AMSER Case of the Month August 2023

18-year-old with headache

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Patient Presentation

- 18-year-old male with worsening headaches and fatigue.
- Physical and neuro exam unremarkable.
- Labs were normal.

What Imaging Should We Order?

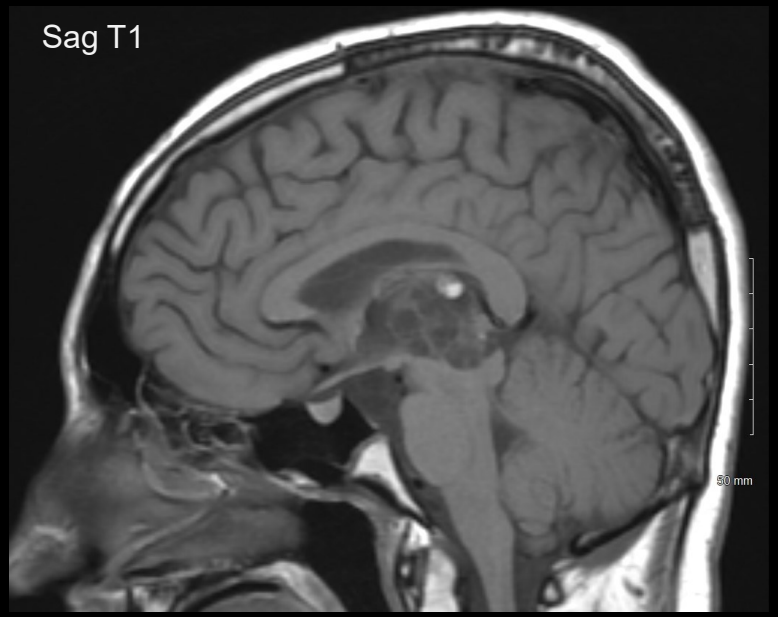
Select the applicable ACR Appropriateness Criteria

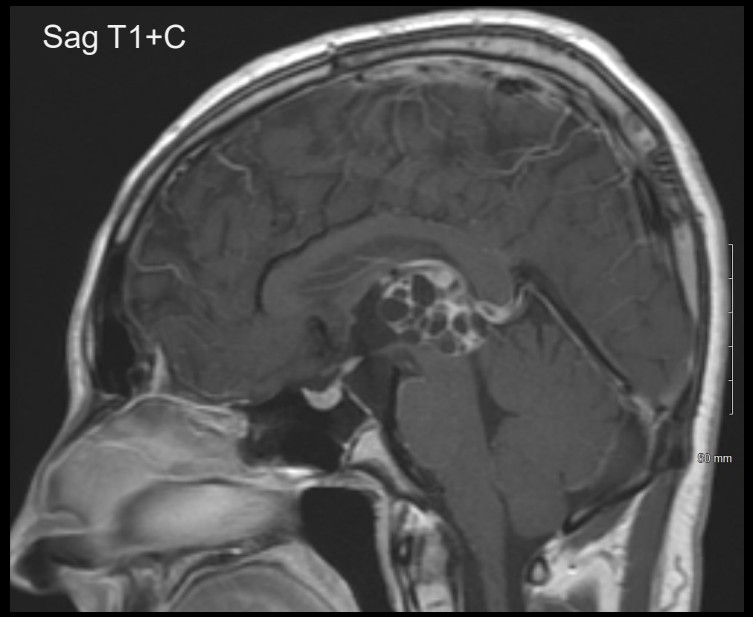
Variant 7: **Headache with one or more of the following "red flags": increasing frequency or severity, fever or neurologic deficit, history of cancer or immunocompromise, older age (>50 years) of onset, or posttraumatic onset. Initial imaging.**

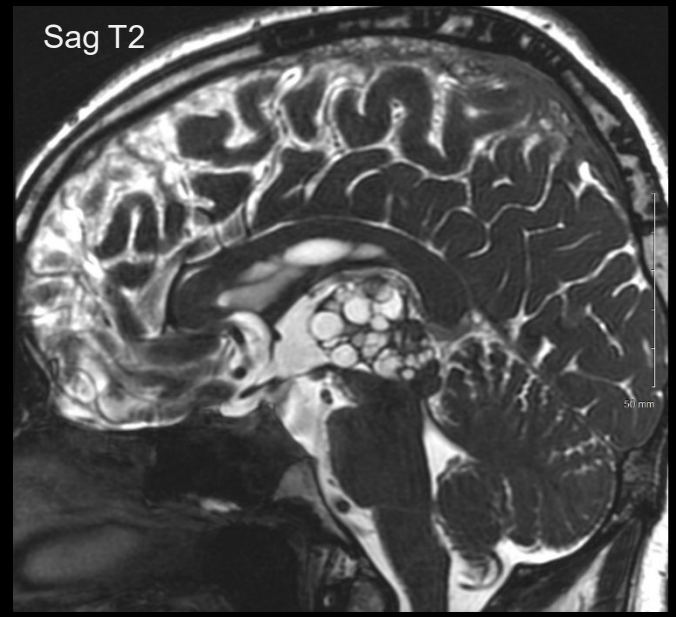
Procedure	Appropriateness Category	Relative Radiation Level
MRI head without and with IV contrast	Usually Appropriate	O
MRI head without IV contrast	Usually Appropriate	O
CT head without IV contrast	Usually Appropriate	☢☢☢
Arteriography cervicocerebral	Usually Not Appropriate	☢☢☢
MRA head with IV contrast	Usually Not Appropriate	O
MRA head without and with IV contrast	Usually Not Appropriate	O

This exam was ordered by the PCP

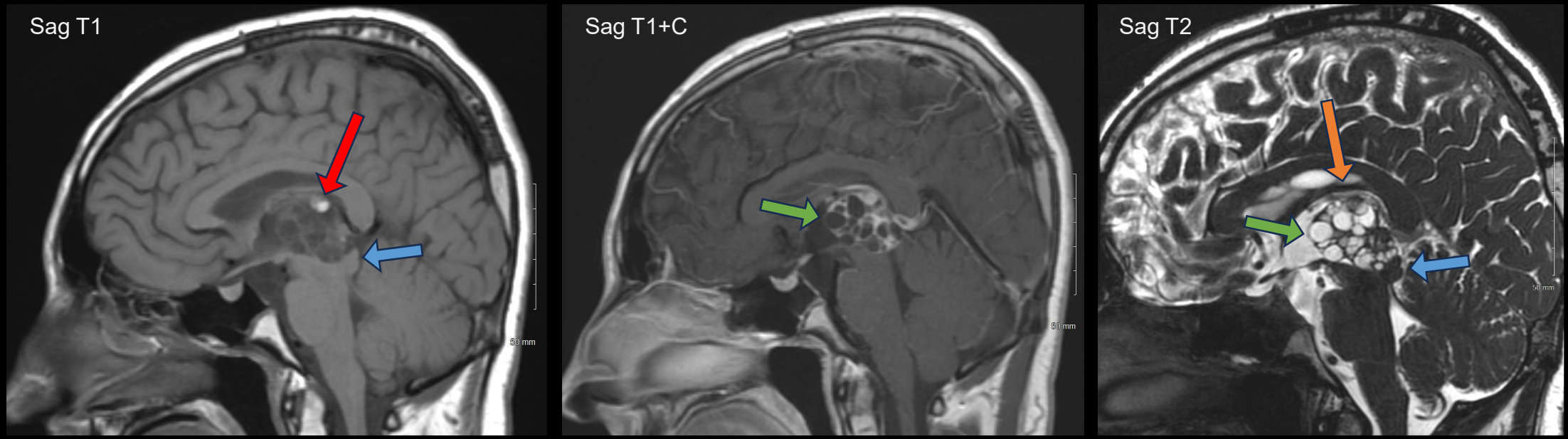
Findings: (unlabeled)





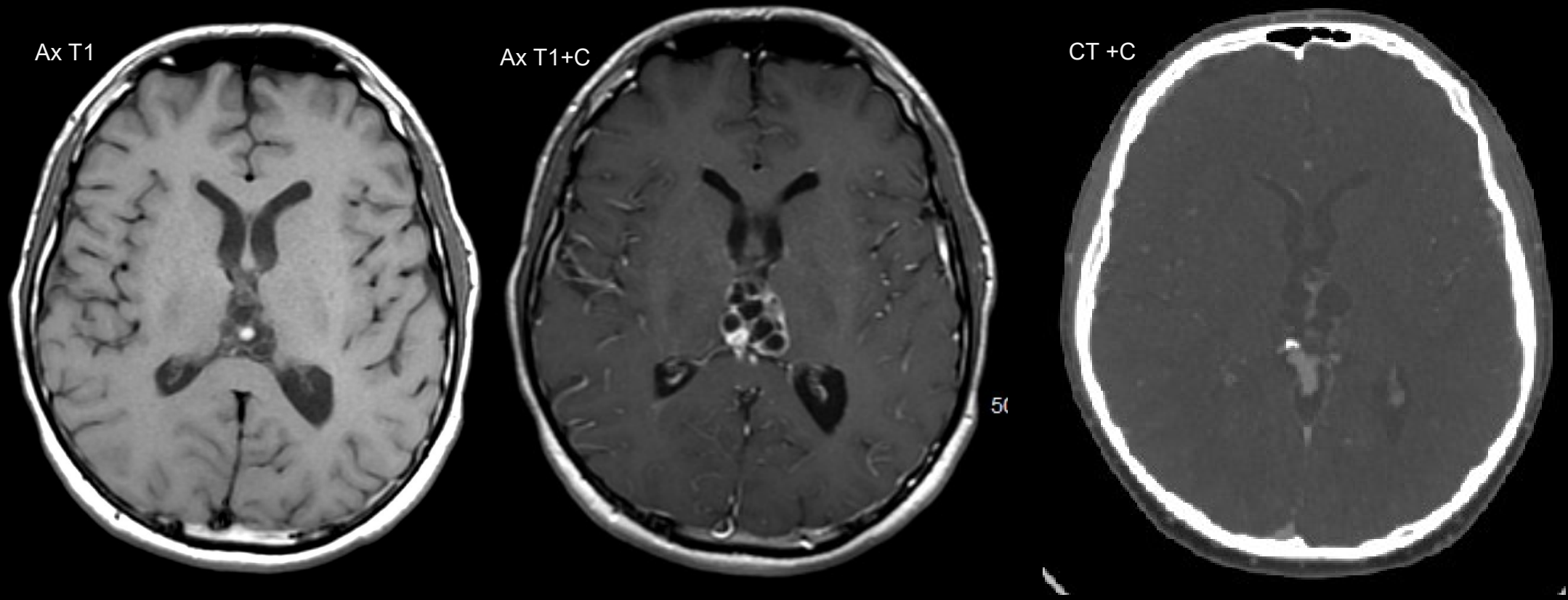


Findings: (labeled)

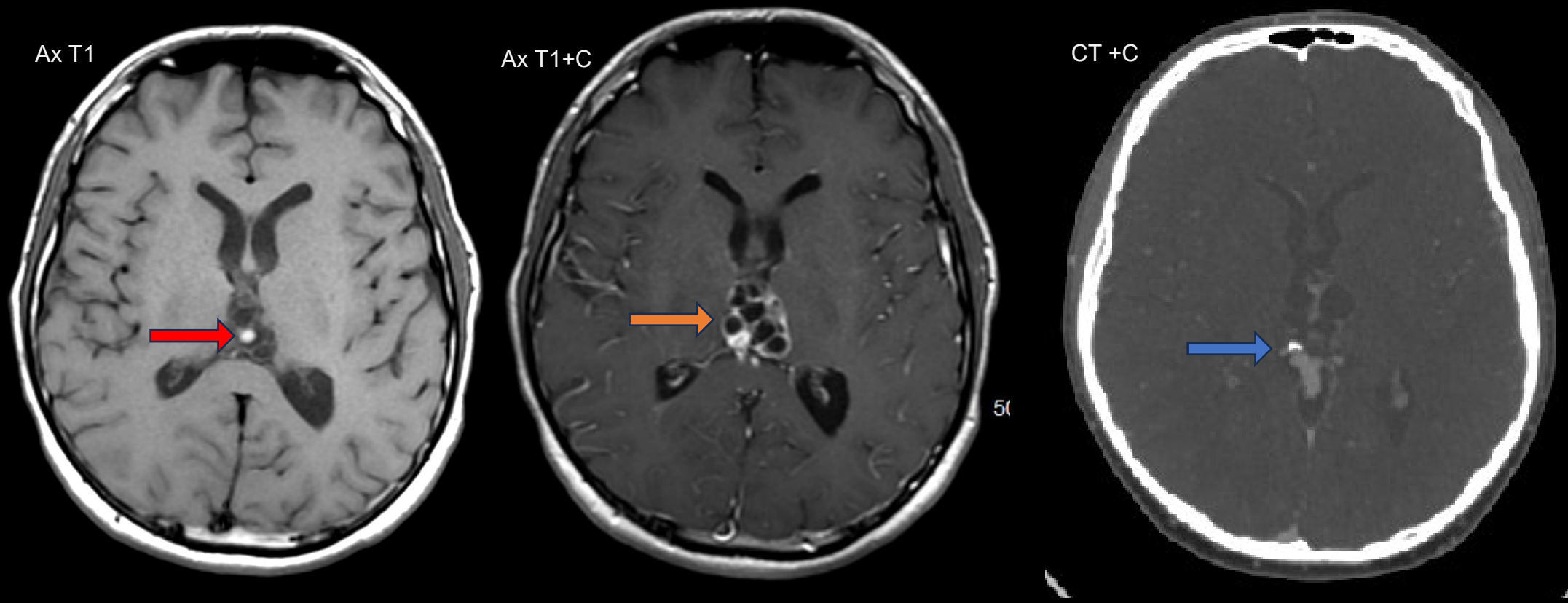


Sagittal MR imaging shows a multicystic enhancing mass in the pineal region containing a small focus of fat.
The mass deforms the tectal plate of the midbrain and superiorly displaces the fornices.

Findings (unlabeled)



Findings (labeled)



Axial MR images again demonstrate the multicystic enhancing mass that deforms the medial thalami and a small focus of fat on T1WI. There is also small peripheral calcification on CT.

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Final Dx:

Nongerminomatous germ cell tumor (NGGCT) - Mature Teratoma

Case Discussion

- Epidemiology
  - 2 types of GCTs are germinoma (60-80%) and non-germinoma (NGGCT).
    - NGGCT include yolk sac, embryonal, choriocarinoma, teratoma, and mixed cell types
  - 90% of pineal NGGCTs occur in males, whereas suprasellar NGGCTs have no definitive gender predilection.
  - The peak incidence of extra-axial teratomas (i.e. pineal) is in adolescence. Intra-axial teratomas tend to present in the neonatal period.
- Pathology
  - Germinomas and NGGCTs contain elements from all 3 embryonic cell layers (ectoderm, mesoderm, and endoderm).
  - Teratomas may be pre-operatively suggested by the presence of serum alpha fetal protein (AFP) and carcinoembryonic antigen (CEA).

Case Discussion

- Clinical Presentation
  - Presenting symptoms for pineal NGGCT are typically related to mass effect.
    - obstructive hydrocephalus at level of cerebral aqueduct → elevated intracranial pressure
    - compression of superior colliculi → upward gaze palsy (Parinaud syndrome)
  - Suprasellar tumors typically present with hypothalamic/pituitary dysfunctions such as delayed or precocious puberty or growth hormone deficiency.
- Non-imaging studies
  - Tumor markers are helpful for pre-operative diagnosis.
    - Mainly AFP, CEA, and human chorionic gonadotropin (hCG)
    - Germinomas typically lack both AFP & hCG, yolk sac tumors have only high AFP, choriocarcinomas have only high hCG, and teratomas have variable results.

Case Discussion

- Radiologic Findings
  - NGGCT are often heterogeneous and cystic which is in contrast to GCT which are typically homogenous and solid.
  - NGGCTs have mixed components such as cysts and calcification. Teratomas will classically show various amounts of fat, calcification, and soft tissue.
  - Germinomas and some NGGCTs are capable of seeding throughout the CNS.
  - Differential diagnoses: Craniopharyngioma, PNET, pineoblastoma, dermoid
- Treatment
  - NGGCTs have a worse prognosis than GCTs and usually require multimodal therapy, including resection, chemotherapy, and radiation.

References:

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