

AMSER Case of the Month

August 2023

21-year-old male with intractable headache
and diplopia with left lateral gaze

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Patient Presentation

- HPI: 21 y/o M presents to his PCP complaining of **worsening daily intractable HAs** x 2 months. **Associated diplopia** especially on left lateral gaze, and difficult reading as well as increased fatigue/lethargy. No other concerning neurologic symptoms: no changes in weakness/sensation/numbness. No dizziness/lightheadedness.
- PMHx: None
- PE:
 - Vitals: **T: 36.9 C (Oral) HR: 69(Peripheral) BP: 143/85 SpO2: 100% on RA**
 - HEENT: Diplopia in the left lateral visual fields, intact extraocular movements, pupils PEERLA
 - Neuro: A&O x3, normal strength/gait. Absent cerebellar signs/pronator drift.
- Pertinent labs: None

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

Clinical Condition:

Headache

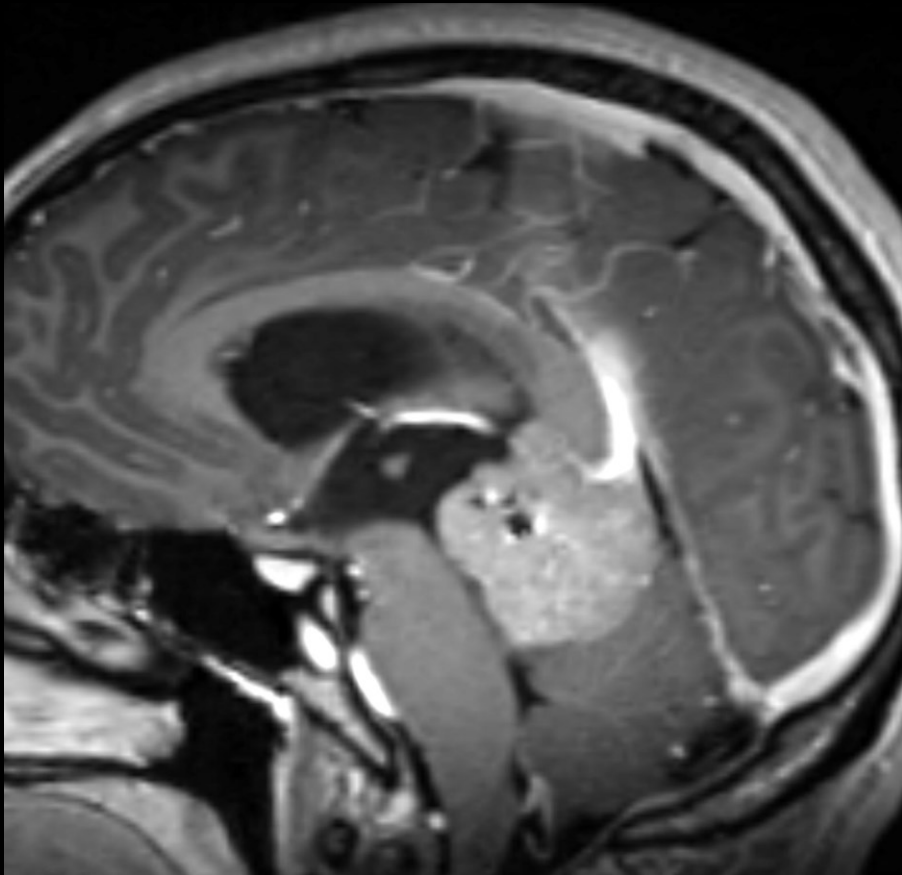
Variant 7:

Headache with one or more of the following “red flags”: increasing frequency or severity, fever or neurologic deficit, history of cancer or immunocompromise, older age (>50 years) of onset, or posttraumatic onset. Initial imaging.

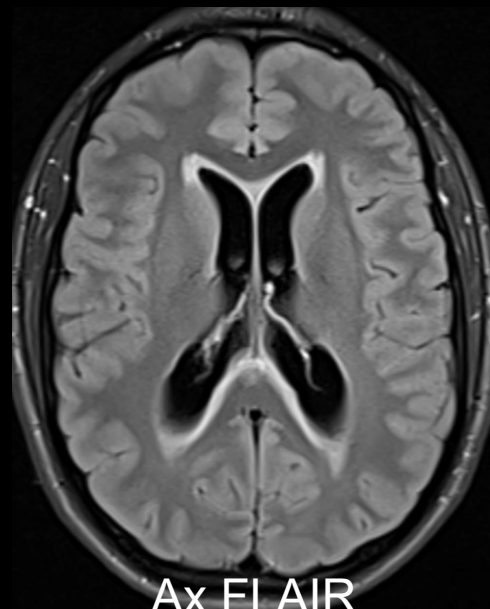
Procedure	Appropriateness Category	Relative Radiation Level
MRI head without and with IV contrast	Usually Appropriate	○
MRI head without IV contrast	Usually Appropriate	○
CT head without IV contrast	Usually Appropriate	⊕⊕⊕
Arteriography cervicocerebral	Usually Not Appropriate	⊕⊕⊕
MRA head with IV contrast	Usually Not Appropriate	○
MRA head without and with IV contrast	Usually Not Appropriate	○
MRA head without IV contrast	Usually Not Appropriate	○
MRI head with IV contrast	Usually Not Appropriate	○
MRV head with IV contrast	Usually Not Appropriate	○
MRV head without and with IV contrast	Usually Not Appropriate	○
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CTV head with IV contrast	Usually Not Appropriate	⊕⊕⊕

This imaging modality was ordered by the PCP

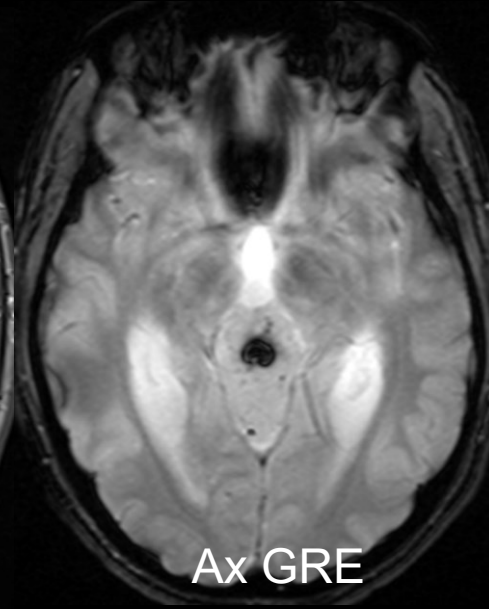
Findings (unlabeled)



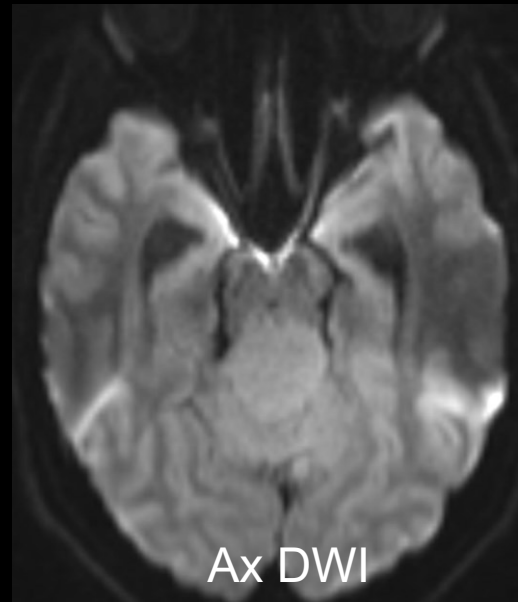
Sagittal T1 MPRAGE post



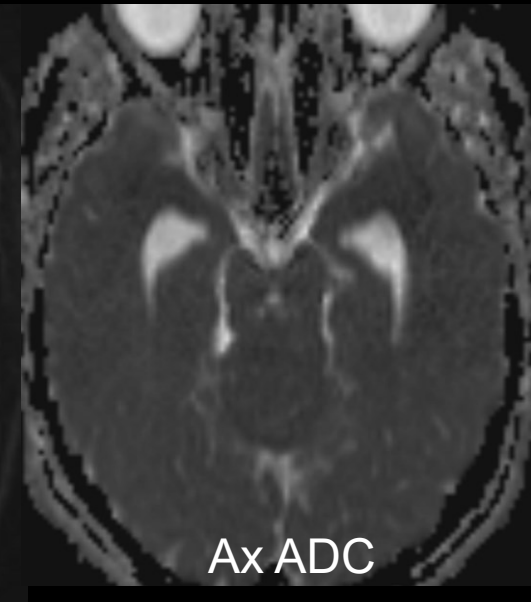
Ax FLAIR



Ax GRE

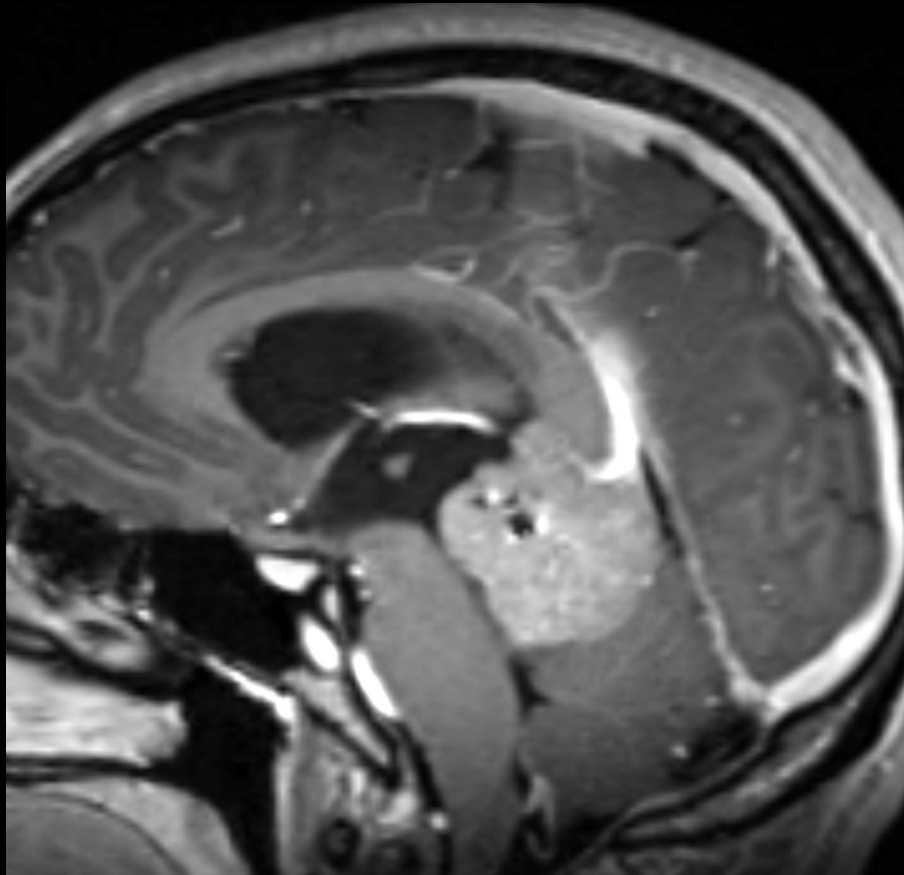


Ax DWI

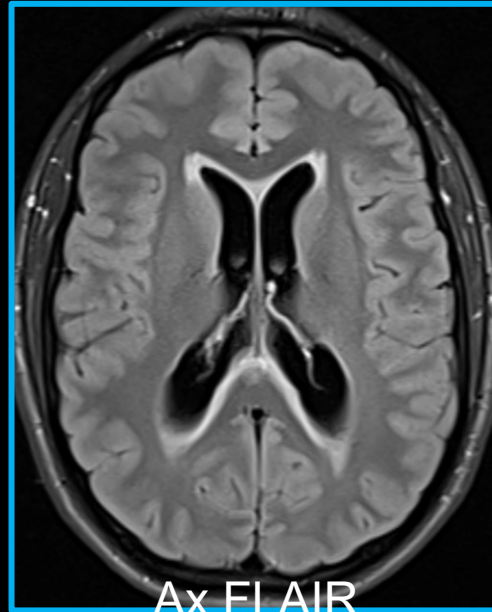


Ax ADC

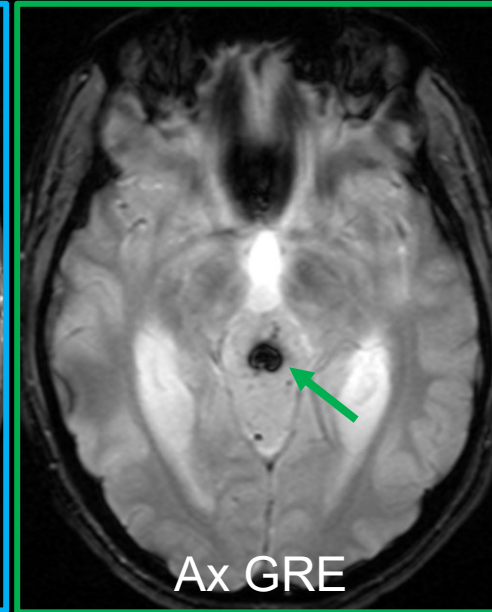
Findings (labeled)



Solidly enhancing mass in the pineal region
Central areas of calcifications



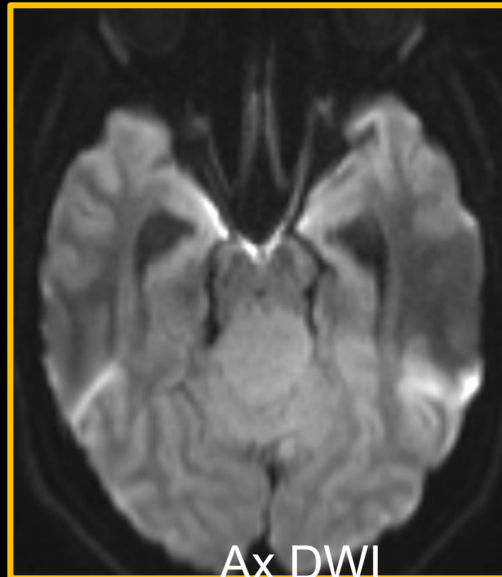
Ax FLAIR



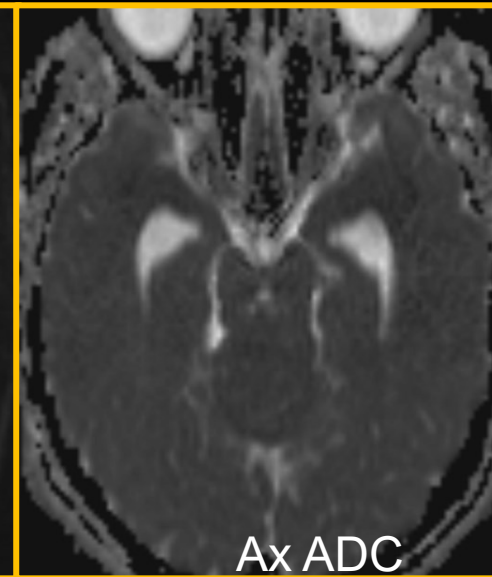
Ax GRE

The mass causes obstructive hydrocephalus with tranependymal flow of CSF

GRE image shows the mass “engulfing” central calcifications



Ax DWI



Ax ADC

Mild restricted diffusion due to increase cellularity of the mass

Final Dx:

Pineal Gland Germinoma

After neurosurgical consult, the patient was scheduled for endoscopic 3rd ventriculostomy and tumor biopsy

However, the patient presented to ER 2 days later with worsening fatigue, nausea, and headaches, concerning for worsening hydrocephalus

Consideration of Additional Imaging

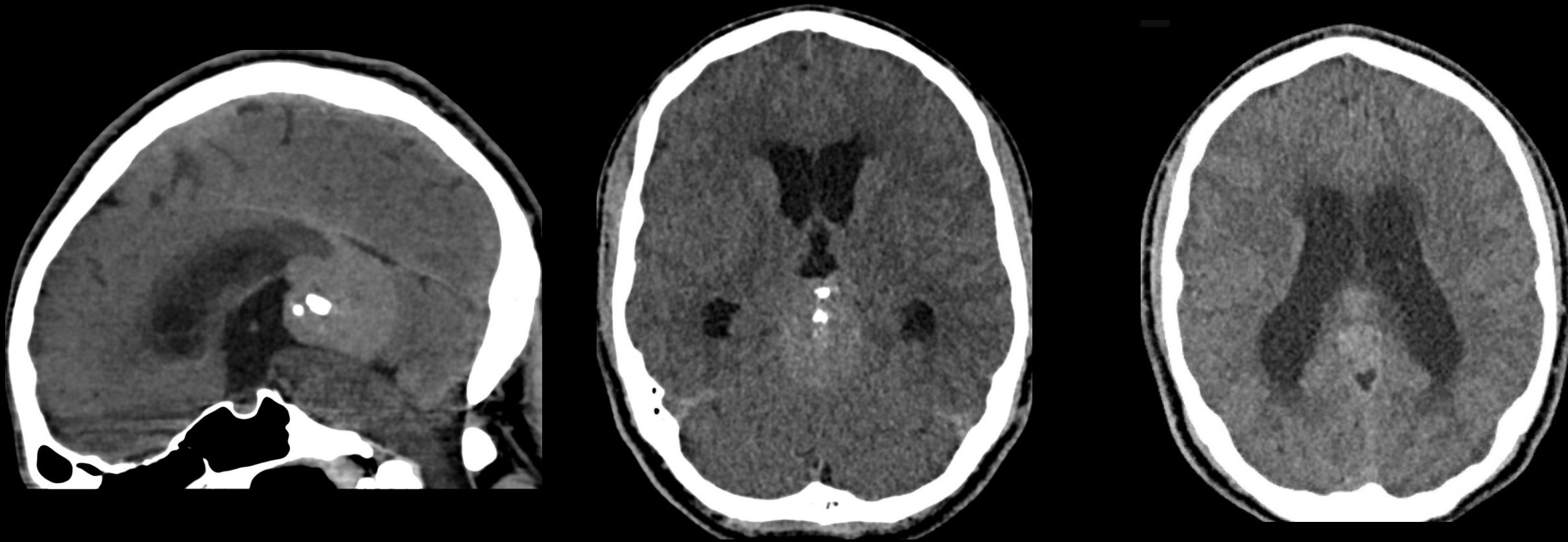
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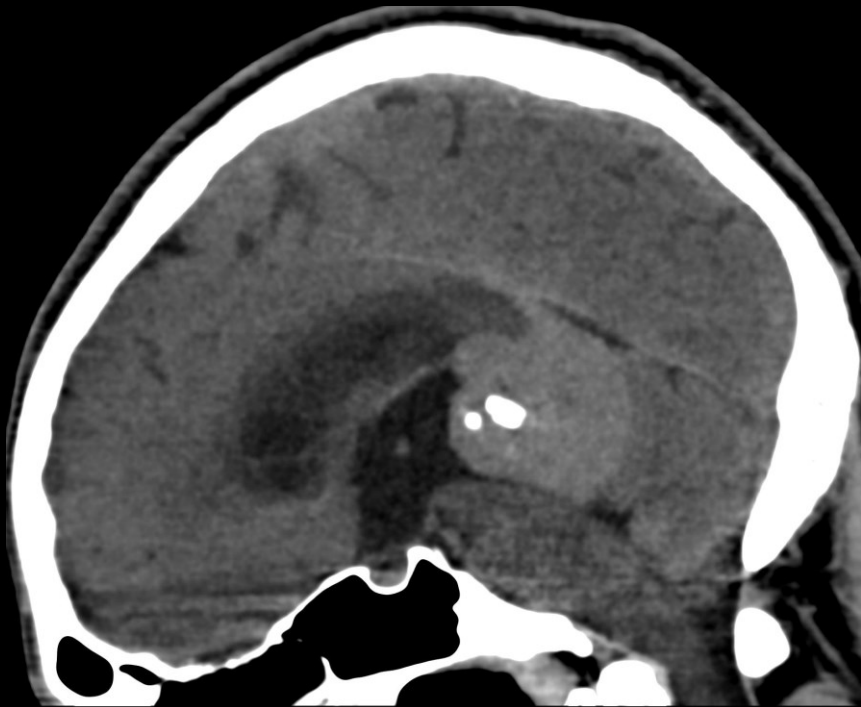
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This imaging modality was ordered by the ER physician

Findings: (unlabeled)



Findings: (labeled)



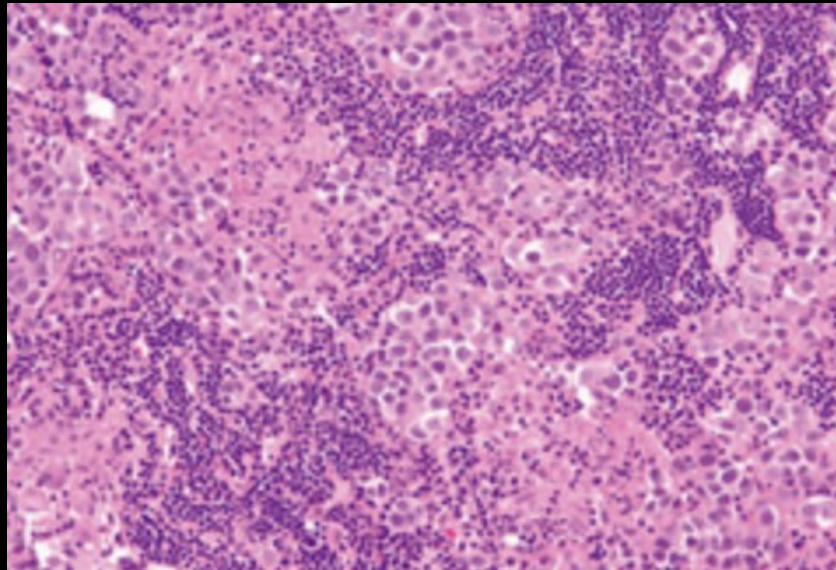
Solid mass in the pineal gland region that is hyperdense due to increased cellularity of the tumor
Mass is “engulfing” central calcifications

Stable obstructive hydrocephalus with transependymal flow of CSF

Intraoperative Pathology (different patient)



Gross pathology:
Smooth and bosselated
external surface, soft and
fleshy interior. Cream colored



Uniform round cells with vesicular
nuclei and clear/finely granular
cytoplasm that is eosinophilic.
Stroma contains lymphocytes.
Sarcoid like granulomas

Immunohistochemistry: CD117,
PLAP, β -HCG (worse prognosis)

Case Discussion

- Pineal gland tumors are less commonly of pineal tissue origin
- Pineal gland germinomas account for 50% of pineal tumors.
- Main differential diagnoses are pineoblastoma and pineocytoma
 - Germinomas “engulf” central calcifications
 - Pineoblastomas and pineocytomas “explode” the calcifications which tend to be at the periphery of the mass
- Tends to affect children/young adults (≤ 20 years at time of dx)
- Males are more commonly affected (M:F 13:1)
- 5-year survival rate is 90%
- Multifocal/disseminated lesions are associated with poorer prognosis
- Left untreated, could lead to seizures, obstructive hydrocephalus and CSF metastasis (leptomeningeal spread)

Case Discussion

- Clinical presentations: varied progression, signs and symptoms of mass effect, neuroendocrine dysfunction, neuro-ophthalmologic dysfunction, compression of nearby structures, tumor invasion, malignancy.
- Abnormal pubertal development in children (early/delayed)
- Central diabetes insipidus (& other hypothalamic pituitary insufficiencies)
- Narcolepsy/other sleep disturbances
- Symptoms of increased intracranial pressure secondary to hydrocephalus: HA, n/v, drowsiness, behavior dysfunction, visual disturbances (double vision)
- Ophthalmologic signs due to brainstem dysfunction: Parinaud syndrome, skew deviation, third/fourth CN palsies
- Associated symptoms: seizures, hearing loss, slowed speech, presyncopal episodes

Case Discussion

- Gold standard for imaging for pineal gland germinomas is MRI: Heterogeneous features, often solid or solid/cystic masses with engulfed calcifications
 - T1: isointense to adjacent brain.
 - T2: isointense to adjacent brain
 - T1 with gadolinium contrast: vivid homogeneous enhancement
 - DWI: restricted diffusion due to high cellularity
- CT scan is also diagnostic of pineal germinoma.
 - Large midline mass in pineal region: hyperdense compared to normal brain, vivid contrast enhancement.
 - Calcification: “engulfed” pineal calcification
 - +/- Obstructive hydrocephalus

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- Themes, U. F. O. “Pineal Region Masses.” *Radiology Key*, 22 Jan. 2016, <https://radiologykey.com/pineal-region-masses/>.
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