

Surgical approaches to pineal region tumors

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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. Dandy⁷³

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

GERM CELL TUMOURS

| | |
|--|--------|
| Germinoma | 9064/3 |
| Embryonal carcinoma | 9070/3 |
| Yolk sac tumour | 9071/3 |
| Choriocarcinoma | 9100/3 |
| Teratoma | 9080/1 |
| Mature | 9080/0 |
| Immature | 9080/3 |
| Teratoma with malignant transformation | 9084/3 |
| Mixed germ cell tumour | 9085/3 |

grade I II III IV

Pineal mass with age

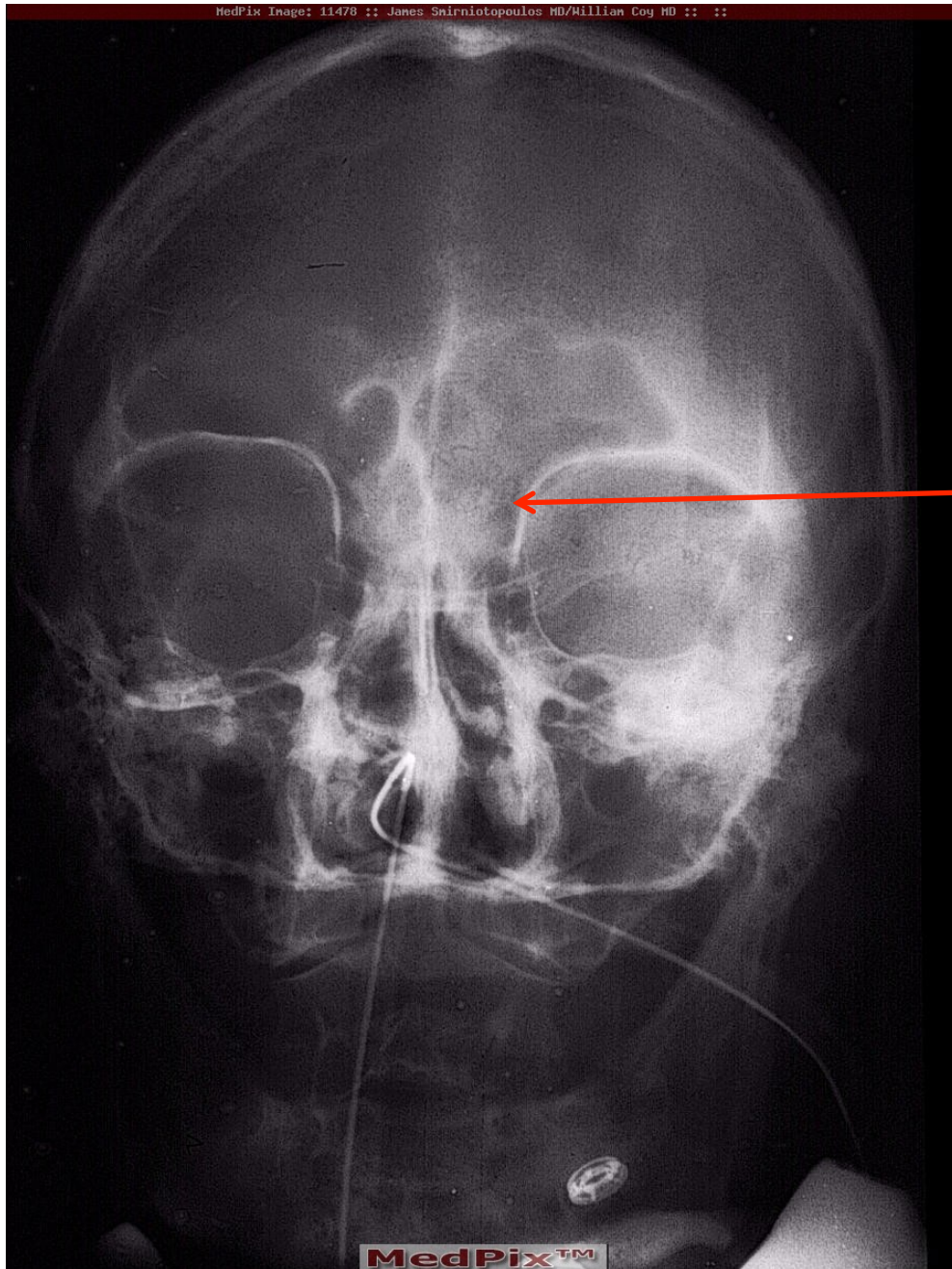
| Age group | Most common | Less common |
|--------------|------------------------------------|--|
| Infants | Pinealoblastoma | Arachnoid cyst Vein of Galen malformation |
| Childhood | Germinoma Glioma Tuberculoma | Pinealoblastoma Pineal cyst |
| Young adults | NGGCT Glioma | Pinealocytoma Pineal cyst |
| Older adults | Pinealocytoma Glioma | Meningioma Epidermoid Metastasis |

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

Mixed germ cell tumor

Pinealoblastoma

Homogenous hyperintense on CT

Exploded (peripheral) calcification

Isointense on T1

Iso – hypointense on T2

Slightly non uniform enhancement

Areas of haemorrhage

Imaging

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

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

| Tumor | β -HCG | AFP | PLAP |
|-------------------|--------------|-----|--------|
| Germinoma | +(CSF) | - | +(CSF) |
| Chorio carcinoma | ++ | - | +/- |
| Yolk sac tumor | - | ++ | - |
| Embryonal Ca | variable | | |
| Mature teratoma | - | - | - |
| Immature teratoma | +/- | +/- | - |
| Mixed GCT | | | |



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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 (Angioplasmic meningioma)

Surgery common approaches

- **Infratentorial supracerebellar**
 - 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 supracerebellar approach

- Exposure

- Incision –inion to C4, spinous process of C2 exposed
- Burrhole – above torcula, lateral aspect of transverse sinus
- Craniotomy – above transverse sinus and torcula
- 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

Combined supra – infratentorial trans
sinus

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 defecits
 - 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 parenchymal 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

| Approach | Advantage | Disadvantage | Indication |
|---|---|---|---|
| Midline infratentorial supracerebellar (Krause) | Midline – orientation Tumor below major veins Gravity assists retraction | Air embolism Narrow corridor Sacrifice of veins – infarction Difficult to reach above incisura | Midline masses No extension laterally/above incisura Tumor < 3 cm |
| Lateral paramedian infratentorial (Van Wagenen) | No sacrifice of veins Possible with steep slope of tent Less air embolism | Only for small tumors SCA and branches at risk Cannot see posterior 3 rd ventricle | Small tumors below tentorial notch with unilateral lateral extension |
| Occipital transtentorial (Poppen) | Good view of structures above and below the tent | Retraction damage to occipital lobe Damage to splenium Cannot see posterior third ventricle | Tumors extending above and below tentorial incisura Tumors with unilateral lateral extension |
| Posterior transcallosal (Dandy) | Lesion above tentorial notch with extension into 3 rd ventricle | ICV in approach Callosotomy – disconnection syndrome Parietal lobe retraction damage | Posterior 3 rd ventricular mass Mass between splenium and venous system |

| Approach | Advantage | Disadvantage | Indication |
|---|---|--|--|
| Posterior transventricular (Van Wagenen) | Exposes atrium and posterior body of lateral ventricle | Fornix section – memory deficit Seizures | Tumor extending into posterior lateral ventricle |
| Anterior transcalsal, transventricular trans vellum interpositum (Sano) | Wider room No fornix section Supine – low air embolism | Increased depth of approach Callosotomy deficits Fornix damage by retraction | Large tumors extending anteriorly in 3 rd ventricle |
| Combined supra – infra tentorial trans sinus (Ziyal and Sekhar) | Access tumor above and below tent Amole room Sinus may be resutured | Division of transverse sinus – venous infarcts / delayed raised ICP | Large meningioma, epidermoid, teratoma |

- **Results of Pineal Region Surgery at the New York Neurological Institute (1990-2008)**

| | |
|---|------------|
| Total Procedures | 128 |
| Benign pathology | 55 (43%) |
| Malignant pathology | 73 (57%) |
| Diagnosis established | 127 (99%) |
| Surgical Morbidity | |
| Death (pulmonary embolism/ cerebellar infarct) | 2 (2%) |
| Permanent major morbidity | 1 (1%) |
| Transient major morbidity (with recovery) | 7 (5%) |