MANAGEMENT OF CRANIOPHARYNGIOMAS

Thesis

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Master Degree in General Surgery

By

Khaled M. Abd Al-Aziz

M.B., B.Ch.

Supervisors

Prof. Dr. Hasanein Alsharif

Professor of Neurosurgery
Faculty of Medicine
Ain Shams University

Prof. Dr. Adel Hussein Al-Hakim

Professor of Neurosurgery
Faculty of Medicine
Ain Shams University

Dr. Magdy M. El-Kalliny

Assist. Prof. of Neurosurgery
Faculty of Medicine
Ain Shams University

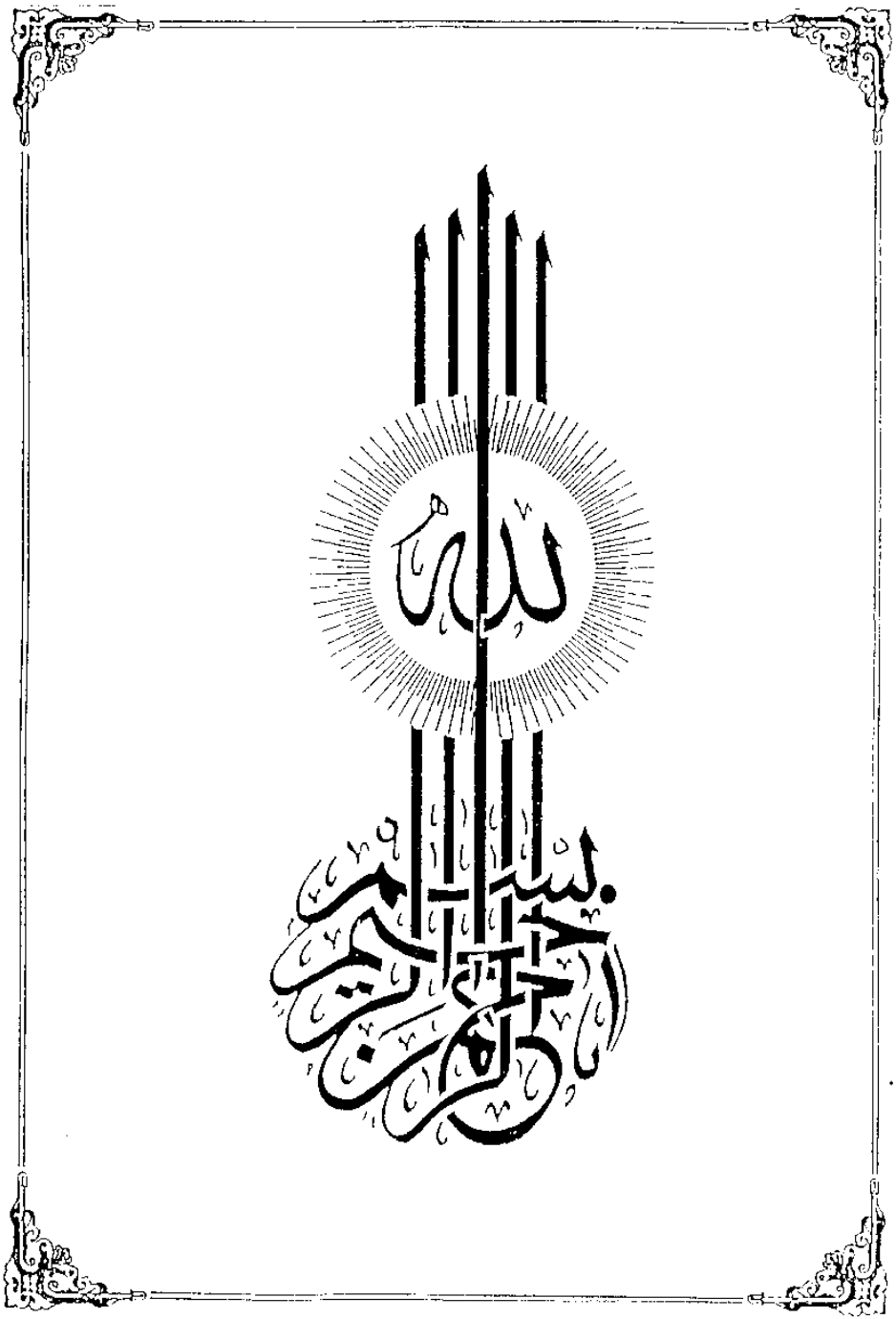
Faculty of Medicine
Ain Shams University
1993

بسم الله الرحمن الرحيم

سُبْحَانكَ لاَ علْمَ لَنَا إلا ما عَلمْتَنَا
إنَّكَ أنتَ العَليمُ الحكيمُ

(سورة البقرة : آية ٣٢)

دار الصداقة



To
My Parents

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INTRODUCTION AND AIM OF THE WORK

INTRODUCTION AND AIM OF THE WORK

Craniopharyngiomas, which are primarily tumors of childhood, are the most common intracranial tumor of nonglial origin in children. Their treatment has always presented a challenge to the neurosurgeon.

Although they are histologically benign, their poximity to the hypothalamus, optic nerves, and internal carotid arteries and their key branches poses an extremely difficult problem even for the experienced and determined neurosurgeon.

Different strategies exist for the primary treatment of craniopharyngiomas.

Proponents of radical removal, conservative surgery, radiation therapy, or a combination of these modalities vigorously espouse their points of view.

The problem is compounded by several factors. First, the growth characteristics of craniopharyngiomas vary considerably. Some patients may lead virtually symptom free lives despite untreated tumor, while others will have tumor that

grow in an uncontrolled manner despite radical surgery and radiation therapy. Second, there may be a difference in behavior of these tumors in the pediatric and adult populations. Finally it is difficult to assess earlier surgical series before the routine use of the operating microscope; it is likely that the morbidity of surgery for this tumor will be much lower with careful microsurgical techniques, combined with advances in preoperative and postoperative endocrine management.

Aim of the work:

The aim of our work is to review embryology, regional anatomy, surgical pathology, clinical picture and investigations for craniopharyngiomas. We also discuss preoperative assessment, evaluate different ways of management, and postoperative complications.

REVIEW OF LITERATURE

Incidence

Craniopharyngiomas are relatively rare tumors, constitute between 1.2-3% of all intracranial tumors (*Zulch K, 1986*) with 0.5-2 new cases/million population occuring each year (*Sorva and Heisknen, 1986*).

They are much more common among children, forming 9% of Matson's series of childhood brain tumors, and making up 54% of neoplasms in the sella-chiasmal region in children (*Matson DD, 1969*). However, one-half of the total cases are found in adults (*Sorva and Heisknen, 1986; Zulch K, 1986*).

There is bimodal age distribution with the first peak at 5-10 years (*Carmel PW, 1985*) and second peak between 55-65 years (*Banna M. et al., 1973*), but the tumour may be symptomatic at any age.

Sex distribtuion

Craniopharyngiomas might show male predominance (*Carmel PW, 1982*). However, some series showed equal sex distribution (*HJ Hoffman, 1977; Dachling Pang, 1993*).

EMBRYOLOGY

The term "craniopharyngioma" was first used in *1931* by *Frazier and Alpers and by Cushing in 1932.*

The first detailed autopsy acount of such tumor was gived in *1857 by Zenker,* further histological evidence for the tumor was given in *1860 by Luscka,* and in *1904 Erdheim* carefully detailed the histological features of craniopharyngiomas and postulated that they roginated from embryonic squamous cell rests of an incompletely involuted hypophyseal-pharyngeal duct.

Embryologic origin: *(Dachling Pang, 1993)*

For several decades, the origin of craniopharyngioma was thought to be related to the embryogenesis of anterior pituitary lobe.

At the end of third gestational week, the stomodeal ectoderm invaginates toward the diencephalon and eventually meets the downwardly projecting infundibular bud. As the sphenoid bone forms ventral of this complex, the stomodeal cleft is pinched off from the pharyngeal epithelium and the cleft becomes a pouch (of Rathke) whose wall later thickens to form the various parts of the anterior pituitary lobe. Ectoblastic cell rests have been found in the pars distalis and tuberalis, and

along the dorsal migration path of the stomodeal cleft, known as the hypophyseopharyngeal duct (craniopharyngeal duct). The frequent occurrence of craniopharyngiomas around the infundibular stalk, their occasional presence along the ventral hypophyseopharyngeal duct (e.g. within the sphenoid bone), and the striking histologic similarities between some craniopharyngiomas and tumors of known ectoblastic origin, such as adamantinomas, led *Erdheim 1904* and others *(Tiberin, 1958; Goldberg, 1960 and Partuiset, 1975),* to propose that craniopharyngiomas are all drived from ectoblastic reminants.

During *1950s,* the embryonic origin of craniopharyngioma was challenged when it was discovered that the pituitary squamous cell rests were rarely present in children under 10 years, but were found with increasing frequency in each succeeding decade, even though the peak incidence of craniopharyngiomas are from age 5-10 years *(Luse SA et al., 1955).* It was postulated that these squamous cell rests, from which craniopharyngiomas originate are products of metaplasia of the mature cells of the anterior pituitary, and not embryonic remnants.

There is evidence to suggest that craniopharyngiomas may indeed have dual origins. The so-called childhood type, which