

CRANIOPHARYNGIOMA



HISTORY

- CRANIOPHARYNGIOMA
- Mclean in 1930,
- Frazier and Alpes in 1931
- Cushing in 1932



- **CUSHING** “MOST FORBIDDING OF THE INTRACRANIAL TUMORS”
- **RUTKA** “THERE IS PERHAPS NO OTHER BRAIN TUMOR THAT EVOKES MORE PASSION, EMOTION, AND CONTROVERSY AS CRANIOPHARYNGIOMA”



- THERAPUETIC GOALS ARE THE CURE OF DISEASE WITH FUNCTIONAL PRESERVATION AND RESTORATION.
- THIS IS OFTEN A WIDELY DEBATED TOPIC WITH CONTROVERSY INCLUDING EXTENT OF SURGICAL RESECTION, SURGICAL APPROACH, AND THE USE OF ADJUVANT THERAPY



OTHER NAME OF THE TUMOUR

- AMELOBLASTOMA
- ADAMANTINOMA
- RATHKE'S POUCH TUMOURS
- CRANIOPHARYNGEAL DUCT TUMOUR
- HYPOPHYSEAL DUCT TUMOUR
EPITHELIOMA



EMBRYOGENESIS

- ARISES FROM SQUAMOUS EPITHELIAL REST ALONG THE INVOLUTED HYPOPHYSEAL RATHKE'S DUCT



GROSS PATHOLOGY

- WELL DELINEATED LOBULATED CYST WITH A MURAL NODULE MOST COMMON
- MIXED CYSTIC AND SOLID OCCURS LESS FREQUENTLY
- CYST CONTENTS RANGE FROM STRAW COLOURED FLUID TO CRANK CASE LIKE OILY MATERIAL RICH IN CHOLESTEROL MURAL NODULES OFTEN CONTAIN GRITY CALCIFIC FOCI



MICROSCOPIC APPEARANCE

- SQUAMOUS EPITHELIUM WITH NECROTIC DEBRIS, CHOLESTEROL CLEFTS, KERATIN PEARLS

TWO DISTINCT PATTERN –

- ADAMANTINOUS PATTERN
- PAPILLARY PATTERN



ADAMANTINOUS PATTERN

- MORE COMMON
- OCCURS IN CHILDREN
- SOLID AND CYSTIC COMPONENT
- CALCIFICATION/KERATIN PEARL FORMATION
- GROWTH IN BRAIN MAY PRODUCE GLIOTIC CAPSULE WITH ROSENTHAL FIBRES
- A LOOSE COLLECTION OF STELLATE CELLS IS SURROUNDED BY A LAYER OF PSEUDO STRATIFIED COLUMNAR CELLS RESTING ON A BASEMENT MEMBRANE.



PAPILLARY TUMOUR

- RARER (10 FOLD RARER)
- OCCURS IN ADULTS
- SOLID
- NO CALCIFICATION/ KERATIN PEARL/
MOTOR OIL
- COMPOSED OF ANASTOMOSING CORDS OF
SQUAMOUS EPITHELIUM



INCIDENCE

- 3-5% OF PRIMARY BRAIN TUMOURS
- 50% OF PAEDIATRIC SUPRA SELLAR TUMOURS
- REPORTS OF INVOLVEMENT OF CHROMOSOME 2 AND 12 (R. RICK BHASIN, MD)



AGE

- >50% IN CHILDREN PEAK AGE 8-12 YEARS
- SECOND PEAK IN ADULTS 40-60 YEARS
- NO GENDER DIFFERENCE



LOCATION

- 70% COMBINED SUPRASELLAR/
INTRASELLAR
 - COMPLETELY INTRASELLAR
- CRANIOPHARYNGIOMAS ARE RARE



BLOOD SUPPLY

INTRASELLAR PORTION -

- TWO BRANCHES FROM THE INTRACAVERNOUS PORTION OF THE IC OR THE INFERIOR HYPOPHYSEAL ARTERIES

SUPRASELLAR PORTION –

- ANTERIOR CEREBRAL, ACoA
- BRANCHES OF THE PCA

**PROXIMAL PCA INTRAVENTRICULAR TUMOUR OR
CLOSE TO THE FLOOR OF THE THIRD VENTRICLE**



VERTICAL TUMOR EXTENSION (CLASSIFICATION BY MADJID SAMII)

- GRADE I: LOCATED PRIMARILY IN INTRASELLAR OR INFRADIAPHRAGMATIC REGION
- II: LOCALIZED IN THE CISTERN WITH OR WITHOUT INTRASELLAR COMPONENT
- III: EXTEND INTO THE LOWER HALF OF THIRD VENTRICLE
- IV: EXTEND INTO THE UPPERHALF OF THIRD VENTRICLE.
- V: TUMORE DOME REACHES THE SEPTUM PELLUCIDUM AND/OR EXTENDS INTO THE LATERAL VENTRICLE.



CLINICAL FEATURES

- SYMPTOMS OF RAISED ICP PREDOMINATE IN CHILDREN
- ENDOCRINOLOGICAL DEFICITS (↓ED THYROID HORMONE, GH, CORTISOL, DI) AND VISUAL SYMPTOMS PREDOMINATE IN ADULT
- ELDERLY PRESENT WITH MENTAL DISTURBANCES
- SHORT STATURE DELAYED PUBERTY NEURO BEHAVIOURAL ABNORMALITY



IMAGING

XRAY- IRREGULAR SPECKLED CALCIFICATION
SEEN JUST ABOVE THE SELLA TURCICA

- THE SEMICIRCULAR SHELL OUTLINING THE WALL OF CYSTIC LESION.
- FINE FLAKY CALCIUM IS ENCOUNTERED WITH FAST GROWING TUMOURS .
- SLOW GROWING TUMOURS SHOW DENSE CALCIFICATION.



IMAGING

- MOSTLY SUPRASELLAR
- CALCIFICATION MAY BE IN CYST WALL AND/OR SOLID COMPONENT
- CT - 90% PARTIALLY CYSTIC
 - 90% PARTIALLY CALCIFIED
 - 90% NODULAR / RIM ENHANCEMENT



MRI

- MOST HETEROGENOUS MR IMAGING SPECTRUM OF ALL SELLAR REGION MASSES.
- MOST COMMON IS HYPO ON T1 AND HYPER ON T2W1.
- ENHANCES STRONGLY/ HETEROGENOUSLY ON CONTRAST ADMINISTRATION.



- MULTILOBULAR, MULTICYSTIC MASSES
- CYSTS MAY HAVE DIFFERENT SIGNAL DEPENDING ON FLUID CONTENT
- OFTEN BOTH CYST WALLS AND SOLID COMPONENTS ENHANCE
- RARELY COMPLETELY SOLID
- DDX- RATHKE CLEFT CYST (NO ENHANCEMENT)



COMPARISON OF COMMON SUPRASELLAR LESIONS

Lesions	Age Group	Clinical Features	CT		MRI		Comments
			Unenhanced	Enhanced	T1W1	T2W2	
Pituitary macroadenoma	Adult tumours (< 10% in Children)	75% endocrinologically active : symptom vary with type of adenoma	Isodense	Modest uniform enhancement	Isointense; enhances strongly,	Isointense	Calcification is rare; displacement rather than invasion of adjacent structures; often lobulated (figure eight); mass indistinguishable from pituitary
Pituitary macroadenoma (hemorrhagic)			Inhomogeneously hyperdense	Hyperdense	Often complex mixed signal	Complex mixed signal	Signal changes with age of clot
Meningioma	40-60 yrs of age	Seizures hemiparesis: symptom depending on site	Slightly hyperdense	Strong uniform enhancement	Isointense (may be inconspicuous) ; enhances strongly	Variable	Smooth well -delineated lesion; calcification is common; dural "tail"; pituitary gland distinct from mass



COMPARISON OF COMMON SUPRASELLAR LESIONS

Lesions	Age Group	Clinical Features	CT		MRI		Comments
			Unenhanced	Enhanced	T1W1	T2W2	
Craniopharyngioma	8-12 yrs; 40-60 yrs of age	Raised ICP, endocrinological deficit; visual symptom; short stature; delayed puberty	Heterogeneous; cystic: 90% partially cystic, 90% partially calcified	Variable: 90% Nodular / Rim enhancement	Variable; Hypo	Hyper	Focal calcification is common (rim, globular) location: 70% intra & suprasellar 20% suprasellar only 10% intrasellar only
Glioma (opticochiasmatic or hypothalamic)	Children	Vary with location	Isodense or slightly hypodense	enhancement	Hypo to Isointense; variable enhancement	Slightly hyperintense (may remain isointense)	May see chiasmal enlargement; calcification rare; retrochiasmatic extension is common (N.B. occasionally compressive lesions such as craniopharyngioma or adenoma can cause hyperintensity in adjacent brain)

COMPARISON OF COMMON SUPRASELLAR LESIONS

Lesions	Age Group	Clinical Features	CT		MRI		Comments
			Unenhanced	Enhanced	T1W1	T2W2	
Aneurysm (patent)	40-60 yrs of age	Of SAH; Cranial neuropathies	Slightly hyperdense	Strong uniform enhancement	Flow void	Flow void	Turbulent flow may give inhomogeneous signal; ICA or ACoA are most common locations; May see rim calcification
Aneurysm (partially thrombosed)			Slightly hyperdense	Nonenhancing in area of thrombus; strongly enhancing patent Lumen; may see rim enhancement	Thrombus variable	Variable	Thrombus may appear heterogeneous (laminated blood products in different stages)
Rathkes Cleft Cyst	Any age but mostly adults (40-60 yrs)	Asymptomatic; visual disturbances ; hypothalamic/ pituitary dysfunction	75% Hypodense non calcified; lack solid component	50% Rim (Capsular enhancement)	Hyper intense	Variable	

RATHKE'S CLEFT CYST

LACK SOLID COMPONENT

CT- 75% HYPODENSE TO BRAIN

NONCALCIFIED

50% RIM (CAPSULAR) ENHANCEMENT

MRI- MOST COMMON HYPER INTENSE ON T1W1
WITH VARIABLE SIGNAL ON T2W1



PITUTARY ADENOMA

- MICROADENOMA – HYPODENSE/HYPOINTENSE COMPARED TO NORMAL PITUTARY ON DYNAMIC CECT OR MRI
- MACROADENOMA-
NECT: ISODENSE, ONLY 1-8% CALCIFY
CECT: ENHANCE INTENSELY
MR: SIGNAL LIKE CORTEX ON T1-, T2 WI IS MOST COMMON PATTERN; VARIABLE SIGNAL IF HAEMORRHAGE, NECROSIS, CYST FORMATION.



CYSTIC HYPOTHALAMIC OPTICO CHIASMATIC GLIOMA

- HYPO TO ISODENSE ON T1W1
- MILD TO MODERATE ENHANCEMENT FOLLOWING CONTRAST



THROMBOSED ANEURYSM

- VARIABLE MRI FINDINGS
 - ACUTELY THROMBOSED:
ISOINTENSE WITH BRAIN PARENCHYMA
 - SUBACUTE THROMBOSIS:
HYPERINTENSIVE ON T1 AND T2 WEIGHTED
STUDIES



TREATMENT

PREOPERATIVE EVALUATION AND MANAGEMENT-

- COMPLETE ENDOCRINOLOGICAL EVALUATION TO UNCOVER HYPOPITUITARISM PARTICULARLY ---
GROWTH HORMONE
-CORTISOL
-THYROID HORMONE



DILEMMAS IN MANAGEMENT DECISIONS

- TOTAL EXCISION
- SUBTOTAL EXCISION
- HORMONAL DISTURBANCES MINIMAL
- RECURRENCE –DO RADIOTHERAPY
- MINIMISE HORMONAL DEPENDENCE AND RECURRENCE
- ILL EFFECTS OF RADIATION NEAR BASE OF BRAIN IN CHILDREN



SURGICAL MANAGEMENT

- 1910 LEWIS DID FIRST SUCCESSFUL EXCISION
- ASSOCIATED HYDROCEPHALUS –MORE COMMON IN RETROCHIASMAL TUMOURS THAN IN PRECHIASMAL VARIETY
- RECURRENT OR RESIDUAL TUMOUR, ASEPTIC MENINGITIS ,CSF RHINORRHEA MAY NECESSIATE A SHUNT INSERTION POST OPERATIVELY/ EVD



OPERATIVE APPROACH

- LOCATION AND EXTENT OF TUMOUR
- CONFIGURATION OF THE VISUAL PATHWAYS
- BLOOD SUPPLY OF THE TUMOUR AND OPTIC APPARATUS
- ENLARGEMENT OF THE SELLA AND TYPE OF SPHENOID SINUS ARE IMPORTANT IF A TRANSSPHENOIDAL APPROACH IS BEING CONSIDERED



FIVE VARIETIES OF CRANIOPHARYNGIOMA ARE RECOGNISED FOR SURGICAL MANAGEMENT

- (A)SELLAR
- (B)PRECHIASMAL
- (C)RETROCHIASMAL
- (D)INTRAVENTRICULAR
- (E)GIANT



(A)SELLAR

- TRANSSPHENOIDAL
- SUBFRONTAL
- TRANSCRANIAL-TRANSSPHENOIDAL



(B)PRECHIASMAL

- SUBFRONTAL- INTEROPTIC
- PTERIONAL



(C)RETROCHIASMAL

- SUBFRONTAL
- PTERIONAL – OPTICOCAROTID
 - TRANSSPHENOIDAL
 - LAMINA TERMINALIS
 - LATERAL CAROTID
- SUBTEMPORAL- INTERPEDUNCULAR CISTERN
- TRANSPETROSAL TRANSTENTORIAL
- CRANIOBASAL MEDIAN SPLITTING



(D)INTRAVENTRICULAR

- TRANSCALLOSAL
- TRANSCORTICAL TRANSVENTRICULAR
- LAMINA TERMINALIS

SUBFRONTAL

INTERHEMISPHERIC



(E)GIANT

- COMBINED APPROACHES IN ONE OR MULTIPLE STAGES



- LAMINA TERMINALIS IS COMPLETELY AVASCULAR AND CAN BE ENTERED WITH SAFETY TO DEAL WITH RETEROCHIASMAL TUMOURS
- PREFIXED CHIASMA MAKES THE SURGICAL TASK MORE DIFFICULT AS THERE IS HARDLY ANY INTEROPTIC SPACE TO APPROACH THE TUMOUR



RADICAL SURGERY VERSUS CONSERVATIVE SURGERY AND RADIATION

- TOTAL EXCISION SHOULD BE THE AIM PROXIMITY AND ADHERENCE OF THE LESION TO THE OPTIC PATHWAYS AND ADJACENT NEUROVASCULAR STRUCTURES OFTEN MAKE TOTAL EXCISION HAZARDOUS
- SMALL OR PRECHIASMATIC CAN BE TOTALLY EXCISED
- RETEROCHIASMATIC, LARGE OR MULTICOMPARTMENTAL- TOTAL EXCISION IS PROBLEMATIC

RADICAL RESECTION

- CURE 85%
- SEVERE COMPLICATIONS 5-10% (VISUAL LOSS/
NEUROLOGIC DYSFUNCTION)
- HYPOPITUITARISM 90-95%
- DIABETES INSIPIDUS 95%
- QUALITY OF LIFE IS THE MAIN ISSUE.
- MORBIC HYPOTHALAMIC OBESITY 50%



RADICAL RESECTION

ADVANTAGE

- ONE TREATMENT THEN ONLY FOLLOW-UP.

DISADVANTAGES

- LIMITED NUMBER OF SURGEONS WITH ADEQUATE EXPERTISE
- DIFFICULT TO ASSESS TRUE RISKS TO INDIVIDUAL CHILD.
- DIFFICULT TO JUDGE THE CHANCE OF SERIOUS CHANGES IN PERSONALITY (IMPAIRED QUALITY OF LIFE).
- DIABETES INSIPIDUS



LIMITED SURGERY + RADIATION

- CURE 85%
- SEVERE COMPLICATIONS 5-10%
- HYPOPITUITARISM 90-95%
- DIABETES INSIPIDUS 5%
- QUALITY OF LIFE IS THE MAIN ISSUE



LIMITED SURGERY + RADIATION

ADVANTAGES

- RARE TO CHANGE PERSONALITY.
- SURGERY CAN BE PERFORMED WITH LIMITED EXPERIENCE

DISADVANTAGES

- DECREASE IN IQ
- CYST MANAGEMENT (OFTEN MULTIPLE CYST PROCEDURES)
- DECOMPRESSION OF CHIASM SOMEWHAT DIFFICULT AND STILL MAINTAIN LIMITED SURGERY GUIDELINES
- COMPLICATIONS OF RADIATION



LIMITED SURGERY + RADIATION

- RADICAL SURGERY FOR ATTEMPTED CURE AFTER FAILURE OF RADIATION IS NOT MORE DIFFICULT THAN ORIGINAL ATTEMPT WOULD HAVE BEEN.
- DESTRUCTION OF ARACHNOID PLANES BY SURGICAL MANIPULATION COMBINED WITH RADIATION PRODUCES THICK SCAR
- IF ARACHNOID PLANES WERE NOT VIOLATED THEN SURGERY NOT MORE DIFFICULT.



GOALS OF LIMITED SURGERY

- DIAGNOSIS
- DRAIN CYSTS
- LIMIT FIELD OF RADIATION
- CONTROL HYDROCEPHALUS
- IMPROVE VISION
- DECOMPRESS CHIASM



LIMITED SURGERY +RADIOTHERAPY

- CYSTS ARE PROBLEMATIC
- MOST ARE EASILY DRAINED WITH STEREOTAXIC PLACED CATHETER AND RESERVOIR. SOME HAVE THICK WALLS AND REQUIRE SURGICAL COLLAPSE.
- NOT UNCOMMON FOR CYST TO PROGRESSIVELY ENLARGE DURING RADIATION THEN SHRINK LATER.
- RARELY REQUIRE ADDITIONAL TREATMENT (P-32).



DIFFICULT EXCISION

- FINGER LIKE PROCESSES OF TUMOUR MAY BURROW INTO HYPOTHALAMUS
- HYPOTHALAMIC DYSFUNCTION RESULTS IN ENDOCRINE, METABOLIC AND PSYCOSOCIAL DISTURBANCES WITH RESULTANT IMPAIRED QUALITY OF LIFE.



- WITH THE USE OF STEROIDS AND MAGNIFICATION THE MORTALITY IS LESS THAN 10%.
- MORTALITY AND MORBIDITY IS HIGHER FOR RECURRENT TUMOURS



- CECT SCAN OR MRI 6-8 WEEKS AFTER OPERATION WILL PICK UP A RESIDUAL LESION THAT MAY BE TACKLED STRAIGHT AWAY BEFORE IT BECOMES LARGE AND DEVELOPS FIRM ADHESIONS TO ADJACENT NEUROVASCULAR STRUCTURES.



- PATIENTS UNDERGOING TOTAL OR SUBTOTAL EXCISION NEED PREOPERATIVE ENDOCRINE THERAPY, ENDOCRINE SUPPORT DURING SURGERY AND SUSTAINED REPLACEMENT THERAPY FOR MANY YEARS FOLLOWING SURGERY.



RADIOTHERAPY

- RESULTS OF SUBTOTAL EXCISION CAN BE BETTERED WITH SUPPLEMENTARY RADIOTHERAPY.
- ENDOCRINOLOGICAL AND PSYCHOSOCIAL DYSFUNCTION
- VASCULAR OR NEUROLOGIC COMPLICATIONS ATTRIBUTABLE TO RADIATION WERE MINIMAL WHEN THE DOSE DID NOT EXCEED 6000GY.



RADIOTHERAPY

- DELAYING OR PREVENTING A RECURRENCE AND IN INCOMPLETELY EXCISED OR RECURRENT TUMOR
- LARGE CYSTIC TUMOURS CAN BE TREATED WITH BETA EMITTING RADIONUCLIDE COLLOIDS SUCH AS ^{90}Y OR ^{32}P



COMPLICATIONS OF RADIOTHERAPY

- OPTIC NEUROPATHY AND RADONECROSIS OF BRAIN
- OCCLUSION OF MAJOR VESSELS MAY RESULT IN ISCHAEMIC ATTACK OR STROKES
- PITUITARY DYSFUNCTION



SURGICAL TECHNIQUE

- 1. UNILATERAL OR BILATERAL FRONTAL APPROACH BE IDEAL FOR EXCISION OF MOST OF THE TUMOURS AS IT GIVES GOOD VIEW OF OPTIC NERVES, CHIASMA AND INTEROPTIC SPACE**
- 2. SUBFRONTAL EXPOSURE IS IDEAL FOR RETROCHIASMATIC.**

TRANSLAMINAR APPROACH WHEN THE CHIASMA IS PREFIXED



INTERHEMISPHERIC APPROACH

- ROUGH LAMINA TERMINALIS FOR EXCISION OF RETROCHIASMATIC INTRAVENTRICULAR LESIONS.
 - BOTH THE OLFACTORY NERVES ARE SPARED
 - MAIN ARTERIES EXPOSED SUFFICIENTLY WITH A WIDE OPERATION FIELD TO RENDER PROCEDURE SAFE.
 - AVOIDS DAMAGE TO LATERAL WALL OF THIRD VENTRICLE.
- 

SUBTEMORAL APPROACH

- GOOD VISUALISATION OF TUMOUR BETWEEN IIIrd NERVE AND PCA INFERIORLY AND OPTIC TRACT SUPERIORLY.
- CAN BE USED IN COMBINATION WITH THE PTERIONAL APPROACH FOR EXCISING SMALL RETROCHIASMATIC TUMOURS OR PARASELLAR LESIONS WITH EXTENSION INTO MIDDLE FOSSAE.



TRANSSPHEROIDAL APPROACH

- FOR TUMOURS THAT ARE INTRASELLAR AND INFRADIAPHRAGMATIC IN LOCATION AND MAINLY CYSTIC WITH A PRIMARILY ENLARGED SELLA .
- PREDOMINANTLY CALCIFIED FIRM FLESHY TUMOUR LEND THEMSELVES POORLY TO REMOVAL BY THIS ROUTE.



- MOSTLY SUBFRONTAL APPROACHES ARE USED FOR LESIONS THAT NEED A MORE LATERAL APPROACH
- TWO STAGE FRONTOTEMPORAL AND POSTERIOR FOSSA CRANIOTOMES ARE NECESSARY FOR EXCEEDINGLY LARGE AND SPRAWLING TUMOR.



- FOR LESIONS THAT NEEDED A MORE LATERAL APPROACH THE PTERIONAL OR SUBTEMPORAL APPROACH WAS USED.
- FOR ESSENTIALLY INTRAVENTRICULAR LESION. EITHER THE TRANSCORTICAL, TRANSVENTRICULAR OR TRANSCALLOSAL APPROACH WAS USED.



- OPERATIVE MORTALITY LIES BETWEEN 0-10 % AND DEPENDS ON THE TUMOUR SITE AND EXTENT OF THE ATTEMPTED REMOVAL



A RECURRENCE RATE OF UPTO 30% WITH IN 10YEARS OF AN APPARENT TOTAL REMOVAL (CAUSED BY RESIDUAL TUMOUR EXTENSIONS LYING BEYOND THE CAPSULE).

WITH SUBTOTAL REMOVAL ,THE RECURRENCE RATE APPROACHES 57% BUT WITH RADIOTHERAPY FALLS IT TO 27% AFTER 5 YEARS



POST OPERATIVE FOLLOW UP

- POST OPERATIVE CT WITH AND WITHOUT CONTRAST TO BE OBTAINED WITH IN 24 HOURS TO DETECT RESIDUAL TUMOUR (WHEN CONTRAST ENHANCEMENT OF THE SURGICALLY TRAUMATIZED BRAIN IS MINIMAL) PRESENCE OF UNSUSPECTED RESIDUAL TUMOUR ON THE NEXT DAY CT (SECOND LOOK OPN WITHIN 1-2WEEKS)BEFORE EXUBERANT SCARRING MAKES REOPERATION DIFFICULT



- IF THE NEXT DAY CT SHOWS NO RESIDUAL OR IF THE SECOND ATTEMPT AT TOTAL RESECTION ACCOMPLISHES ITS GOAL ,THE NEXT CT AND MRI CAN BE OBTAINED IN 3-6 MONTHS. THEREAFTER YEARLY CT AND MRI ,COMPLETE ENDOCRINE EVALUATION SHOULD BE REPEATED 6 MONTHS AFTER SURGERY



HORMONAL DISTURBANCES

- DI - TRI PHASIC COURSE
10-15% WILL RESUME
PARTIAL PRODUCTION OF ADH WITHIN
3 YEARS
- HYPOTHYROIDISM: REPLACEMENT IN 60-80%
- CORTICO STEROIDS: TAPERED TO MAINTENANCE
DOSES AFTER 4-5 DAYS



- ESTROGEN, TESTOSTERONE REPLACEMENT IN PUBERTY
- FSH/LH DEFICIENCIES ARE PERMANENT
- GH THERAPY IS STARTED 6-9 MONTHS AFTER SURGERY.
- HYPOTHALMIC DYSFUNCTION SYNDROME



RECURRENCE

- EXTENT OF SURGICAL RESECTION THAN ON HPE
- > 4 CM OF TUMOR
- CYSTIC NATURE OF TUMOR



RECURRENT CRANIO

- REOPERATION

IF THE PATIENT IS A CHILD AND GOOD MEDICAL
CONDITION

RECEIVED RADIATION AND SOLID COMPONENTS
REGROWS

IN UNRADIATED CASES RECURRENT TUMOR
DENSILY FUSED WITH THE DURAL WALL OF THE
SELLA AND CAVERNUOUS SINUS.



- POST RADIATION CASES TUMOR FRAGMENTS ARE ADHERENT TO VESSEL WALL AND HYPOTHALAMOUS
- CYSTIC TUMOR: INTRACAVITARY BLEOMYCIN OR 90 γ
- INTERFERON α 2A – WHEN ALL CONVENTIONAL THERAPY FAILS



Craniopharyngiomas remain associated with significant morbidity. Gross total removal provide favourable result in terms of recurrences. If it can not be achieved safely, adjuvant radiotherapy is beneficial in preventing tumour re-growth. Childhood and adult onset lesions generally behave similarly.

[Raravitaki N, Brufani C, Warner JT et al Clin Endocrinol (Oxf), 2005 Apri; 62(4): 397-409]



Tumor recurrences/progressions are frequent and occur early after initial treatment of childhood craniopharyngioma. A radical resection preserving the integrity of hypothalamic structures appears optimal at original diagnosis. Irradiation was efficient in preventing recurrences/progressions. GH treatment has no impact on the low 3-years event free survival.

[Muller HL, Gebhardt U, Schroder s, Pohl F et al. Horm Res Paediatr. 2010; 73(3): 175-80.



CYSTIC CRANIOPHARYNGIOMA

TOTAL
EXCISION

SUBTOTAL
EXCISION

BIOPSY AND
ASPIRATION

STEREOTACTIC
ASPIRATION +

INTRA CAVITORY

- IRRADIATION
- BLEOMYCIN
- CHEMICAL AGENTS

RECURRENCE

RADIATION
RECURRENCE

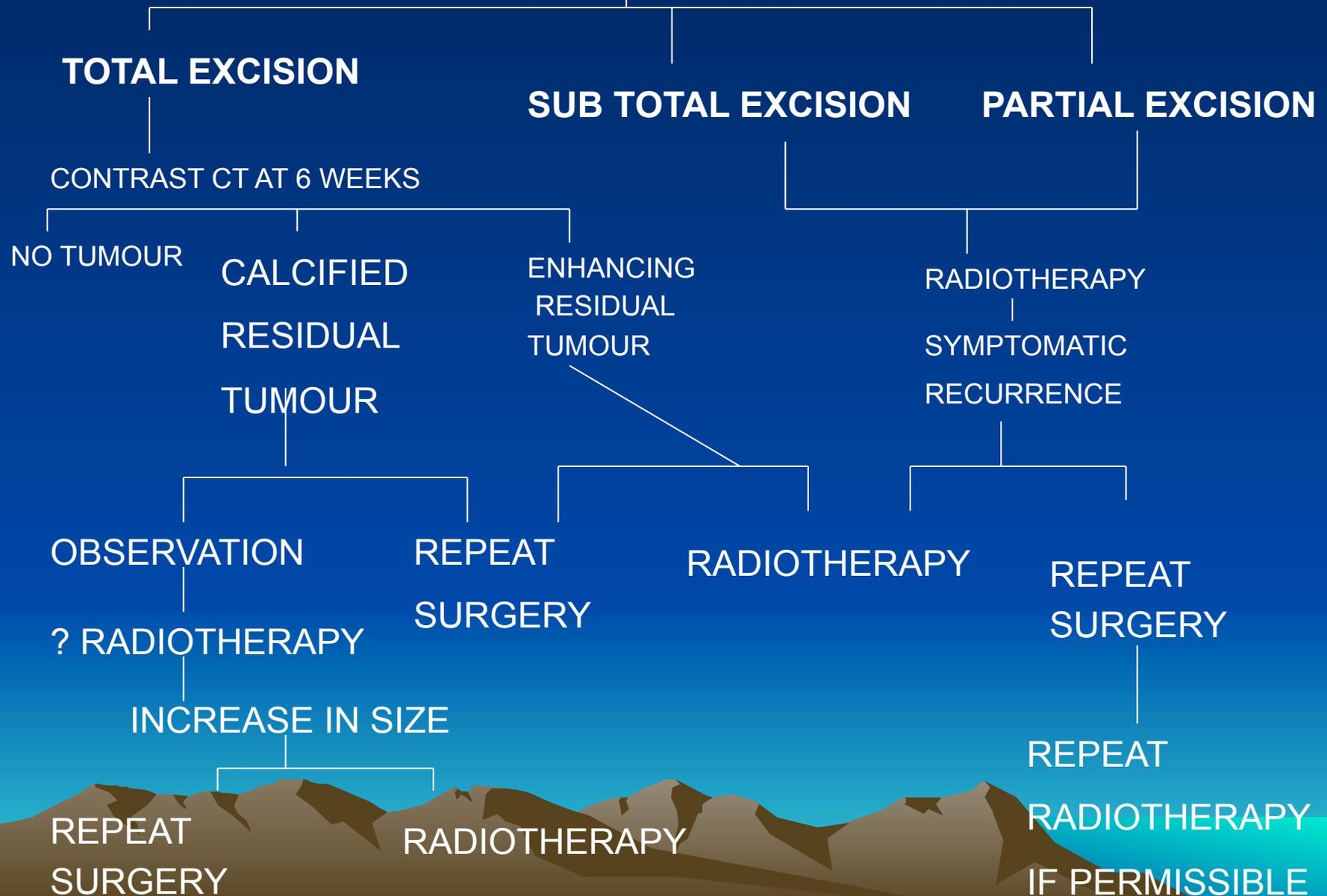
REPEAT
SURGERY

RADIATION

REPEAT
SURGERY

RESERVOIR
INSERTION

SOLID CRANIOPHARYNGIOMA



MIXED CRANIOPHARYNGIOMA

TOTAL EXCISION

SUBTOTAL EXCISION

CYST DRAINAGE+
PARTIAL EXCISION

CONTRAST CT AT 6 WEEKS

NO TUMOUR

CALCIFIED
RESIDUAL
TUMOUR

ENHANCING
RESIDUAL
TUMOUR

RADIODTHERAPY

SYMPTOMATIC
RECURRENCE

OBSERVATION

REPEAT SURGERY

RADIODTHERAPY

? RADIODTHERAPY

PREDOMINANTLY
CYSTIC

MIXED
SOLID

ASPIRATION
DRAINAGE

REPEAT SURGERY
? RADIODTHERAPY

GIANT CRANIOPHARYNGIOMA

EXCISION IN ONE OR MULTIPLE STAGES



AS MUCH EXCISION AS POSSIBLE



SUMMARY

- A POLICY OF ATTEMPTED TOTAL RESECTION WHERE POSSIBLE AND SUBTOTAL REMOVAL ALONG WITH ADJUVANT RADIATION IN CASES WHERE TOTAL RESECTION IS DEEMED UNSAFE IS RECOMMENDED AS A SOFTER AND MORE EFFECTIVE MODE OF THERAPY THAN AGGRESSIVE TOTAL RESECTION .
- SIMILAR APPROACH MAY BE USED IN DEALING WITH CP THAT RECCUR AFTER APPARENT TOTAL EXCISION



THANK YOU

