SURGICAL APPROACHES TO PINEAL REGION TUMORS
History

• Pinealis – pine cone
• Vedas – one of the 7 centers of vital energy
• Herophilus first described the pineal gland
• Descartes – seat of the human soul
• Studnicka 1905 – glandular function
• Lerner 1958 – melatonin discovered
• Derlincort 1717 – first pineal tumor
History of surgery of pineal tumors

• Horsley – 1910  first attempted resection
• Krause – 1913  first successful surgery
• Dandy – 1921  parieto occipital transcallosal approach
• Van Wagenen – 1931  transcortical transventricular approach
• Poppen – 1960  occipital transtentorial approach
• Stein – 1971  popularized infratentorial supracerebellar approach
Introduction

- Deep seated
- Difficult to access
- Diverse pathologies
- Surrounded by important structures
- The depth to the pineal region is the same from all approaches

“Personally, I have never succeeded in exposing pineal region tumor sufficiently well to justify an attempt to remove it “

_Cushing (1932)_

Pineal tumors are perhaps the most dangerous of all intracranial tumors to attack surgically.

—Walter E. Dandy
Arterial supply

• **P1**
  – Quadrigeminal artery
    – superior colliculus

• **P2**
  – Medial posterior choroidal artery
    – Pineal body, corpora quadrigemina, tela choroidea, thalamus
  – Lateral posterior choroidal artery
    – Choroid plexus lat ventricle, LGB, Thalamus

• **P3, P4**
  – Medial occipital artery
    – Calcarine artery – calcarine sulcus
    – Parieto-occipital artery – parieto-occipital sulcus
  – Posterior pericallosal artery

• **SCA**
  – Inferior colliculus
- **Pineal parenchymal tumor**
  - Pinealocytoma
  - Pinealoblastoma
  - Pineal parenchymal tumor of intermediate differentiation
  - Papillary tumor of pineal region

- **Germ cell tumors**
  - Germinoma
  - Non germinomatous germ cell tumor
    - Embryonal carcinoma
    - Yolk sac tumor (endodermal sinus tumor)
    - Choriocarcinoma
    - Teratoma (mature, immature, malignant)
    - Mixed germ cell tumor

- **Glial cell tumors**
  - Astrocytoma
  - Oligodendroglioma
  - Ependymoma
  - Choroid plexus papilloma
  - Anaplastic astrocytoma/GBM

- **Mesenchymal cell tumors**
  - Meningioma
  - Cavernoma/haemangioblastoma

- **Other tumors**
  - Epidermoid
  - Craniopharyngioma
  - Ganglioglioma
  - Lipoma

- **Metastasis**
- **Lymphoma**
- **Non neoplastic mass**
  - Pineal cyst
  - Arachnoid cyst
  - Cysticercosis
  - Tuberculoma
  - Sarcoidosis
  - Aneurysm of vein of Gallen
Pineal tumors WHO 2007

<table>
<thead>
<tr>
<th>GERM CELL TUMOURS</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Germinoma</td>
<td>9064/3</td>
</tr>
<tr>
<td>Embryonal carcinoma</td>
<td>9070/3</td>
</tr>
<tr>
<td>Yolk sac tumour</td>
<td>9071/3</td>
</tr>
<tr>
<td>Choriocarcinoma</td>
<td>9100/3</td>
</tr>
<tr>
<td>Teratoma</td>
<td>9080/1</td>
</tr>
<tr>
<td>Mature</td>
<td>9080/0</td>
</tr>
<tr>
<td>Immature</td>
<td>9080/3</td>
</tr>
<tr>
<td>Teratoma with malignant transformation</td>
<td>9084/3</td>
</tr>
<tr>
<td>Mixed germ cell tumour</td>
<td>9085/3</td>
</tr>
</tbody>
</table>

grade I II III IV
## Pineal mass with age

<table>
<thead>
<tr>
<th>Age group</th>
<th>Most common</th>
<th>Less common</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infants</td>
<td>Pinealoblastoma</td>
<td>Arachnoid cyst</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Vein of Galen malformation</td>
</tr>
<tr>
<td>Childhood</td>
<td>Germinoma</td>
<td>Pinealoblastoma</td>
</tr>
<tr>
<td></td>
<td>Glioma</td>
<td>Pineal cyst</td>
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<tr>
<td></td>
<td>Tuberculoma</td>
<td></td>
</tr>
<tr>
<td>Young adults</td>
<td>NGGCT</td>
<td>Pinealocytoma</td>
</tr>
<tr>
<td></td>
<td>Glioma</td>
<td>Pineal cyst</td>
</tr>
<tr>
<td>Older adults</td>
<td>Pinealocytoma</td>
<td>Meningioma</td>
</tr>
<tr>
<td></td>
<td>Glioma</td>
<td>Epidermoid Metastasis</td>
</tr>
</tbody>
</table>
Presentation

- Hydrocephalus
- Brainstem compression
  - Parinaud’s syndrome
  - Downgaze palsy
  - Dorsal midbrain compression/ infiltration – Lid retraction/ ptosis
  - Rarely IV palsy
  - Inferior colliculus compression – hearing disturbance
- Cerebellar signs
  - Superior peduncle – ataxia, dysmetria
- Endocrine disturbance
  - Diabetes Insipidus
  - Precocious puberty: β-HCG secretion. In chorio ca./Germinoma with NSGCT – androgen secretion by Leydig cells
- Pineal apoplexy
  - In vascular tumors: Pineal cell tumors/Choriocarcinoma
Imaging

- **X ray**
  - Calcification below 10 years is abnormal

- **CE MRI**
  - Spine should be imaged in all
    - Size and extent
    - Relation to surrounding structures
    - MRV

- **CT**
  - Rarely required
  - Augments information from MRI
    - Calcification
    - BBB breakdown
    - Vascularity
Germ cell tumor

- CT - Hyperdense
  - Sharp borders
  - Intrinsic calcification
- T1 - Hypo
- T2 - Hypo
  - Uniform intense enhancement

Young
M>F
Choriocarcinoma - haemorrhage
Teratoma - calcification
Pinealoblastoma

Homogenous hyperintense on CT

Exploded (peripheral) calcification

Isointense on T1

Iso – hypointense on T2

Slightly non uniform enhancement

Areas of haemorrhage
<table>
<thead>
<tr>
<th>Tumor</th>
<th>CT</th>
<th>T1</th>
<th>T2</th>
<th>CMRI</th>
<th>Others</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pineal cyst</td>
<td>Hypodense Rim calcification</td>
<td>Hypo</td>
<td>Hyper</td>
<td>Peripheral enhancement</td>
<td></td>
</tr>
<tr>
<td>Germ cell tumors</td>
<td>Hyperdense Sharp borders</td>
<td>Hypo</td>
<td>Hypo</td>
<td>Uniform intense enhancement</td>
<td>Young M&gt;F Choriocarcinoma-Haemorrhage Teratoma - calcification</td>
</tr>
<tr>
<td>Pineal parrenchymal tumors</td>
<td>Hyperdense Blastoma- homogenous Cytoma non homogenous Exploded calcification</td>
<td>Iso- hypo</td>
<td>Blastoma – iso/ hypo Cytoma hyper</td>
<td>blastoma – slightly non uniform Cytoma – non uniform</td>
<td>Haemorrhage Non uniform borders</td>
</tr>
<tr>
<td>Papillary tumor of pineal region</td>
<td>Variable</td>
<td>Marked hyper</td>
<td></td>
<td></td>
<td>Cystic areas</td>
</tr>
<tr>
<td>Glioma</td>
<td>Hypodense Calcification rare</td>
<td>Iso/ hypo</td>
<td>hyper</td>
<td>Variable non homogenous</td>
<td>Adults</td>
</tr>
</tbody>
</table>
Imaging

– ANATOMICAL relationships
  • Involvement of 3rd ventricle/ position within 3rd ventricle
  • Superolateral extension into ventricular trigone
  • Location of deep venous system and its relation to the tumor
  • Supratentorial spread of lesion
Tumor markers

• Presence indicates malignant germ cell tumor, converse not true
• More significance in follow up/ recurrence
• Help avoid unnecessary surgery
# Tumor markers

<table>
<thead>
<tr>
<th>Tumor</th>
<th>β-HCG</th>
<th>AFP</th>
<th>PLAP</th>
</tr>
</thead>
<tbody>
<tr>
<td>Germinoma</td>
<td>+(CSF)</td>
<td>-</td>
<td>+(CSF)</td>
</tr>
<tr>
<td>Chorionic carcinoma</td>
<td>++</td>
<td>-</td>
<td>+/-</td>
</tr>
<tr>
<td>Yolk sac tumor</td>
<td>-</td>
<td>++</td>
<td>-</td>
</tr>
<tr>
<td>Embryonal Ca</td>
<td></td>
<td></td>
<td>variable</td>
</tr>
<tr>
<td>Mature teratoma</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Immature teratoma</td>
<td>+/-</td>
<td>+/-</td>
<td>-</td>
</tr>
<tr>
<td>Mixed GCT</td>
<td></td>
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</tr>
</tbody>
</table>
CSF analysis

• Cytology for cells
• Tumor markers
  • Non secreting tumors – CSF level increased, not detectable in blood
  • Germinoma β HCG and PLAP only in CSF
Management

- Hydrocephalus
  - EVD
  - Shunt
  - ETV (+/- biopsy)

- Tissue diagnosis
  - ETV + biopsy
  - Stereotactic biopsy
  - Open surgery

- Tumor control
  - Radiotherapy
  - Surgery
Hydrocephalus

• Present in almost all cases
• Must be addressed prior to tumor surgery
• Stable patient, complete resection likely, temporary EVD at time of surgery
• Symptomatic raised ICP
  • ETV +/- biopsy
    » Gradual reduction of ICP
    » Avoids peritoneal seeding
    » Avoids shunt related complications
  • VP shunt
SURGICAL ANATOMY

• Most tumors arise from or attached to undersurface of velum interpositum
• Tumors rarely extend above velum
• Blood supply comes from within velum mainly from M P.ch & L P.ch with anastomoses to pericallosal & quadrigeminal artery
• Most tumors are centered at pineal gland, some extend to Foramen of Monroe
SURGICAL ANATOMY

• Mostly, ICV, Galen, Rosenthal & precentral cerebellar veins surround or cap the periphery of these tumors.

• Rarely, ICV are ventral to tumor.

• Highly vascular tumors
  – Pineoblastomas
  – Hemangioblastomas
  – Hemangiopericytomas (Angioplastic meningioma)
Surgery common approaches

- **Infratentorial supracerебellar**
  - Approach to centre of tumor
  - Minimizes risk to veins
  - Good exposure
  - No violation of normal tissue

- **Occipital transtentorial / Transcallosal interhemispheric**
  - Tumors extending superiorly
  - Extending laterally
  - Displaces veins ventrally
  - Large tumors
  - Greater exposure
Choice of approach

- Location of tumor (tentorial incisura)
- Tumor morphology (lateral extent)
- Displacement of great veins
- Probable diagnosis on imaging
- Angle of tentorium/ posterior fossa size
- Surgeons preference
Infratentorial supracerebellar approach

• Position
  • Sitting preferred
    – Can also be done in Concorde position
  • Large ventricle/ <3 years – 3 quarter prone
  • Table should be able to go low
  • Head flexed to keep tentorium parallel to floor
  • Patient tilted forward
Infratentorial supracerbellar approach

• Exposure
  • Incision – inion to C4, spinous process of C2 exposed
  • Burrhole – above torcular, lateral aspect of transverse sinus
  • Craniotomy – above transverse sinus and torcular
  • Bone edges waxed
  • If dura tense release CSF (ventricular tap)
  • Dural incision – curved between lateral most aspect of transverse sinus
  • Dura retracted avoid excess retraction – sinus occlusion
Infratentorial supracerebellar approach

• Surgical technique
  – Cauterize and divide adhesions and veins between cerebellum and tentorium
  – Retract vermis postero – inferiorly
  – Open arachnoid over the tumor (opaque white), midline precentral cerebellar vein may be divided
  – Small branches of choroidal and SCA over tumor divided
  – Trajectory of dissection changed towards the tumor
  – **Internal debulking of tumor**
  – **Lateral** walls dissected, vessels on it are choroidal and may be sacrificed
  – Dissection of **inferior** tumor from brainstem – most dangerous part, assistant retracts capsule upwards
  – Final dissection – **superior** along velum interpositum, great veins at risk
Infratentorial supracerebellar approach

• Mortality 3-4%

• Complications
  • Transient ocular dysfunction
  • Ataxia
  • Cognitive impairment, akinetic mutism – brainstem handling
  • Bleed in incompletely resected tumor
# Infratentorial supracerebellar

## Advantage
- Gravity aided drainage of blood/CSF
- Gravity aided cerebellar retraction
- Midline – orientation easy
- No neural structures en route

## Disadvantage
- Air embolism
- Surgeon fatigue
- Difficult in very young and old
- Quadriplegia from excessive flexion in elderly
- Hypotension
Lateral paramedian infratentorial

• INDICATIONS
  • Biopsy
  • Small quadrigeminal area tumor

• ADVANTAGE
  • Minimal damage to neural tissues
  • Useful in steep tentorium
  • Reduced risk of air embolism (lateral position)

• DISADVANTAGES
  • Narrow space
  • Difficult to reach tumor portion extending to infero posterior part of 3rd ventricle
Lateral paramedian infratentorial

• POSITION
  – On the side: usually right side down
  – Upper part of trunk raised 30˚
  – Head flexed with neck stretched & rotated 45˚ face down

• SURGICAL TECHNIQUE
  – S-shaped incision behind mastoid
  – Oval craniectomy close to sigmoid sinus laterally & transverse sinus superiorly
  – Durotomy : cruciate
  – Bridging veins divided, petrosal & precentral cerebellar veins preserved.
  – Tentorial incisura reached, preserving SCA.
Occipital transtentorial approach

• Commonest supratentorial approach

• Indications
  – Predominantly supratentorial
  – Corpus callosum extension
  – Lateral extension into cerebral hemisphere
  – Thalamic extension
  – Predominantly third ventricular mass

• Advantage
  – Extensive tumor view
  – Managing bleeding is easier
  – Working distance is smaller
  – Access to pineal, third ventricle, midbrain, superior vermis

• Disadvantages
  – View obstructed by Galenic venous system
  – Restricted view of opposite side
Occipital transtentorial approach

• Position
  – Lateral decubitus with
    • rt side down
    • Midsagittal plane 30’ above horizontal
  – Three quarter prone
  – Prone
  – Sitting

• Craniotomy
  – Incision: U-shape
  – Craniotomy: 6 burr holes: 2 on left, 2 on right of sag. Sinus, 1 just rostral to trans. Sinus & 1 parietal.
  – Durotomy: T- shape & reflected along sinuses
  – Retractor on inferior surface of occipital lobe
Occipital transtentorial approach

• Surgical steps
  – Occipital retraction to be kept minimum
  – CSF release (from posterior callosal/ dorsal mesencephalic cisterns)
  – Opening of arachnoid (venous system lies in it)
  – Yasargil – positively identify vein of Rosenthal – Galen junction (Vein of Rosenthal may be mistaken for darkly colored dorsal mesencephalic cistern)
  – Tentorium incised 5 – 10 mm from the midline, medial flap sutured to falx
  – Identify and preserve IV nerve when manipulating tent
  – Precentral cerebellar vein may be sacrificed
Occipital transtentorial approach

- Cleavage plane found in small tumor
- Debulking in large tumor
- For hypervascular tumor: feeding arteries identified & coagulated prior to debulking.
- To avoid venous injury, total resection is not necessary & should not be attempted.
- Immaculate haemostasis, water-tight dura closure.
Transcallosal interhemispheric

• Indications
  • Predominantly supratentorial tumor

• Position
  • Sitting/prone preferred
  • Lateral / 3 quarter prone

• surgery
  – U shapes skin flap across the midline
  – Bone flap across the midline
  – Position of bone flap depending on centering of the tumor
  – Wide craniotomy for alternate corridors to avoid bridging veins
  – Avoid sacrifice of more than 1 bridging vein
  – Pericallosal retracted
  – Callosotomy <2 cm centered over the tumor bulge
  – Identify deep veins early
Transcortical transventricular

• Indication
  – Tumor extending into lateral ventricle

• Disadvantage
  • Limited exposure
  • Cortical incision required
  • Stereotactic guidance may be required
Complications of supratentorial approach

• Hemiparesis
  – Brain retraction
  – Sacrifice of bridging veins

• Sensory stereognostic deficits
  – Parietal lobe retraction injury

• Visual field defects
  – Occipital lobe retraction injury

• Disconnection syndrome
  – Corpus callosum section

• Memory defects
  – Fornix injury

• Bleed in residual tumor

• Venous infarction
Stereotactic biopsy

• Indications
  – Invasive disseminated tumor at diagnosis
  – Multiple medical problems
  – Selected cases with very large tumors
  – Neonate with large tumor (highly malignant, poor prognosis)
  – Presentation suggestive of infectious/ metastatic disease with diffuse systemic disease

• Target selection
  – Avidly enhancing tumor, preferably from the centre
  – Multiple sites
Trajectories

- **Orthogonal lateral (orange)**
  - Traverses the temporalis muscle
  - Technically difficult using a stereotactic frame

- **Oblique anterolateral (green)**
  - Most preferred
  - Low frontal trajectory below the plane of the internal cerebral veins

- **Posterolateral (pink)**
  - Lesions with significant lateral extension
Radiotherapy

• Primary
  – Germinoma

• Adjuvant
  – Pinealoblastoma (55 Gy to bed, 35 Gy to spinal axis)
  – Pinealocytoma (NO EFFECT on survival in incompletely excised tumors)
  – All malignant germ cell/ pineal cell neoplasm
  – CAN BE withheld for HISTOLOGICALLY benign COMPLETELY resected pinealocytoma, ependymoma
GKRS

- Histologically confirmed
- Maximum experience with pinealocytoma
- Used as an adjuvant therapy
- Possibly primary therapy for pinealocytoma
- Indications still evolving
- Current possible indications
  - Pineal par enchymal tumors
  - Germinoma
  - NGGCT
  - Astrocytoma
ADJUVANT THERAPY

• CHEMOTHERAPY
  – Indications
    • Non germinomatous malignant germ cell tumors
    • Germinoma with syncytiotrophoblastic giant cells
    • Recurrent /disseminated pineal cell tumors
  – Cisplatin/carboplatin + Etoposide
  – Others: vincristine/lomustine/cyclophosphamide
<table>
<thead>
<tr>
<th>Approach</th>
<th>Advantage</th>
<th>Disadvantage</th>
<th>Indication</th>
</tr>
</thead>
<tbody>
<tr>
<td>Midline innfratentorial supracerebellar (Krause)</td>
<td>Midline – orientation Tumor below major veins Gravity assists retraction</td>
<td>Air embolism Narrow corridor Sacrifice of veins – infarction Difficult to reach above incisura</td>
<td>Midline masses No extension laterally/ above incisura Tumor &lt; 3 cm</td>
</tr>
<tr>
<td>Lateral paramedian infratentorial (Van Wagenen)</td>
<td>No sacrifice of veins Possible with steep slope of tent Less air embolism</td>
<td>Only for small tumors SCA and branches at risk Cannot see posterior 3rd ventricle</td>
<td>Small tumors below tentorial notch with unilateral lateral extension</td>
</tr>
<tr>
<td>Occipital transtentorial (Poppen)</td>
<td>Good view of structures above and below the tent</td>
<td>Retraction damage to occipital lobe Damage to splenium Cannot see posterior third ventricle</td>
<td>Tumors extending above and below tentorial incisura Tumors with unilateral lateral extension</td>
</tr>
<tr>
<td>Posterior transcallosal (Dandy)</td>
<td>Lesion above tentorial notch with extension into 3rd ventricle</td>
<td>ICV in approach Callosotomy – disconnection syndrome Parietal lobe retraction damage</td>
<td>Posterior 3rd ventricular mass Mass between splenium and venous system</td>
</tr>
<tr>
<td>Approach</td>
<td>Advantage</td>
<td>Disadvantage</td>
<td>Indication</td>
</tr>
<tr>
<td>-----------------------------------------------</td>
<td>-----------------------------------------------------</td>
<td>------------------------------------------------------------</td>
<td>------------------------------------------------</td>
</tr>
<tr>
<td>Posterior transventricular (Van Wagenen)</td>
<td>Exposes atrium and posterior body of lateral ventricle</td>
<td>Fornix section – memory defecit Seizures</td>
<td>Tumor extending into posterior lateral ventricle</td>
</tr>
<tr>
<td>Anterior transcallosal, transventricular trans vellum interpositum (Sano)</td>
<td>Wider room No fornix section Supine – low air embolism</td>
<td>Increased depth of approach Callosotomy defecits Fornix damage by retraction</td>
<td>Large tumors extending anteriorly in 3rd ventricle</td>
</tr>
<tr>
<td>Combined supra – infratentorial trans sinus (Ziyal and Sekhar)</td>
<td>Access tumor above and below tent Amole room Sinus may be resutured</td>
<td>Division of transverse sinus – venous infarcts / delayed raised ICP</td>
<td>Large meningioma, epidermoid, teratoma</td>
</tr>
</tbody>
</table>
Results of Pineal Region Surgery at the New York Neurological Institute (1990-2008)

<table>
<thead>
<tr>
<th>Total Procedures</th>
<th>128</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benign pathology</td>
<td>55 (43%)</td>
</tr>
<tr>
<td>Malignant pathology</td>
<td>73 (57%)</td>
</tr>
<tr>
<td>Diagnosis established</td>
<td>127 (99%)</td>
</tr>
</tbody>
</table>

Surgical Morbidity

| Death (pulmonary embolism/cerebellar infarct) | 2 (2%) |
| Permanent major morbidity                  | 1 (1%) |
| Transient major morbidity (with recovery)   | 7 (5%) |