Efficacy of Endoscopic Surgery Management of Craniopharyngiomas in Terms of Tumor Control and Post-operative Morbidity

Systematic Review Submitted for Partial Fulfillment of Master's degree in Neurosurgery

By

Shady Kamal Kamel Beniamen

M.B.B. Ch; Faculty of medicine, Cairo University

Neurosurgery resident at Dar Elshfa hospital

Under supervision of

Dr. Khaled Mohamed El bahy

Professor of Neurosurgery

Department of Neurosurgery, Ain Shams University

Dr. Mohamed Samir Mohamed Kabil

Professor of Neurosurgery

Department of Neurosurgery, Ain Shams University.

Dr. Abdelrahman Hesham Elgayar

Lecturer of Neurosurgery

Department of Neurosurgery, Ain Shams University.

Faculty of Medicine

AIN SHAMS UNIVERSITY

-2020-

Acknowledgment

First of all, I would like to express my deep gratitude and thanks to ALLAH, the most merciful.

I had great honor to work under the supervision of ***Prof/ Khaled El bahy*****,** professor of Neurosurgery, Ain Shams University. I would like to express my profound gratitude and sincere appreciation for his kind supervision, continuous encouragement and unlimited support in every step throughout this work.

I would like to express my deepest thanks and sincere appreciation to ***Prof/ Mohamed Kabil***, Professor of Neurosurgery, Ain Shams University for his continuous support, great encouragement, great help, his patience and valuable advice in every step throughout this work.

I would like also to thank ***Dr. / Abdelrahman Elgayar,*** Lecturer of Neurosurgery, Ain Shams University for his valuable advice and encouragement throughout this work.

Last but not the least; I would like to express all my feelings of love and appreciation to all my senior staff and my colleagues in Neurosurgery department for their lovely help throughout this work.

Shady Kamal

List of Figures

List of Tables

List of Abbreviations

Abbreviation	Meaning
BMI	Body mass index.
CI	Confidence interval.
CSF	Cerebrospinal fluid
CT	Computed Tomography
EVD	External ventricular drain
FESS	functional endoscopic sinus surgery
FIESTA	fast imaging employing steady-state acquisition
GCS	Glasgow Coma Scale
ICH	Intracerebral hemorrhage
IFE,	Immunofixation electrophoresis.
IHC	Immunohistochemistry
IIH	idiopathic intracranial hypertension
IVH	Intraventricular hemorrhage
LTR	Lateral pterygoid recess.
MD	Mean difference
MMP	matrix metalloproteinase
MPR	multiplanar reformation
MPRL	Macroprolactinoma
MRI	magnetic resonance imaging
NFA	nonfunctioning pituitary adenoma
PAR1	protease-activated receptor 1.
RR	Risk Ratio
TRF	Transferring
TSA	transsphenoidal adenectomy
VP	ventriculoperitoneal shunt.
EFS	event-free survival
QOL	quality of life.
CBCL	Child Behavior Checklist
ESS	Epworth Sleepiness Scale
FMH	Fertigkeitenskala Münster-Heidelberg Scale of daily living
HRQoL	Health-related quality of life
PedsQL	Pediatric Quality of Life Inventory
YSR	Youth Self Report

Table of Contents

Introduction

Introduction

Craniopharyngiomas are histologically benign tumors of embryonic epithelial origin, believed to arise from ectodermally derived epithelial cell remnants of Rathke's pouch and the craniopharyngeal duct. Epidemiologically, they have a bimodal age distribution pattern with a peak between 5 and 14 years in children and 50 and 74 years in adults, although the tumor has been reported in all age groups. They account for 2%–5% of primary intracranial neoplasms overall and 6%–13% of intracranial tumors in children. These tumors, while rare, occur much more commonly in the pediatric population than in the adult population, with an annual incidence of 5.25 cases per million. The population-adjusted incidence rates suggest that craniopharyngioma has a slightly higher incidence in black persons. There is an even distribution between genders **(Karavitaki, 2014).**

These tumors arise within the sellar and suprasellar regions, where they directly involve critical structures, including the pituitary gland, optic chiasm and nerves, third ventricle, and hypothalamic nuclei, as well as multiple major intracranial vessels traversing this region. Despite its benign histological aspect it can be considered a great challenge for the neurosurgeon and can be associated with a poor prognosis for the patient, even though significant improvements in the therapeutic management of these lesions have been developed. Treatment should focus on the relief of symptoms, avoidance of treatment-related morbidity, preservation of quality of life, and prevention of recurrence with extension of survival. Increased

recurrence rates usually necessitate multimodality treatments (surgery, radiotherapy, stereotactic radiosurgery, intracystic irradiation, local/intracystic chemotherapy, or systemic chemotherapy) **(Larkin and Ansorge, 2013).**

Ideally, radical resection is the therapy of choice at any age for primary craniopharyngioma. However, the location and significant size of these tumors, together with the involvement of critical neurovascular structures and frequent associated calcific components, limit the extent of a resection in many cases. Moreover, surgery can provoke visual impairment, endocrinological disturbances such as diabetes insipidus (DI), and other hypothalamic disturbances, important adverse events that make a radical surgical approach difficult to recommend, because the future life of these patients may be seriously impaired by social and behavioural disturbances. Several authors have reported high recurrence rates despite apparently complete tumor resection and negative postoperative brain imaging **(Singh et al., 2013)**.

The surgical routes used to remove craniopharyngiomas have previously been transcranial approaches, whereas the transsphenoidal approach, in accordance with the indications defined by Guiot and Derome in the early 1960s, was proposed initially for infradiaphragmatic lesions, with an enlarged sella, preferably in patients who had already developed panhypopituitarism. Traditional transcranial approaches require some degree of brain retraction and manipulation of cerebrovascular structures that lay between the surgeon and the pathology. The transsphenoidal approach offers a more direct route to the sellar/suprasellar region and if performed with the

operating microscope provides only a limited field of view, which often precludes safe and complete resection of these lesions. The use of the transsphenoidal corridor was broadened with the development of expanded approaches, which offered a safe alternative method for reaching suprasellar craniopharyngioma. The incorporation of the endoscope was the next step on the evolutionary ladder. Attempting to remedy these issues, in 1979 Halves and Bushe recognized the facilitatory role of the endoscope when accessing suprasellar lesions (**Cappabianca et al., 2004**).

By virtue of an improved field of view and superb illumination, this allowed for further expansion of the endonasal corridor. Finally, the development of a variety of complementary multilayer techniques to repair the skull base minimized the risk of postoperative cerebrospinal fluid (CSF) leak. Several groups have now published their series incorporating these innovations into a fully endoscopic, endonasal, extended transsphenoidal approach for sellar and suprasellar craniopharyngioma(**Hofstetter et al., 2011**).

The introduction of the endoscopic technique confirmed the effectiveness of the transsphenoidal route for the treatment of craniopharyngiomas, providing further advantages in terms of visualization. The widespread use of the endoscope in sinus surgery was brought to transsphenoidal surgery for the treatment of pituitary tumors. The wider panoramic view offered by the endoscope increased the versatility of the transsphenoidal approach and permitted it to be expanded to different parts of the skull base, allowing the removal of different “pure” supradiaphragmatic lesions. Because craniopharyngiomas often are

infrachiasmatic midline tumors, the endonasal route provides the advantage of accessing the tumor immediately after suprasellar dural opening, without brain or optic nerve retraction and with direct visualization through a straight surgical route(**Komotar et al., 2012**).

Aim of the Work

The purpose of this study is to evaluate the efficacy of a minimally invasive/endoscopic approach to Craniopharyngiomas given that the surgical aim was a complete excision of the tumor as a single stage procedure and to analyze outcome.

We also discuss the surgical success of the endonasal approach, compare its results with those for open approaches, and provide a review of the literature. We highlight the feasibility of this technique and evaluate its advantages and limits compared with the transcranial approaches.

The objective of the study is to systematically review the literature published discussing the endoscopic management of craniopharyngioma as regard tumor control, symptomatic relief and post-operative morbidity.

Chapter I: Anatomy and Embryology

A- Definition and Embryogenesis:

Craniopharyngioma has been defined by the World Health Organization (WHO) as "a benign, partially cystic epithelial tumor of the sellar region, presumably derived from Rathke pouch epithelium". Despite a WHO Grade I designation, the tendency of craniopharyngiomas (especially the adamantinomatous type) to densely adhere to and invade surrounding structures such as the infundibular stalk, hypothalamus, and basal vessels creates significant clinical challenges (**Perondi et al., 2013**).

The adamantinomatous craniopharyngioma accounts for about 85-90% of all craniopharyngiomas, is most common in childhood, and has bimodal peaks of incidence between 5-10 years of age and 50-60 years. Histological and molecular genetic similarities between the adamantinomatous craniopharyngioma and tumors of the jaw, such as ameloblastoma and calcifying odontogenic cyst, including expression of enamel proteins by this variant of craniopharyngioma, suggests an origin from odontogenic epithelium. In contrast, papillary craniopharyngiomas make up only 11-14% of tumors and occur almost exclusively in adults. The papillary variant shows histologic features reminiscent of Rathke cleft epithelium. Although rare, congenital/neonatal craniopharyngiomas comprise about 7% of all congenital brain tumors **(Lazaridis et al., 2010).**

Literature Review

Adamantinomatous craniopharyngiomas are believed to arise from neoplastic transformation of embryonic rests that occur during development and involution of Rathke's pouch. During development, the adenohypophysis is formed by an out pouching of ectodermal epithelium that forms the roof of the stomadeum ("Rathke's diverticulum").This epithelium comes into contact with a downward evagination of neuroepithelium from the infundibular region of the third ventricular floor. The latter neuroepithelium gives rise to the neurohypophysis and infundibular stalk.

By the fifth gestational week in humans, Rathke's pouch has elongated and becomes constricted at the oral epithelium, forming a pharyngo-hypophyseal stalk/duct. This Rathke's pouch remnant becomes separated from the oral epithelium during weeks 6-8. Involution of Rathke's cleft occurs during the 7th week of gestation, with the pharyngo-hypophyseal duct persisting in about 33% of cases and extending from the floor of the sella to the vomer. Remnants of this duct may give rise to craniopharyngiomas. Also, the location of pharyngo-hypophyseal duct remnants helps to explain the suprasellar location of most craniopharyngiomas. While an "embryogenetic" theory currently is used to explain the origin of adamantinomatous craniopharyngiomas, a "metaplastic" theory of tumorigenesis has been suggested for the papillary variant. In the latter, the differentiated squamous epithelium found in papillary craniopharyngiomas is derived from metaplastic transformation of epithelium that forms part of the anterior pituitary gland or pituitary stalk.