

AMSER Case of the Month

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67-year-old female presenting with complex partial seizure and subsequent word finding difficulty, memory difficulties, generalized weakness, and falls

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Patient Presentation

- **Chief Complaint:** Initially presented to ED with **seizure and neck jerking lasting one minute in duration**. Non-contrast CT head obtained, Keppra was started, and patient lost to outpatient follow-up. Over the course of two years had **begun to develop generalized weakness, falls, and word finding difficulties**.
- **Past medical history:** **Breast cancer (IDC), squamous cell carcinoma of the nose**, obstructive sleep apnea, hypertension, hyperlipidemia, transient ischemic attack,
- **Past surgical history:** Left breast lumpectomy with axillary lymph node biopsy
- **Medications:** Tamoxifen, Keppra, Lisinopril, pantoprazole, atorvastatin

Pertinent Labs

- Basic Metabolic Panel (BMP) had no abnormal findings
- Complete Blood Count (CBC) had no abnormal findings
- Vitamin B1/B12/E are within normal limits
- Microbiological workup was negative

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria [1]

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	○
MRI head without and with IV contrast	May Be Appropriate	○
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	○
MRI functional (fMRI) head without IV contrast	Usually Not Appropriate	○
HMPAO SPECT or SPECT/CT brain ictal and interictal	Usually Not Appropriate	⊗⊗⊗

This imaging modality was ordered by the ED physician

Select the applicable ACR Appropriateness Criteria [2]

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.

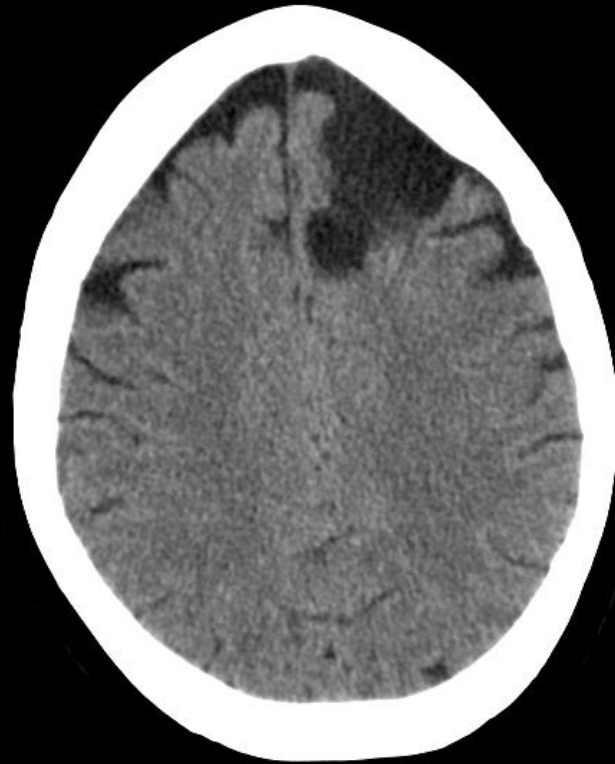
Procedure	Appropriateness Category	Relative Radiation Level
MRI head without and with IV contrast	Usually Appropriate	0
MRI head without IV contrast	Usually Appropriate	0
CT head without IV contrast	Usually Appropriate	☼ ☼ ☼
CT head without and with IV contrast	May Be Appropriate	☼ ☼ ☼
CT head with IV contrast	Usually Not Appropriate	☼ ☼ ☼

This imaging modality was ordered by the neurologist



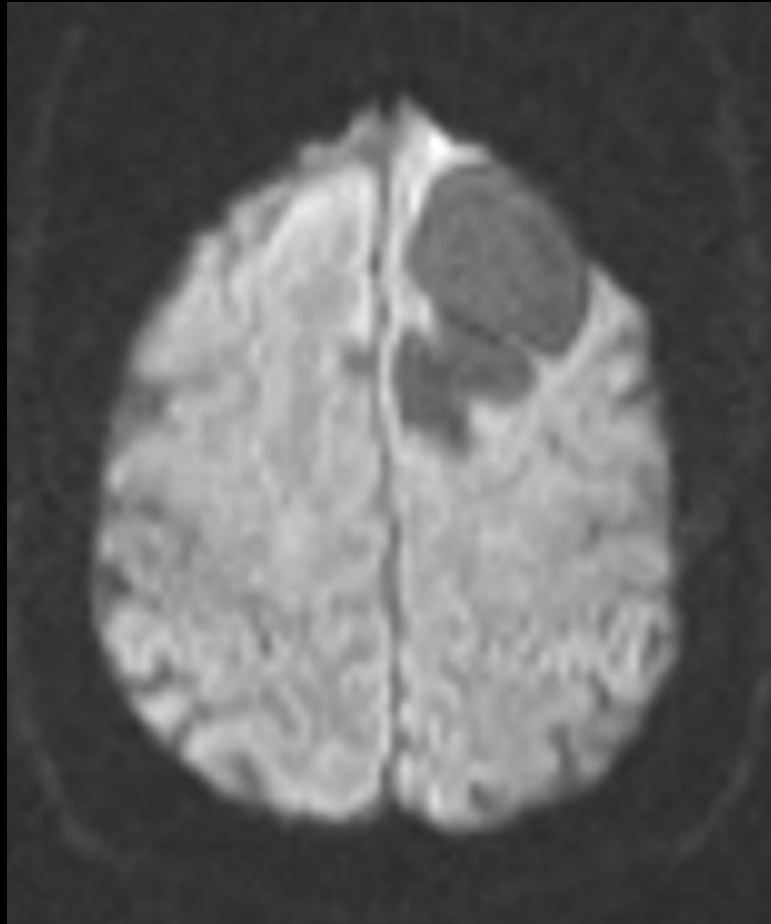
Findings (unlabeled)

Head CT Without IV
Contrast

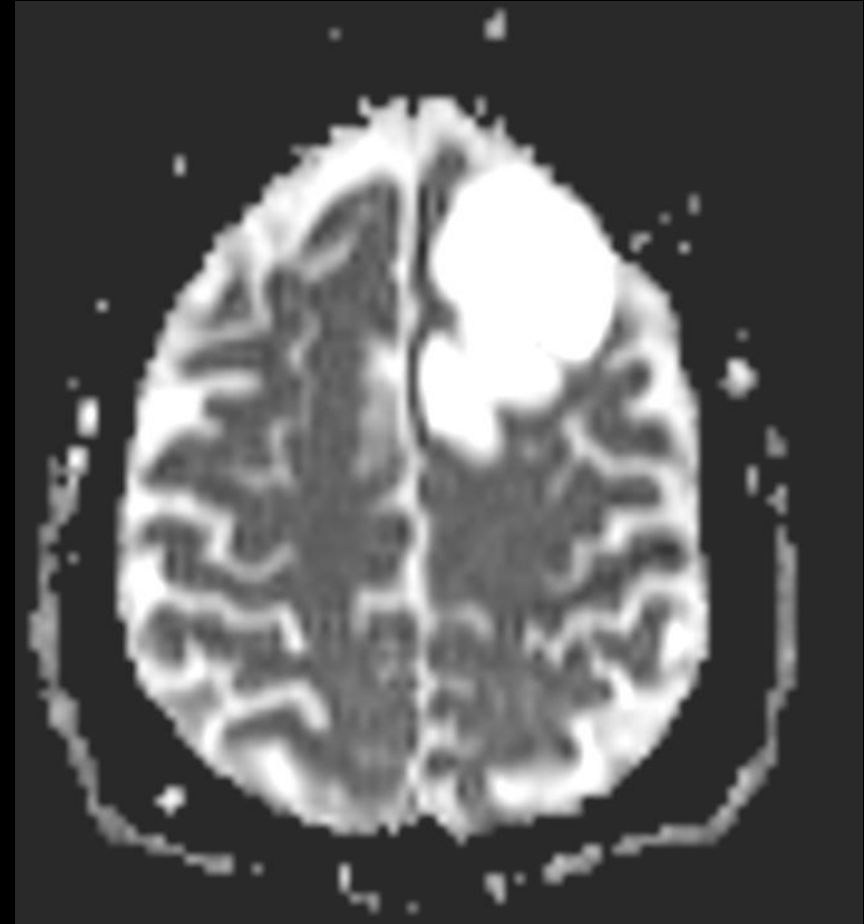


Findings (unlabeled)

DWI – b1000

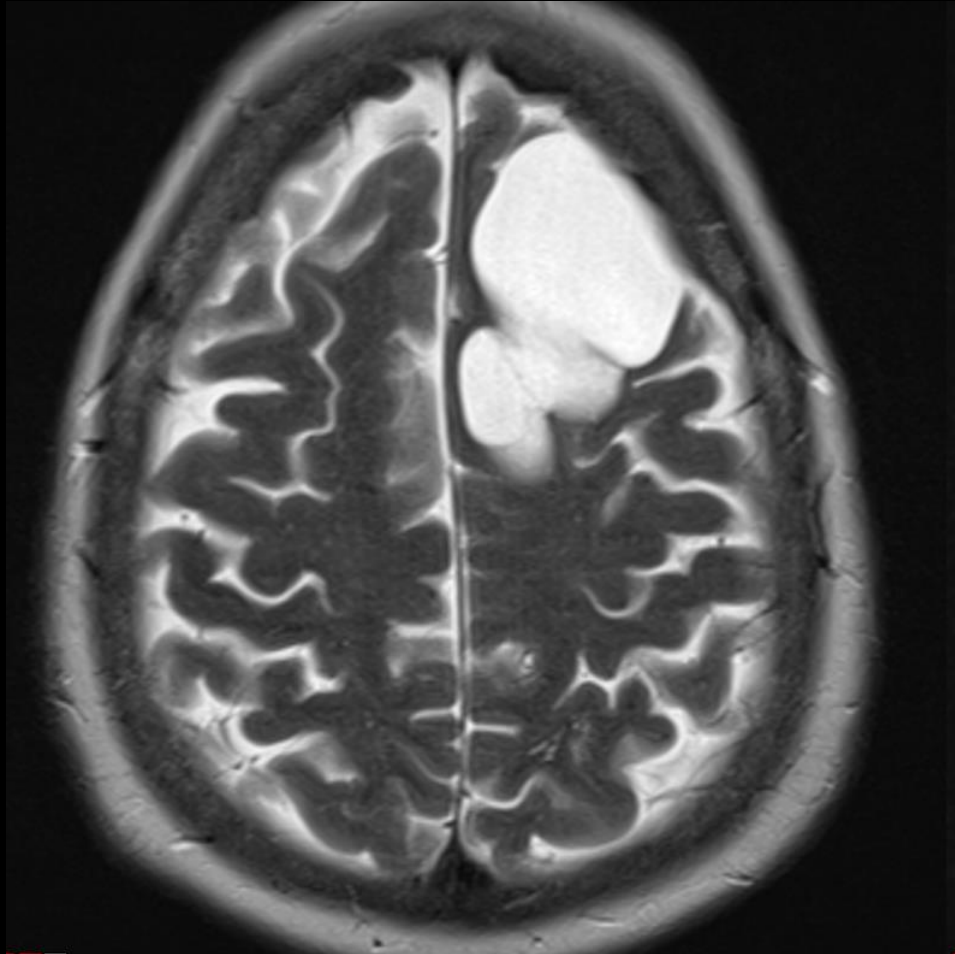


ADC

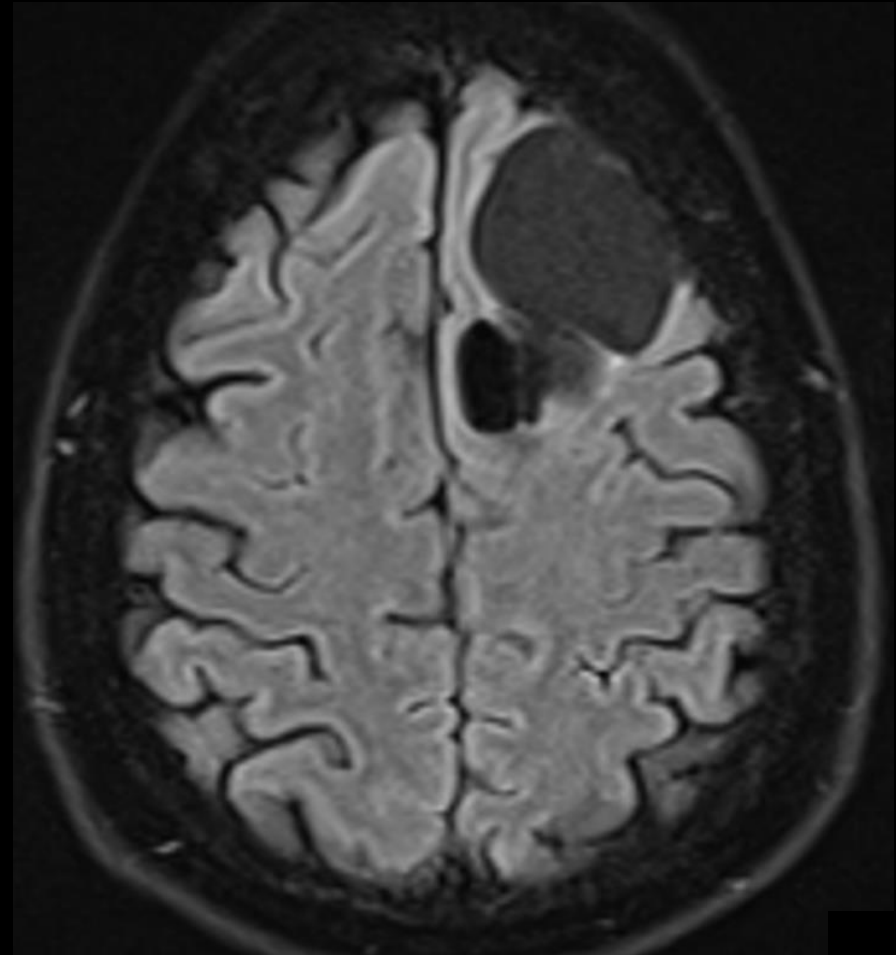


Findings (unlabeled)

T2 Axial

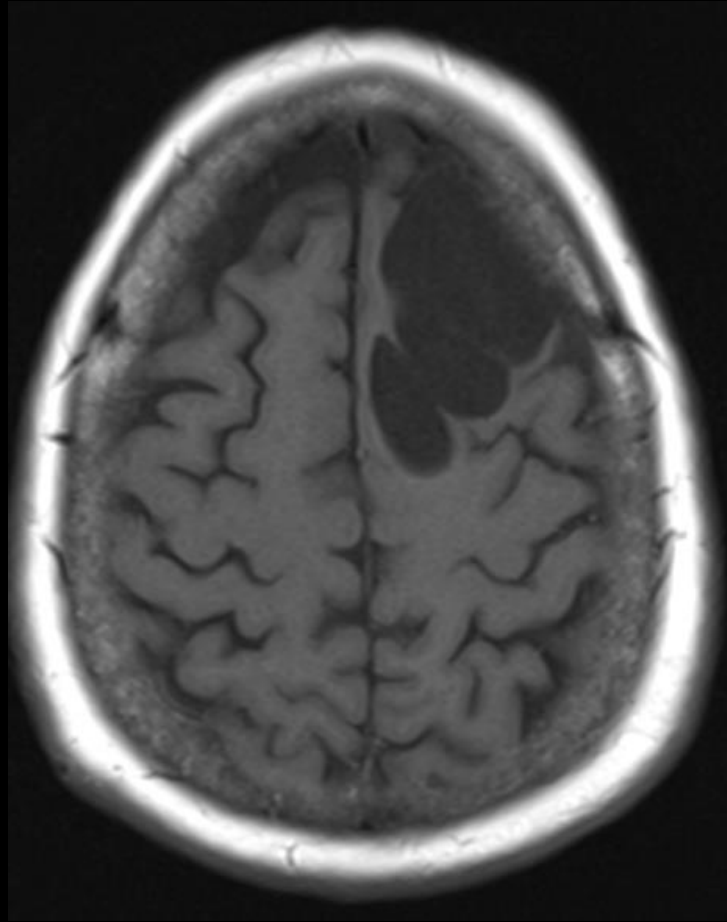


T2 FLAIR

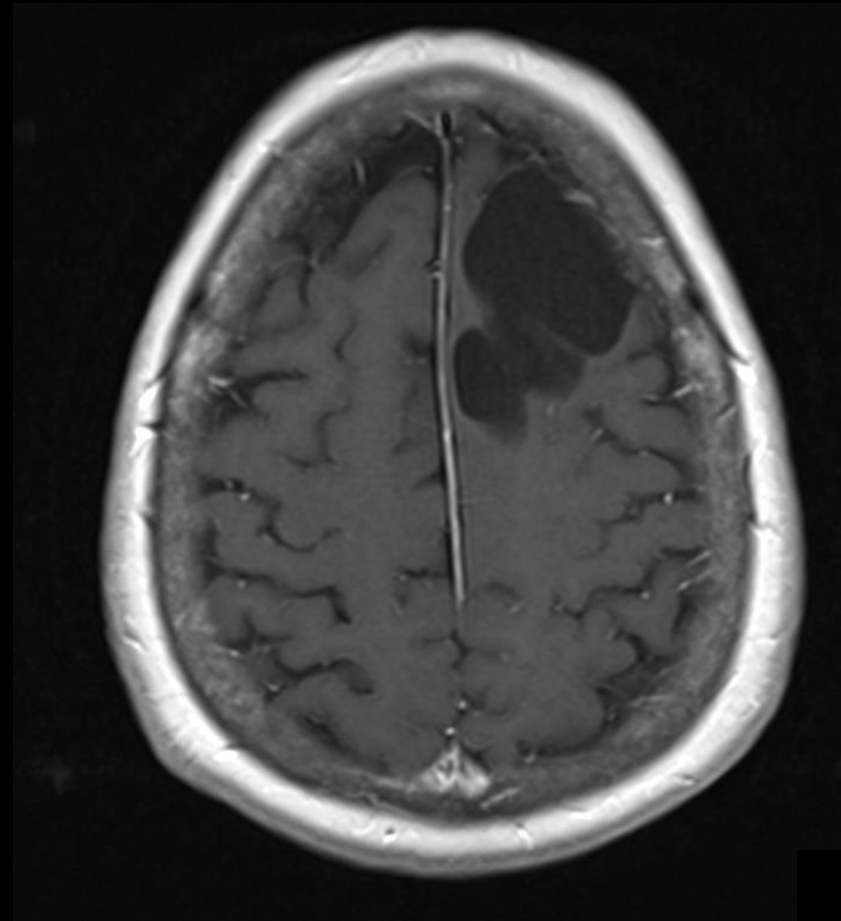


Findings (unlabeled)

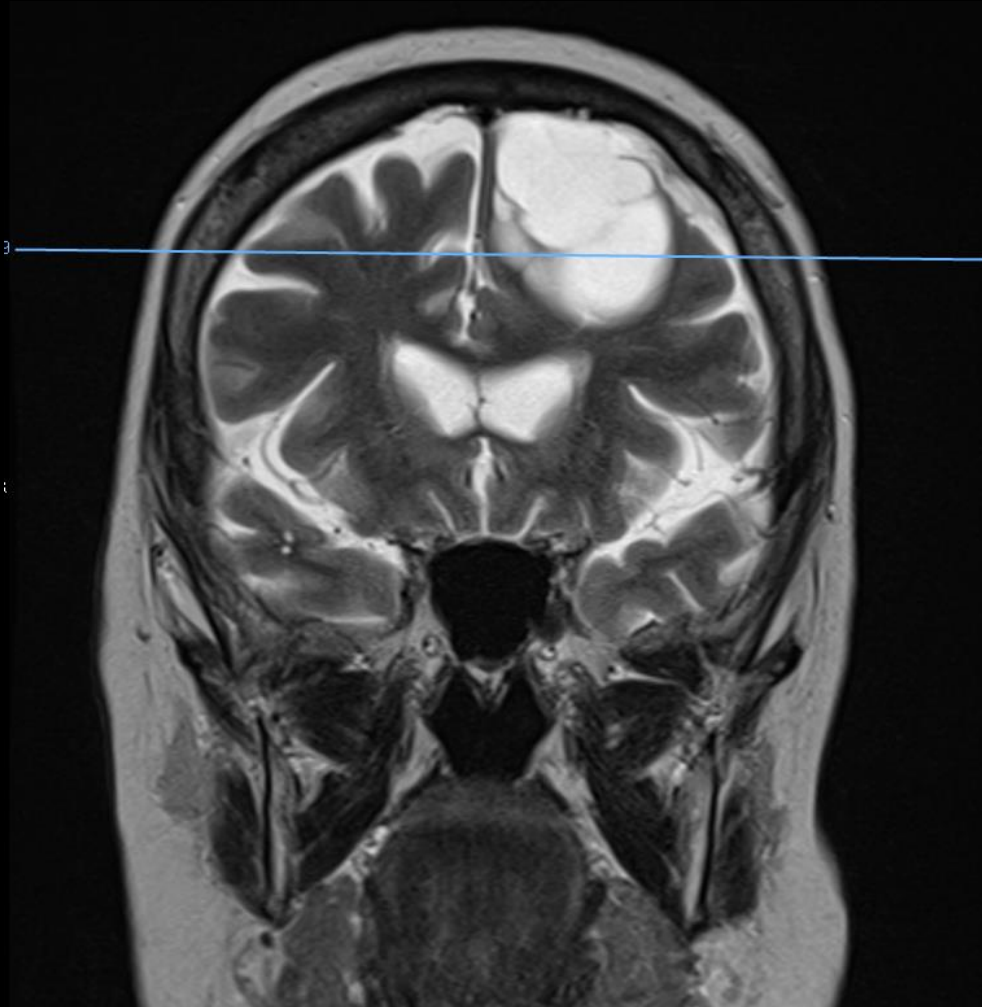
T1 Pre-contrast



T1 Post-contrast

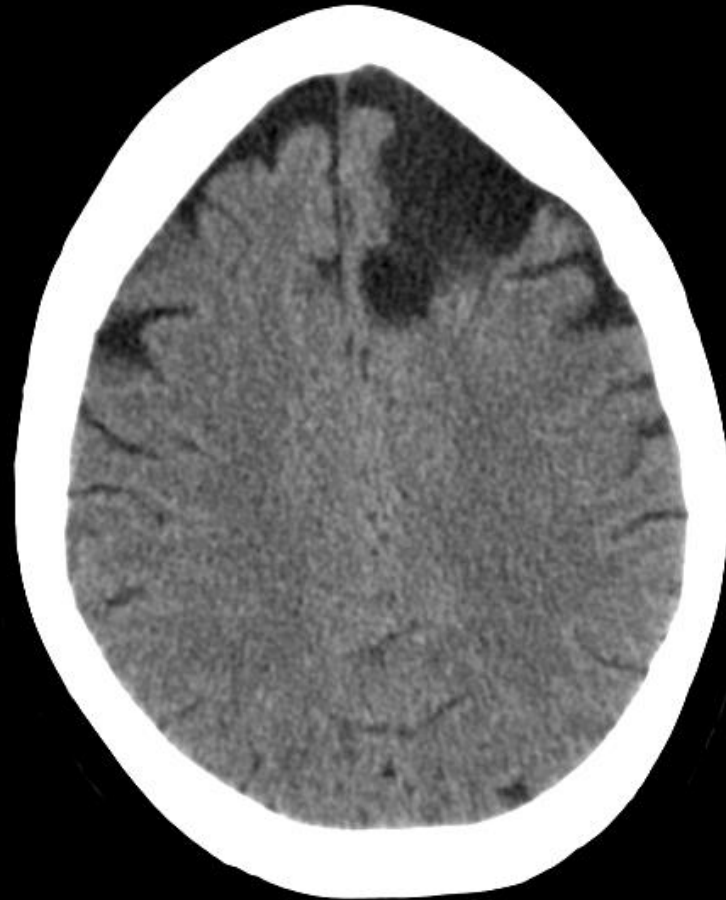


Findings (unlabeled)



Findings: (labeled)

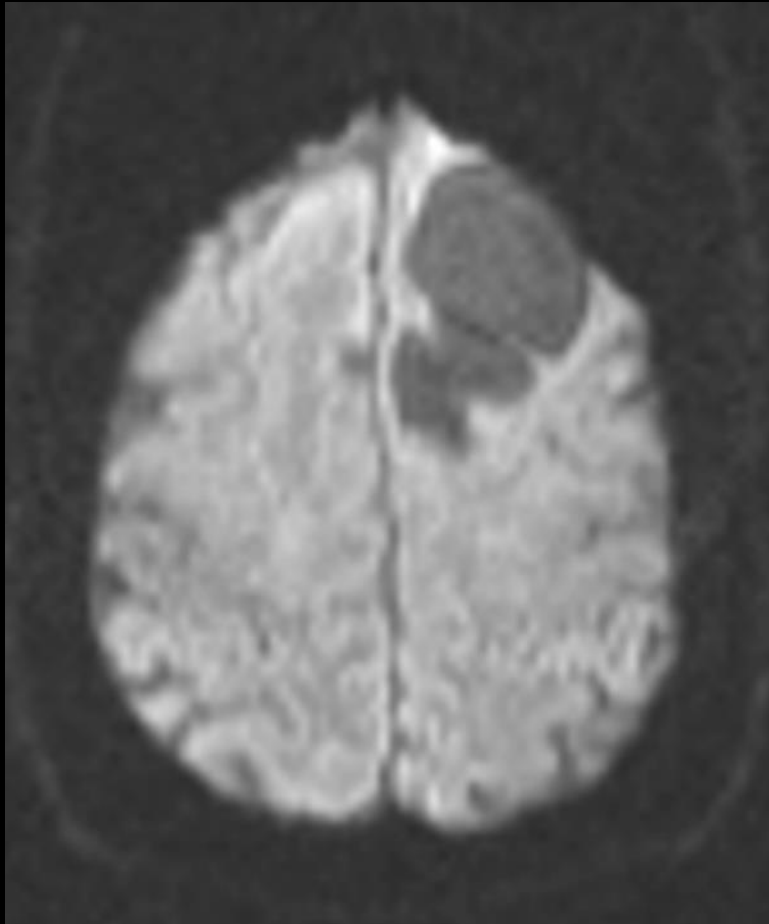
Head CT Without IV
Contrast



Low density, extra-axial cystic mass present in the left superior frontal region with mild mass effect on underlying brain parenchyma and sulcal effacement. No IV contrast was used so cannot comment on degree of enhancement.

Findings: (labeled)

DWI – b1000



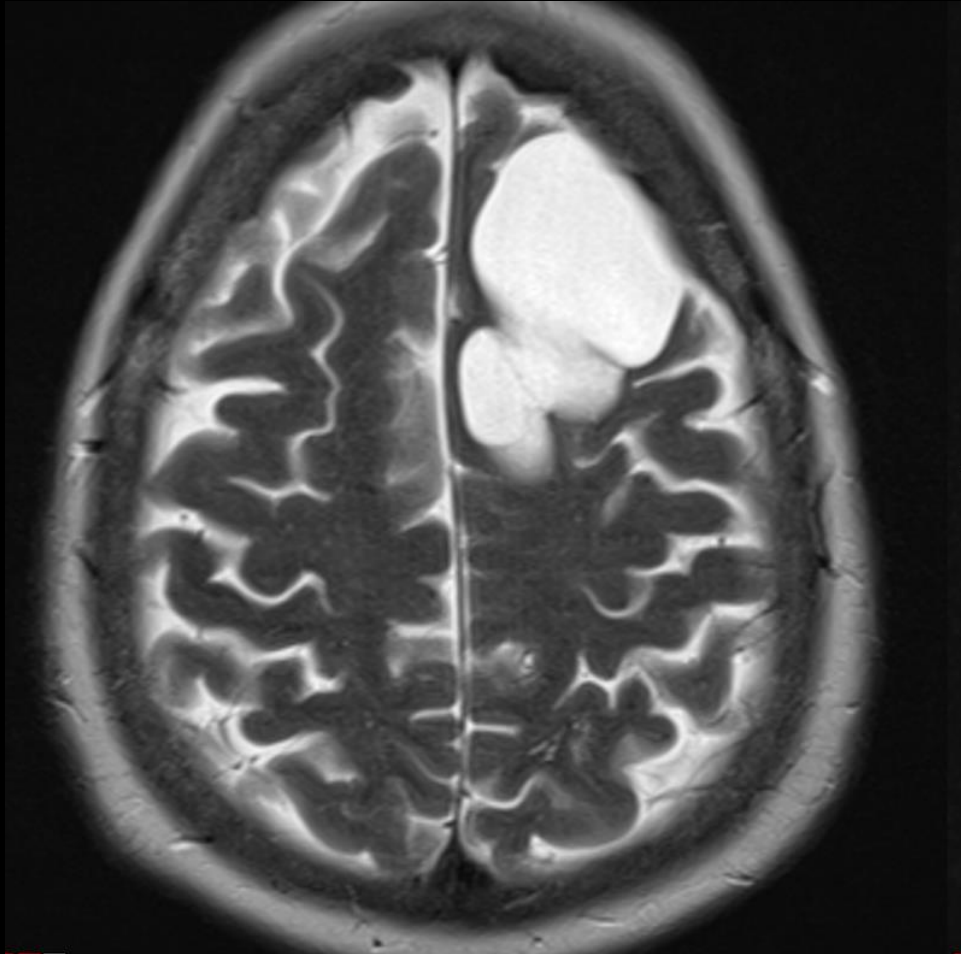
No diffusion restriction seen on DWI (b1000) and ADC Map.

ADC



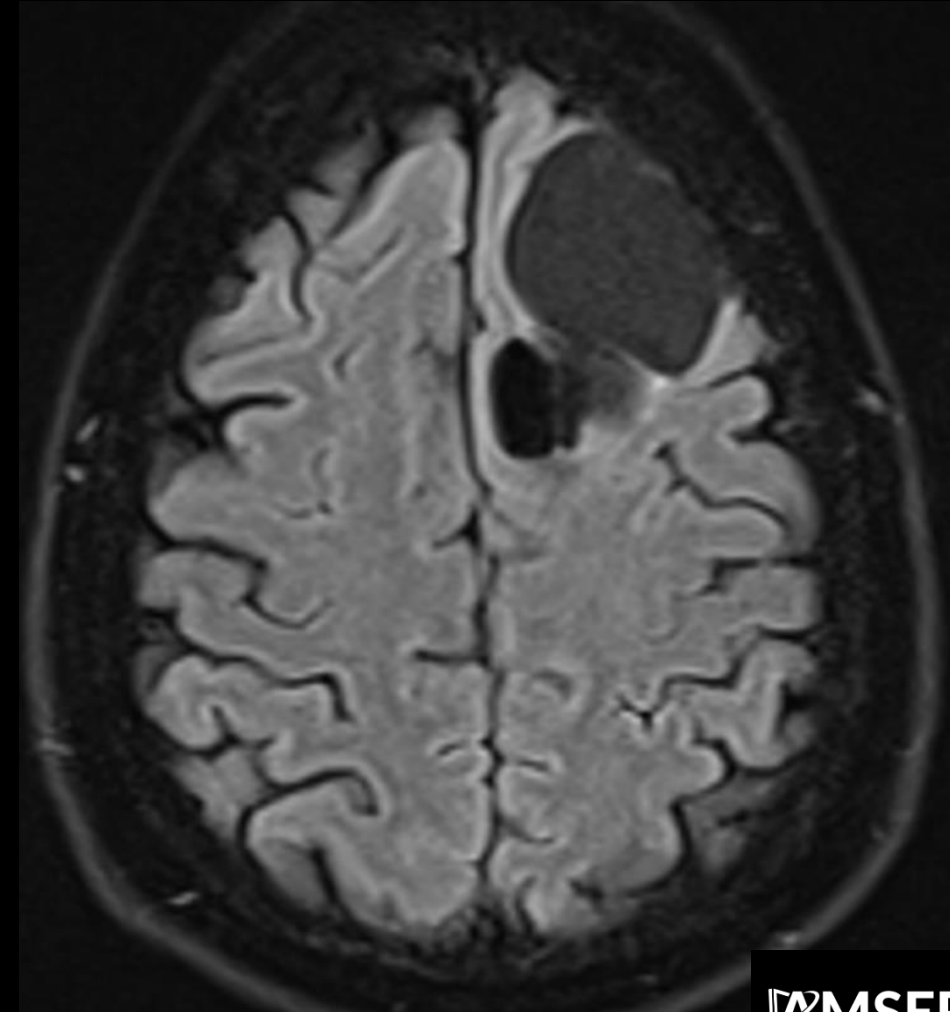
Findings: (labeled)

T2 Axial



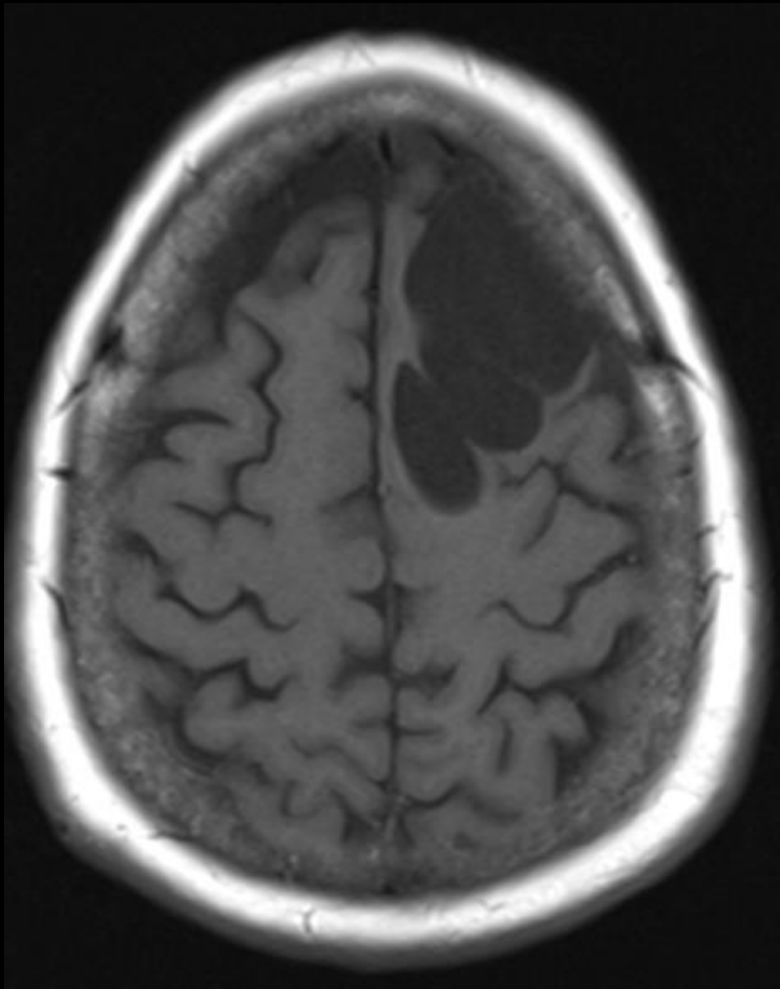
Multi-septate
cystic extra-axial
mass with
incomplete intra-
cystic FLAIR
suppression.

T2 FLAIR



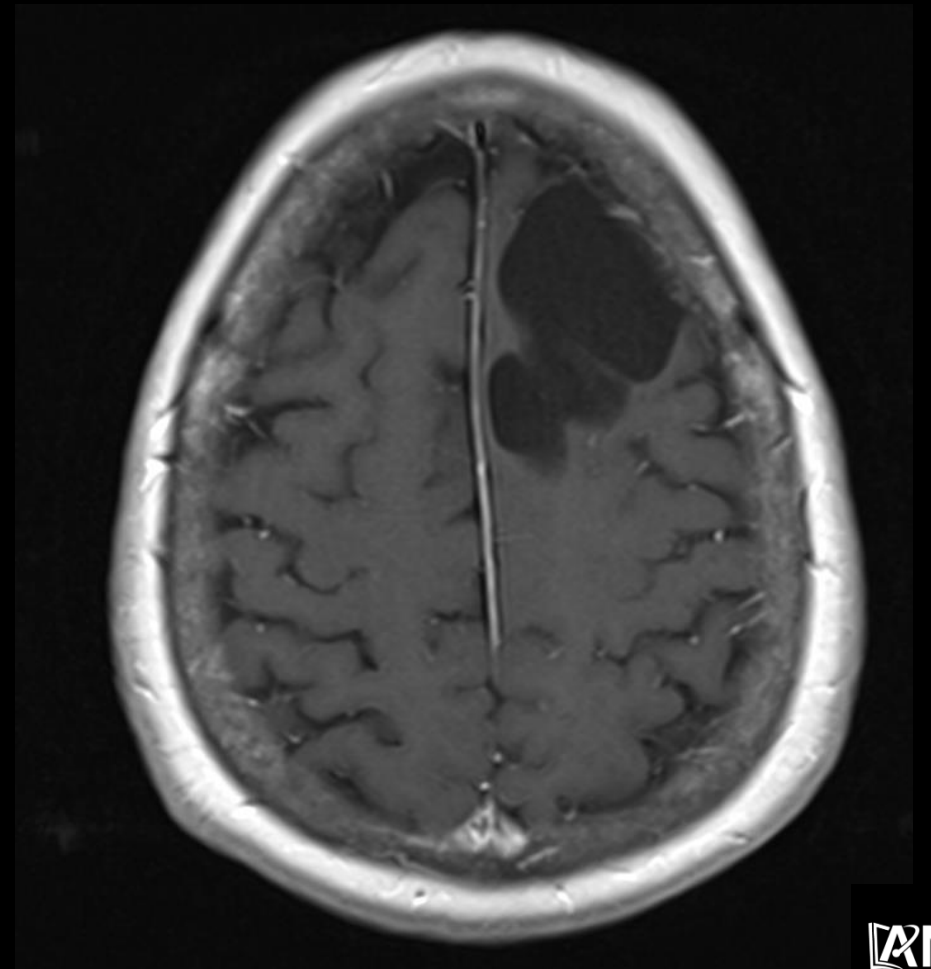
Findings: (labeled)

T1 Pre-contrast

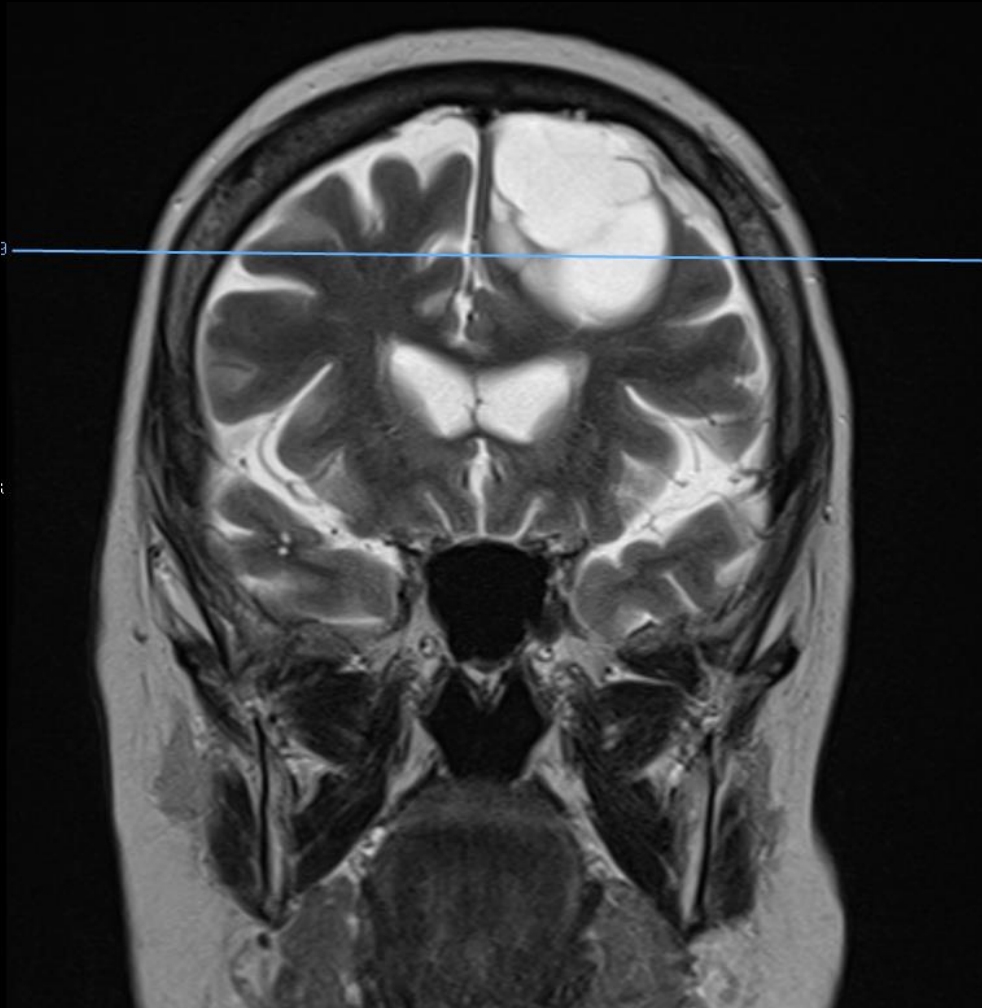


Multi-septate cystic mass with no evidence of intra-cystic or rim-enhancement on T1 contrast image.

T1 Post-contrast



Findings: (labeled)



T2 Coronal again demonstrates an extra-axial, multi-septate cystic mass displacing the gray-white matter junction and with midline shift.

Final Dx:

Neurenteric (Endodermal) Cyst

Case Discussion (1)

Lesion was biopsied!

FINAL DIAGNOSIS AND ATTENDING SIGNATURE

- A. BRAIN, "CYST WALL", EXCISION:
ENDODERMAL CYST. (See comment.)
- B. BRAIN, "FLARE CHANGE", BIOPSY:
ENDODERMAL CYST.
- C. BRAIN, THICKENED TISSUE OVER CORTEX, BIOPSY:
ENDODERMAL CYST.
- D. BRAIN, THICKENED TISSUE OVER CORTEX, BIOPSY:
ENDODERMAL CYST.

FINAL DIAGNOSIS COMMENT

Histological examination of the biopsies received in this case reveal cyst walls lined by a single layer of low cuboidal cells, best visualized in block A1. Immunohistochemical staining is performed on this block (A1) for cytokeratin 7 (CK7) and epithelial membrane antigen (EMA), demonstrating strong immunoreactivity in the cells lining the cyst (positive) for both stains. In the setting of a differential diagnosis of endodermal cyst versus subarachnoid meningotheial cyst, this staining pattern confirms the diagnosis of an endodermal cyst.

Case Discussion (2)

- Neurenteric cysts are rare congenital lesions with endodermal origin¹
 - Typically occur in the subdural space of the spinal cord
 - Very rarely can occur in an intra-parenchymal location
- Supratentorial cystic lesion differential diagnosis²
 - Intra-axial
 - Dysembryoplastic neuroepithelial tumor (DNET)
 - Ganglioglioma
 - Multinodular and vacuolating neuronal tumor
 - Abscess
 - Pleomorphic xanthoastrocytoma
 - Extra-axial
 - Epidermoid cyst
 - Arachnoid Cyst
 - Schwannoma (cystic)

Case Discussion (3)

- Imaging findings²
 - **CT Scan**: will demonstrate hypo to hyperdense mass with no calcium or hemorrhage
 - **MRI**
 - T1 Weighted Imaging will demonstrate a **cystic, extra-axial mass iso or hyperintense to CSF**
 - T2 Weighted Imaging and T2 Flair will typically show a cystic mass with **hyperintensity to CSF**
 - DWI: Demonstrates **no diffusion!**
 - T1 Weight Imaging with Contrast **may show rim enhancement but usually does not enhance**

References:

1. Kalfas F, Scudieri C. Endodermal Cysts of the Central Nervous System: Review of the Literature and a Case Report. Asian J Neurosurg. 2020;15(4):989-996. Published 2020 Dec 21. doi:10.4103/ajns.AJNS_322_19
2. Simon JA, Olan WJ, Santi M. Intracranial neurenteric cysts: a differential diagnosis and review. Radiographics. 1997;17(6):1587-1593. doi:10.1148/radiographics.17.6.9397466