Slow growing
Extra axial
Epithelial (Squamous)
Calcified cystic tumor occupying supra sellar region
Arising out of craniopharyngeal duct and or Rathke’s pouch

BENIGN HISTOLOGY WITH MALIGNANT BEHAVIOUR

often associated with serious long term health consequences.
INCIDENCE & CHARACTERISTICS

Most common non-glial brain tumors in children

3rd most common intracranial brain tumor in children, after medulloblastomas and gliomas

<table>
<thead>
<tr>
<th>Incidence among children</th>
<th>5 - 10% (of all pediatric brain tumors)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Incidence among adults</td>
<td>1.2 - 4% (of all adult tumors)</td>
</tr>
<tr>
<td>Most common location</td>
<td>Suprasellar region</td>
</tr>
<tr>
<td>Proportion of suprasellar tumors</td>
<td>50%</td>
</tr>
<tr>
<td>that are craniopharyngiomas</td>
<td></td>
</tr>
<tr>
<td>Peak age of incidence in children</td>
<td>5-14 Years</td>
</tr>
<tr>
<td>Peak age of incidence in adults</td>
<td>50- 60 years</td>
</tr>
<tr>
<td>Gender bias</td>
<td>None</td>
</tr>
<tr>
<td>Racial bias</td>
<td>None</td>
</tr>
</tbody>
</table>
Determ ines the presentation.

* 70% tumor involves both infra and suprasellar region
Mixed type is radiologically and clinically similar to adamantaneous type.
## PATHOLOGY

<table>
<thead>
<tr>
<th>LOCATION</th>
<th>EXTRA AXIAL</th>
</tr>
</thead>
<tbody>
<tr>
<td>MACROSCOPIC</td>
<td>LARGELY CYSTIC/SOLID-CYSTIC</td>
</tr>
<tr>
<td>MICROSCOPIC</td>
<td>Chords of columnar cells</td>
</tr>
<tr>
<td></td>
<td>Specific findings:</td>
</tr>
<tr>
<td></td>
<td>Stellate reticulum</td>
</tr>
<tr>
<td></td>
<td>Wet keratin</td>
</tr>
<tr>
<td></td>
<td>Rosentheal fibres</td>
</tr>
<tr>
<td>CALCIFICATION</td>
<td>Yes</td>
</tr>
<tr>
<td>BENIGN/MALIGNANT</td>
<td>Benign</td>
</tr>
<tr>
<td>CARCINOMATOUS AREA</td>
<td>Yet not described</td>
</tr>
</tbody>
</table>
Gliial tissue adjacent to craniopharyngioma with obvious signs of gliosis, hemosiderin deposition secondary to chronic hemorrhage, infiltration by chronic inflammatory cells, and presence of Rosenthal fibres and eosinophilic granular bodies (EGBs).

20X photomicrograph of craniopharyngioma with adjacent brain. There are multiple wet keratins, focal areas of stellate reticulum, baseloid-appearing cells – all classic histological features of the adamantinomatous variant of craniopharyngioma.
TUMOR GROWTH AND SPREAD
determines clinical behaviour and surgical approach

- **PRECHIASMATIC:**
  - originate between the optic nerves and push them posteriorly
  - more accessible and less adherent to vital suprasellar structures.

- **RETROCHIASMATIC:**
  - usually extend superiorly
  - may grow into the 3rd ventricle, Posterior fossa, producing hydrocephalus, compress the optic tracts, or grow into the hypothalamus.

Both types may reach large sizes before being diagnosed.
Recurrences usually occur at the primary site.

Ectopic and metastatic recurrences are extremely rare and have been reported after surgical removal.

In a large retrospective review, histopathologic type of craniopharyngioma and/or brain invasion did not correlate with risk of recurrence.
Insidious onset, symptomatic when tumor attains large size

Most common presentation

<table>
<thead>
<tr>
<th>Headache (retrochiasmatic &amp; intrasellar)</th>
<th>55- 86%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Visual loss (prechiasmatic)</td>
<td>37- 68%</td>
</tr>
<tr>
<td>Endocrine dysfunction (intrasellar)</td>
<td>66 – 90%</td>
</tr>
</tbody>
</table>

Symptoms mechanism:

1. Due to increased ICP: (in retrochiasmatic lesion causing hydrocephalus)
   - headache (55- 86%)
   - vomiting (projectile)
   - visual loss (papilloedema, prechiasmatic lesion also optic atrophy)
### CLINICAL PRESENTATION

2. compression to adjacent structures

<table>
<thead>
<tr>
<th>Structures</th>
<th>Presentation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypothalamus/ pituitary axis</td>
<td>endocrinopathies</td>
</tr>
<tr>
<td>Optic pathway</td>
<td>Visual field deficits (often bitemporal hemianopia).</td>
</tr>
<tr>
<td></td>
<td>Optic atrophy and visual loss</td>
</tr>
<tr>
<td>Frontal lobes &amp; thalamus</td>
<td>Dementia, Apathy, hyperphagia, obesity, short term memory loss, psychomotor retardation</td>
</tr>
<tr>
<td>Temporal lobes</td>
<td>Seizures, Amnesia</td>
</tr>
</tbody>
</table>
### ENDOCRINOPATHIES

<table>
<thead>
<tr>
<th>HORMONAL EFFECTS</th>
<th>INCIDENCE (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypothyroidism</td>
<td>40</td>
</tr>
<tr>
<td>adrenal failure</td>
<td>25</td>
</tr>
<tr>
<td>Diabetes insipidus</td>
<td>20</td>
</tr>
<tr>
<td>Short stature</td>
<td>23 - 45</td>
</tr>
<tr>
<td>Sexual dysfunction</td>
<td>80 - 90</td>
</tr>
</tbody>
</table>
WORK UP

<table>
<thead>
<tr>
<th>IMAGING</th>
<th>OPHTHALMIC EVALUATION</th>
<th>NEURO-PSYCHOLOGICAL TESTING</th>
<th>ENDOCRINE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre contrast &amp; contrast CT</td>
<td>Neuro ophthalmic evaluation</td>
<td></td>
<td>Hormone levels</td>
</tr>
<tr>
<td>MRI</td>
<td>Visual field testing</td>
<td></td>
<td>Serum electrolytes</td>
</tr>
<tr>
<td>MR Angiography</td>
<td></td>
<td></td>
<td>Serum &amp; urine osmolality</td>
</tr>
</tbody>
</table>

**CT:**
Calcification (in non contrast also)
Cystic & solid appearance
Contrast enhancement
Sellar erosion

**MRI:**
Relationship with neuro vascular structures
**Variant**

(Left) Axial NECT shows a predominantly solid, minimally calcified (arrow), suprasellar craniopharyngioma. (Right) Sagittal T1 C+ MR shows a principally cystic, sellar/suprasellar mass with rim-enhancement (arrow).
Typical

(Left) Sagittal T1WI MR shows a complex predominantly cystic suprasellar mass. T1 shortening within the cyst due to machine oil-like proteinaceous fluid (arrow).

(Right) Coronal T2WI MR shows a mixed signal, cystic suprasellar mass with internal hypointense elements representing calcification (arrow).
Fig. 5.43a: MRI in lateral view. Large cystic craniopharyngioma in an adult with severe hormonal deficiency, drowsiness and visual deficit. The yellow arrow points to the small solid part, the red to the large cyst.
Fig. 5.42a: Typical craniopharyngioma at frontal and lateral MRI scan. Yellow arrows point to the cyst, the red to the solid part of the tumor at blue to a dilated lateral ventricle (hydrocephalus).

Fig. 5.42b: Same case as in Fig. 5.46 after complete removal with a minimvasive supraorbital approach. The green arrow points to the optic chiasm.
The prognosis in this disease is very variable. Some people live progression-free lives without treatment, whereas others have continued tumor progression despite extensive treatment.

- craniopharyngiomas tend to shorten life, and can more aptly be called low-grade malignancies.

One of the most important prognostic factors is recurrence

- Presence of calcification
- Incomplete tumor resection
- Severe hydrocephalus
- Adverse intraoperative events
- Age of less than 5 years
1. GROSS TOTAL RESECTION (GTR)

2. SUBTOTAL RESECTION (STR) + RADICAL RADIATION THERAPY

OTHER OPTIONS:

- intracystic injection of bleomycin
- intra cavity radiotherapy
- systemic therapy ??
- systemic biological therapy (under trial Interferon alfa 2a)
no consensus………. controversial

Successful management is determined by the ability to maintain independent social functioning, symptomatic recurrence, and survival.

Neuropsychological deficits represent the major limiting factor of independent social functioning.
Gross Total Resection (GTR)
treatment of choice
## GROSS TOTAL RESECTION (GTR)

- Favourable outcome. Local control is 85-100%.
- Mostly suitable for small tumor, only 50-80% cases.

### Surgical Technique vs. Indications

<table>
<thead>
<tr>
<th>Surgical Technique</th>
<th>Indications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bifrontal and subfrontal approach</td>
<td>Suprasellar tumors (prechiasmatic and large retrochiasmatic lesions)</td>
</tr>
<tr>
<td>Trans-sphenoidal approach</td>
<td>Cystic infradiaphragmatic lesions</td>
</tr>
<tr>
<td></td>
<td>Symmetrical and well-defined suprasellar and retrosellar lesions with enlarged sella</td>
</tr>
<tr>
<td></td>
<td>Tumors without calcification</td>
</tr>
<tr>
<td></td>
<td>Lowers surgical morbidity and postoperative visual loss</td>
</tr>
<tr>
<td>Stereotactic aspiration of cyst</td>
<td>Cystic tumors</td>
</tr>
</tbody>
</table>

Approach depends on the anatomical location.
Endocrinopathy is common.

Recurrence/progession following failed gross total or subtotal resection is common and occurs in 75% of patients.

<table>
<thead>
<tr>
<th>Condition</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Residual calcification or tumor after resection</td>
<td>15-20%</td>
</tr>
<tr>
<td>Clinical recurrence</td>
<td>10-30%</td>
</tr>
<tr>
<td>Diabetes insipidus</td>
<td>50-80%</td>
</tr>
<tr>
<td>Panhypopituitarism</td>
<td>75-100%</td>
</tr>
<tr>
<td>Neuropsychologic and behavioural disturbances</td>
<td>36-60%</td>
</tr>
<tr>
<td>Morbid hypothalamic obesity</td>
<td>50%</td>
</tr>
<tr>
<td>Visual deterioration</td>
<td>2-45%</td>
</tr>
<tr>
<td>Fusiform dilation of internal carotid artery</td>
<td>15%</td>
</tr>
</tbody>
</table>
MAXIMAL SAFE RESECTION
THE ULTIMATE KEY PRINCIPLE OF SURGICAL MANAGEMENT
MODIFIED APPROACH

SUBTOTAL RESECTION (STR) with postop RT

* Goals of this approach

(1) pathologic confirmation of the tumor
(2) Less chance of damage of near by structures
## GTR VS STR + PORT

<table>
<thead>
<tr>
<th>Modality</th>
<th>Local control (%)</th>
<th>Recurrence rate (%)</th>
<th>10 yr survival (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>GTR + OBSERVATION</td>
<td>70</td>
<td>32</td>
<td>24- 100</td>
</tr>
<tr>
<td>STR + PORT</td>
<td>75</td>
<td>NIL</td>
<td>62 - 86</td>
</tr>
</tbody>
</table>

Lesser chance of hypothalamic and pituitary dysfunction. Negetive impact on IQ is less in STR+PORT arm

children’s Memorial Hospital , Chicago experience
### RECOMMENDED MANAGEMENT

**Risk factors**

<table>
<thead>
<tr>
<th>Good</th>
<th>Poor</th>
</tr>
</thead>
<tbody>
<tr>
<td>Small tumour (2–4 cm)</td>
<td>Large (retrochiasmatic) tumour (&gt;2–4 cm)</td>
</tr>
<tr>
<td>No hydrocephalus</td>
<td>Hydrocephalus</td>
</tr>
<tr>
<td>No hypothalamic symptoms</td>
<td>Hypothalamic symptoms</td>
</tr>
<tr>
<td>No breach third ventricle floor</td>
<td>Breach third ventricle floor</td>
</tr>
</tbody>
</table>

**Consideration**

<table>
<thead>
<tr>
<th>Good</th>
<th>Poor</th>
</tr>
</thead>
<tbody>
<tr>
<td>Consider GTR + OBSERVATION/ RT</td>
<td>Consider STR + adjuvant RT</td>
</tr>
</tbody>
</table>

**SOURCE:** European Journal of Endocrinology (2011)
RADIATION THERAPY

SCOPE:
- Sub total resection (STR)
- Progressive disease after GTR
- Recurrent disease after GTR

Principles:
- Radioisotope Therapy
- External-Beam Radiation Therapy
**RADIOISOTOPE THERAPY**

- Stereotactic or endoscopic instillation of colloidal therapeutic radioisotopes e.g. yttrium 90, phosphorous 32.

- Beta particles have short penetrance, no damage to surrounding structures.

- In Cystic tumor as primary modality or recurrent tumor after conventional EBRT

- No effect on solid part of tumor
EBRT

- Target Volume/Critical Structures
- Patient positioning
- Beam Arrangement
- Dose
- Sequelae

TECHNIQUE IS SIMILAR AS IN PITUTARY TUMORS
2 cms anterior and 2 cms superior to tragus is the landmark for pituitary gland clinically.

- Setting a 5x5 cms square field around the pituitary point gives the target volume.

- Fields: bilaterally opposing

- Dose: 50-54 Gy @ 1.8-2 Gy/fr

- Beam energy: < 6MV photons
ORGAN(S) AT RISK

- Lens (10Gy)
- Retina (50Gy)
- Optic nerve (60Gy)
- Tumor tissue
- Temporal lobe (50Gy)
- Mid brain (54Gy)

Others:
1. Optic chiasma (54Gy)
2. Pituitary gland (50Gy)
3. Middle ear (55Gy)
Pt’s head immobilisation and positioning is essential part.

PATIENT IS SUPINE AND HEAD IS FLEXED

Head is flexed extremely to neck (around 60 degree). Ant beam avoids OAR eg optic chiasma, vital brain tissue. Thermoplastic mask with this head support fulfill the purpose
TRADITIONAL 3 FIELD APPROACH:

1. Wedged opposed lateral beams
2. Vertex beam superior to the eye

Problem - high temporal lobe dose

True vertex beam is avoided
5-10% gantry rotation so that beam does not exit through body (specially in female)
Couch rotation so that beam becomes supero lateral
Conventional RT
MODERN METHOD OF PLANNING & TREATMENT

3DCRT
SRS
SRT
IMRT
ARC THERAPY
PLANNING STEPS

- Positioning
- Immobilization
- Localisation
- Target volume delineation.
- Treatment
POSITIONING AND IMMOBILISATION

- Supine position
- Head flexed with neck
- Immobilisation with thermoplastic mold limits error to 3-5 mm.

- For SRS and SRT:
  Immobilisation with fixed frame [Brown Robert Wells] decreases motion to 0.3 mm
  Use of re-locatable device [Gill Thomas Cosman System] decreases motion to 1.5-2 mm.
Patient Positioning with NATURAL NECK REST
FIGURE 5.11. A: A set of Timos head and neck supports indexed to recreate a specific head height or neck slant day after day. Timos are typically either letter or color coded. B: Hollow clear plastic Timos to reduce scatter and potential skin reaction. (Courtesy of Bionix, Akron, Ohio.)

FIGURE 5.18. Combination of head and neck supporting foam Timo and tilt board provides immobilization and patient positioning. (Courtesy of Nuclear Associates, Carle Place, NY.)
FIGURE 5.29. A: Relocatable head immobilization device (Gill-Thomas-Cosman system) for stereotactic radiotherapy. B: The same device in place on a patient. This system makes use of a dental mold and an occipital tray with a cast of the occiput for stabilization. (Courtesy of Radionics, Burlington, MA.)

FIGURE 5.2. A: The BRW/CRW (Brown-Roberts-Wells/Cosman-Roberts-Wells) head ring assembly for stereotactic radiosurgery. B: Head ring assembly attached to the treatment couch for accelerator-based radiosurgery. (Courtesy of Radionics, Burlington, MA.)
HEAD POSITIONING
LOCALISATION

- CT based.
- Patient lies supine on CT couch in the same position with head rest and thermoplastic immobilisation.
- Take CT (CECT) cuts 3-5mm.
- CT data of patient is networked to treatment planning system.
**Targets**

* **GTV:** Both cystic & solid components of any residual/recurrent disease; Entire surgical bed, disease left at the stalk

* **CTV = GTV + 0-5 mm margin**

* Contour GTV CTV and OAR

**PTV** is additional margin given for geometric uncertainties of immobilisation, motion and set up variation.

* a margin of 5 mm around CTV is given.

**PTV ALSO DEPENDS ON TREATMENT TECHNIQUES**
Planning Aim:
Achieving a homogenous dose distribution in PTV and a high conformal dose around it.

Field arrangements: NON COPLANAR

3 field non coplanar -- one vertex
  two oblique

5 field non coplanar -- 2 supero-lateral non opposed
  3 oriented along sagittal planes
3 FIELD NON COPLANAR TECHNIQUE

* LEFT POSTERIOR OBLIQUE
* RIGHT POSTERIOR OBLIQUE
* VERTEX
Left Posterior Oblique

Gantry=100
Collimator=0
Couch=10
Right Posterior Oblique

Gantry = 260
Couch = 350
Collimator = 0
Vertex field

Gantry = 20
Couch = 90
Collimator = 0
MODERN TECHNIQUES

- 5 non co-planer shaped static beams.
- 2 supero-lateral non opposed
- 3 oriented along sagittal planes

Suggested beam arrangements: LT15S, RT15S, SG30A, SG60P, P

IMRT:
Improved dose distribution specially in irregular shaped lesion
Four T2 weighted MRs are shown. The first is the baseline MR that was taken 10 days before the start of treatment, the second was taken 6 days after the start of treatment, the third was 14 days after the start of treatment, and the last was taken 35 days after treatment. Also shown are the contour for the baseline GTV and the contour of the day 6 adaptive GTV on each image.

Some times cyst expansion occurs changing the size of GTV during treatment course.

It may be critical to monitor tumor size and to create an adaptive plans when target volume increases.
Challenges in long-term survivors

<table>
<thead>
<tr>
<th>Challenge</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cognitive dysfunction</td>
<td>38%</td>
</tr>
<tr>
<td>Motor deficit</td>
<td>25%</td>
</tr>
<tr>
<td>Visual impairment</td>
<td>20%</td>
</tr>
<tr>
<td>Hormonal dysfunction</td>
<td>20%</td>
</tr>
<tr>
<td>Psychological-emotional problems</td>
<td>14%</td>
</tr>
</tbody>
</table>

Second malignancies
Cerebrovascular events
STEREOTACTIC RADIATION:

- Very precise delivery of radiation to a brain tumor with sparing of the surrounding normal brain
- Special tumor localisation technique

TWO TYPES

- 1. SRS
- 2. SRT
STEREOTACTIC CONFORMAL RADIATION THERAPY

- SCRT is a high precision fixed field technique using multiple beams and customized shielding.
- **Immobilisation** in BrainLAB mask/frame system
- **Imaging** Planning CT with mask/frame and MR image
- CT MR fusion in Brain Lab
- PTV is 2mm with CTV
- **Planning technique**: 6-8 non-coplanar fields, individually shaped with microMLC of the BrainLAB.
Stereotactic Conformal Radiotherapy (SCRT)

Accurate immobilisation
MRI planning
Tight conformation

High Quality Assurance
Precise treatment delivery
Dose distribution
SCRT: Heidelberg experience
(n=40)

- Median dose: 52.2 Gy @ 1.8 Gy/#
- Median FU: 98 months
- Median PFS at 5 & 10 yrs: 97% & 89%
- OS at 5 & 10 yrs: 100%
- No pts had visual deterioration after RT

SCRT provides acceptable local control & toxicity

Coombs et al Cancer 2007
STEREOTACTIC RADIO SURGERY

- Highly specific, allows PTV up to 2mm +CTV
- Radiosurgery is having a limited role. Results not good.
- This option should be confined only in tumors confined to pituitary fossa, away from chiasma and hypothalamus
- LINAC: X KNIFE
- COBALT60: GAMMA KNIFE
- CYBER KNIFE
OTHER MODERN TECHNIQUES

PRTON BEAM THERAPY

VOLUME MODULATED ARC THERAPY (VMAT)

TOMOTHERAPY
**ENDOCRINE MANAGEMENT**

- HORMONE REPLACEMENT THERAPY

**Disturbances of thirst and appetite are the most challenging aspects of management**

- POST OPERATIVE DIABETES INSIPIDUS
  Careful water and electrolyte management
  DDAVP

- HYPOTHALAMIC OBESITY
  Bariatric surgery
  Octreotide
  Sibutramine
FOLLOW UP
multi team approach

* Neuro cognitive assessment
* Neuro psychological functioning

* Physical activity functioning
* Quality of life

* Neurological, radiological, endocrinal, ophthalmological assessment
THANK YOU