Imaging of Pineal Masses

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Clinical Presentation: Patient 1

• A 26 year-old man with a past history of chronic headaches, previous Percocet addiction and asthma presented to the ED at BIDMC, after transfer from an outside hospital, with an increasing headache and nausea over a period of several days.

• His chronic headaches began 8 years prior to presentation.

• Vital signs were stable, and physical exam was unremarkable.
Outside Hospital Findings

• At the outside hospital, he had received a CT head due to concern for acute head processes.

• A calcified pineal mass and hydrocephalus in the lateral ventricles and the 3\textsuperscript{rd} ventricle were found.
Outside Hospital Imaging: Patient 1

Patient 1. Non-Contrast Axial CT Head. Image taken day before presentation. Key Finding is a calcified mass measuring 1.6 cm x 1.1 cm at region of pineal gland. (From PACS, BIDMC)

Patient 1. Non-Contrast Axial CT Head. Image taken day before presentation. Key finding is hydrocephalus in both lateral ventricles. (From PACS, BIDMC)
BIDMC Imaging Work-Up: Patient 1

• MR imaging was performed at BIDMC to better characterize the pineal mass.
Imaging on Day 1: Patient 1

Patient 1. Non-Contrast Sagittal T1-Weighted MR Head. Image taken on day 1 of presentation. There is hydrocephalus, with dilated lateral and 3rd ventricles but a normal 4th ventricle. Also, a hypo-intense to iso-intense signal to gray matter pineal mass along the posterior margin of the 3rd ventricle narrowing the Sylvian aqueduct is seen. The mass is estimated to be 2 cm x 2 cm x 1.1 cm (4400 mm$^3$), which far exceeds the normal dimensions and volume of the pineal gland.

(From PACS, BIDMC)
Anatomy: Normal Pineal Gland

• Pineal Gland Facts:
  – Originates embryologically as part of the brain
  – Receives innervation from peripheral sympathetic nerves
  – Makes melatonin which regulates sleep and circadian cycles

• Pineal Gland Imaging
  – Can appear crescent-like, nodule-like, or ring-like
  – Iso-dense to gray matter on T1
  – Several mm in height, length, and width
  – Volume varies, but about 60 mm³ is common for young adult

Comparison Patient 2. Non-Contrast Sagittal T1-Weighted MR Head. All structures, including pineal gland, are normal. (From PACS, BIDMC)
Additional View of Day 1 Imaging: Patient 1

Patient 1. Non-Contrast Axial T2-Weighted MR Head. Image taken on day 1 of admission. Key finding is an iso-intense to hyper-intense mass in the region of the pineal gland. (From PACS, BIDMC)
Immediate Treatment: Patient 1

- In order to treat the patient’s hydrocephalus caused by the pineal mass, a ventriculoperitoneal (VP) shunt was placed.

- This resulted in improving hydrocephalus over a number of days.
VP Shunt Placement: Patient 1

Patient 1. Non-Contrast Axial CT Head. Image taken on day of presentation. Dilated 3rd ventricle measuring 10 mm at maximum diameter is seen. (From PACS, BIDMC)

Patient 1. Non-Contrast Axial CT Head. Image taken 2 days after presentation. Interval improvement in hydrocephalus after VP shunt can be seen. 3rd ventricle now measures 5 mm at maximum diameter. (From PACS, BIDMC)
Further Imaging Work-Up: Patient 1

• In order to further characterize the pineal mass, MR head with contrast enhancement was performed.

• The pineal mass was found to have punctate contrast enhancement and mild overall enhancement.
Additional Imaging 3 Days After Presentation: Patient 1

Patient 1. Non-Contrast Axial T1-Weighted MR Head. Image taken 3 days after presentation. **Mass** is iso-dense to gray matter but somewhat heterogeneous. (From PACS, BIDMC)

Patient 1. Post-Contrast Axial T1-Weighted MR Head. Image taken 3 days after presentation. There is punctate contrast enhancement of **mass** and overall mild enhancement (compare to image on left). (From PACS, BIDMC)
Calcification Distribution: Pineal Masses

- Two types of Calcifications:
  - Exploded to periphery
    - Pineal parenchymal tumors
  - Engulfed
    - Germinomas

Comparison Patient 3. Calcifications Exploded to the Periphery (white arrows) in a Pineoblastoma. Non-Contrast Axial CT.

Comparison Patient 4. Engulfed Calcifications (black arrow) in a Germinoma. Non-Contrast Axial CT.

(Images taken from Smith et al. “Lesions of the Pineal Region: Radiologic-Pathologic Correlation”)
Calcification Distribution: Patient 1

- For patient 1, calcification distribution was indeterminate.

Patient 1. Post-Contrast Axial CTA Head. Image taken 2 days after presentation. Calcifications present heterogeneously throughout mass.
(From PACS, BIDMC)
Summary of Imaging Findings: Patient 1

• CT:
  – Well-demarcated, iso- to hyper-attenuated mass in region of the pineal gland
  – Obstructive hydrocephalus at Sylvian aqueduct, improved after VP shunt placement
  – Equal to or < 2 cm in length in any dimension
  – Heterogeneous calcifications of mass, not in periphery or engulfed centrally

• MR:
  – On T1-weighted images, hypo- to iso-intense to gray matter
  – On T2-weighted images, iso- to hyper-intense to gray matter
  – Post-contrast punctate enhancement and some mild overall enhancement
Symptoms and Signs: Pineal Masses

- Related to mass effect on adjacent structures or invasion of tissues
- Headache/Nausea/Vomiting
  - Occurs from ↑ICP from hydrocephalus
- Parinaud syndrome
  - Invasion of tectal plate
  - Symptoms:
    - Failed conjugate vertical eye movements
    - Mydriasis
    - Failed ocular convergence
    - Blepharospasm
- Precocious Puberty
  - Germ Cell Tumors (increased beta-HCG)
- Pineal Apoplexy
  - Hemorrhage into tumor/cyst
  - Sudden decrease in consciousness associated with a headache
- Secondary Parkinsonism
  - Cause unknown
Differential Diagnosis: Pineal Masses

• Pineal Parenchymal Tumors
  – Pineocytoma
  – Pineal Parenchymal Tumor of Intermediate Differentiation
  – Pineoblastoma

• Pineal Cysts

• Non-Pineal Parenchymal Tumors
  – Metastasis
  – Glioma
  – Germinoma
  – Lipoma
  – Meningioma
  – Astrocytoma
  – Trilateral Retinoblastoma
  – Ependymoma
Epidemiology: Pineal Masses

- Pineal tumors less than 1% of all brain tumors
  - More common in children (3%)
  - More common in Asia (increased risk of pineal germ cell tumors)

- Male 3:1 Predominance for Pineal Tumor

- Pineal cysts present in 25-40% of some autopsy series

- Germ cell neoplasms about 40% of pineal region neoplasms

- Parenchymal Tumors (14-27% of pineal neoplasms):
  - Pineocytomas: 14-60%
  - Pineal Parenchymal Tumors of Intermediate Differentiation: 20%
  - Pineoblastomas: 40%
Pineocytoma: Comparison Patient 5

Comparison Patient 5. Post-Contrast Sagittal T1-Weighted MR Image. **Pineocytoma** is an enhancing mass in the pineal region, resulting in hydrocephalus.

(Image above from Smith et al. “Lesions of the Pineal Region: Radiologic-Pathologic Correlation”)

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Pineal Parenchymal Tumor of Intermediate Differentiation: Comparison Patient 6


(Image above from Smith et al. “Lesions of the Pineal Region: Radiologic-Pathologic Correlation”)

Pineoblastoma: Comparison Patient 7

Comparison Patient 7. Post-Contrast Axial T1-Weighted MR Image. Pineoblastoma is the ill-defined enhancing mass.

(Image above from Smith et al. “Lesions of the Pineal Region: Radiologic-Pathologic Correlation”)

Pineal Cyst: Comparison Patient 8

Comparison Patient 8. Post-Contrast Axial T1-Weighted MR Image. A round, low-intensity cyst (white arrow) with an incompletely-enhancing rim is shown. No nodularity or hydrocephalus is present.

(Image above from Smith et al. “Lesions of the Pineal Region: Radiologic-Pathologic Correlation”
Mature Teratoma: Comparison Patient 9

Comparison Patient 9. Post-Contrast Axial T1-Weighted Image. Pineal teratoma is a heterogeneously enhancing mass with honeycomb-like cystic components.

(Image above from Korogi et al. “MRI of Pineal Region Tumors”)
Pineal Germinoma: Comparison Patient 10

Comparison Patient 10. Non-Contrast Sagittal T2-Weighted MR Image. **Pineal Germinoma** has a solid component iso-dense with gray matter and a cystic component iso-dense with CSF.

(Image above from Liang et al. “MRI of Intracranial Germ-Cell Tumours”)
Meningioma: Comparison Patient 11


(Image above from Korogi et al. “MRI of Pineal Region Tumors”)

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<th>Diagnosis</th>
<th>Summary of Selected Pineal Mass Imaging Findings</th>
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<tr>
<td>Pineocytoma</td>
<td>CT: well-demarcated, iso- to hyperattenuating lesion, peripheral calcifications&lt;br&gt;MR: well-circumscribed, hypo- to iso-intense on T1 and hyper-intense on T2. Avid, homogeneous enhancement post-contrast</td>
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<tr>
<td>PPTID</td>
<td>No specific findings separate PPTID from pineoblastoma or pineocytoma</td>
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<tr>
<td>Pineoblastoma</td>
<td>No specific findings separate pineoblastoma from pineocytoma or PPTID&lt;br&gt;CT: may be large (&gt;3 cm), lobulated and hyperattenuated&lt;br&gt;MR: Heterogeneous enhancement with solid portion hypo- to iso-intense on T1 and iso- to hyper-intense on T2. Heterogeneous enhancement post-contrast</td>
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<tr>
<td>Germ Cell Tumor</td>
<td>CT: sharply circumscribed, hyperattenuating lesion, engulfing calcifications&lt;br&gt;MR: solid mass with cystic components. Iso- to hyper-intense on T1 and T2 images. Avid, homogeneous enhancement post-contrast</td>
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<tr>
<td>Teratoma</td>
<td>CT: multi-loculated, lobulated lesion with foci of fat attenuation, calcification, and cyst&lt;br&gt;MR: foci of T1 shortening due to fat and variable signal intensity from calcification. T2 images have soft-tissue iso- to hypo-intensity. Soft tissue has post-contrast enhancement</td>
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<tr>
<td>Pineal Cyst</td>
<td>CT: round or oval, thin-walled, well-circumscribed&lt;br&gt;MR: Intensity similar to CSF on T1&amp; T2. Post-contrast: incomplete cyst wall enhancement</td>
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<td>Meningioma</td>
<td>CT: hyperattenuating, calcifications in 15-20%. Avid enhancement post-contrast. Dural tail.&lt;br&gt;MR: Hypo- to iso-intense on T1 and iso- to hyper-intense on T2</td>
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*(Information from Smith et al. “Lesions of the Pineal Region: Radiologic-Pathologic Correlation”)*
Diagnosis: Patient 1

• No diagnostic imaging findings for the mass were seen.

• For any suspected pineal parenchymal tumor, a biopsy is necessary to diagnose tumor type.
Diagnosis: Brain Biopsy Results for Patient 1

• Highly cellular tumor, monomorphic cells in sheets and rosettes, with mild atypia and rare prominent nucleoli

• No Mitotic Figures

• Tumors positive for neuronal tumor markers

• Tumor classified as a Pineal Parenchymal Tumor of Intermediate Differentiation

• However, given small pathology specimen, pineocytoma or pineoblastoma could not be ruled-out
Pathology Specimens: Grading of Pineal Parenchymal Tumors

Pineocytoma. Lowest-grade tumor. Note small, uniform cells resembling normal pineocytes.

Pineal Parenchymal Tumor of Intermediate Differentiation. Intermediate-grade tumor. Moderate levels of nuclear atypia and mitotic activity are seen.

Pineoblastoma. High-grade tumor. Note sheets of cells with scant cytoplasm.

(Images above from Smith et al. “Lesions of the Pineal Region: Radiologic-Pathologic Correlation”)

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Treatment: Pineal Parenchymal Tumors

• Acute Hydrocephalus
  – VP Shunt

• Pineocytoma/PPTID
  – Surgical Resection +/- Radiotherapy
  – Radiotherapy often only recommended if tumor appears to behave like pineoblastoma

• Pineoblastoma
  – Usually Surgical Resection + Radiotherapy + Chemotherapy
  – Craniospinal irradiation important to reduce risk of metastasis or potential to seed CSF
  – Radiotherapy not universally recommended
  – Chemotherapy used to reduce common leptomeningeal seeding
Emerging Treatment: Stereotactic Radiosurgery

- Emerging as alternative to surgical resection and traditional radiotherapy for pineocytomas
- Precise radiation fields defined by MRI or CT to reduce damage to surrounding brain are used
- Many non-parallel beams of radiation converge on a small area of brain to target tumor
- Craniotomy and general anesthesia avoided
- Preliminary results suggest higher survival rates than using surgical resection and/or traditional radiotherapy
Clinical Course Update: Patient 1

- Following diagnosis with PPTID:
  - CSF found to be negative for malignant cells
  - Clinical correlation is with an intermediate grade pineal tumor – unlikely to be pineoblastoma
  - Surgical risk deemed too high given patient’s young age
  - Stereotactic radiosurgery also considered risky given patient’s young age, as there is risk of damage to tectum and for late sequelae
  - Stereotactic radiotherapy suggested
Prognosis: Pineal Tumors

• In SEER Data from 1973-2005:
  – 5 year survival for all tumors 65%
  – Germ Cell Tumors 79%
  – Gliomas 61%
  – Pineal Parenchymal Tumors 47%

(Information taken from Al-Hussaini et al., “Pineal Gland Tumors: Experience from the SEER Database”)
Summary

• Pineal masses are rare lesions (<1% of primary brain tumors)
• Pineal tumors can have various presentations, including headaches, nausea/vomiting, and visual defects (Parinaud syndrome)
• MRI is most helpful for characterizing brain lesions like pineal masses, although CT can help for calcification distribution
• No pathognomonic imaging findings to differentiate between parenchymal pineal masses
• Pineal parenchymal tumors usually iso- to hyperattenuating on CT with calcifications “exploded to the periphery”
• Pineal parenchymal tumors usually hypo- to iso-intense on T1, iso- to hyper-intense on T2, and enhancing post-contrast
• Correlation with other data can narrow differential diagnosis
• Brain masses are difficult to evaluate given limited biopsy potential
• Stereotactic radiosurgery is increasingly being used for pineal masses
References

Acknowledgements

- Dr. Gul Moonis, Staff Neuroradiologist, Beth Israel Deaconess Medical Center

- Dr. Gillian Lieberman, Core Radiology Clerkship Director, Beth Israel Deaconess Medical Center

- Emily Hanson, Radiology Clerkship Medical Student Education Coordinator, Beth Israel Deaconess Medical Center