



Treatment modalities of Craniopharyngioma

Review of literature

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By

Mohammad Al-Sayed Ali Yusuf

M.B.B.Ch

Ain Shams University

Supervised by

Professor Dr. Mohammad Sayed Ismael

Professor of Neurosurgery

Faculty of Medicine - Ain Shams University

**Assistant Professor Dr. Mohammad Abdullah
Al-Wardany**

Assistant Professor of Neurosurgery

Faculty of Medicine - Ain Shams University

Dr. Ahmed Al-Sayed Abdelbar

Lecturer of Neurosurgery

Faculty of Medicine - Ain Shams University

Faculty of Medicine

Ain Shams University

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بِسْمِ اللَّهِ الرَّحْمَنِ الرَّحِيمِ

Dedication

*To the soul of my father
You are the one to whom I owe everything.
You encouraged me all the way long.
I wish you were here to see your dream come true.*

*To my mother,
The sun of my life,
The giver of endless love, deep faith and prayers,
Without your blessings, I'm nothing!*

*To my brothers and sister
Ahmad, Osama and Eman
You are my heroes.
Your words of inspiration and encouragement have been always
"the wind beneath my wings".*

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INDEX

Nm	Chapter	Pages
1	<i>Introduction and aim of the work</i>	1
2	<i>Embryology</i>	5
3	<i>Surgical anatomy of the sellar and suprasellar</i>	9
4	<i>Pathology of Craniopharyngioma</i>	55
5	<i>Diagnosis of Craniopharyngioma</i>	70
6	<i>Treatment modalities of Craniopharyngioma</i>	119
7	<i>Summary & Conclusion</i>	234
8	<i>References</i>	238
9	<i>Arabic Summary</i>	252

LIST OF FIGURES

Figure No.	Title	Page
2-1	<i>Embryogenesis of pituitary gland</i>	6
3-1	<i>Osseous relationships of the anterior and middle cranial base</i>	10
3-2	<i>Superior view of the osseous relations of the sphenoid bone</i>	11
3-3	<i>Posterior view of sphenoid bone</i>	13
3-4	<i>The bony part of the sella turcica and its surrounding bony structures</i>	14
3-5	<i>Lateral view of osseous relations of the sphenoid bone</i>	15
3-6	<i>Three types of sphenoid sinuses</i>	16
3-7	<i>multiseptated sphenoid sinus and the anterior wall of the sella</i>	18
3-8	<i>Coronal section in the sella and the cavernous sinus</i>	22
3-9	<i>superior view of the suprasellar region</i>	24
3-10	<i>Vascular relationships of the suprasellar</i>	29
3-11	<i>Circle of Willis</i>	31
3-12	<i>Six sagittal sections of the sellar region showing variations in the intercavernous venous connections within the dura</i>	34
3-13	<i>The optic nerves enclosed in the optic sheath within the optic canal</i>	36
3-14	<i>The optic nerve and chiasma and the surrounding vessels</i>	38
3-15	<i>Types of the optic chiasm</i>	39
3-16	<i>Lateral aspect of the brainstem and parasellar area showing the origin and course of cranial nerves</i>	42
3-17	<i>Anatomy of the third ventricle through midsagittal section</i>	45
3-18	<i>The floor of the third ventricle is located medial to the uncus and anterior perforated substance and above the</i>	47

Figure No.	Title	Page
	<i>midbrain</i>	
3-19	<i>Olfactory, carotid, chiasmatic, interpeduncular cistern</i>	49
3-20	<i>parasagittal section to the left of the midline to show Liliequist's membrane and the cisterns</i>	51
3-21	<i>Midsagittal section show the chiasmatic and lamina terminalis cistern</i>	52
4-1	<i>Sagittal gross pathology shows classic Adamantinomatous craniopharyngioma with mixed solid, cystic components</i>	58
4-2	<i>Low magnification of adamantinomatous craniopharyngiomas</i>	59
4-3	<i>Low magnification view of papillary craniopharyngioma</i>	60
4-4	<i>Histopathology of craniopharyngioma</i>	61
4-5	<i>Drawing represents the grading of craniopharyngiomas. The main focus is on the vertical tumor extension</i>	63
4-6	<i>Diagrammatic representation of the most common locations of craniopharyngiomas</i>	64
4-7	<i>Preoperative and postoperative magnetic resonance imaging (MRI) classification of pediatric craniopharyngiomas</i>	65
4-8	<i>Wnt signaling pathway</i>	67
4-9	<i>Blood supply of craniopharyngioma</i>	69
5-1	<i>Skull x-ray film: craniopharyngioma causing enlarged sella with sellar destruction and suprasellar flocculonodular calcification</i>	78
5-2	<i>Axial unenhanced and contrast-enhanced CT demonstrating an inhomogeneously enhancing soft-tissue mass in the suprasellar cistern extending into the third ventricle</i>	80
5-3	<i>Axial CT brain demonstrating a suprasellar lesion with coarse calcification and dilatation of the temporal horns of the lateral ventricles</i>	80
5-4	<i>Coronal enhanced CT. There is a heterogeneous sellar and suprasellar mass with large areas of calcification</i>	81

Figure No.	Title	Page
	<i>within the sella</i>	
5-5	<i>MRI showing normal pituitary gland, sella turcica, and parasellar region</i>	82
5-6	<i>Nonenhanced and contrast-enhanced coronal T-weighted MRIs demonstrating an intra-/suprasellar craniopharyngioma extending into the third ventricle with multiple calcifications</i>	83
5-7	<i>MRIs demonstrating a hypointense suprasellar tumor with peripherally enhancing cystic areas and an inhomogeneously enhancing solid tumor part</i>	84
5-8	<i>MRIs showing an intra-/suprasellar craniopharyngioma with a hyperintense cystic peripherally enhancing mass and a small solid inhomogeneously enhancing portion</i>	84
5-9	<i>MRI demonstrating measurement guidelines to predict visual impairment</i>	86
5-10	<i>Coronal T1-weighted Gd-enhanced MR images demonstrating typical Grade 1 and Grade 2 hypothalamic compression</i>	87
5-11	<i>Axial FLAIR image showing cysts more uniformly hyperintense</i>	88
5-12	<i>(A) normal visual field, (B) Example of visual fields in a patient with anterior chiasmal syndrome demonstrating extensive ipsilateral loss and a superotemporal field cut in the fellow eye, (C) Visual fields demonstrating bitemporal hemianopia</i>	102
5-13	<i>Fundus oculi examination showing moderate papilledema, atrophic papilla</i>	103
5-14	<i>MRI showing chiasmatic astrocytoma</i>	107
5-15	<i>Axial CT showing craniopharyngioma</i>	107
5-16	<i>MRI showing Large multicystic craniopharyngioma</i>	108
5-17	<i>MRI showing suprasellar germinoma with extension inferiorly along the pituitary stalk</i>	108
5-18	<i>MRI showing hypothalamic hamartoma</i>	109
5-19	<i>Coronal enhanced T1-weighted image of Langerhans' cell histiocytosis</i>	109
5-20	<i>Pituitary adenoma filling the sella and extending into the suprasellar space. (coronal T1-WI with contrast)</i>	110

Figure No.	Title	Page
5-21	<i>Rathke's cleft cyst (coronal T1-WI with contrast)</i>	110
5-22	<i>Imaging and histopathological examples of typical RCCs</i>	111
5-23	<i>Imaging and histopathological examples of a suprasellar epidermoid tumor</i>	114
5-24	<i>lymphoma of hypothalamus; post-contrast, T1-weighted image and DWI</i>	115
5-25	<i>Coronal enhanced T1-weighted image of a chondrosarcoma</i>	115
5-26	<i>Sagittal enhanced T1-weighted image of a suprasellar meningioma</i>	116
5-27	<i>Sagittal contrasted T1 MRI shows meningioma</i>	116
5-28	<i>Coronal T1WI MR shows a classic Rathke's cleft cysts that elevates and drapes the optic chiasm</i>	117
5-29	<i>Sagittal T1MRI of the suprasellar arachnoid cyst</i>	117
5-30	<i>Coronal T1WI MR shows a classic pituitary macroadenoma with "snowman" shaped suprasellar extension</i>	118
5-31	<i>Coronal T1 enhanced MR shows Microadenoma in a patient with galactorrhea</i>	118
6-1	<i>Surgical corridors that may be useful for the approach to a craniopharyngioma, depending on the size and pattern of the tumor and preference of the surgeon</i>	129
6-2	<i>Approaches to the anterior fossa and sellar region include the trans-sphenoidal, extended transsphenoidal, bifrontal, interhemispheric, pterional, and supraorbital</i>	130
6-3	<i>A: Intraoperative photograph of the bicoronal incision that extends behind the hairline. B: The vascularized pedicled pericranial flap that is elevated as a separate layer</i>	132
6-4	<i>Bifrontal and extended bifrontal approaches</i>	134
6-5	<i>A: Intraoperative photograph showing the bifrontal transbasal bone the inferior margin of the osteotomy is at the nasofrontal suture and extends laterally over both orbital rims. B: 3D reconstructed CT scan of the skull demonstrating the bifrontal transbasal bone flap</i>	135
6-6	<i>A: Intraoperative photograph after the bone flap has</i>	136

Figure No.	Title	Page
	<i>been removed. B: the bifrontal dural opening, cottonoid patties are used to protect the frontal lobes during ligation and division of the superior sagittal sinus and falx cerebri</i>	
6-7	<i>Intraoperative photographs of intradural exposure and tumor removal achieved using the transbasal subfrontal approach</i>	138
6-8	<i>The bifrontal interhemispheric approach</i>	139
6-9	<i>Right lateral dissection within carotico-optic space</i>	140
6-10	<i>Detaching the upper pole of the tumour from the hypothalamus</i>	142
6-11	<i>The subfrontal approach subdivided into several different routes</i>	145
6-12	<i>Lamina terminalis approach</i>	147
6-13	<i>The skin incision for a frontolateral craniotomy & the bone flap for a right frontolateral craniotomy</i>	151
6-14	<i>The supraorbital approach</i>	155
6-15	<i>Cadaveric dissection showing the steps of the supraorbital approach</i>	156
6-16	<i>The stepwise development of supraorbital approach, showing the reduction in size of the skin incision and craniotomy</i>	157
6-17	<i>Pterional approach</i>	160
6-18	<i>Intraoperative photograph showing the initial exposure during a left-side approach via the opticocarotid and carotid oculomotor triangles</i>	163
6-19	<i>Artist's illustration showing a schematic of a modified orbitozygomatic craniotomy</i>	165
6-20	<i>Orbitozygomatic approach</i>	168
6-21	<i>Axial view in a cadaveric dissection show the angles of exposure to the anterior and middle fossae gained through the orbitozygomatic craniotomy</i>	169
6-22	<i>Preoperative CT and MR images and sagittal and coronal T1-weighted post-Gd MR images of a large craniopharyngioma in a 4-year-old girl that was resected using a modified orbitozygomatic approach</i>	170
6-23	<i>Interhemispheric transventricular approach</i>	174

Figure No.	Title	Page
6-24	<i>The position of the patient and the skin incision for a right-sided petrosal approach</i>	175
6-25	<i>Initial exposure via a presigmoid route</i>	176
6-26	<i>Illustration of a retrochiasmatic craniopharyngioma exposed via the petrosal approach</i>	177
6-27	<i>The trans-sphenoidal approach used for craniopharyngiomas that are primarily intrasellar or when the suprasellar component is cystic</i>	180
6-28	<i>Examples of craniopharyngioma extension (and recommended approach)</i>	186
6-29	<i>The transventricular endoscopic approach</i>	189
6-30	<i>Neuronavigational image showing the catheter trajectory and the cystic craniopharyngioma</i>	190
6-31	<i>Types of craniopharyngiomas</i>	192
6-32	<i>A case of a type 1 (preinfundibular) craniopharyngioma</i>	196
6-33	<i>A case of type 2 (infundibular) craniopharyngioma</i>	198
6-34	<i>A case of a type 3 (retroinfundibular) craniopharyngioma</i>	200
6-35	<i>A, Leksell gamma knife, Model C. B, helmet and robotic device for migration through Cartesian coordinates in radiosurgical device</i>	218
6-36	<i>Photograph of CyberKnife</i>	219
6-37	<i>A, close-up view of CyberKnife robot arm and LINAC. B, close-up view of treatment couch and floor-mounted orthogonal amorphous silicon detectors. C, patient with fitted mask on treatment couch</i>	220
6-38	<i>Dose planning with the CyberKnife treatment planning software</i>	220
6-39	<i>CT/MR fusion planning images</i>	221
6-40	<i>MR images obtained before treatment of a cystic CP and the 4 years after the IFNα2A treatment</i>	227
6-41	<i>Treatment algorithm for craniopharyngiomas</i>	233

LIST OF TABLES

Table No.	<i>Title</i>	Page
5-1	Differential diagnosis of sellar mass	105
5-2	Sellar and suprasellar tumors	106
5-3	Differential diagnosis of craniopharyngioma in CT or MRI	112,113
6-1	Selection criteria used for radical surgery	128
6-2	Highest urinary osmolality after pitressin in water deprivation test	209

Introduction

"There is perhaps no other primary brain tumour that evokes passion, emotion, and as a result, controversy than does the craniopharyngioma" (*Gleeson et al, 2008*).

Craniopharyngiomas are slow-growing, benign, locally invasive intracranial tumours that can generate considerable morbidity and recurrences are often difficult to manage. As reliable morphologic criteria for accurately predicting the clinical outcome of these tumours is lacking, it has challenged the neurosurgeon and his skills for years. These tumours have a very intricate relation with the hypothalamus, pituitary stalk and optic apparatus, which have caused excessive problems regarding optimal management. Even now, no consensus exists in the optimal management of these patients (*Kato et al, 2008*).

One of the earliest descriptions of a craniopharyngioma is credited to *Zenker*, who in an 1857 autopsy study recognized a suprasellar lesion containing cholesterol crystals. Extensive study by *Luschka* of the squamous epithelial cells in the adenohypophysis followed in 1860. The significance of these findings was not initially recognized, and for many decades they remained overlooked (*Karavitaki et al., 2006*).

In 1892, *Onanoff* coined the term *pituitary adamantinoma* after appreciating the similarities between tumors of the jaw and tumors of the pituitary region. In 1899, pathologists *Mott* and *Barrett* began to investigate a group of epithelial-type tumors that occupied the sellar region.

They postulated that these tumors arose from either Rathke's pouch or the hypophyseal duct. In the next few years, these tumors were reported by both *Babinski* and *Frohlich* as suprasellar lesions without acromegaly (*Mehta and Black, 2004*).

In 1902, *Saxer* reported a tumor consisting of these cells. Two years later, *Erdheim*, after a systematic study of the squamous epithelial cells in the adenohypophysis, described them only in the glands of adult patients, usually on the anterior surface of the infundibulum and in groups or islets of variable size, shape, and number. Because a few of these groups of cells contained small cysts similar to some pituitary tumors unnamed at that time, he was convinced that both lesions had the same origin and called them *hypophyseal duct neoplasms*. Interestingly, he did not find any cell rests along the route of the regressed craniopharyngeal duct, a discrepancy explained by *von Mihalkovitcs'* theory that the developing adenohypophysis underwent a forward and upward rotation carrying with it the cranial insertion of the gland. Similar observations on clumps of cell rests were later published by *Duffy, Kiyono*, and *Carmichael*, but it wasn't until 1932 that squamous epithelial cells were also detected in the pituitary glands of childhood populations by *Susman* (*Karavitaki et al., 2006*). Different terminologies were used for these tumors (including hypophyseal duct or craniopharyngeal duct or Rathke's pouch tumors, interpeduncular or dysontogenetic or suprasellar or craniobuccal cysts, suprasellar epitheliomas and adamantinomas), until 1932, when the name "craniopharyngioma" was introduced by *Cushing*. Commenting on the new terminology, *Cushing* wrote:

"This admittedly somewhat cumbersome term has been employed for want of something more brief to include the kaleidoscopic tumors, solid and cystic, which take their origin from epithelial rests ascribable to an imperfect closure of the hypophyseal or craniopharyngeal duct" (*Karavitaki et al., 2006*).

This term is now well entrenched in the neurosurgical literature, although embryologically, these tumors are remnants of the primitive stomodeum and not the pharynx (*Mehta and Black, 2004*).

The first attempt for surgical removal of craniopharyngioma was credited to *Halstead* who performed transsphenoidal surgery for a patient with symptoms of a sella mass, in 1909 (*Karavitaki et al., 2006*).

The surgical philosophy regarding the treatment of craniopharyngiomas has vacillated significantly over the last 5 decades. Early operative series demonstrated an extremely high mortality rate of 40%, with only 15% of patients undergoing total removal. By the early 1960s, many felt that aggressive surgery should be abandoned in favor of cytoreduction combined with radiotherapy. In mid-1970s, with improvements in both postoperative endocrinologic care and overall surgical technique, there was renewed support for an aggressive surgical approach. Even today, controversy exists between those who advocate aggressive surgical resection and those who support a more conservative approach (*Mehta and Black, 2004*).