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
THERAPEUTIC 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%

Radiation Therapy

- GTR not possible
  - Conventional RT
  - Intracavitary radiation
  - Fractionated radiotherapy
  - Stereotactric 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)

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)


- 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.

Outcomes

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

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

- 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%
- 10-year local control rate = 83%
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