CRANIOPHARYNGIOMA: MANAGEMENT PRINCIPLES AND RECENT ADVANCES

Introduction

- 3-5% of primary brain tumors
- 50% of paediatric supra sellar tumors
- No gender difference

- 70% combined suprasellar/ intrasellar
- Completely intrasellar craniopharyngiomas are rare.

Preoperative evaluation and management

- Complete endocrinological evaluation to uncover hypopituitarism particularly

  Growth hormone

  Cortisol

  Thyroid hormone deficiencies.

Imaging

X ray:

- Irregular speckled calcification seen just above the sella turcica.
- The semicircular shell outlining the wall of cystic lesion.
- Fine flaky calcium -fast growing tumours.
- Dense calcification -slow growing tumours.
- Mostly suprasellar.
- Calcification may be in cyst wall and/or solid component.

Imaging

MRI:

- Hypo on T1,hyper on T2WI
- Multilobular
- Multicystic
- Enhances strongly/ heterogenously
- Often both cyst walls and solid components enhance
- Completely solid (rare)

Classification

- Grade I (intrasellar or infradiaphragmatic)
- Grade II (occupying the cistern with or without an intrasellar component)
- Grade III (lower half of the third ventricle)
- Grade IV (upper half of the third ventricle)
- Grade V (reaching the septum pellucidum or lateral ventricles)

Samii M, Tatagiba M, Neurol Med Chir, 1997;37:141

THERAPUETIC GOALS

- Cure of disease with functional preservation and restoration.

Surgical Approaches

- Ideal approach – Varies.
- Influenced by the tumour location with respect to the sella, chiasm and third ventricle.

Anterior Midline Approach

Trans-sphenoidal

- Grade I and II
- Decreased risk of visual injury
- Difficult in young children (non-pneumatised sphenoid sinus)
- CSF leak

Anterior Midline Approach

Subfrontal

- Grade III and IV
- Pre-chiasmatic dissection of the tumour
- Potential violation of the frontal sinus
- Damage to the olfactory tract
- Technically more complicated (pre-fixed chiasm)

Anterolateral Approach

Pterional

- Facilitating the resection of intrasellar, suprasellar, pre-chiasmatic and retrochiasmatic tumours.
- Restricted view of the contra lateral opticocarotid triangle, the contralateral retrocarotid space and the ipsilateral hypothalamic wall.

Anterolateral Approach

Orbitozygomatic

- Expands on the pterional approach
- Significant suprasellar extension

Transpetrosal Approach

- Large retrochiasmatic tumors

Intraventricular Approaches

Transcallosal –transventricular

- Foramen of Monro is dilated by a tumour projecting into the lateral ventricle
- Frontal lobe retraction injury

Intraventricular Approaches

Transcortical –transventricular

- Seizures
- Large ventricles and tumour extending to the dorsal surface of the frontal lobe

Intraventricular Approaches

Trans lamina Terminalis

- Intraventricular tumors
- Pterional or a subfrontal approach to access the lamina terminalis

Combined Approaches

- **Subtemporal –transpetrosal** –
  - Primarily retrochiasmatic unilateral tumors extending to the posterior fossa along the clivus.

- **Pterional –transcallosal** –
  - Aid removal of adherent and calcified tumor within the third ventricle.

- In transcallosal + pterional approach, intraventricular portions of the tumour should be removed first, with the pterional approach only being performed if basal portions of the tumor remain inaccessible.

Radical surgery

- Possible in:
  - Small or prechiasmatic
- Difficult in:
  - Proximity and adherence of the lesion to the optic pathways and adjacent neurovascular structures
  - Reterochiasmatic
  - Large
  - Multicompartmental

Radical surgery

Advantage

- One treatment then only follow-up

Disadvantages

- Limited number of surgeons with adequate expertise
- Difficult to assess true risks to individual child
- Impaired quality of life
- Diabetes insipidus (95%)

Limited surgery

Goals

- Diagnosis
- Drain cysts
- Limit field of radiation
- Control hydrocephalus
- Improve vision
- Decompress chiasm

Limited surgery + radiation

Advantages

- Surgery can be performed with limited experience

Disadvantages

- Decrease in IQ
- Cyst management (often multiple cyst procedures)
- Complications of radiation
- Diabetes insipidus (5%)

Endoscopy

- Grade 1 and 2 tumors
  - Transnasal
  - Trans -sphenoidal
  - Transethmoidal
  - Transmaxillary
- Advantages :
  - No brain retraction and the cosmetic deficit
  - Less invasive
- Not appropriate
  - When the lateral extent of the tumor passes more than 1cm beyond the lateral limits of the exposure
  - Epicentre of the tumour does not lie within the midline
- GTR rate 100%

Schwartz TH, Fraser JF, Brown S, et al., Neurosurgery,2008;62:991

Radiation Therapy

- GTR not possible
  - Conventional RT
  - Intracavitary radiation
  - Fractionated radiotherapy
  - Stereotactic radiosurgery

Stereotactic radiosurgery

- Better control rates with single type tumors
- Mean morbidity rate 4%
- Mortality rate 0.05%
- Favourable quality of life outcome with tumours that decreased in size following GKS, while poor outcomes associated with tumour progression
- Limitation : Radio-sensitivity of the adjacent visual pathways(<8Gy)

Gopalan R, Dassoulas K, Rainey J, et al., Neurosurg Focus,2008;24:E5

Stereotactic radiosurgery

- Advantages over conventional fractionated radiation therapy
  - Greater precision
  - Reducing the volume of irradiated brain tissue
  - Delivery of higher radiation doses with less damage to adjacent neurological structures

- Concerns :
  - Vasculitis
  - Neuropsychological changes
  - Increased visual deficits

Intracavitary radiation

- Beta -emitting isotopes (Yttrium -90 ,Phosphorus -32)
- Control rates
  - 96% for cystic tumours
  - 88% for partially cystic tumours
  - Not effective for solid tumours (progression)

Gopalan R, Dassoulas K, Rainey J, et al., *Neurosurg Focus,*2008;24:E5

- Complications
  - Panhypopituitarism
  - Diabetes insipidus
  - CNS and visual dysfunction

- Combination of GKS and intracavitary irradiation with yttrium-90 or phosphorus-32 isotopes as primary therapy for mixed cystic–solid tumours.

Hasegawa T, Kondziolka D, Hadjipanayis CG, et al.,*Neurosurgery, 2004;54:813*

Outcomes

- Five -year progression-free survival rate
  - fractionated stereotactic radiosurgery 92%
  - complete excision 80–90%
  - partial resection 50–60%

Minniti G, Saran F, Traish D, et al., *Radiother Oncol,*2007;82:90

- 10-year recurrence-free survival rate
  - GTR 74–81%
  - partial removal 41–42%
  - surgery and radiotherapy 83–90%

Duff JM, Meyer FB, Ilstrup DM, et al., *Neurosurgery,*2000;46:291

- Overall survival = 80 to 91% at five- year follow-up (regardless of treatment modality)

- Best predictor of survival : an absence of recurrence

Recurrence

- Within 1 - 4.3 years
- Peri -operative mortality significantly increased
- Radiotherapy +/- surgery = significantly prevents further tumour progression
- 15-year progression-free survival =72%

  Hakuba A, Nishimura S, Inoue Y, *Surg Neurol, 1985;24:405*
- 10-year local control rate = 83%

  Stripp DC, Maity A, Janss AJ, et al., *Int J Radiat Oncol BiolPhys, 2004;58:714*

Morbidity and Management of Complications

- Increased intra- and post-operative morbidity rates
  - Diabetes insipidus
  - Hypoadrenalism
  - Hypothyroidism

    Hypopituitarism - requires lifelong treatment

- Visual fields/visual acuity improved or stabilized = 74%
- Long-term major visual field defects 48% at 10-year follow-up
- Short -term memory loss
- Personality changes
- Cranial nerve deficits
- Epilepsy
- Anosmia
- Position -dependent vertigo

- Hypothalamic dysfunction
  - Appetite changes
  - Apathy
  - Sleep disorders
  - Memory deficits
  - Hyperphagia and obesity =26–52% =
- Long -term mortality rates in adult patients five-fold higher (cardiovascular mortality).

Conclusions

- Conclusive treatment remains a matter of debate.
- When GTR is not an option, STR combined with radiotherapy becomes the therapeutic option of choice.
- Most importantly, the treatment of craniopharyngiomas is complicated both surgically and medically, necessitating a multidisciplinary approach involving neurosurgery, neurology, endocrinology, ophthalmology and neuropsychology.

Cystic tumor

- Intracavitary bleomycin / 90 Y
- Interferon alpha 2a (when all conventional therapy fails)

Thank you