# SURGICAL ANATOMY & APPROACHES TO BRAINSTEM GLIOMA

### Introduction

- Brainstem comprises of
  - o Midbrain (Mesencephalon),
  - Pons and
  - o Medulla
- Highly complex neural structure both anatomically and functionally
- Cranial nerve nuclei and numerous fascicles and pathways as well as reticular formation- all playing important roles in securing normal central nervous function and regulation of bodily homeostasis

### Historical considerations

- Because of its difficult access and functional importance, in the past, the brainstem was seldom explored by neurosurgeons, with its injury often conducive to deep coma
- For many years, a tumor growing inside the brainstem was considered malignant in itself and managed empirically as a homogeneous group with radiation therapy as well as adjunctive chemotherapy

### Historical considerations

- Bailey et al (1939)- 'BSG are a hopeless problem for treatment'
- Dandy (1962)- 'There is little indication for attempting any enucleation of the tumour in this region
- Baker (1964)- published a series of pts with 'subependymal gliomas'
- Pool(1968)- operated BSG , some of them having a long-term survival

### Historical considerations

- Gradual advancement in microsurgical technique, sophisticated imaging technology, most importantly availability of MRI
- Identification of subcategories of tumours which appear to have low- grade pathologies and offer a better prognosis
- Different series on BSG since then



# Surgical anatomy - Dorsal aspect

### • Floor of IV ventricle –

- Rhomboid
- Pons- rostral 2/3<sup>rd</sup>
- Medulla- caudal 1/3<sup>rd</sup>

#### SURGICAL ANATOMY-DORSAL ASPECT



#### Three parts-

- Superior/ pontine
- Intermediate/juctional
- Inferior / medullary part

Rhoton

#### SURGICAL ANATOMY-POSTERIOR ASPECT

- **o** Median sulcus
- o Sulcus limitans
  - Median eminence(M)
  - Vestibular area(L)
- Median eminence-
  - Facial colliculus
  - Hypoglossal triangle
  - Vagal triangle
  - Area postrema
- Striae medullares



Rhoton

### Brainstem glioma(BSG)-Epidemiology

- Approx. 1 % of all primary brain tumours, 10-20% of pediatric brain tumours
- 75% occur in children, 25 % in adults
- Median age at presentation-6.5 yrs, adults- 3<sup>rd</sup> -4<sup>th</sup> decade
- o M=F
- Approx. 75% diffuse, 25 % focal
- Most focal tumours occur in midbrain
- Pontine tumours are usually diffuse and high grade

#### **BRAIN STEM TUMOUR CLASSIFICATION**

#### <u>SYSTEM</u>

AUTHOR AND YEAR	METHOD USED TO CREATE SYSTEM	CLASSIFICATION SYSTEM
Epstein, 1985	СТ	Intrinsic Diffuse Focal Cervicomedullary Exophytic Anterolat into cerebellopontine angle Posterolat& into brachium pontine Disseminated Positive cytological findings Positive myleographic finding
Epstein & McCleary , 1986	CT, MRI, SURGICAL OBSERVATION	Diffuse Focal Cerebellomedullary
Stroink et al, 1987	СТ	Group 1:-dorsal exophyticglioma Group 2:-intrinsic brainstem tumours 2 a hypodense, no enhancement 2 b hyperdense, contrast enhancing, exophytic Group 3:-focal cystic tumour w/contrast enhancement Group 4:- focal intrinsic isodense lesion w/ contrast enhancement
Barkovich et al, 1990	MRI	Location (midbrain, pons, medulla) Focality (diffuse or focal) Direction and extent of tumour growth Degree of brainstem enlargement

		Exophyticgrowth Hemorrhage and necrosis Evidence of hydrocephalus
Albright, 1996	MRI	Focal (midbrain, pons, medulla) Diffuse
Fischbein et al, 1996	MRI	Midbrain Diffuse Focal Tectal Pons Diffuse Focal Medulla Diffuse Focal Dorsal exophytic
Choux et al, 2000	CT & MRI	Type i:-diffuse Type ii:- intrinsic, focal Type iii:- exophytic, focal Type iv:- cervicomedullary

#### **Brain stem tumour classification**

Type of brainstem	Features	Comparison with other	Surgery
glioma		classifications	
Intrinsic only	The tumour is well within the brainstem axis without any breech of parenchyma		Yes Radical excision
Expanding variety	<ul> <li>Well delineation on Gd-MRI</li> <li>Slow progression of clinical symptoms (&gt; 6 months)</li> <li>Good preservation of motor function with independent daily living</li> <li>Size may be &gt;2cm</li> </ul>	Epistein et al. Focal{<2cm}, cervicomedullary Stroink et al. Group iii, iv Choux et al. Type ii, iv Tumour>2cm could still be expanding variety ** based on contrast enhancement and radiology only, clinical features could not be taken into consideration *** based more on anatomical localization, clinical features not mentioned again.	No
Diffuse infiltrative variety	<ul> <li>No margin of delineation on Gd-MRI</li> <li>Rapid progression of symptoms</li> </ul>	Epistein et al: diffuse Stroink et al :lla Choux et al: type i Tumourlokkin focal but >2cm are still consider diffuse	No
Ventrally located	Pure ventral location	May be kept as focal in other classifications, but author prefer not to operate in view of difficult location ans associated high risk of complications.	

Not that the classification for exophytic tumour is the same as proposed byearler authors but has been included here to provide completeness.

Mehta VS, Chandra PS, Singh PK, Garg A, Rath GK. Surgical considerations for 'intrinsic' brainstem gliomas: Proposal of a modification in classification.Neurol India 2009;57:274-81

### **BSG-** Pathogenesis

• Molecular biology-

- Mutation of p53, a tumour suppressor gene
- Amplification of mutated EGFR gene
- Trisomy 1q, deletion of chr 19

**○** NF – I –

• More indolent course



**o** CT-

 Diffuse - hypodense lesion on NCCT that enlarge the pons
 (diffuse pontine hypertrophy) and displace IVth ventricle posteriorly, inhomogenous postcontrast enhancement

# Imaging



#### CT-

- Focal midbrain tumours (tectal plate glioma) may not be seen on NC + CECT head , leading to a false diagnosis of late onset aqueductal stenosis
- MRI is an accurate and noninvasive method of diagnosis that can be indicated in all cases of late onset hydrocephalus and aqueductal obstruction, especially in adults.

# Imaging

### o MRI-

### Imaging modality of choice

- Precise localization
- Together with clinical picture, suggest the microscopic pathology of tumour, with a relatively high degree of probabaility





#### o MRI-

#### **Diffuse BSG-**

- Hypo on T1, hyper on T2, with hyperintensity extending into adjacent midbrain/medulla, inhomogenous contrast enhancement within or around the tumour
- Contrast enhancement in only 1/3<sup>rd</sup> cases
- No significant difference in prognosis with/without contrast enhancement





#### o MRI-

- Focal BSG-
  - Well circumscribed, of limited size, may be partially cystic, without associated oedema/infiltration
  - o Midbrain >Medulla>Pons
  - Hypo on T1, hyper on T2, nidus of focal enhancement
  - Usually pilocytic astrocytomas



#### o MRI-

- Dorsally exophytic BSG-
  - Intra-IVth ventricular
  - Resemble vermian astrocytoma with involvement of IVth ventricular floor

# Practical decisions regarding treatment of BSG



# Practical decisions regarding treatment of BSG



Brainstem tumour location and surgical			
approach			
Location Approach			
Dorsal midbrain	Supracerebellar		
(tectum	infratentorial		
mesencephalii)			
Ventral midbrain	Pterional trans-		
	sylvian		
Lateral midbrain	Subtemporal		
	transtentorial		
Ventolateral pons	Retromastoid		
(cerebellopontine	retrosigmoid		
angle)			
Dorsal pons and	Midline suboccipital		
medulla oblongata	transventricular		
Lower medulla	Midline suboccipital &		
oblongata &	c1 laminectomy		
cervicomedullary			
junction			

### Intraoperative monitoring

- o Cranial nerves-
  - EMG monitoring III,IV,V,VI,VII, IX,X,XI,XII
  - **o** BAEP
- SSEP and MEP

### Anaesthesia for brainstem surgery

 Multimodal monitoring – SpO2 & EtCO<sub>2</sub> monitoring, CVP line, arterial line, transesophageal echocardiography,

etc.

### Anaesthesia for brainstem surgery

- During brain stem surgery, traction of cranial nerves and stimulation of nuclei and connecting pathways may cause severe alterations in blood pressure and heart rate, sudden respiratory drive despite the surgical level of anesthesia.
- Extreme bradycardia and ventricular arrhythmia can be life-threatening and must be treated promptly by immediate interruption of surgical stimulation before any pharmacological intervention

# Surgical technique-

- Almost all BSTs are dorsally located, therefore should be approached through posterior fossa
- Position-prone(preferred)/sitting
- Midline skin incision
- Suboccipital craniotomy±cervical laminotomy
- Y shaped dural opening

## Surgical technique-

- Vermis coagulated and split at appropriate level
- Cerebellum held to the sides using self- retaining retractors( \* avoid excessive side retraction – pseudobulbar palsy )
- IV<sup>th</sup> ventricle approached after division of medullary velum

# Surgical technique-

- Pontine bulge in IVth ventricular floor
- o Medullary- medulla will be ballooned
- Midbrain precentral cerebellar vein and arachnoid over vein of galen complex may need to be divided

### Safe entry zones to brainstem -Rationale

- The brain stem is densely composed of important neural structures such as nuclei and neural tracts
- Causes of morbidity following brainstem surgery-
  - Direct damage during removal of the lesion,
  - Selection of an entry route into the brain stem, and
  - The direction of brain stem retraction
- In most cases, the optimal surgical route can be established by use of the 2-point method, in which an imaginary line drawn from the center of the lesion to the point nearest the surface of the brain defines the least disruptive approach
- Where critical neural structures are sparse and no perforating arteries are present.

### Safe entry zones to brainstem

### **o** Suprafacial triangle-

- MLF medially,
- VII nerve caudally
- SCP & ICP laterally
- The brain stem can be retracted either laterally or rostrally with relative safety

### Safe entry zones to brainstem

#### o Infrafacial triangle

- MLF medially,
- Striae medullares caudally,
- Facial nerve laterally

#### • The brain stem can be retracted only laterally

#### <u>Structures potentially damaged</u> <u>by brainstem retraction</u>

Position	Superfacial triangl	e ir	nterfacial triang	e
relative	Structure	Symptom	Structure	Symptom
to				
surgery			5 I I	5 I I
Lateral	Superior	Hemiataxia	Facial	Facial nerve
	cerebellar	Sensory motor	nerve(deeper)	paisy
	Trigominal nuclei	face	vestibular	nystagmus
Medial	MIF	Gaze	MIE	Nystagmus
IVIEUIAI	IVILI	nalsy nystagmus	IVILI T	Nystaginus
Rostral	Sup. Cerebellar	Hemiataxia	Nucleus of 6 <sup>th</sup>	Abducent palsy
	paduncle	Occulomotor&	nerve	
	3 <sup>rd</sup> & 4 <sup>th</sup> nerves	trochlear palsy	PPRF	Lateral-gaze
	and nuclei		Facial nerve	palsy
				Facial nerve
				palsy
Caudal	Nucleus of 6 <sup>th</sup>	Abducens nalsy	Nuclei of	Swallowing
	nerve	, inducerio puio,	lower cranial	impairment.
	PPRF	Lateral gaze	nerve	dysarthria
		palsy		·
	Facial nerve	Facial nerve		
		palsy		
ventral	Medilaleminiscus	Ataxia, depth	Medial	Ataxia, depth
		perception	leminiscus	perception
	Lateral	impairment		impairment
	spinothalamic	Analgesia,	Lateral	Analgesia,
	tract	thermanesthesia	spinothalamic	thermanesthesia
	Corticospinal	Matar	tract	IVIOTOr
	tract	NIOTOR	Corticospinal	impairment
		impairment	tract	

Safe entry zones to brainstem-Anterolateral aspect

 Midbrain- lateral mesencephalic sulcus

- Pons- peritrigeminal area
- o Medulla- retro-olivary sulcus

Recalde R. MICROSURGICAL ANATOMY OF THE SAFE ENTRY ZONES ON THE ANTEROLATERAL BRAINSTEM RELATED TO SURGICAL APPROACHES TO CAVERNOUS MALFORMATIONS.

Neurosurgery, 2008.

### **Tumour decompression**

- Conventional suction technique frequently causes brainstem dysfunction manifested by bradycardia & arrhythmia
- CUSA causes movement of adjacent structures only within 1mm of vibrating tip, allowing for extensive and quick dissection adjacent to or within the substance of brainstem

### Surgical technique-Focal tumour

- Essential that rostral & caudal pole of the tumor be completely exposed
- Incise the lower vermis to obtain adequate separation of tonsils to view the entire posterior surface of IVth ventricle
- Important to view the median raphe, calamus scriptorius and the obex

### Surgical technique-Focal tumour

- Incision at an area where tumor is most superficial
- It also must be away from the midline and at least 1.5cm rostral to the obexavoids injury to cranial nerve nuclei X-XII
- Incision <1cm

### Surgical technique-Focal tumour

- Use of plated bayonet(very small plates at the tip) as 'microretractor'
- CUSA at a low setting
- Careful identification of white matter interface
- Minimal manipulation of adjacent normal tissue

- Suboccipital craniotomy + osteoplastic laminotomy
- Expose both rostral and caudal extent of the tumor.
- USG guidance to know extent of tumor prior to opening the dura - entire tumor should be within the confines of the operative exposure

- The rostral end of a benign cervicomedullary tumor invariably expands posteriorly at the obex
- Tumor is, in fact, displacing the medulla rostrally rather than extending into it.
- This explains why these tumor present with cervical myelopathy rather than LCN dysfunction
- Conceptually, these tumor should be regarded as ' intramedullary spinal cord tumours'

- o Midline myelotomy
  - 'True' midline to be identified
  - Identify DREZ bilaterally
- If tm is solid-cystic, myelotomy to be palced first at tumor-cyst junction→cyst removed prior to tumor excision.
- If tumor is non-cystic, myelotomy where tumor is most voluminous & closest to the pial surface.

- Myelotomy to be terminated 1 cm proximal to the caudal pole of the tumor→tumor is least voluminous here, removed by gradual upward dissection
- At the rostral pole,tumor invariably subpial and bulging posteriorly at the obex

- USG to guide the extent of tumor excision- to confirm bulk of tumour is removed
- Don't chase small questionable fragments
- If deterioration of SSEP/MEP during the procedure, interrupt the dissection and move to another area

### Surgical technique-Cystic tumour

- Bulge into the IVth ventricle
- Collapse" of the cyst cavity and surrounding neural tissue following cyst evacuation → difficulty in identifying the solid nodule
- 'Hand-held' retractor compared to fixed
   Avoid frequent manipulation of retractor
   Use of LASER

### Surgical technique-Dorsally exophytic tumour

- Mostly benign, arising from subependymal tissue and grow posteriorly in the area of 'least resistance' -through the floor of IVth ventricle
- Major technical complication-injury to neural structures immediately below the ependymal lining

### Surgical technique-Dorsally exophytic tumour

- Remove tumor "flush" with the floor of IVth ventricle.
- Do not pursue tumour inside the brainstem.
- Low grade astrocytoma, ganglioglioma.
- Facial colliculus injury.

### Complication avoidance & management– Cervicomedullary tumour surgery

Complication	Avoidance	Management
Kyphoscoliosis	-Osteoplastic laminotomy -Conservative extent of bone removal based upon USG guidance	Correction & fusion( late post-op)
Sensory (posterior column) deficit	<ul> <li>-True midline myelotomy</li> <li>-SSEP</li> <li>-Initiation of myelotomy at the most bulky portion of the tumor using USG guidance</li> <li>-Myelotomy to end 1 cm short of tapering caudal end of the tumor</li> </ul>	Physiotherapy, Rehabilitataion

### Complication avoidance & management – Cervicomedullary tumour surgery

Complication	Avoidance	Management
Motor deficit	<ul> <li>Avoid chasing small questionable fragments in ventrolateral aspect of the resection cavity</li> <li>USG guidance</li> <li>MEP</li> </ul>	Physiotherapy Proper nursing Rehabilitataion
Cardiovascular instability	Close anaesthetic monitoring and prompt discontinuation of manuever	

### Complication avoidance & management – Focal BSG surgery

Complication	Avoidance	Management
Cr nv V palsy	- Careful inspection of IVth ventricular floor to detect area of greatest bulge/tumour erosion to be used as entry into the tumour	Corneal lubrication, Tarsorrhaphy
VI,VII	<ul> <li>-Careful inspection of erosion site</li> <li>- Localize median raphe and incise away from midline</li> <li>- Safe entry zone landmarks</li> </ul>	Corneal lubrication, Tarsorrhaphy, Corrective surgery for LR palsy
VIII	BAER	Hearing aid

### Complication avoidance & management – Focal BSG surgery

Complication	Avoidance	Management
IX-XII palsy	- Identify obex and incise floor 1.5 cm rostral to it if no ependymal erosion is present	Prolonged ventilation and tracheostomy
Cr nv palsies	<ul> <li>Ependymal incision &lt; 1cm</li> <li>Use of plated bayonet</li> <li>Stay within the tumour, inspect carefully for the interface</li> </ul>	
Post-op hypoventilation, hypercarbia & brainstem hypoxia		Persistent mechanical ventilation, slow weaning, tracheostomy

### Complication avoidance & management – Cystic BSG surgery

Complication	Avoidance	Management
Retraction injury	<ul> <li>Avoid excessive retractor manipulation</li> <li>Hand-held reatactor</li> <li>Laser</li> <li>Avoid CUSA</li> <li>Don't chase questionable fragments</li> </ul>	

### Complication avoidance & management – Dorsally exophytic BSG surgery

Complication	Avoidance	Management
Brainstem nuclei injury in general	<ul> <li>-Good visualization of ependyma above and below the tumour</li> <li>- Avoid resection below the ependymal floor</li> </ul>	

### Peri-operative care

- Perioperative steroids( methylprednisolone)
- Elective ventilation for at least 48 hours
- Mechanical ventilation till recovery of ventilation & normal cough reflex
- LCN paresis- NG/feeding gastrostomy
- V,VII nv paresis- temporary tarsorrhaphy
- Good nursing care
- Physiotherapy
- Post-op brainstem injury mostly reversible if surgical technique is proper

### Role of stereotactic biopsy

- Diffuse glioma is an infiltrative, highly aggressive lesion which is always malignant regardless of the histology at the time of biopsy, associated with a very poor prognosis, MR appearance is reliable → No role of biopsy for these lesions (Epstein,McCleary,1986)
- No role of open surgery/stereotactic biopsy in diffuse tumor because of typical MRI characteristics and clinical presentation (Isamat, 1999)
- Risks of biopsy far outweigh the remote possibility of diagnosing something other than a glioma

### Role of stereotactic biopsy

 Majority of focal, dorsally exophytic and cervicomedullary BSG are benign and resectable by direct surgery with low morbidity and good outcome

> New york symposium on Brainstem surgery, 1996. Epstein, Constantini ,Hoffman, A Bricolo

# Role of stereotactic biopsy

#### • Reserved to

- When the diagnosis is uncertain, to rule out inflammatory pathology like TB
- Focal intrinsic endophytic lesion- well limited masses within the brainstem surrounded by neural tissue and therefore do not reach the surface

### Role of GKRS

<u>Yen CP, Sheehan J, Steiner M, Patterson G, Steiner L</u>. **Gamma knife surgery for focal brainstem gliomas.** J Neurosurg. 2007 Jan;106(1):8-17.

20 patients
10-18 Gy
Median follow up- 78 months
Tm disappeared in 4 pts, decreased in size in 12 pts
Minimal peri & post- procedural morbidity

# Take home message

- BSG are a heterogenous group of neoplasm
- Importance of MRI in diagnosis and planning of treatment
- Minimize complications by operating upon 'benign' lesions in the presence of minimal neurological dysfunction
   Knowledge of 'safe entry zones'

### Take home message

- Diffuse tumor almost invariably malignant and should not be operated upon→ Direct RT + CT
- Focal medullary tumor
  - Likely to be benign
  - Surgery associated with significant morbidity
  - If laterally located & appears to be approachable with acceptable risks, resection is appropriate. If more centrally located→ Stereotactic biopsy + Irradiation
  - Role of primary radical excision still unclear

### Take home message

#### • Dorsally exophytic tumor-

- Likely to be benign
- Radical excision
- Do not enter brainstem
- Cervicomedullary tumor-
  - Likely to be benign
  - Radical excision
- Cystic tumor
  - Radical excision
- o Focal pontine tumor-
  - Radical excision if tm is close to the surface

### Thank you