AMSER RAD PATH CASE OF THE MONTH November 2018

28 year old female with olfactory/auditory hallucinations

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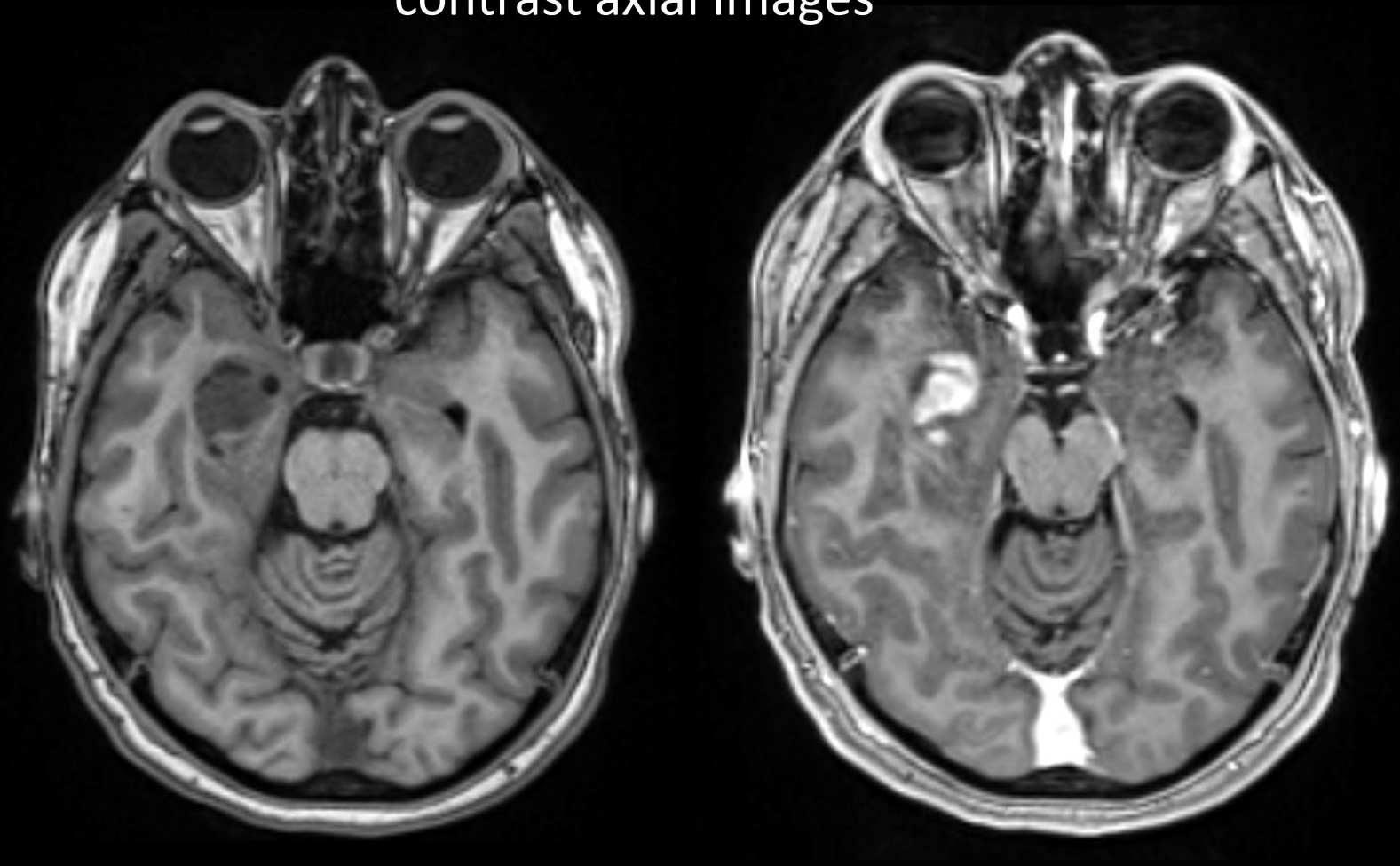
Patient Presentation

- CC/HPI: 28 year old female with history of a right sided temporal brain mass presented to her primary care physician with nausea and worsening olfactory and auditory hallucinations. The mass was previously imaged 8 years prior, and was known to be stable until at least 5 years prior. The hallucinations were believed to be seizures so an EEG and MRI were obtained.

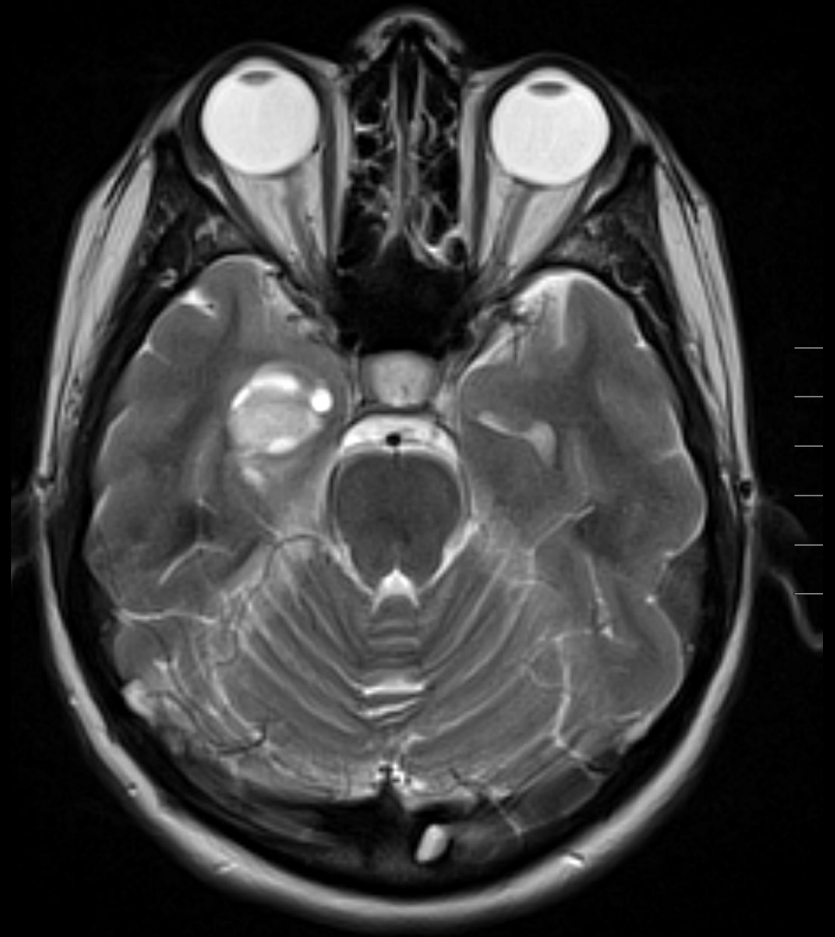
- Targeted physical exam: CN 3-12 intact, muscle strength 5/5 and reflexes 2/4 throughout
- Medical Hx: Anxiety, polycystic ovarian disease
- Past Surgical Hx: none
- Past Medications: none

MRI Results

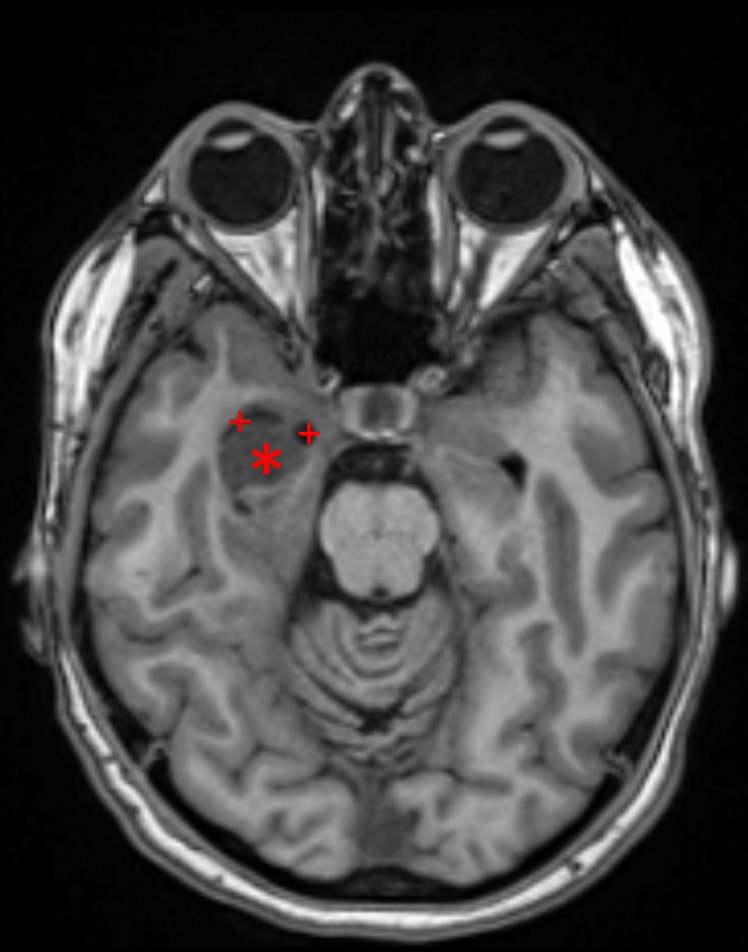
T1 weighted pre- and post-contrast axial images



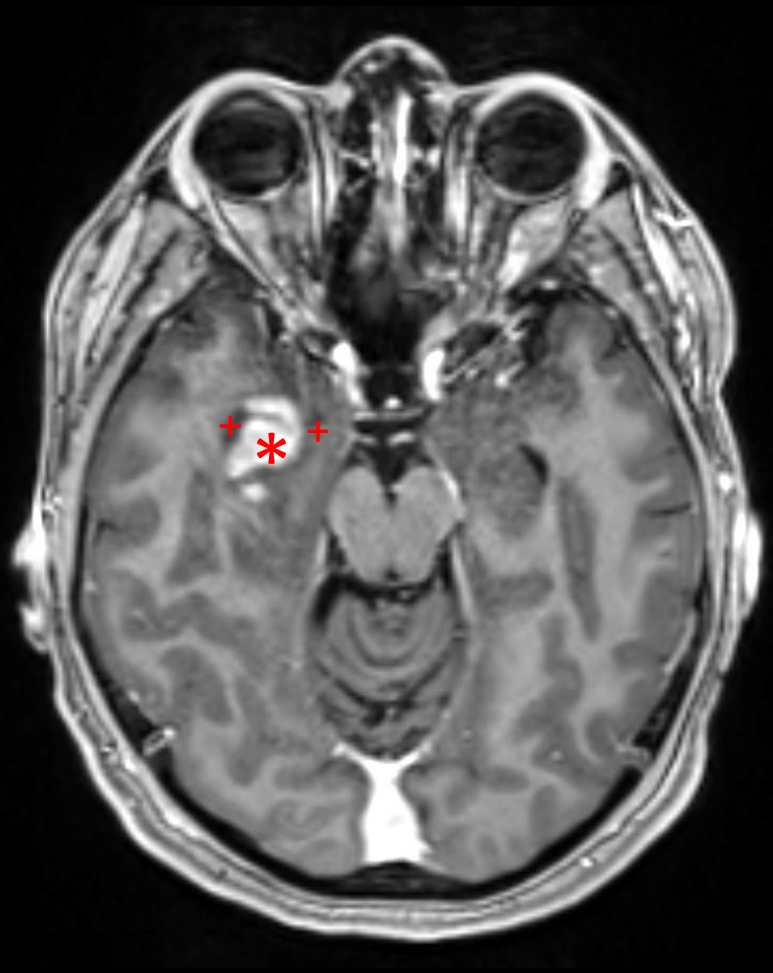
T2 weighted axial image



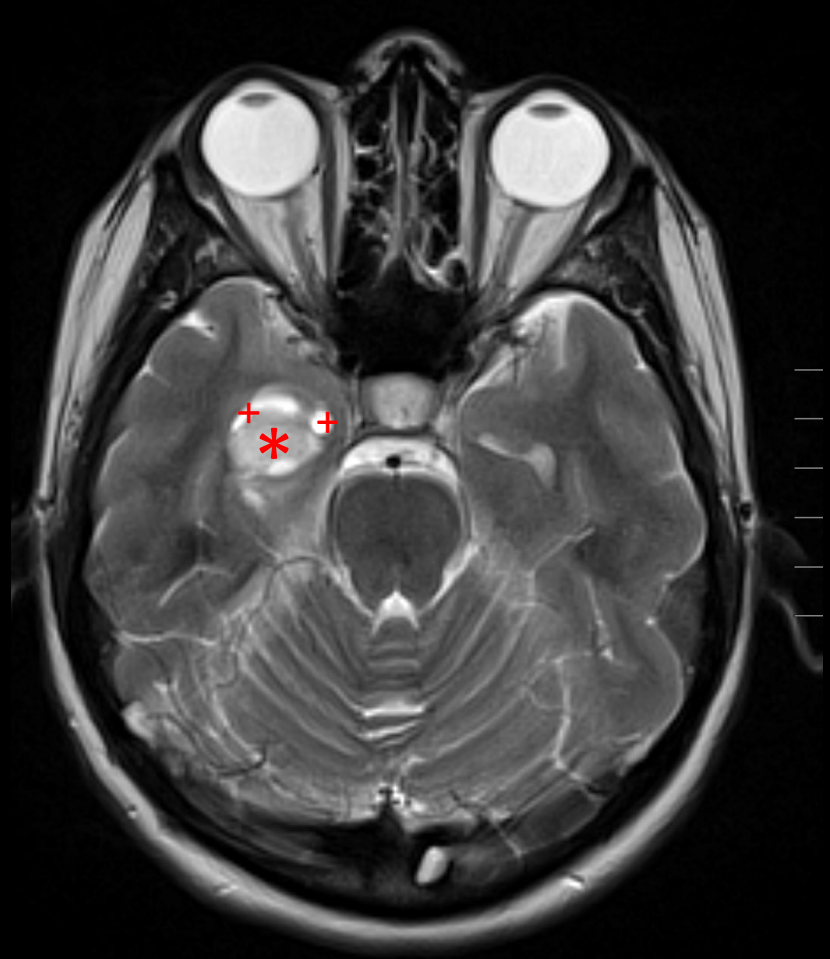
Low signal mass component (*) with cystic components (+) on the pre-contrast.



Enhancing mass component (*) and nonenhancing cystic components (+) post-contrast.



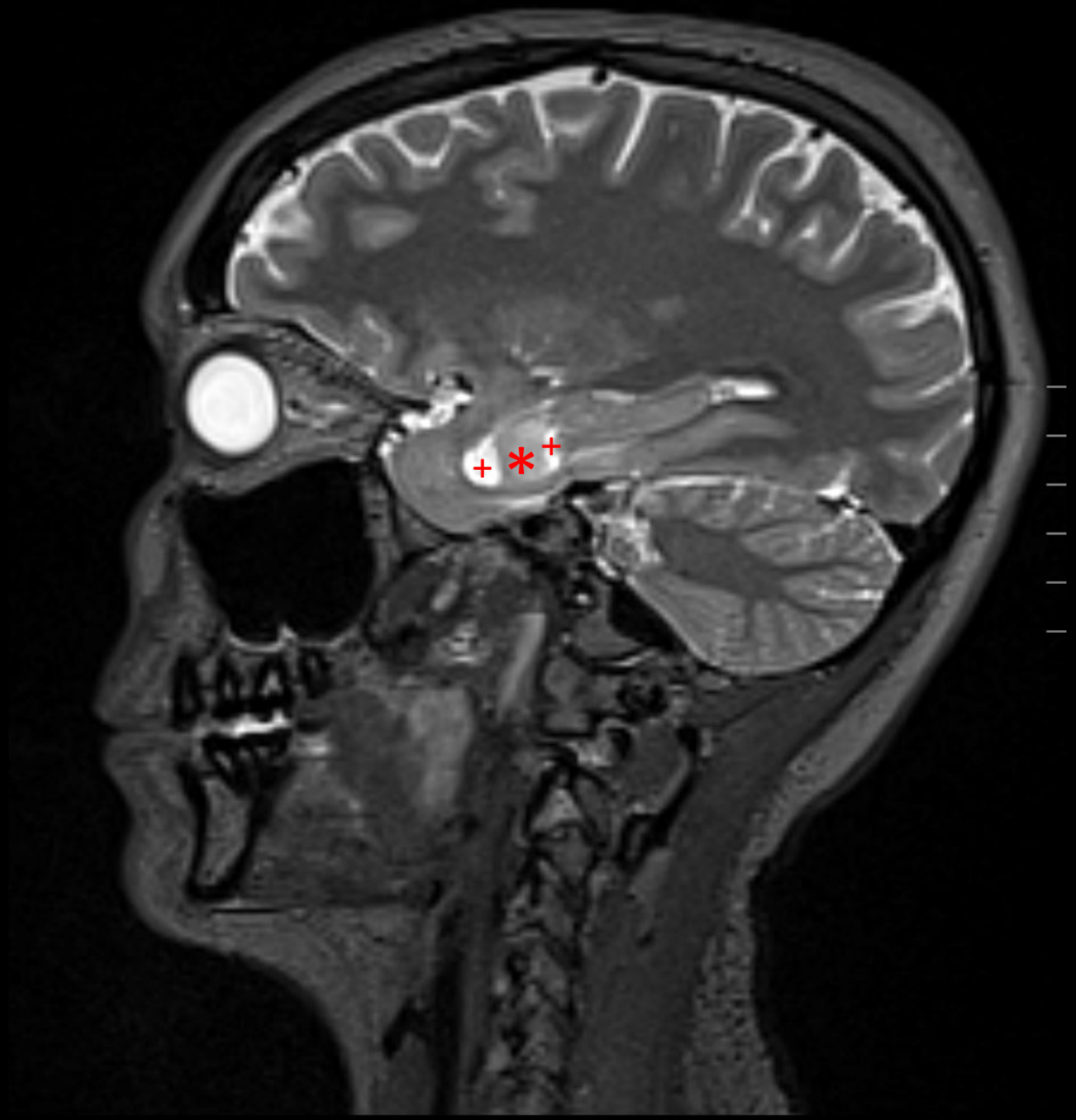
Mass component isointense to gray matter (*) with high signal cystic components (+).



MRI Results

T2 weighted sagittal image

Mass component (*) iso-intense to gray matter, with high signal cystic components (+)



Differential Diagnosis:

- Ganglioglioma
- Dysembryoplastic neuroepithelial tumor (DNET)
- Pleomorphic xanthoastrocytoma (PXA)
- Ependymoma

The Patient's Workup

- First EEG was negative.
- MRI showed enlargement of the right temporal mass.
- At this time, she was put on clonazepam .25mg PRN and lamotrigine 25mg titrated up to 100mg BID.
  - These medications did not improve her symptoms so she was sent to an epilepsy monitoring unit.
- The EMU did not demonstrate new seizures but it was decided that it would be best for the tumor to be removed due to its recent growth.
- Wada testing showed bilateral memory areas in the hippocampus.

The Surgery and Pathology Report

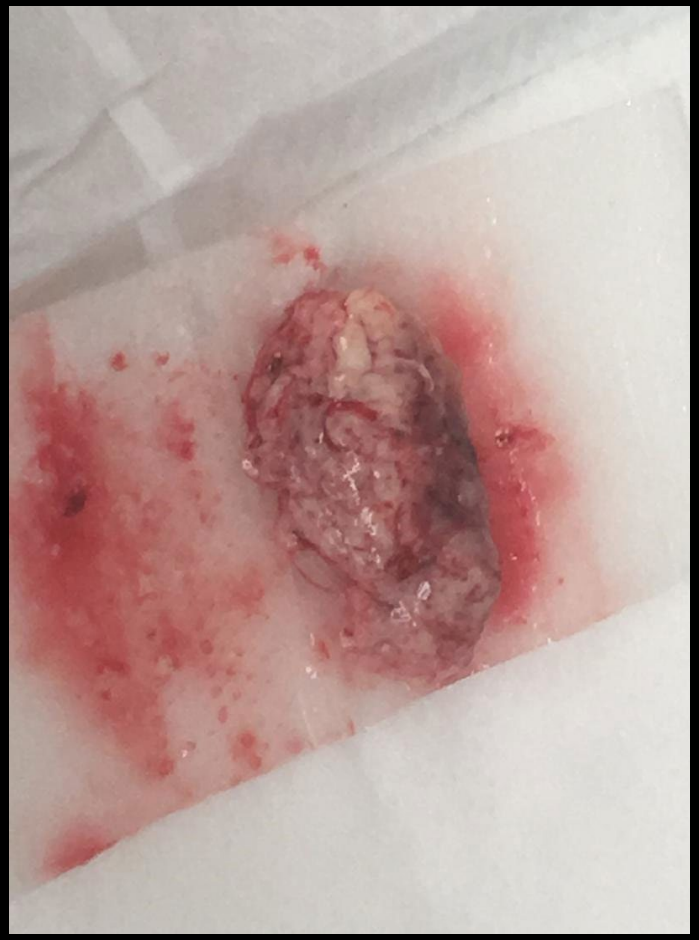
- The patient underwent craniotomy with excision of the mass and right hippocampus.

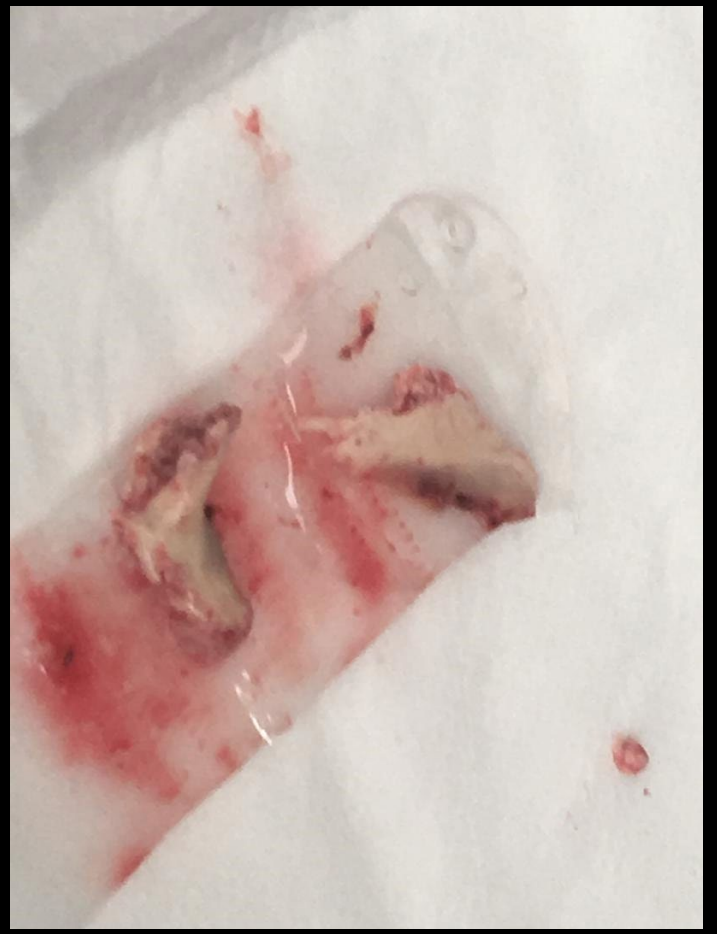
- Pathologic analysis described a focally densely calcified neuroepithelial neoplasm with both glial and neuronal differentiation. It also contained a glioneuronal component consisting of oligodendrocyte-like cells and floating neurons. Cortical dysplasia was also present. These findings were consistent with a dysembryoplastic neuroepithelial tumor (DNET).

Gross Pathology

This tumor measured 2.5 x 1.2 x 0.4 cm.

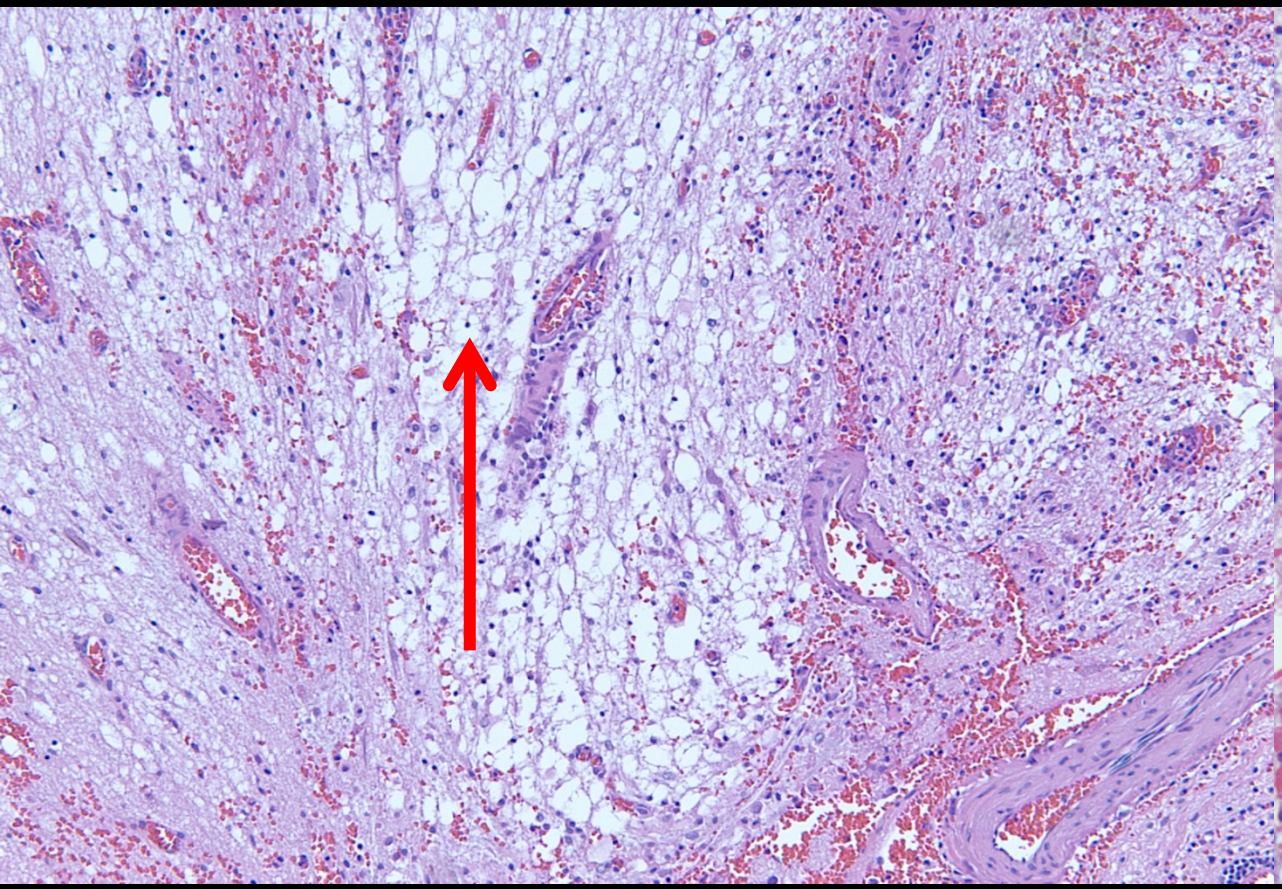
On the cut image, portions of gray and white matter are demonstrated.





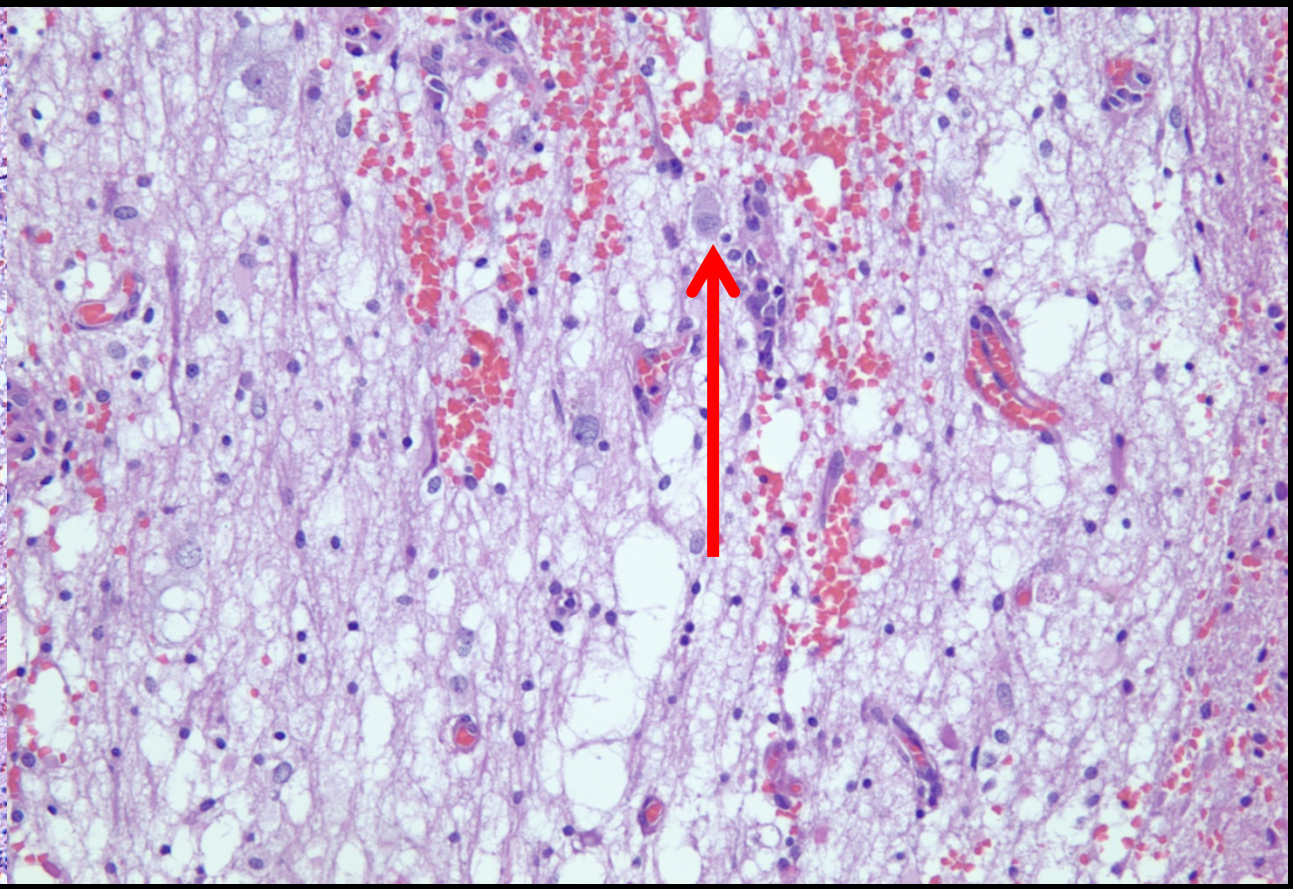
Histopathology

Cystic area with background mucin located centrally which give a “fried egg” appearance of the oligodendrocyte-like cells.



H&E 10x

Eosinophilic granular bodies in the background.



H&E 20x

Dysembryoplastic Neuroepithelial Tumor (DNET)

- This is a benign tumor which grows very slowly. It is typically diagnosed in children and young adults following a seizure work-up.
- It is usually found in the temporal lobe (65%) with the frontal lobe being the second most common location (20%).
- They are usually visible on the brain intra-operatively and when sectioned may have a heterogeneous or gelatinous appearance with nodules of firm tissue.
- There is typically focal cortical dysplasia present.

Dysembryoplastic Neuroepithelial Tumor (DNET)

- On microscopy, they are observed to be mixed glioneural neoplasms with a multinodular architecture and a heterogeneous cellular composition.
  - There will often be “bundles” of axons from oligodendrocyte-like cells arranged at right angles to the overlying cortex.
    - These cells are usually S100 and OLIG2 positive.
  - Between the columns of axon “bundles,” floating neurons and stellate astrocytes are seen.
    - These stain positive for GFAP and NeuN respectively.
- To distinguish from low grade astrocytomas and oligodendrogliomas, we stain for IDH, TP53, and 1p19q co-deletion mutations, which are negative in DNET.

Radiologic Findings

- CT scans will typically show a low density mass with possible calcifications adjacent to hemorrhagic areas.
  - It typically does not enhance.
  - If growing near bone, there may be scalloping without erosion.
- MRI scans will show:
  - T1 hypointense lesion that may enhance (about 30% of the time).
  - T2 will show a high signal with “soap bubble” appearance.
  - FLAIR will show mixed signal with bright ring sign and attenuation of some of the “soap bubbles.”
  - There is no restricted diffusion.

References

- https://app.statdx.com/document/dnet/a7e18979-334e-47cc-9f92-b61bcd6a5a12?searchTerm=DNET
- https://www-uptodate-com/contents/uncommon-brain-tumors
- http://www.pathologyoutlines.com/topic/cnstumorDNET.html