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[73]

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 parrenchymal 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

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			

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 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 transvetricular

- 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 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 parrenchymal 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 innfratentorial 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 3rd 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 3rd ventricle	ICV in approach Callosotomy – disconnection syndrome Parietal lobe retraction damage	Posterior 3rd 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 defecit Seizures	Tumor extending into posterior lateral ventricle
Anterior transcallosal, transventricular trans vellum interpositum (Sano)	Wider room No fornix section Supine – low air embolism	Increased depth of approach Callosotomy defecits Fornix damage by retraction	Large tumors extending anteriorly in 3rd 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%)