AMSER Case of the Month
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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

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
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<tbody>
<tr>
<td>MRI head without and with IV contrast</td>
<td>Usually Appropriate</td>
<td>0</td>
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<tr>
<td>MRI head without IV contrast</td>
<td>Usually Appropriate</td>
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<td>CT head without IV contrast</td>
<td>Usually Appropriate</td>
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<td>Arteriography cervicocerebral</td>
<td>Usually Not Appropriate</td>
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<tr>
<td>MRA head with IV contrast</td>
<td>Usually Not Appropriate</td>
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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)

Ax T1

Ax T1+C

CT +C
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.
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, choriocarcinoma, 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:


