



Cairo University
Faculty of Medicine

***RECENT MODALITIES IN TREATMENT OF
CRANIOPHARYNGIOMA***

An Essay

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General Surgery

By

Walid Eissa El Halaby

MBBCh, Cairo University

Supervised by

Prof. Dr.

Faheem Aly El Bassiony

Professor of General Surgery, Cairo University

Prof. Dr.

Ahmed Zohdi Mostafa Zohdi

Professor of Neurosurgery, Cairo University

Dr.

Amr El Samman

Lecturer of Neurosurgery, Cairo University

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”قالوا سبحانك لا علم لنا إلا

ما علمتنا إنك أنت العليم الحكيم”

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ABSTRACT

There are two main management pathways with regards to the treatment of the tumour. The first involves attempted gross total resection of the tumour, the second approach is for more limited surgery, aimed at debulking the tumour to reduce the mass effect on the optic pathways and/or to re-establish the cerebrospinal fluid pathways, followed by external beam radiotherapy or stereotactic radiosurgery (gamma knife); which show very promising results.

The recent progress in high resolution imaging equipment (especially the MRI) showed the extent of the hypothalamic involvement which preoperatively guided the surgeon which patient is candidate for gross total removal and who is not.

The extended endonasal approach is a very promising minimally invasive approach with many advantages but needs an experienced hand as well as some complications need to be solved as CSF leak.

KEYWORDS

Craniopharyngioma, sellar/suprasellar tumours, brain tumours, endonasal approach, visual impairment.

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My mother,
my wife
And all my
family

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Abbreviations

A1	First part of anterior cerebral artery
ACA	anterior cerebral artery
ACTH	adreno corticotropic hormone
ACoA	Anterior communicating artery
ADH hormone	Antidiuretic hormone
CrN	Cranial nerve
CSF	Cerebrospinal fluid
CT	Computerized tomography
DI	Diabetes insipidus
GKS	Gamma knife radio surgery
ICA	Internal carotid artery
ICP	Intra cranial pressure
MRI	Magnetic resonance imaging
RT	Radiotherapy
SIADH	Syndrome of inappropriate ADH secretion
TSH	Thyroid stimulating hormone

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Introduction and aim of work

Craniopharyngiomas are benign, slowly growing tumours that are located within the sellar and supra-sellar region of the central nervous system. These tumours grow by expansion, although glial reaction and small papillary tumour projections into the glial under surface may falsely lead to the impression of tumour invasion. The point prevalence of this tumour is approximately 2/100000. Distribution by age is bimodal with the peak incidence in children at 5-14 years and in adults at 65-74 years of age. In children, Craniopharyngiomas account for 5% of all tumours and 50% of all sellar/supra-sellar tumours. There are two histological phenotypes seen in Craniopharyngiomas: i) the adamantinomatous tumours, seen in children and ii) the squamous papillary form, predominantly seen in adults. (*Haupt R et al, 2006*)

The onset of symptoms is normally insidious with most patients and there is often a delay of 1-2 years between symptoms onset and diagnosis. The usual symptoms on presentation are :i)raised intracranial pressure(headache, nausea and vomiting...etc.) either from mass effect from the tumour itself or from secondary hydrocephalus, ii)visual disturbance (classically bitemporal hemianopia but homonymous hemianopia, scotoma and post papilloedemic optic atrophy may occur, iii)endocrine dysfunction(normally suppressed endocrine function, e.g. hypothyroidism, short stature, Diabetes insipidus, impotence and amenorrhea, but exaggerated endocrine function can occur, e.g. precocious puberty in children and obesity in adults.)(*Karavitaki, 2006*)

The diagnosis of a patient with Craniopharyngiomas is based on clinical and radiological findings and is then confirmed by characteristic histopathological findings. The classical appearance of Craniopharyngiomas by imaging is a sellar/supra sellar, partly solid, partly cystic calcified lesion. Calcification is best delineated on CT. MRI with and without contrast more accurately delineate the extent of the tumour and, in particular, its involvement with the hypothalamus. It is the investigation of choice to plan the surgical approach. MRA is useful not only to delineate the course of the vessels, which can be through the tumour, but also to help differentiate a tumour from a possible vascular malformation. Histologically the tumour cells are small and have an epithelial appearance with numerous micro cystic spaces formed. (**Rossi A, 2006**).

There are two main management plans for the treatment of Craniopharyngiomas. The first involves attempted gross total resection of the tumour. The second is for more limited surgery, aimed at debulking the tumour to reduce the mass effect on the optic pathways and/or to reestablish the C.S.F. pathway, followed by radiotherapy. The second pathway was developed because of the high morbidity experienced with the gross total resection of tumours invading the hypothalamus. Occasionally, a patient presenting with a purely cystic tumour, the management options include stereotactic placement of a catheter to allow repeated aspiration, furthermore, the use of intracystic radiotherapy or chemotherapy has had some success. Residual tumours confirmed by post-operative MRI, is generally treated by beam radiotherapy, however stereotactic radio surgery has been used. (**Puget S, 2007**).

Craniopharyngioma could be approached via different routes according to its location and extension e.g. Sub frontal, pterional, interhemispheric and recently several successful attempts through trans-nasal.

Aim of work

Reviewing the literature and recent publications concerning pathology, clinical picture, diagnosis, and management of Craniopharyngiomas.

Chapter 1

Embryology

❖ **Origin of craniopharyngioma.**

❖ **Nomenclature of craniopharyngioma.**

ORIGIN OF CRANIOPHARYNGIOMAS

Craniopharyngioma is a slowly growing, extra-axial, squamous, calcified, partly solid, partly cystic tumor. It grows at the site of the development of the craniopharyngeal duct i.e., in the pharynx, sphenoid sinus, intrasellar and suprasellar regions. But mostly in the sellar suprasellar region. Two main hypotheses, that complement each other, explain the origin of craniopharyngioma. (*Bobustuc et al., 2002*).

The embryogenic theory

Craniopharyngioma was thought for long time to arise only from epithelial rests along the embryological migration of the anterior pituitary lobe (Rathke's pouch). (*GlodBerg & Eshbaugh, 1960*).

In 1899, Mott and Barrett postulated that Craniopharyngiomas might arise from the hypophyseal duct or Rathke's pouch. (*Mott & Barrett, 1899*).

In 1904, Erdheim described the histologic criteria of these tumors and pointed to the similarities with adamantinomas. He and others thought that this resemblance proved that craniopharyngiomas arise from ectoblastic remnants of Rathke's duct. In addition that Rathke's pouch shares in the formation of the anterior lobe of pituitary, the most common site for craniopharyngiomas. (*GoldBerg & Eshbaugh, 1960*).

The metaplastic theory

Some investigators have found histologic differences between adult and childhood craniopharyngioma. They found that half of the adult craniopharyngioma are formed of squamous epithelium in a papillary form without adamantinomatous criteria.

These squamous epithelial rests are found in 3% only of neonates with increasing frequency with age. This theory suggests that squamous epithelial rests may undergo metaplasia causing the development of craniopharyngioma. (*carmicheal 1931, luse &kernohan 1955 and Carmel, 1996*).

The dual theory

Explains the craniopharyngioma spectrum attributing the adamantinomatous type (most prevalent in childhood) to embryonic remnant, and the adult type (squamous papillary) to metaplastic foci derived from mature cells of the anterior hypophysis (prevalence of the adult type increases with each decade of life and is almost never found in children). (*Bobustuc et al., 2002*).

NOMENCLATURE

The term craniopharyngioma was widely popularized by McKenzie and Sosman (1924), McLean (1930), and by Cushing (1934).

Cushing wrote, commenting on the new terminology, "This admittedly somewhat cumbersome term has been employed for need of something briefer to include the kaleidoscopic tumors, solid and cystic, which take origin from epithelial rests ascribable to the partial closure of the hypophyseal or craniopharyngeal duct. From a developmental stand point, they are variously called hypophyseal duct tumors, cranipharyngeal duct tumors and Rathke's pouch tumors; from anatomical basis they are called interpeduncular tumors or suprasellar tumors and from a histopathological basis, adamantinomas which is still unfortunate".

This term was considered unsatisfactory by Russel and Rubenstien (1977), as some are infra as well as suprasellar. They stated that the term craniopharyngioma is convenient but inaccurate because Rathke's pouch is an invagination from the primitive stomodeum and not from the pharynx, the objection is academic and the name is widely used and is likely to persist.