



MANAGEMENT STRATEGY OF CRANIOPHARYNGIOMAS



ELGOHARY MOHAMED ELGOHARY

PROF. OF NEUROSURGERY

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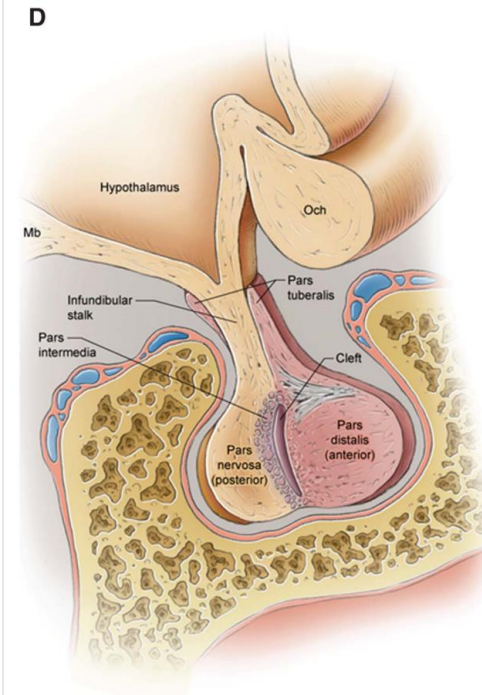
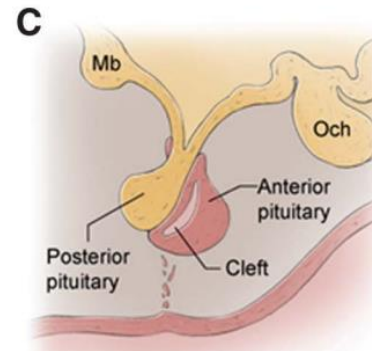
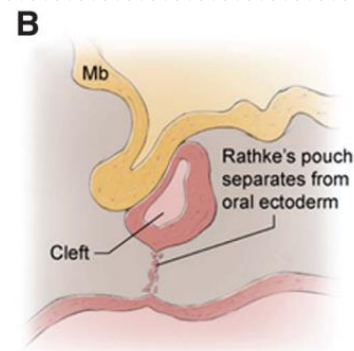
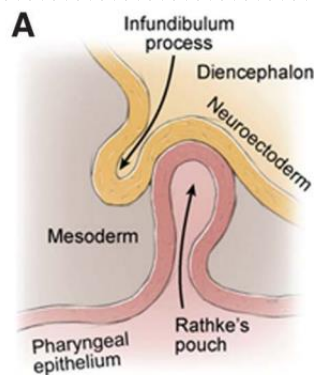
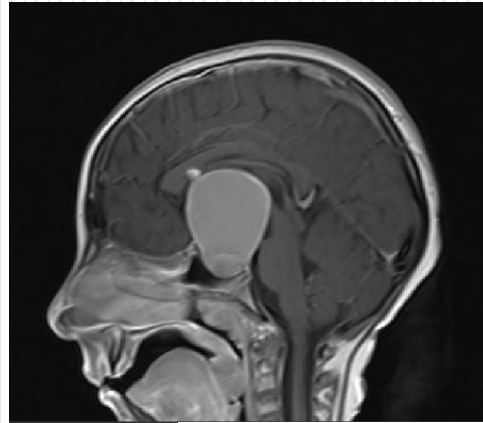
MEDICINE

CAIRO UNIVERSITY

EGYPT

CRANIOPHARYNGIOMAS

Are rare solid or mixed solid-cystic tumors that arise from remnants of Rathke's pouch along a line from the nasopharynx to the diencephalon



EPIDEMIOLOGY

Craniopharyngiomas comprise about **5 to 10 percent** of brain tumors in **children**

Craniopharyngiomas are approximately equally common in males and females

Histological types

craniopharyngiomas are divided into two categories, **adamantinomatous** craniopharyngiomas (Children) and **papillary** craniopharyngiomas (Adult)

these two histologic subtypes behave similarly with respect to resectability, sensitivity to radiation, and overall survival

Presentation

Growth failure, which can be caused by either hypothyroidism or growth hormone deficiency, is the most common presentation in children

DIAGNOSIS

A cystic calcified parasellar lesion is very likely to be a craniopharyngioma

Craniopharyngiomas must be distinguished from other tumors in the parasellar area, including :

- pituitary macroadenoma
- meningioma
- optic glioma
- germinoma
- teratoma
- lymphoma
- metastasis
- from nonneoplastic cysts (Rathke's, and arachnoid)
- and from infiltrative disorders such as sarcoidosis and systemic histiocytosis



Pretreatment evaluation

Endocrine testing, particularly of **adrenal** and **thyroid** function should be done to establish a presurgical baseline.

A detailed neuroophthalmologic examination including visual field testing helps to determine whether there is compression of the optic pathways.

History

- The first surgical techniques for craniopharyngioma originated in 1891 with the first trepanation done by Selke, then the first successful transcranial approach performed by Horsley in 1907.
- The first successful resection of craniopharyngioma through a transsphenoidal approach was done by Eiselsberg in 1910, which was improved by Halstead as a sublabial transsphenoidal resection in the same year.
- Harvey Cushing created the foundations of craniopharyngioma patient treatment with hormone replacement therapy.
- He also, in 1919, developed and performed the first successful resection through the trans-laminaterminalis approach of a retrochiasmatic craniopharyngioma.
- Later, in 1924, he performed a transcallosal resection of a craniopharyngioma.

TREATMENT

The optimal treatment of craniopharyngiomas has been controversial and included two basic approaches:

➤ **Aggressive surgery** with an attempt to achieve complete resection at diagnosis

versus

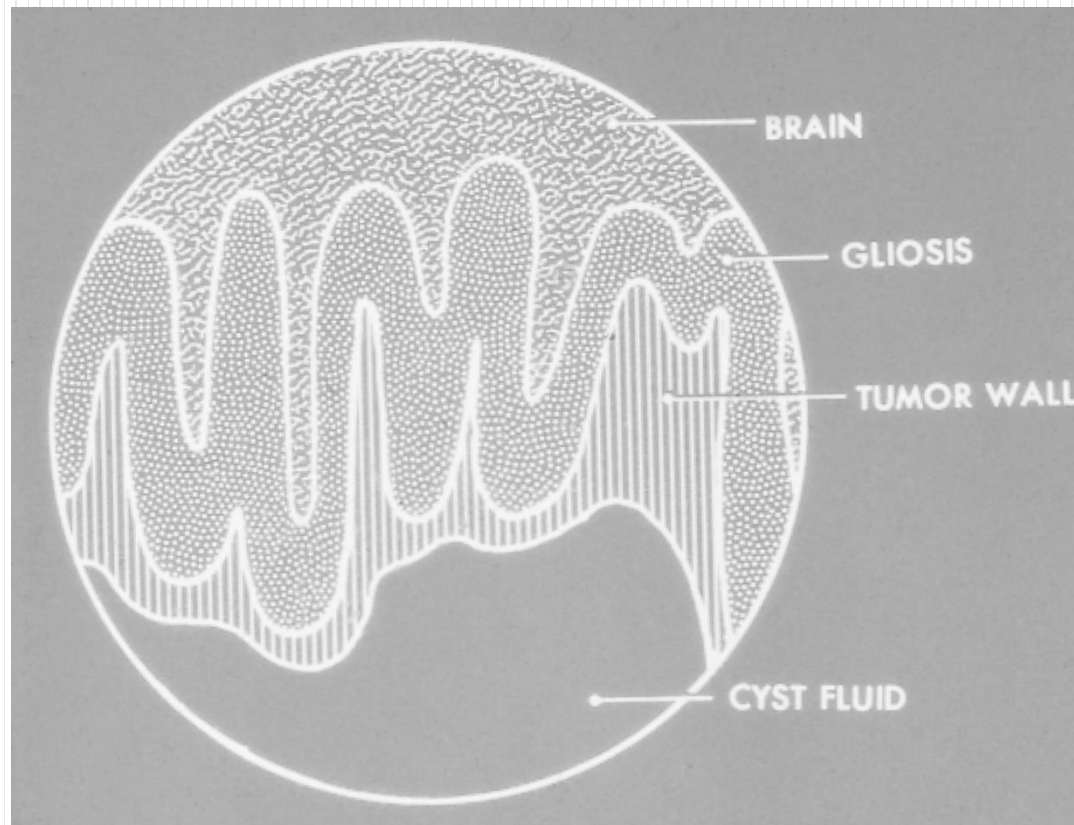
➤ **Conservative surgical** approach that used radiation therapy (RT) to treat residual disease

➤ Major advances in neurosurgical techniques have significantly decreased the morbidity and mortality associated with resection, making aggressive resection feasible in more cases.

➤ At the same time, improvements in RT techniques have permitted more accurate delivery of radiation to the tumor target, while minimizing radiation damage to normal structures.

Surgery

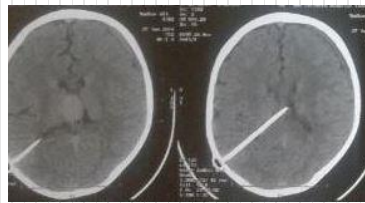
is indicated in almost **all cases**.



Preoperative management



- Endocrine function should be assessed and significant abnormalities corrected prior to surgery if possible.
- Peritumoral edema and increased intracranial pressure should be controlled.
- Hydrocephalus may require either a temporary or permanent shunt.



- Patients with a large cystic component to their tumor may require (aspiration) prior to surgery.

surgical approach

Many craniopharyngiomas are currently approached by a **pterional craniotomy**.

Far superior extension of a cyst into the third ventricle may require opening of the **lamina terminalis** or an approach from above, across the corpus callosum, through the **foramen of Monro**.

Intrasellar tumors may be removed **transsphenoidally**

An **extended transsphenoidal** approach, now commonly performed using the **endoscope**, can remove tumors with **suprasellar extension**

Goal of Surgery

Radical surgery, while capable of providing long-term tumour control, can in certain cases be responsible for an unacceptable degree of hypothalamic damage.

What has been learned from the experience in the literature that might help us in the future is to achieve the ultimate goal of decompressing the visual apparatus and preventing further tumour growth, while at the same time preserving not only hypothalamic function but also, if possible, pituitary function?

Radiation therapy(RT)

Radiation therapy can be used to treat patients with **residual disease** who have undergone a partial surgical resection.

Treat disease that has recurred following what **was initially thought to be a gross total resection**

RT techniques

Recent RT techniques (**Stereotactic techniques**) permit greater treatment precision. These approaches decrease long-term toxicity by limiting the exposure of surrounding normal tissues to ionizing radiation:

- Stereotactic radiotherapy (**SRT**)
- Stereotactic radiosurgery (**SRS**)

They both utilize a stable frame that establishes a patient specific coordinate system in order to localize the target volume within the fixation device coordinates.

Once the target is identified and the desired dose chosen, the treatment planning will then determine the field size, coordinates and relative weights to be delivered for coverage of the target.

➤ **SRS** uses a **single fraction** of radiation and is an alternative to fractionated radiotherapy for small tumors or small focal postoperative residuals

➤ **SRT** uses a **fractionated treatment** schedule to minimize damage to normal structures.

In both care must be taken to limit dose to the tolerance of the optic nerves, chiasm, and tracts.

Cyst management

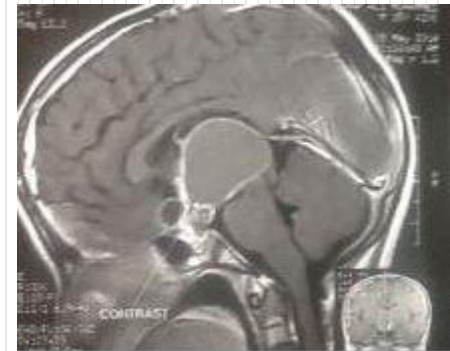
Techniques that decrease cyst size are generally indicated when a cyst compresses visual or hypothalamic structures or causes symptomatic obstruction of the third ventricle.

➤ Aspiration :

- **Percutaneous aspiration** of cyst contents has been used to alleviate symptoms, and intermittent aspiration may be recommended whenever total excision is not feasible.
- An alternative is placement of an **Ommaya reservoir** system.

➤ Intracavitary irradiation.

➤ Intracavitary chemotherapy using bleomycin.



Disease control

Complete surgical resection is the goal of initial treatment. However, the benefits of surgery must be balanced against treatment related morbidity.

RT is used as an adjuvant following subtotal resection, and this approach significantly reduces the risk of a local recurrence .

RECURRENT DISEASE

Most recurrences are local; **surgery**, whether used initially or not, may be indicated, as may be radiosurgery, if **enlarging focal solid disease rather than cyst expansion** is the problem

Review of reports from large series:

Stripp DC, Maity A, Janss AJ, Belasco JB, Tochner ZA, Goldwein JW, Moshang T, Rorke LB, Phillips PC, Sutton LN, Shu HK in 2004 reported:

75 patients with craniopharyngioma over a 27 year period.

All patients underwent an attempt at gross total excision.

Postoperative RT was given to 18 of the 27 in whom only a subtotal resection was possible.

At a median followup of 7.6 years, the 10 year local control rate was significantly better for the patients with a subtotal resection plus RT compared to the patients treated with surgery alone.

Karavitaki N, Brufani C, Warner JT, Adams CB, Richards P, Ansoorge O, Shine B, Turner HE, Wass JA 1n 2005 reported:

121 patients treated between 1963 and 2002.

19 underwent gross total removal, 84 partial resection, and nine cyst evacuation.

All of the patients with gross total resection were free of recurrence at 10 years.

Among those managed with partial resection, the 10year recurrence free rates were 77 and 38 percent, with and without postoperative RT respectively.

Combs SE, Thilmann C, Huber PE, Hoess A, Debus J, Schulz-Ertner D in 2007 reported:

40 patients treated with stereotactic RT (28 with recurrent disease, 12 as an adjuvant after surgery). At a median followup of 98 months, local control was 100 percent at ten years and overall survival was 89 percent.

Niranjan A, Kano H, Mathieu D, Kondziolka D, Flickinger JC, Lunsford LD in 2010 reported:

46 patients with craniopharyngioma underwent 51 courses of treatment with SRS for residual or recurrent craniopharyngioma (median tumor volume 1.0 cm³). At a mean followup of over five years, the five-year overall and progression free survival rates were 97 and 92 percent, respectively.

Treatment complications

➤ **Endocrine** — Endocrine abnormalities are due in part to the original tumor but can be exacerbated by treatment

Panhypopituitarism is present in the majority of cases and can be manifested by hypogonadism, hypothyroidism, adrenal insufficiency, and/or growth hormone deficiency

Hypothalamic dysfunction can cause disabling obesity, secondary type 2 diabetes, disorders of temperature regulation, sleep disorders, or diabetes insipidus.

➤ **Neurologic**

Impaired intellectual functioning

Hypothalamic obesity

Sleep disorders

Disrupted circadian rhythm

Behavioral problems

➤ **Visual**

Most patients have visual deficits prior to treatment. These may be exacerbated by either surgery or radiation therapy.

➤ **Vascular abnormalities**

A variety of vascular abnormalities can follow radiation of a craniopharyngioma in a child:

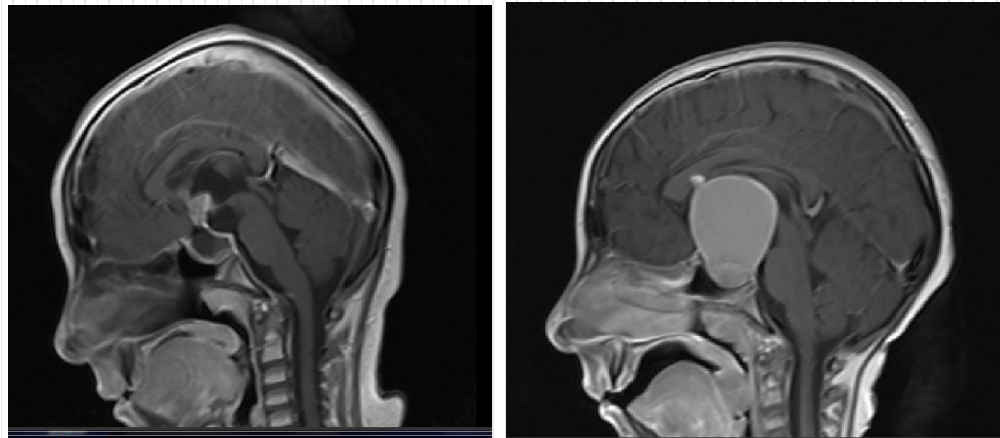
- temporal cavernomas
- moyamoya syndrome
- aneurysms
- and decreases in arterial caliber

➤ **Secondary malignancies**

The use of RT to treat craniopharyngiomas has been associated with the secondary development of meningioma and malignant glial tumors

POST TREATMENT FOLLOWUP

- Neuroimaging with MRI, on an annual basis. The duration of imaging follow-up depends upon the extent of initial surgery, the presence or absence of residual tumor, and symptomatology.
- Monitoring of endocrine function with replacement hormone therapy as needed.
- Formal assessment of visual function including visual field testing postoperatively and annually thereafter.



PROGNOSIS

The long-term prognosis following treatment is influenced both by the **ability to control tumor** and the **treatment of the related complications**.

Individuals with craniopharyngioma had a three- fivefold increase in expected mortality compared with the general population.

The major contributors to excess mortality are:

Uncontrolled diabetes insipidus

Pontine infarction

Panhypopituitarism

liver failure.

Cerebrovascular disease

Type 2 diabetes mellitus

Myocardial infarction

Severe infection

SUMMARY AND RECOMMENDATIONS

- Management of the patient with a craniopharyngioma involves a balance between **controlling the disease** and **minimizing the side effects** of treatment.
- Patients should be managed by a **multidisciplinary team**. The team should include neurosurgery, radiation therapy, neuro-oncology, endocrine, and ophthalmologic expertise.
- The initial management approach is **neurosurgical resection**, with the goal of removing as much of the tumor as possible, while avoiding severe treatment-induced deficits.
- For patients who are not able to undergo a complete resection of their tumor, **postoperative radiation therapy rather than observation** with salvage RT if a recurrence occurs is recommended