AMSER Case of the Month
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HPI: 18 year old male with intractable epilepsy

Mara Jindeel, MS4
St. George's University School of Medicine

Alexander Scott, MD PGY4
Deborah Brahee, MD
Cleveland Clinic Foundation
Department of Radiology

Cleveland Clinic

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Patient Presentation

- HPI: 18-year-old male presented with recurrent seizures that began at age 16 after d/c topiramate (used for migraine prophylaxis). The seizures continued despite being placed on four anti-seizure medications.
- PMHx: Migraines
- FHx: Maternal history of migraines
- Labs: Noncontributory

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

Variant 1: **New-onset seizure. Unrelated to trauma. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
CT head without IV contrast	Usually Appropriate	☢☢☢
MRI head without IV contrast	Usually Appropriate	O
MRI head without and with IV contrast	May Be Appropriate	O
CT head with IV contrast	Usually Not Appropriate	☢☢☢
CT head without and with IV contrast	Usually Not Appropriate	☢☢☢
FDG-PET/CT brain	Usually Not Appropriate	☢☢☢
MEG	Usually Not Appropriate	O
MRI functional (fMRI) head without IV contrast	Usually Not Appropriate	O
HMPAO SPECT or SPECT/CT brain ictal and interictal	Usually Not Appropriate	☢☢☢

This imaging modality was ordered by the ED physician

Findings (unlabeled)

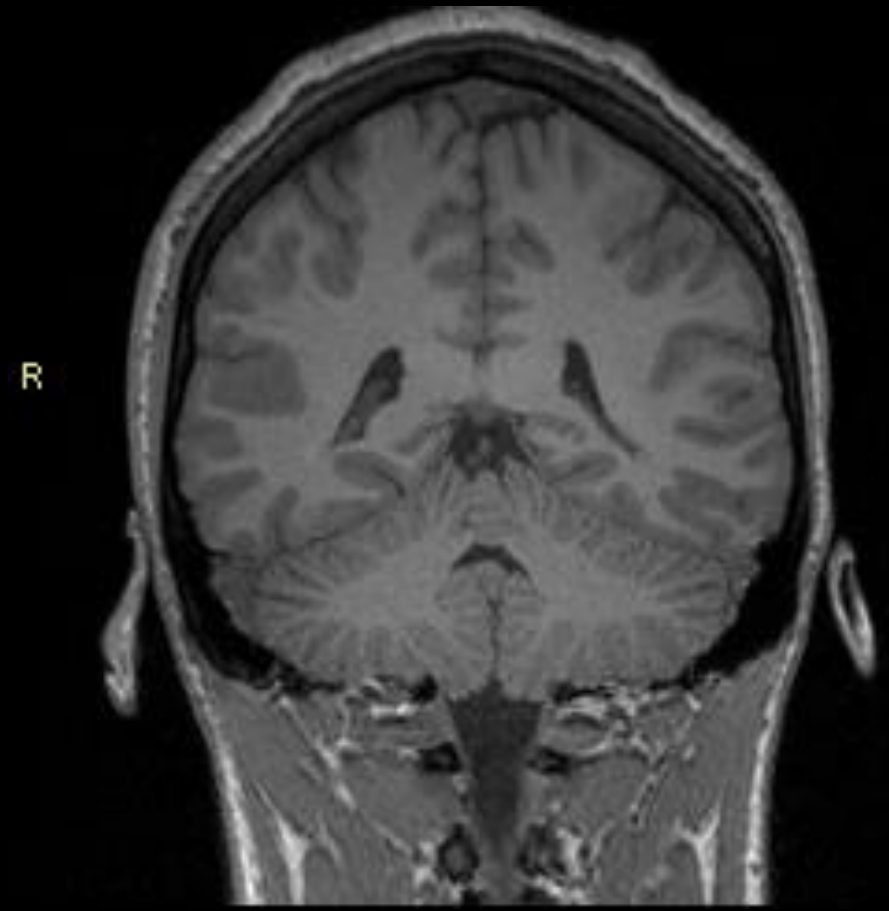
Axial

Discussion of initial Findings

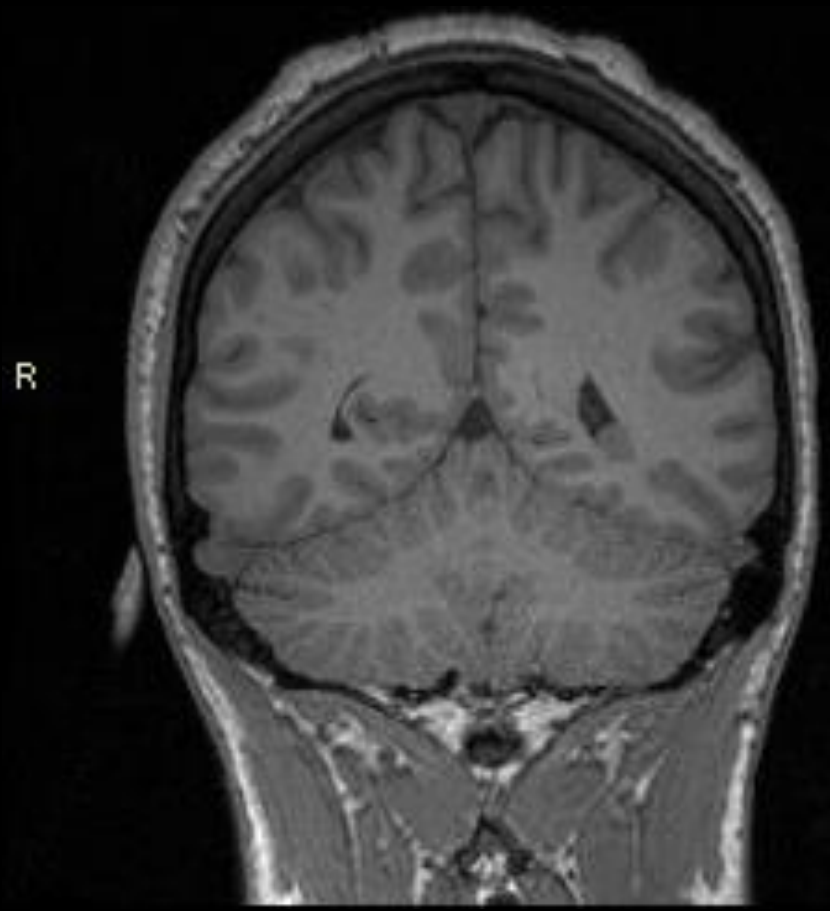
- CT read as normal, so MRI was ordered to evaluate for underlying structural abnormalities

Axial

Findings: (unlabeled)—Initial Imaging



Coronal T1



Coronal T1

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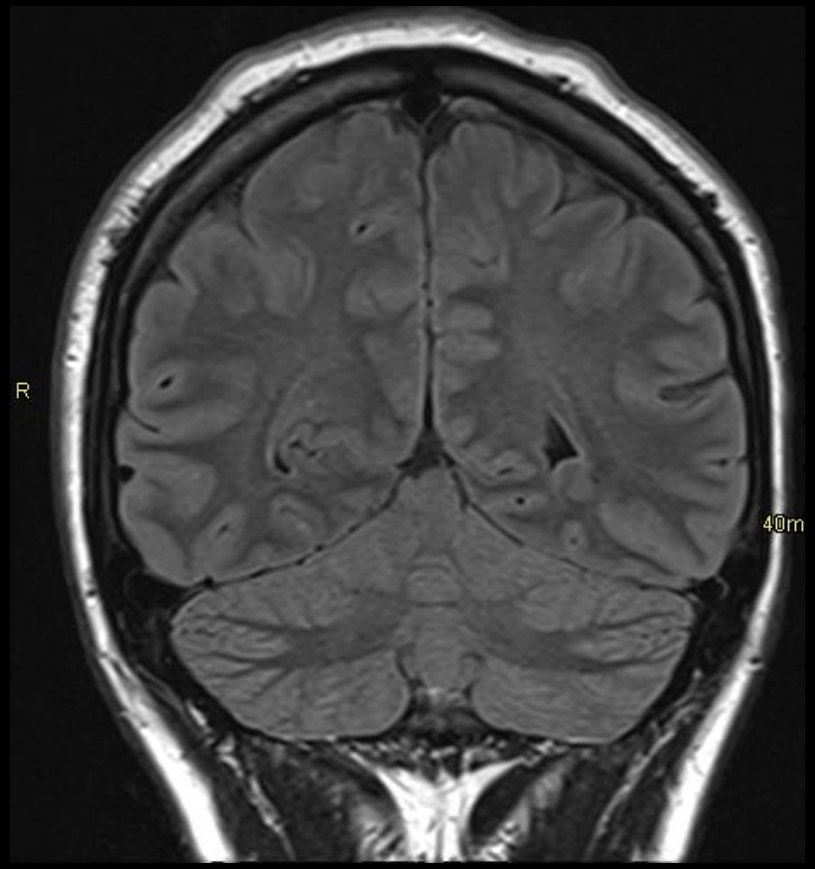
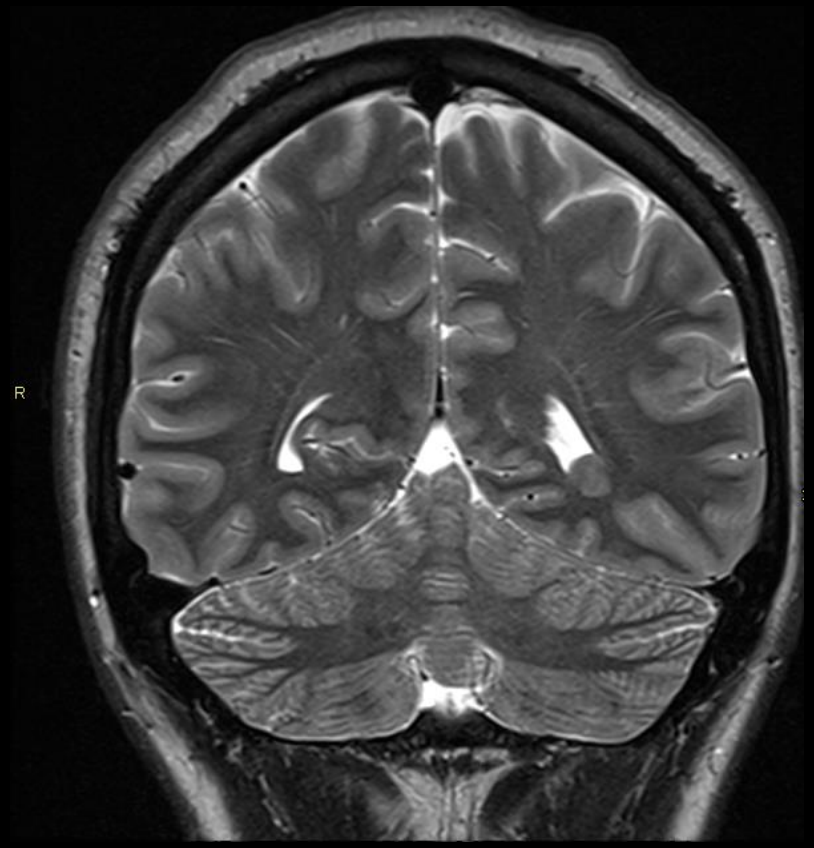
Findings: (unlabeled)—Initial Imaging

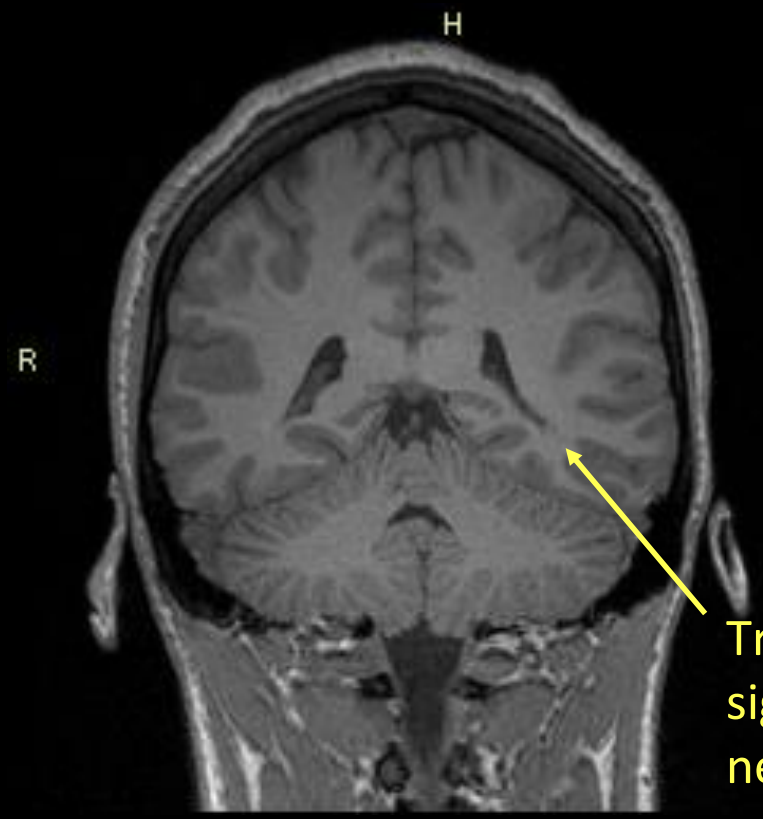


FLAIR



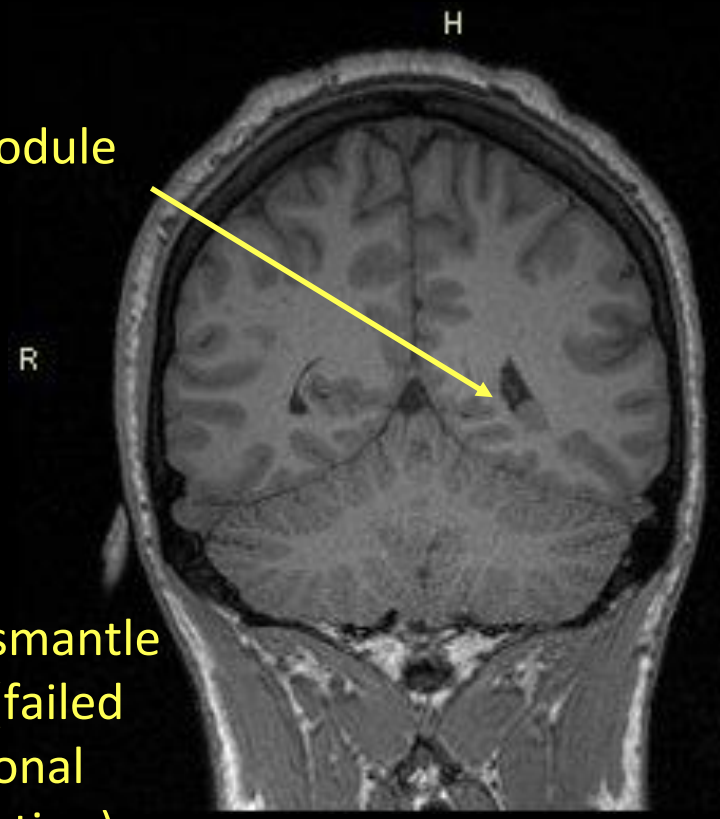
T2

Findings: Labeled—Initial Imaging



Transmantle sign (failed neuronal migration)

Coronal T1 MRI



Nodule

Coronal T1 MRI

Isodense region corresponds with the finding

Axial CT

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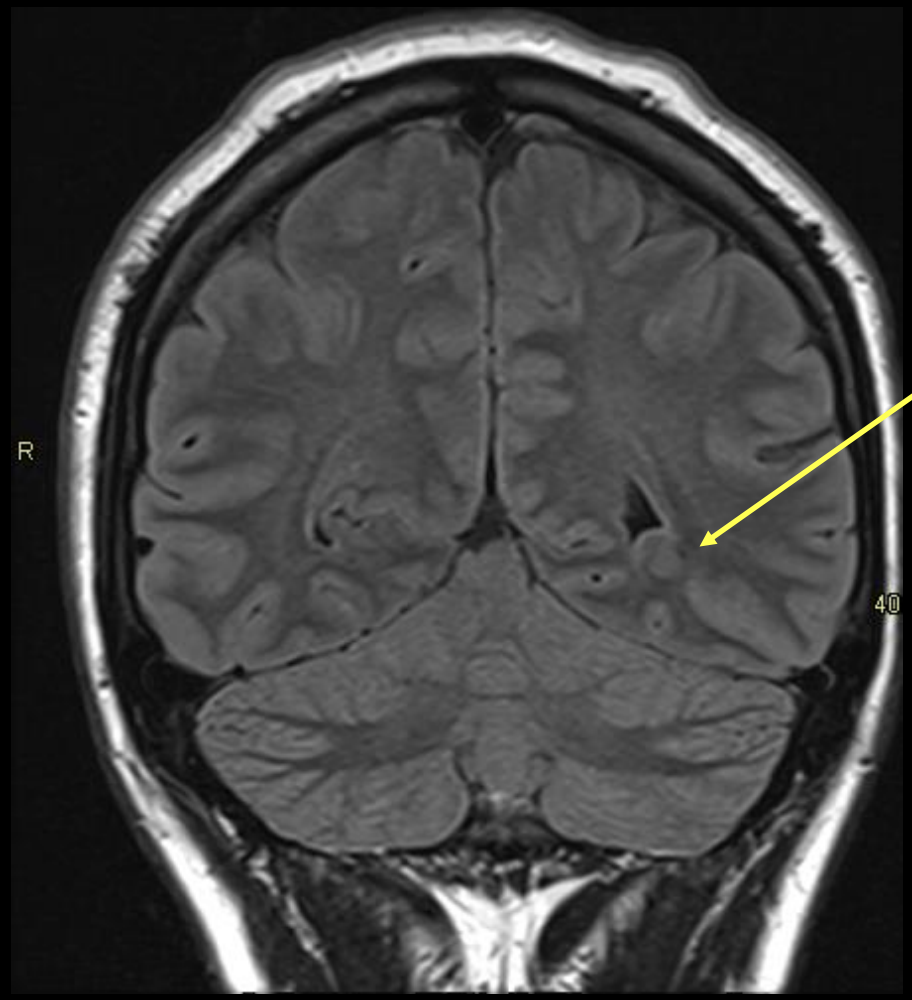
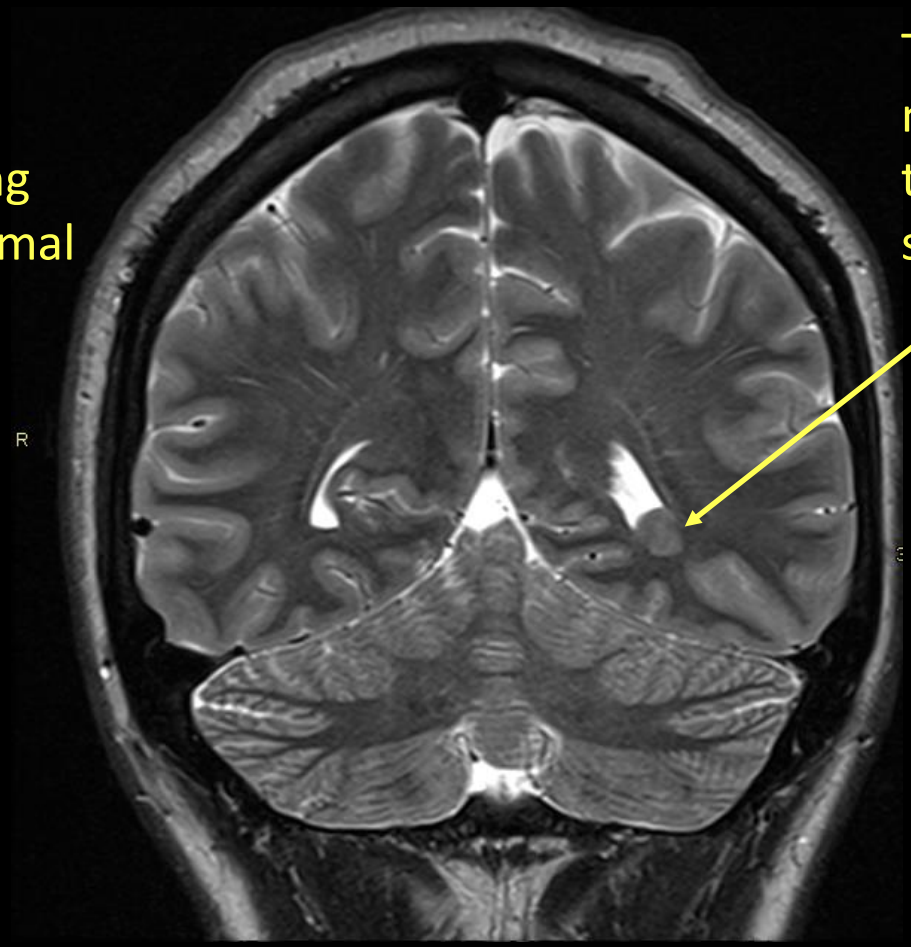
Findings: Labeled—Initial Imaging



FLAIR
Isointense nodule along the ependymal surface

FLAIR



T2 Isointense nodule along the ependymal surface

T2

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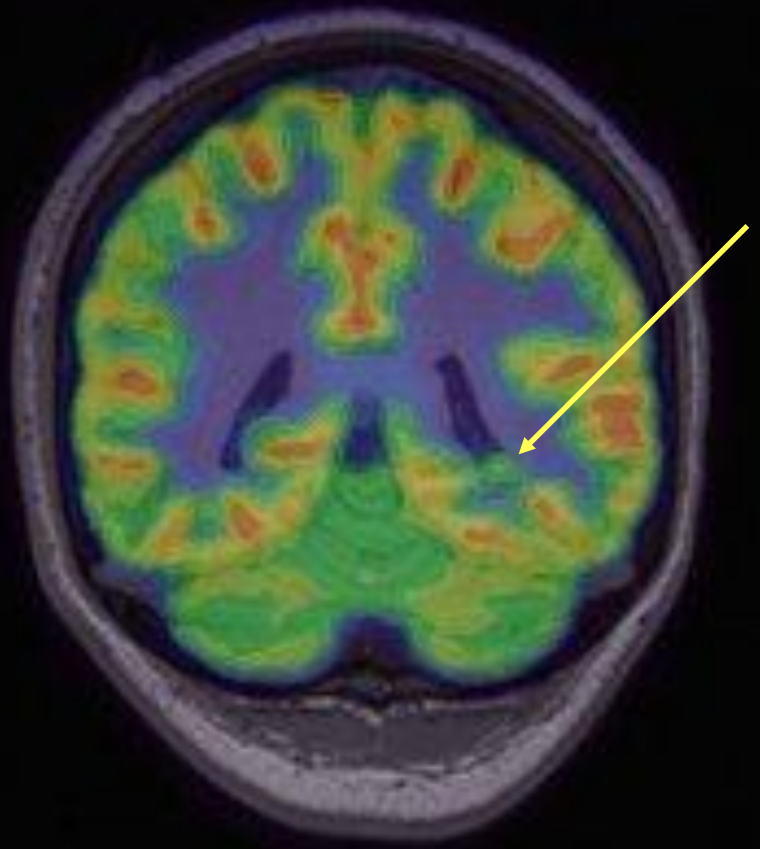
Additional Pre-op Planning Image



FDG PET/MR Fusion

- Nodule located along the ependymal surface
- Exhibits similar metabolic activity to gray matter

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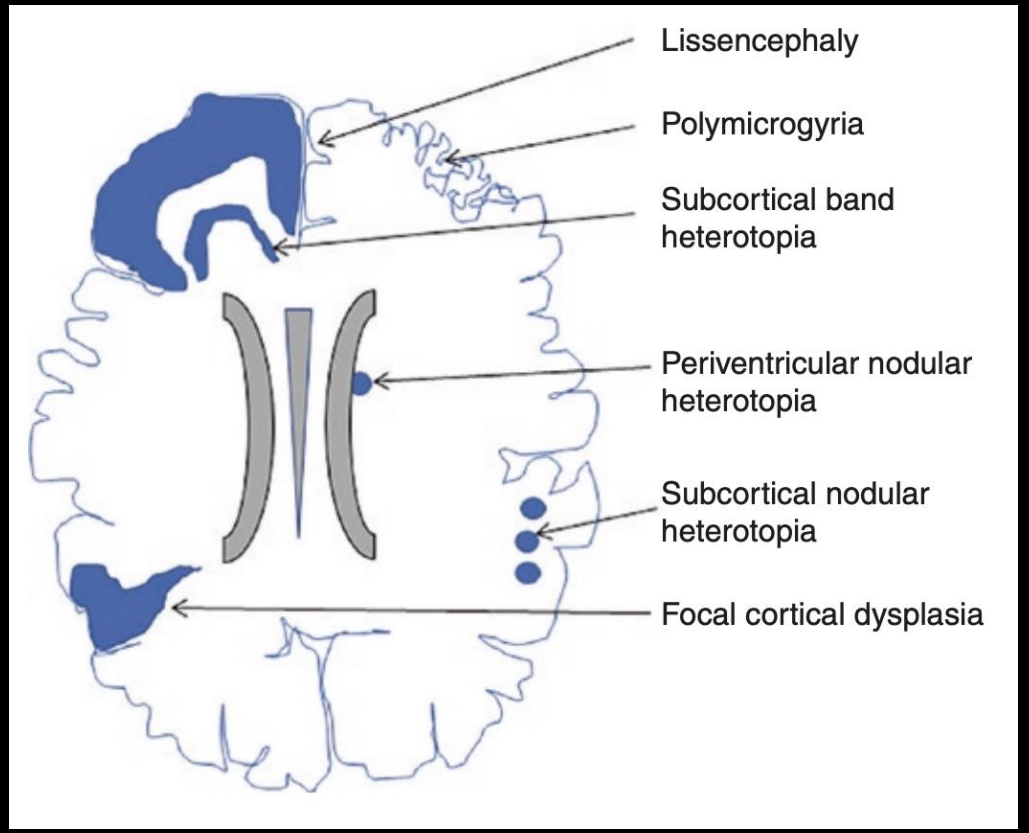
Final Dx:

Periventricular Nodular Heterotopia (PVNH)

Case Discussion

- PVNH is a developmental brain anomaly
- caused by *impaired migration of normal neurons* from the ependymal surface to the cortex
- Results in ectopic clumps of neurons (heterotopia) near the ventricles
- Multiple possible outcomes for failed migration and organization (see image)
- Severity of clinical manifestations may be associated with the number of malformations



Rajeswaran[1]

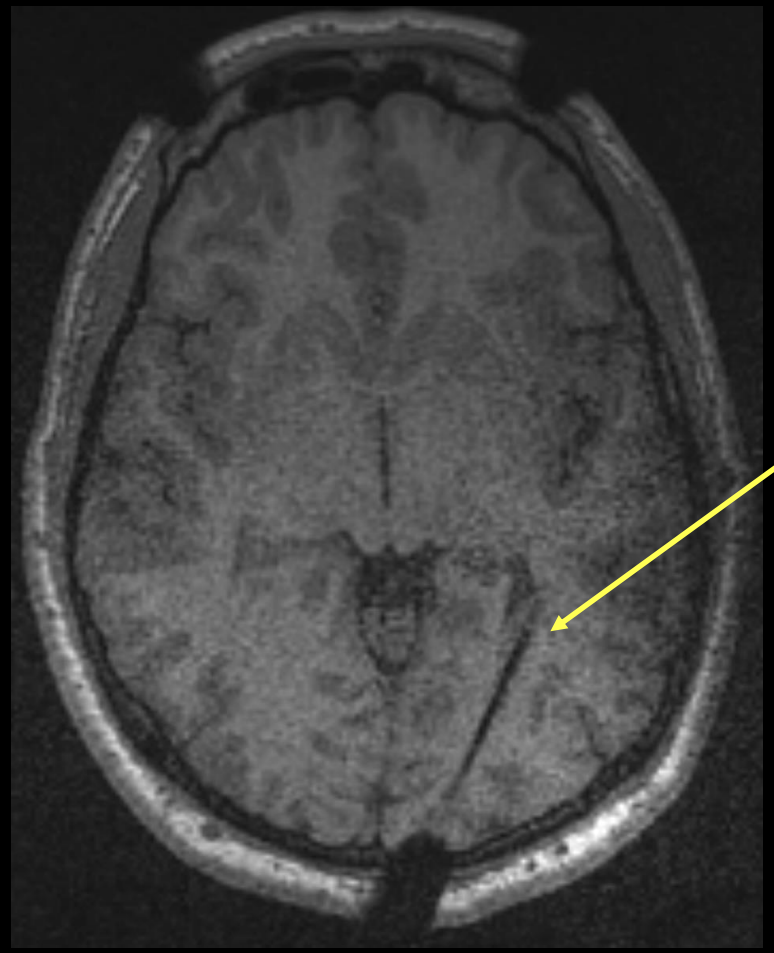
Case Discussion

- Female predominance, with seizures generally beginning in the teenage years
- Medically intractable epilepsy is a hallmark of the condition (as in our patient)
- May be associated with a higher risk of cardiovascular abnormalities and coagulopathy
- Can be inherited or occur de novo with several gene associations
  - Filamin A (FLNA) gene on X-chromosome most common

Treatment—Laser Interstitial Thermal Therapy (LITT)

- Our patient was treated with Laser Interstitial Thermal Therapy (LITT), a promising treatment for PVNH
- Involves inserting a thermal laser into the brain under MRI guidance to ablate the nodules
- Has been shown to be as effective as resection, while being less invasive, and with lower morbidity[2]

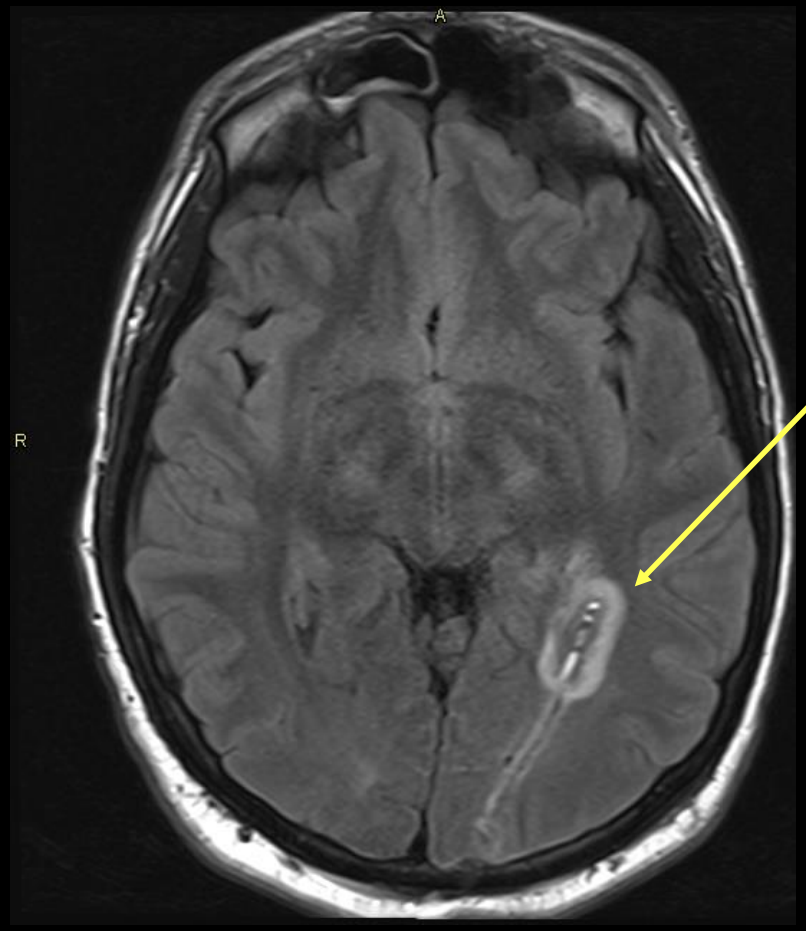
LITT Procedure - Intraoperative



Ablation probe extending to the focus of heterotopia.

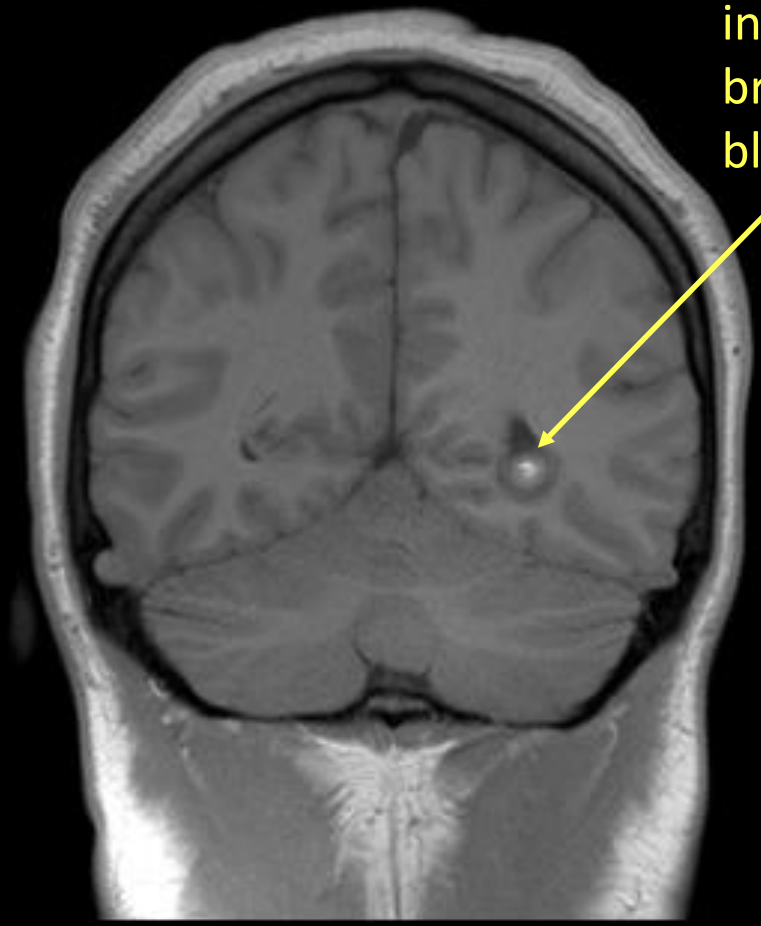
Axial T1

Post-LITT Procedure



Focal hyperintensity along the tract (cytotoxic edema)

Axial FLAIR



intrinsic T1 hyper intensity— breakdown of blood products

Coronal T1

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Case Resolution

- Prior to LITT procedure, the patient had intractable epilepsy despite being on four anti-seizure medications
- Patient reported no seizures at routine 6-month follow-up, with medication reduction after treatment

References:

1. Rajeswaran, R. (2022). Abnormalities of Proliferation, Neuronal Migration, and Cortical Organization. In: MR Imaging of the Fetus. Springer, Singapore. https://doi.org/10.1007/978-981-16-9209-3_9

2. Shukla ND, Ho AL, Pendharkar AV, Sussman ES, Halpern CH. Laser interstitial thermal therapy for the treatment of epilepsy: evidence to date. *Neuropsychiatr Dis Treat*. 2017;13:2469-2475. Published 2017 Sep 26. doi:10.2147/NDT.S139544

3. Direk MC, Komur M, Ozgur A, Okuyaz C. A Rare Cause of Refractory Epilepsy: Posterior Periventricular Nodular Heterotopia. *J Pediatr Neurosci*. 2018;13(3):340-342. doi:10.4103/JPN.JPN_178_17

4. Gaillard F, Sharma R, Chieng R, et al. Focal cortical dysplasia. Reference article, Radiopaedia.org (Accessed on 12 Aug 2024) https://doi.org/10.53347/rID-10164

5. Kim SA, Ivanov AO, Halepa AA, Sysoeva AA, Gunenko GA. Surgical treatment of epilepsy in a patient with bilateral periventricular nodular heterotopia: A case report. *Surg Neurol Int*. 2023;14:283. Published 2023 Aug 11. doi:10.25259/SNI_478_2023

6. Tandon, N., and Lhatoo, S. Periventiricular Nodular Heterotopia. PowerPoint Presented at Texas Institute of Restorative Neurotechnologies. May 21, 2021. Houston, Texas. Accessed on August 11, 2024.

7. Whiting AC, Bingaman JR, Catapano JS, et al. Laser Interstitial Thermal Therapy for Epileptogenic Periventricular Nodular Heterotopia. *World Neurosurg*. 2020;138:e892-e897. doi:10.1016/j.wneu.2020.03.133

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