Papillary Tumor of the Pineal Region and the Differential Diagnosis of Papillary Tumors

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Dr. Brent Orr declares he/she has no conflict(s) of interest to disclose.

Papillary Tumor of the Pineal Region

• Rare pineal tumor
• First Described by Anne Jouvet in 2003
• Mostly an adult disease with rare pediatric presentation
• Believed to be derived from the specialized ependymal cells of the subcommissural organ in the roof of the 3rd Ventricle
• histologic overlap in some regions
• ultrastructure demonstrating microlumens
• WHO does not designate definitive grade (grade II/III)

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2015
2000
2005
2010
Jouvet 2003 AJISP
First description of 6 cases
Hasselblatt 2006 Neuropath App Nuinclbl
Copy number data in 15 cases
Fevre-Montagne 2006 JNEN
First description of prognosis
31 total cases
OS 73%; PFS 27%
Fevre-Montagne 2006 JNEN
Unique gene expression pattern compared to other pineal tumors
Heim 2014 AJISP (Prognostic factors - pathology)
2007 WHO Classification of tumors of the CNS (all site sections)
Fauche 2013 J Neurol Onc
Role of surgery, RT, and chemotheraphy
Ham 2016 Brain Path
Methylation clustering

Papillary Tumor of the Pineal Region

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Median age at diagnosis for 181 published cases of PTPR is 35 years (range 1-71 years).

**Presentation of PTPR**

- Symptoms of hydrocephalus
  - Headache
  - Gait instability
  - Short term memory deficits
- Parinaud syndrome
  - Impairment of upward gaze

**Imaging characteristics of PTPR**

- Well circumscribed
- Solid and cystic mass in the pineal region

**PTPR is proposed to derive from the subcommissural organ of 3rd Ventricle**
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Papillary and pseudopapillary growth in PTPR

Solid growth pattern of PTPR

Perivascular pseudorosettes in PTPR

Vascular Hyalinization in PTPR

Geographic Necrosis in PTPR

Strong immunoreactivity for CK in PTPR

CAM5.2

CK18
GFAP expression is typically focal in PTPR

Synaptophysin is often focally expressed

Other pertinent positive and negative stains

Papillary Ependymoma

Prognostic features and Treatment of PTPR

Pineal Parenchymal Tumor of Intermediate Differentiation

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Choroid Plexus Tumors

- Dominated by true papillary growth
- CK7 positive
- Kir7.1 immunopositive

Germ Cell Tumors

- Look for other germ cell components (teratoma)
- Germ cell markers can help distinguish ambiguous cases
- Carcinomas arising in GCT frequently express CK7 or CK20

Endolymphatic sac tumor

- Arises from the inner ear, may grow into the cerebellopontine angle
- Histologically can resemble choroid plexus tumors
- Focally GFAP+, EMA, S-100 positive
- Erodes temporal bone

Papillary meningioma (WHO grade III)

- Typically not in the pineal region
- EMA more diffusely positive
- Rarely express cytokeratin and not diffusely as in PTPR

Metastatic carcinoma

- History and imaging can help—often multifocal
- Lung and breast most common origin
- CK7 and CK20 immunoreactivity can help

Papillary Glioneuronal tumor

- Tumor of cerebral hemisphere (temporal lobe)
- Biphasic, expressing both synaptophysin and GFAP
- SLC44A1-PRKCA fusion
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