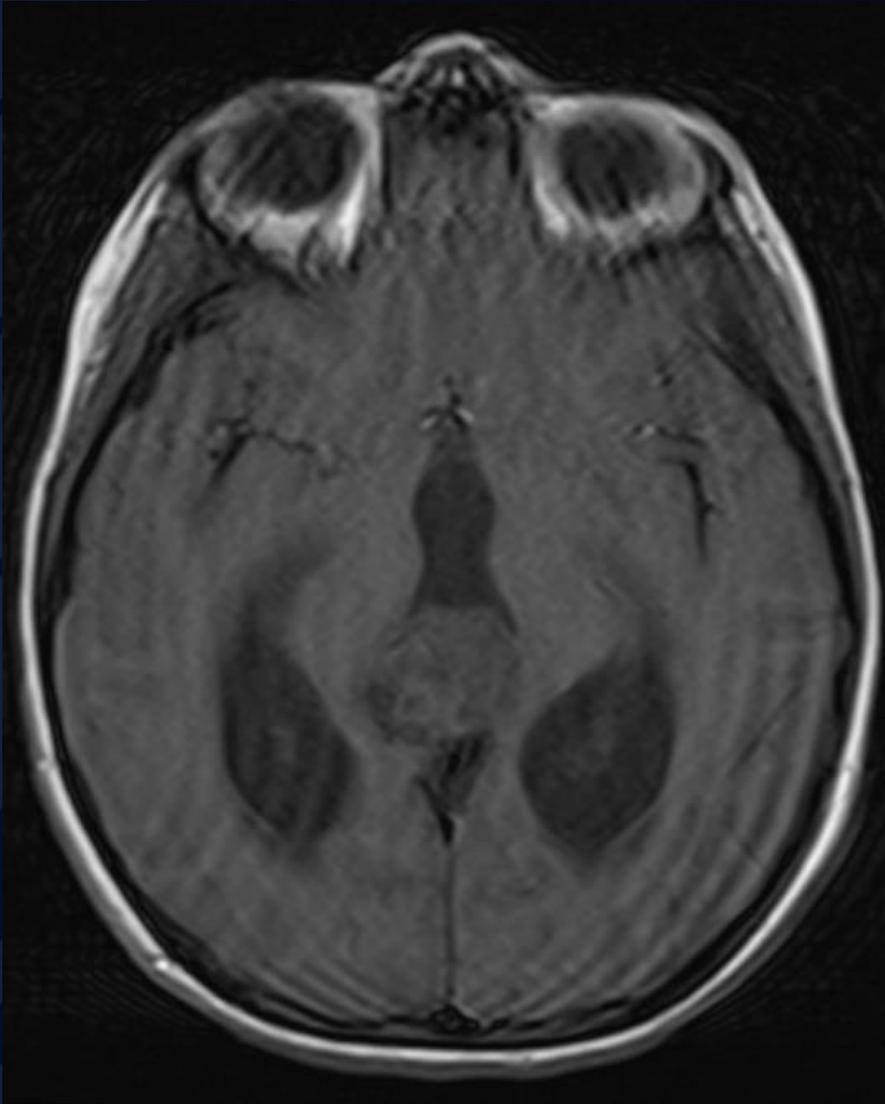
Joseph Masison

Connecticut Children's Medical Center

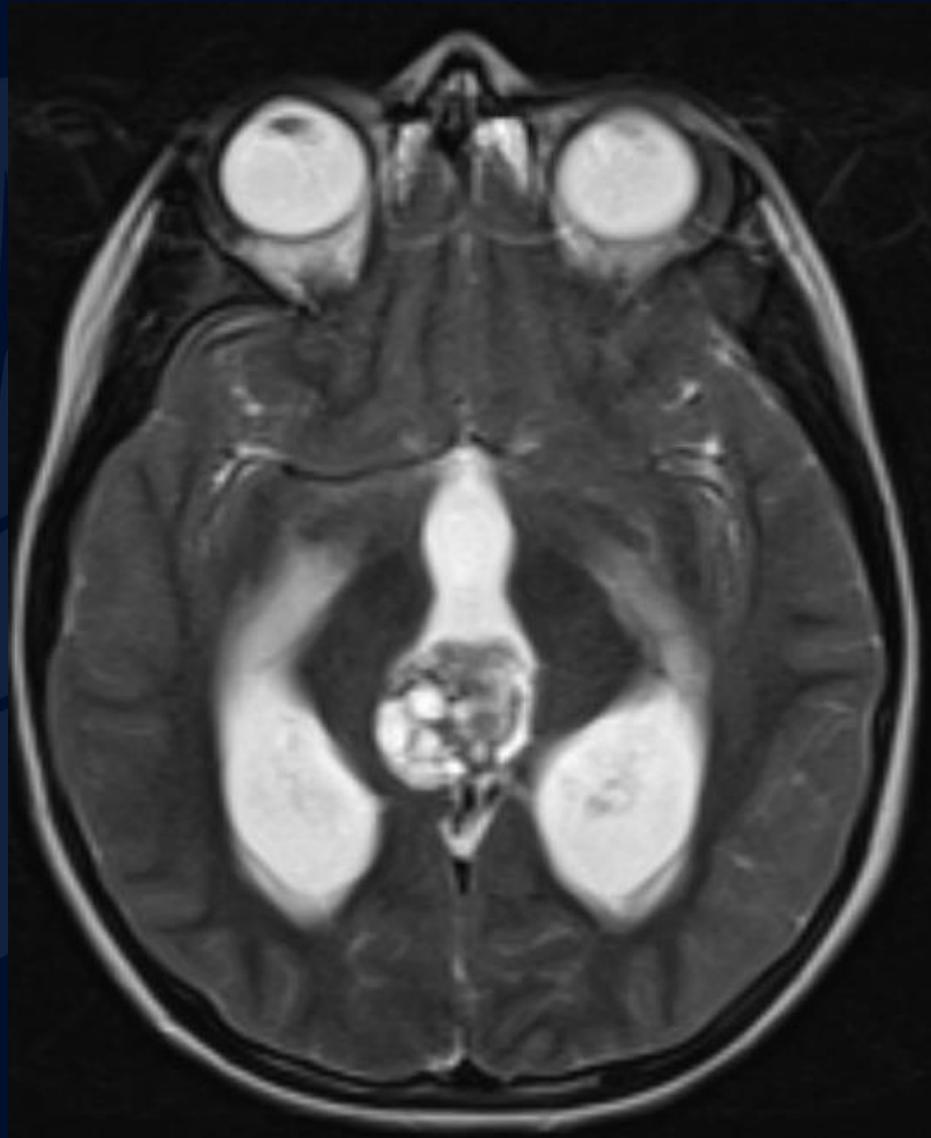
# MR Sagittal T1



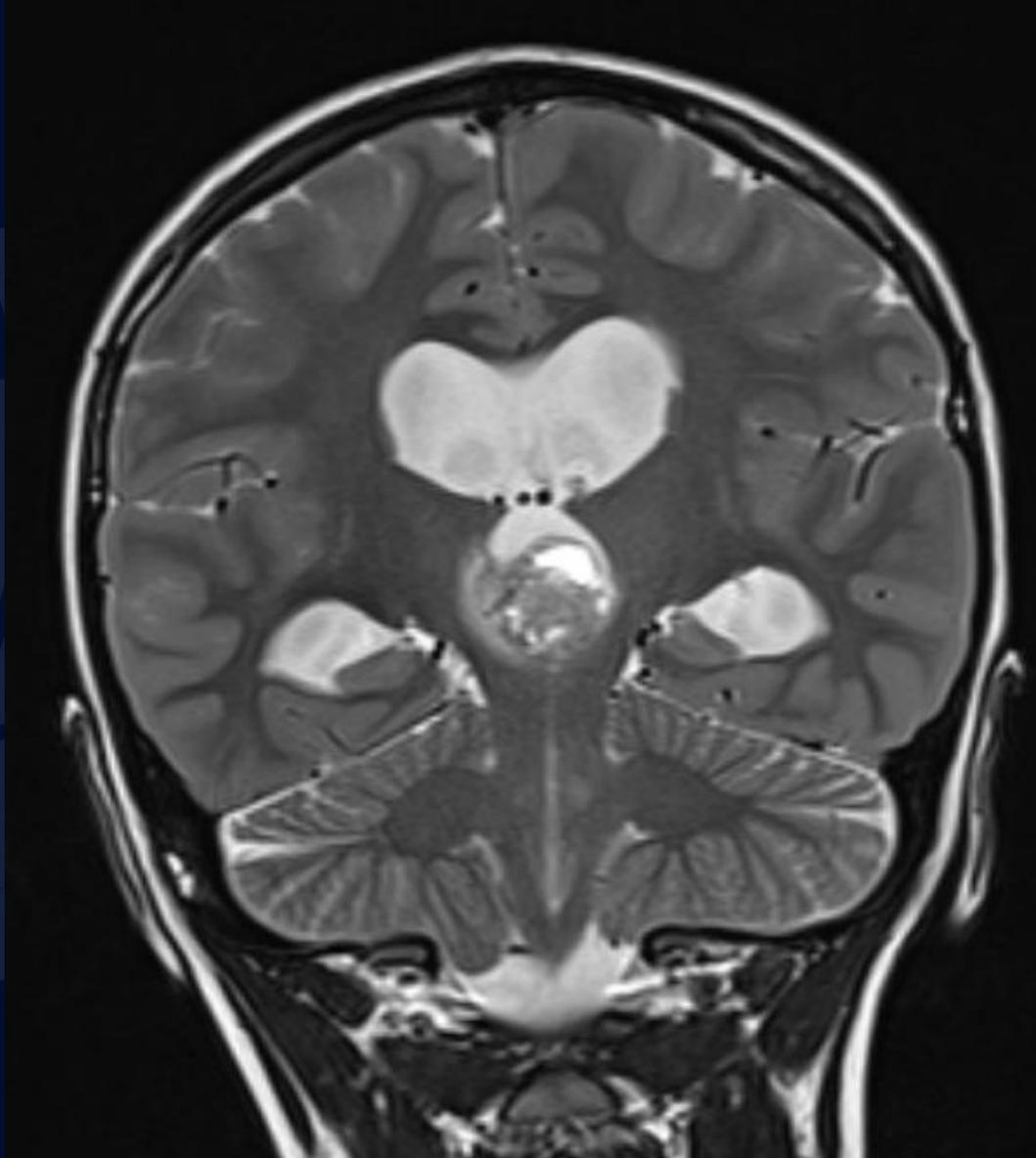
# MR Axial T1 w/o + w/ gad



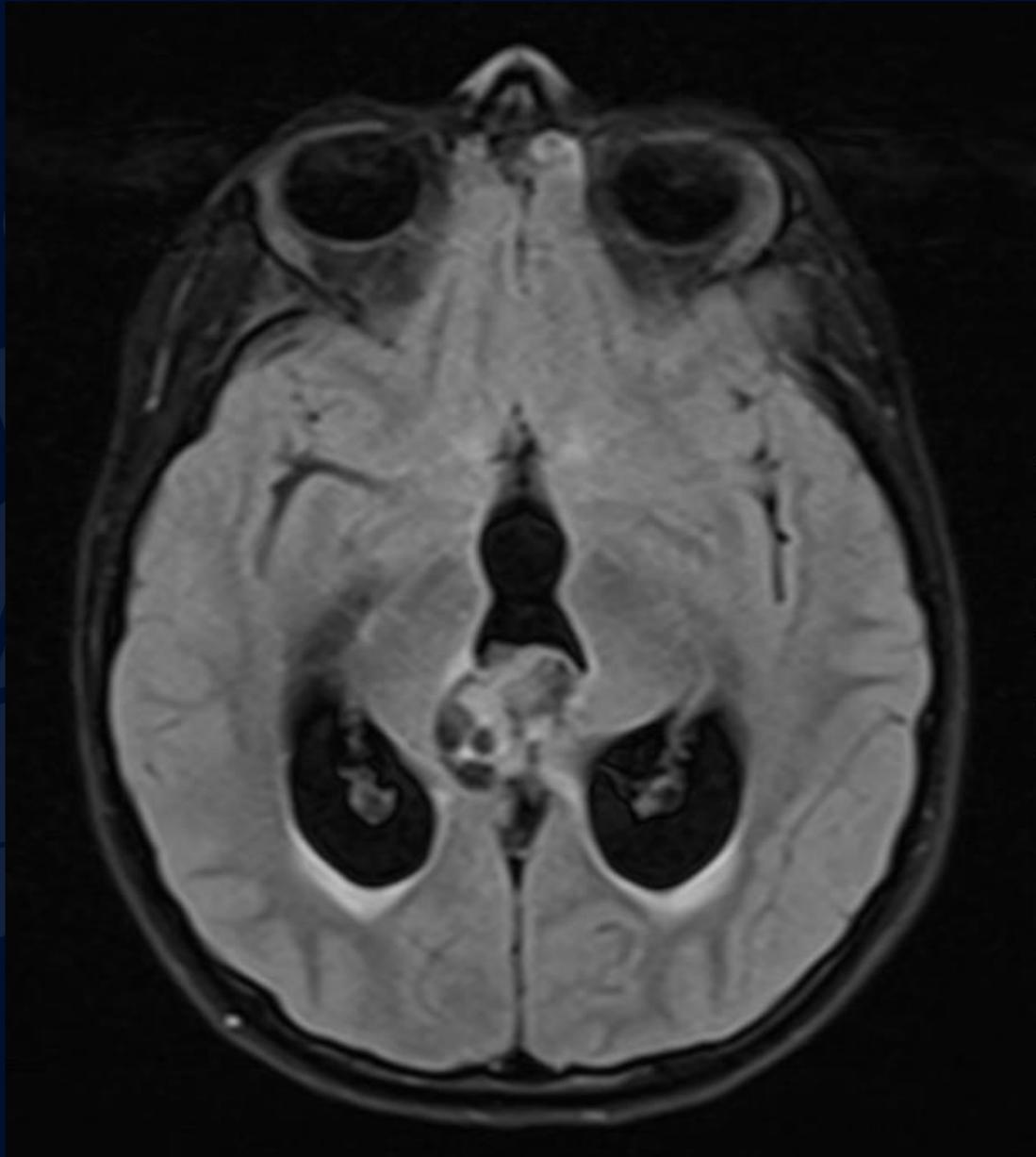
# MR Axial T2



# MR Coronal T2



# MR Axial FLAIR

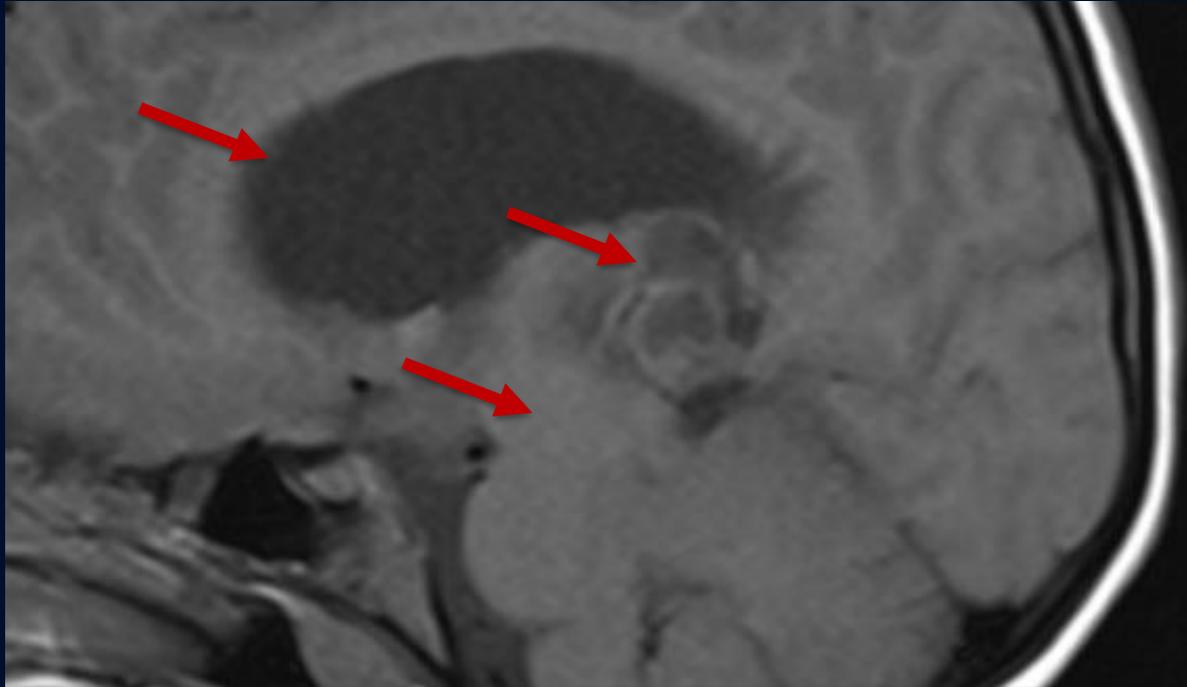


A large, stylized oak leaf graphic in a dark blue color, positioned on the left side of the slide. The leaf has a prominent central vein and several smaller veins branching off it. The leaf's edge is serrated.

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# CNS non-germinomatous germ cell tumor

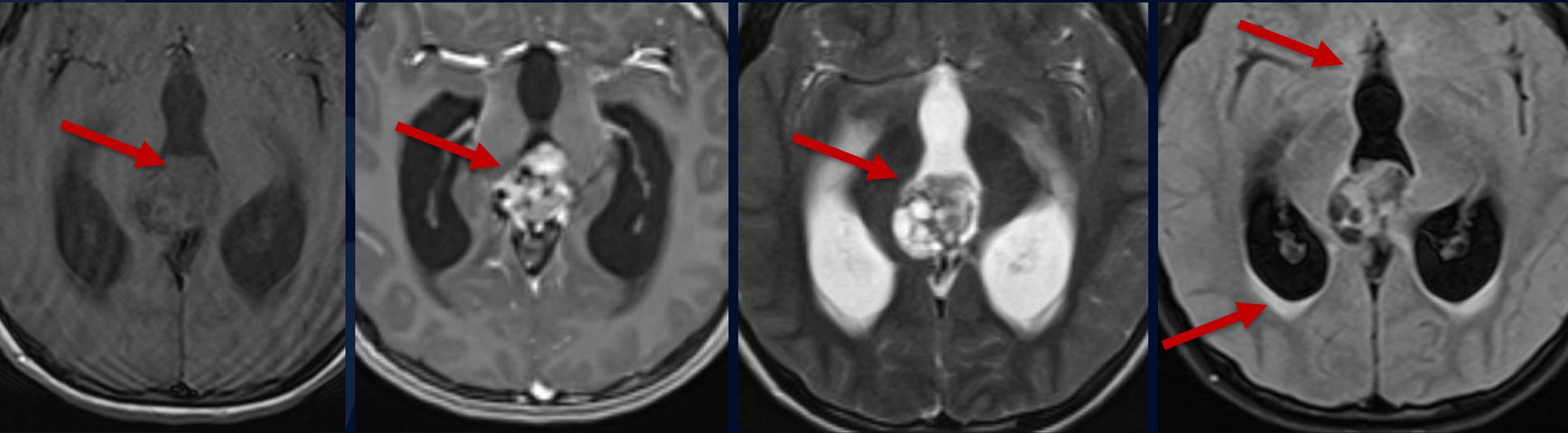
## MR Sagittal T1 w/o gad



Heterogeneously enhancing lobulated mass centered within the pineal gland demonstrating solid and cystic components. Some of these cystic components demonstrates T1 hypointensity

Marked mass effect on the tectal plate and midbrain, causing 1) severe narrowing of the cerebral aqueduct and 2) dilation of the lateral ventricle

## MR T1 w/wo Gad, T2, FLAIR

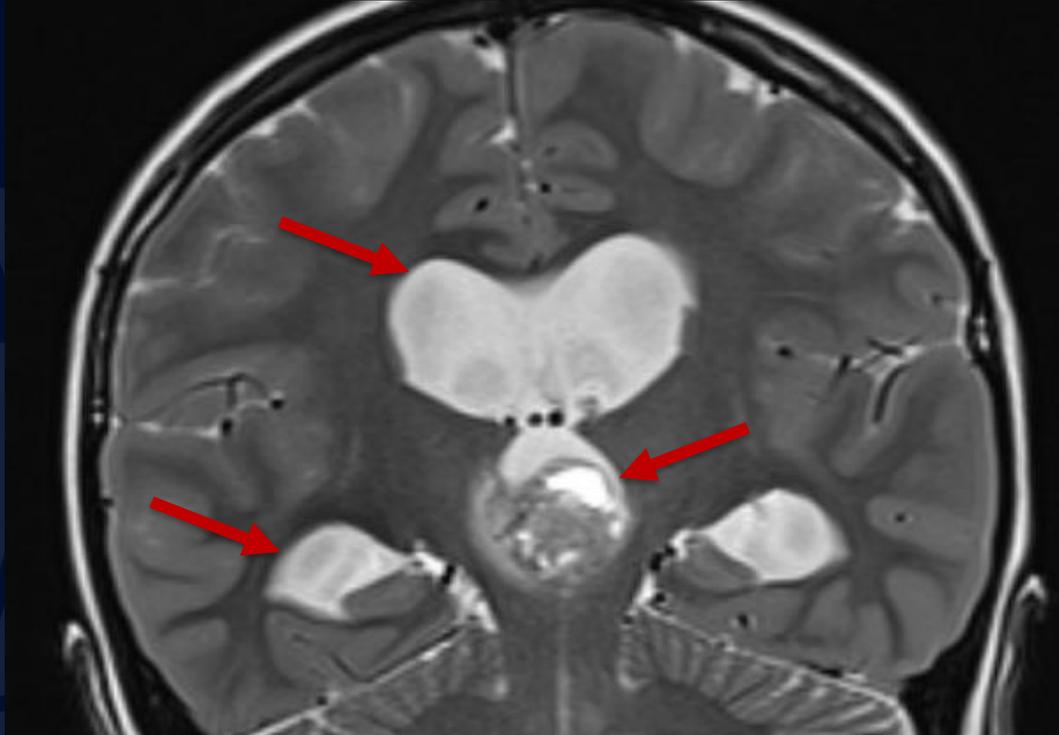


Heterogeneously enhancing lobulated mass centered within the pineal gland demonstrating solid and cystic components. Some of these cystic components demonstrate T1 hypointensity and are hyperintense on T2. The mass enhances with GAD on T1

Marked dilation of the third and lateral ventricles

Periventricular FLAIR hyperintensity, consistent with transependymal CSF flow

## MR Coronal T2



Heterogeneously enhancing lobulated mass centered within the pineal gland demonstrating solid and cystic components. Some of these cystic components demonstrate T2 hyperintensity

Marked dilation of the third and lateral ventricles

# CNS non-germinomatous germ cell tumor

Intracranial germ cell tumors (iGCTs) account for approximately 0.9% of all pediatric tumors and 3.7% of pediatric brain tumors, divided into Germinomatous germ cell tumor and Non-germinomatous germ cell tumor (**NGGCT**) subgroups. NGGCT account for 30-50% of iGCT cases.

## **NGGCTs :**

- Predominantly seen in pineal/suprasellar regions (midline structures)
  - Without evidence of gonadal primary tumors
- Etiology unknown, possibly from germ cell precursors that erroneously arrested in their midline migration during embryogenesis or there may be occult or remised GC tumor in gonads

## **Clinical presentation, Diagnosis:**

- Signs and symptoms result from mass effect or invasion of adjacent structures:
  - Obstructive hydrocephalous leads to signs of increased ICP
  - Impaired upward gaze, convergence retraction nystagmus, and pupillary hyporeflexia (Parinaud Syndrome, compression of the rostral midbrain )
- Diagnosed using a combination of histology and tumor markers (+AFP, +hCG, vs germinoma: usually neither AFP or hCG)

## **Treatment / management:**

- Chemotherapy followed by radiation therapy (e.g. carboplatin, etoposide alternating with ifosfamide, etoposide × 6 cycles).
- Surgery, for tumors that partially respond to chemotherapy or that increase in size during or after therapy.

# CNS non-germinomatous germ cell tumor

## Imaging findings

### CT

- Nodular or lobulated, slightly hyperdense lesions
- Weakly enhances with contrast

### MR

- T1: hypointense to isointense
- T2: isointense to hyperintense
- T1 C+ (GAD): heterogenous enhancement
- FLAIR: Periventricular hyperintensity suggests transependymal CSF flow

### Differential

- Germinomatous germ cell tumor
- Craniopharyngioma
- Glioma
- Meningioma

# References

1. Morana, Giovanni & Alves, César & Tortora, Domenico & Finlay, Jonathan & Severino, Mariasavina & Nozza, Paolo & Ravegnani, Marcello & Pavanello, Marco & Milanaccio, Claudia & Maghnie, Mohamad & Rossi, Andrea & Garre, Maria. (2018). T2\*-based MR imaging (gradient echo or susceptibility-weighted imaging) in midline and off-midline intracranial germ cell tumors: a pilot study. *Neuroradiology*. 60. 10.1007/s00234-017-1947-3.
2. Fujimaki T, Matsutani M, Funada N, Kirino T, Takakura K, Nakamura O, Tamura A, Sano K. CT and MRI features of intracranial germ cell tumors. *J Neurooncol*. 1994;19(3):217-26. doi: 10.1007/BF01053275. PMID: 7807172.
3. Aizer AA, Sethi RV, Hedley-Whyte ET, Ebb D, Tarbell NJ, Yock TI, Macdonald SM. Bifocal intracranial tumors of nongerminomatous germ cell etiology: diagnostic and therapeutic implications. *Neuro Oncol*. 2013 Jul;15(7):955-60. doi: 10.1093/neuonc/not050. Epub 2013 May 2. PMID: 23640532; PMCID: PMC3688019.
4. Yang M, Wang J, Zhang L, Liu J. Update on MRI in pediatric intracranial germ cell tumors-The clinical and radiological features. *Front Pediatr*. 2023 May 4;11:1141397. doi: 10.3389/fped.2023.1141397. PMID: 37215600; PMCID: PMC10192609.
5. National Cancer Institute. "Childhood Central Nervous System Germ Cell Tumors Treatment (PDQ®)—Health Professional Version." PDQ Cancer Information Summaries, National Cancer Institute, 21 Feb. 2024, <https://www.cancer.gov/types/brain/hp/child-cns-germ-cell-treatment-pdq>.