Transcranial Approach in Craniopharyngioma Surgery: Results from Tertiary Care Center in Nepal

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Abstract

Introduction: Management of craniopharyngiomas poses a significant challenge in neurosurgery, because of their involvement with the hypothalamus and visual systems. Complete resection attempts a higher mortality while incomplete surgery has high recurrence rates.

Materials and Methods: This is a prospective study from the period of January 2016 to April 2024 at the National Neurosurgical Referral Center (NNRC), National Academy of Medical Sciences (NAMS) Bir Hospital. Inclusion criteria consists of all the histopathological proven cases of craniopharyngioma who underwent transcranial surgery during the study period.The analysis encompassed clinical, ophthalmological, imaging, endocrinological, neuropsychological, and surgical complication data from medical records.

Results: A total of 52 craniopharyngioma cases were operated during the study period. It had a classical bimodal age distribution, with an increased incidence rate in 5 to 14 years and 50 to 74 years of age. Predominantly affecting young females, the common manifestations were visual problems ,hydrocephalus, features of raised intracranial pressure(ICP) and endocrinopathy. Most of the tumors were larger than 4 cm size with Samii and Tatagiba Grade IV.The orbitozygomatic approach was used in all cases. Gross-total resection was achieved in 80% of patients, near-total in 12%, and partial in 8%. Postoperative complications included Diabetes Insipidus and hormonal insufficiency. Most cases involved adamantinomatous grade 1 type craniopharyngioma, with a mortality rate of 6%.

Conclusion: Craniopharyngioma patients may remain asymptomatic until they acquire very large size and can present with features of raised intracranial pressure(ICP), visual disturbances and endocrinopathy. While there are multimodal treatment options, surgery stands as the cornerstone in managing this tumor.

Keywords: Craniopharyngioma Sellar Tumor Adamantinomas Myxopapillary

Introduction

Craniopharyngiomas are rare brain tumors with a global incidence of 0.5–2 cases per million annually and account for 1–3% of all brain tumors. They present most frequently in children aged 5–14 and adults aged 50–70. While specific data for Nepal is limited, the incidence is likely consistent with global trends. These tumors originate from embryonic remnants of Rathke's pouch and are often associated with CTNNB1 or BRAF gene mutations.[1]

Clinically, craniopharyngiomas commonly present with headaches, visual disturbances and hormonal deficiencies due to their proximity to the hypothalamus and pituitary gland. Craniopharyngioma patients also present with cognitive and psychological issues, such as memory problems and depression. They are usually diagnosed on neuroimaging (MRI or CT) and histopathological evaluation.[2]

Management involves surgical resection, often supplemented by radiotherapy to reduce recurrence risks. However, complete removal can be challenging due to the tumor's proximity to critical structures. Long-term follow-up is essential, as recurrences occur in approximately 25–30% of cases. Patients may require lifelong hormonal replacement therapy and management of metabolic, neurological, and psychological complications.3Prognosis is generally good, with a 5-year survival rate of 80–90%. However,

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quality of life may be impacted by chronic issues, necessitating multidisciplinary care. The purpose of our study was to evaluate the demographics, clinical presentation, radiological findings, extent of surgery, histological findings, overall outcome and recurrence rates over the past 8 years at our institution.

Materials and Methods:

This is a prospective study of craniopharyngiomas, carried out at National Neurosurgical Referral Center (NNRC), National Academy of Medical Sciences (NAMS) Bir hospital. Institutional review board (IRB) approval was taken from the hospital for the study. Consent was taken from the patients if they were able to communicate and from the next of kin if they were not able to give consent. All histopathological proven craniopharyngiomas in this department were included in this study. The duration of the study was from January 2016 to April 2024 (for 100 months). Age, sex, clinical presentations and neurological manifestations were noted. Outcome measured on the basis of extent of tumor excision, histopathological types, recurrence and Glasgow Outcome Scale (GOS) at 6 months.

Results:

A total of 52 craniopharyngioma cases were operated during the study period. It had a classical bimodal age distribution, with an increased incidence rate in 5 to 14 years (58%)and 50 to 74 years of age (Table 1). Predominantly affecting young females(52%) (Table 2). The common manifestations were features of raised intracranial pressure in almost half of the cases followed by visual disturbances and disturbance of hypothalamic-pituitary axis.(Table 3). Decreased visual acuity and visual field defects were common ophthalmological findings but only papilledema shows a statistical significant improvement post operatively(P=0.031)(Table 4).

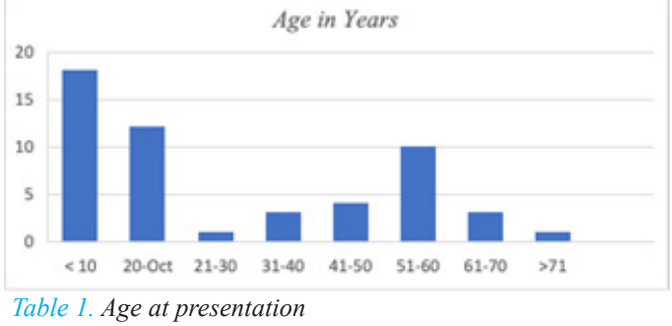

Table 1. Age at presentation

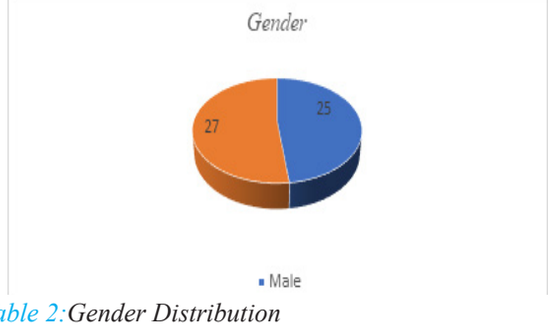

Table 2:Gender Distribution

Table 3: Clinical Presentation

Clinical Presentation	Number of Patients	
Raised ICP	26(50%)	
Visual Disturbances	Decreased visual acuity	14(27%)
	Blindness	5(10%)
	Visual Field Cut	12(23%)
	Optic atrophy	8(15%)
	Pappiledema	5(10%)
	Oculomotor Palsies	5(10%)
	RAPD	1(2%)
Disturbances of Hypothalamic-Pituitary Axis	9(18%)	

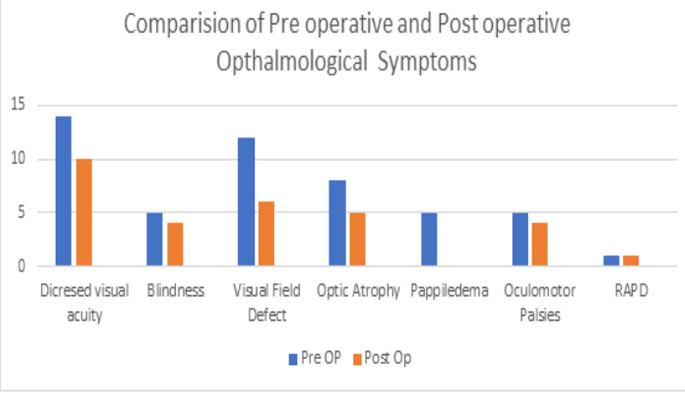

Table 4: Ophthalmological Symptoms

Group	Condition	Count(%)
Adults	Diabetes Insipidus(DI)	4(14%)
	Thyroid Insufficiency	3(12%)
	High Prolactin	26%)
	Adrenal Insufficiency	1(4%)
	Gonadal Insufficiency	1(4%)
Children	Obesity	6(23%)
	Anorexia	4(15%)
	Delayed/Precocious Puberty	4(15%)
	Diabetes Insipidus	1(6%)
	Thyroid Insufficiency	2(4%)
	High Prolactin	1(2%)
	Adrenal Insufficiency	1(2%)
	Gonadal Insufficiency	1(2%)

Table 5: Endocrine Dysfunction

Grade	Description	Number of Patients	Hydrocephalus
I	Intrasellar or Intra-diaphragmatic	None	None
II	Occupying the cistern with or without an intrasellar component	4(8%)	None
III	Lower half of the third ventricle	24(46%)	3(12.5%)
IV	Upper half of the third ventricle)	14(27%)	5(36%)
V	Reaching the septum pellucidum or lateral ventricles	10(19%)	10(100%)

While diabetes insipidus was major endocrinal finding in adults, obesity was commoner in pediatric group (Table 5). Most of the tumors were larger than 4 cm size with Samii and Tatagiba Grade IV, and hydrocephalus was seen in 35% of the cases and was directly related to increased size of the tumor (Table 6). The orbitozygomatic approach was used in all cases. Gross-total resection was achieved in 87% of patients, near-total in 8%, and partial in rest of the cases. Almost all pure cystic and pure solid tumor irrespective of tumor size had greater extent of resection (Table 7)

Table 6:Size of the tumor according to Samii et al classification system and association with Hydrocephalus

	Nature of Tumor	Number of Tumor(%)	Extent of Resection
I	Cystic	None	-
	Solid	None	-
	Cystosolid	None	-
II	Cystic	1(2%)	Total
	Solid	None	-
	Cystosolid	3(6%)	Total
III	Cystic	4(8%)	Total
	Solid	4(8%)	3 Total, 1Near Total
	Cystosolid	16(31%)	14 Total ,1Near Total, 1 Subtotal
IV	Cystic	3(6%)	Total
	Solid	2(4%)	Total
	Cystosolid	9(17%)	8 Total, 1Subtotal
V	Cystic	3(6%)	2 Total 1,Near Total
	Solid	None	-
	Cystosolid	7(13%)	5 Total , 1Near Total, 1Subtotal

Table 7.Extent of resection

Types	**Subtotal**	**Near Total**	**Total**	**Overall**	**p-value**
Cystic	0(0.0%)	1(25.0%)	10(21.73%)	11(21.15%)	0.646
Solid	0(0.0%)	1(25.0%)	5(10.86%)	6(11.53%)	0.690
Cysto-Solid	2(100.0%)	2(50.0%)	31(67.39%)	35(67.3%)	0.445
Total	2(5.8%)	4(7.7%)	46(86.5%)	52(100.0%)	

P-value for each type of cystic craniopharyngioma was 0.646, for solid 0.690 and for cysto-solid was 0.445, indicating there was no statistically significant difference among the groups with regards to extent of resection (Table 8). However, there was trend of total excision (86.5%) in all types with majority of total excision in cystosolid craniopharyngiomas(67.4%). Postoperative complications included Diabetes Insipidus and hormonal insufficiency. Most cases (67%)involved adamantinomatous grade 1 type craniopharyngioma, with a mortality rate of 6%.

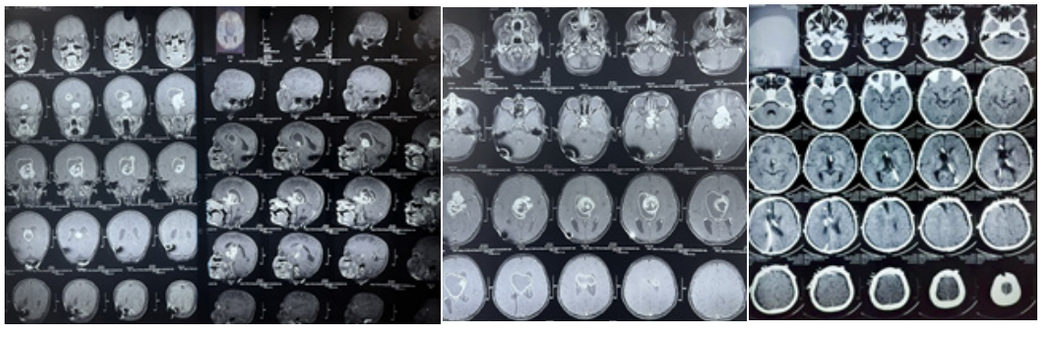

Figure 1.Pre Op and post op images

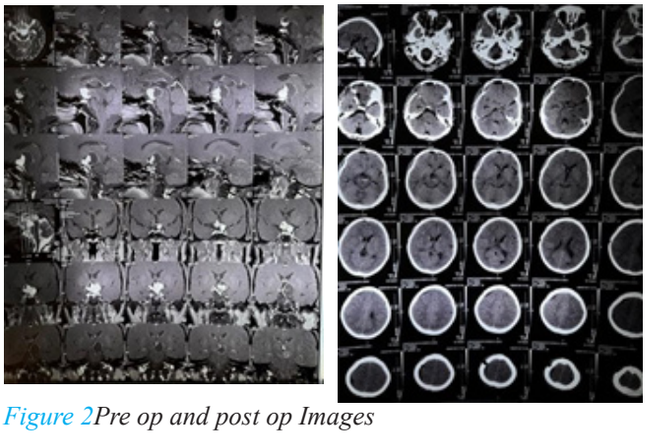

Figure 2Pre op and post op Images

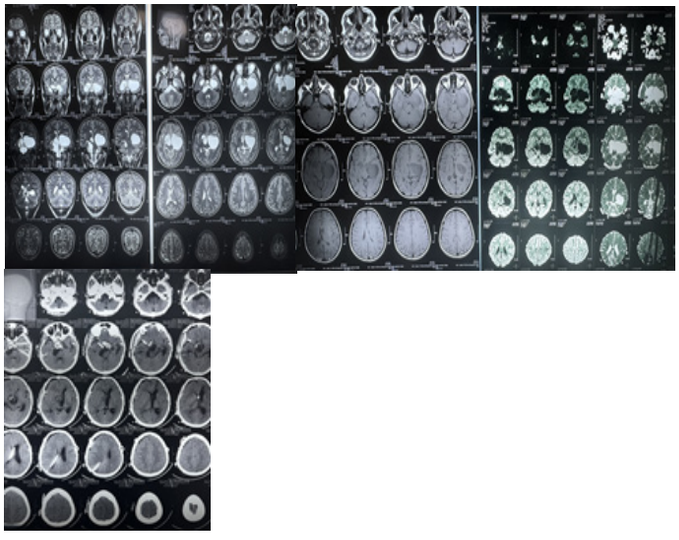

Figure 3:Pre op and Post op images

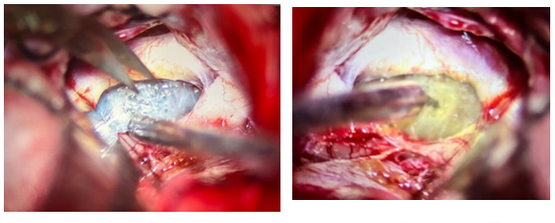

Figure 4&5 Tumor seen at Optico-Carotid Triangle &Tumor containing Xanthochromic fluid

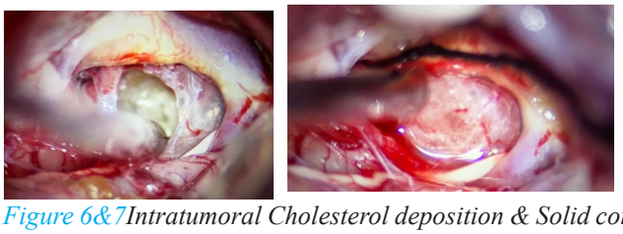

Figure 6&7Intratumoral Cholesterol deposition & Solid content of the tumor

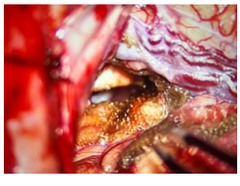

Figure 8 Total excision of the tumor

Discussion:

Craniopharyngiomas are rare tumors arising near the pituitary gland and pose intricate challenges in the realm of neurosurgery. Craniopharyngiomas are typically benign but locally behaves aggressive which affect the hypothalamus and can impact vital neurological and endocrinological functions. This comprehensive exploration delves into the intricacies of craniopharyngioma, encompassing its clinical presentation, diagnostic modalities, treatment approaches, and the associated complexities.

Craniopharyngiomas often presents subtly which makes their diagnosis challenging. Headaches and visual disturbances are the common symptoms, reflecting the tumor's proximity to the optic nerves and hypothalamus. The location of these tumors adds complexity, as they can affect hormonal regulation, leading to endocrinological abnormalities. The clinical presentation may vary, with some patients specially in children, remaining asymptomatic for extended periods, underscoring the importance of vigilant monitoring.

Accurate diagnosis is paramount for effective management. Clinical and ophthalmological examinations provide crucial initial insights, while imaging studies such as magnetic resonance imaging (MRI) and computed tomography (CT) scans offer detailed anatomical information. CT scan is superior to MR imaging in the detection of calcification. MR imaging technique was superior to CT for determining tumor extent and provides valuable information about the relationships of the tumor to surrounding structures. Thus, CT and MR imaging have complementary roles in the diagnosis of craniopharyngiomas (Figure 1,2,3). In cases of possible craniopharyngioma, noncontrast sagittal T1-weighted images may enable the identification of the normal pituitary, possibly leading to the correct diagnosis.[4]These modalities aid in determining the tumor's size, location, and potential involvement with adjacent structures. Additionally, endocrinological studies contribute valuable data on hormonal imbalances, guiding the comprehensive understanding of the tumor's impact.[5]

Microsurgical resection remains the mainstay of treatment for craniopharyngiomas. The choice of surgical approach is tailored to each case, with common techniques including the orbitozygomatic and transsphenoidal approaches. The former provides access to the tumor from above, while the latter involves reaching it through the sphenoid sinus. The surgical challenge lies in achieving a balance between radical resection and minimizing damage to critical surrounding structures, particularly the hypothalamus. Craniopharyngiomas are histologically benign World Health Organization (WHO) grade I tumors. There are two histologic subtypes of craniopharyngiomas: adamantinomatous and papillary. Adamantinomatous craniopharyngioma (ACP) is primarily seen in pediatric cases but can be seen in adults. They have solid and cystic parts (Figure 5,7). The solid part is characterized by dense nodules and trabeculae of squamous epithelium bordered by a palisade of columnar epithelium, sometimes referred to as a "picket fence." These nests of squamous epithelium are surrounded by loose aggregates of squamous epithelium known as stellate reticulum. The cystic part has a yellow-brown, cholesterol-rich fluid. Nodules of “wet keratin” represent desquamated cells that

form large, pale, eosinophilic masses that occasionally contain calcium (Figure 6).Piloid gliosis with abundant Rosenthal fibers is suggestive of invasion of surrounding brain tissue.Papillary craniopharyngioma (PCP) is commonly seen in adults. They are characterized as well-differentiated squamous epithelium lacking surface maturation, with occasional goblet cells and ciliated epithelium. Calcifications are rare in the papillary type. The papillary craniopharyngiomas are well-circumscribed compared to the adamantinomatous type, and invasion of surrounding brain tissue is much less common.6 Craniopharyngioma is a rare brain tumor of uncertain behavior that occurs at a rate of 1.3 per million person years.[7]

While surgery forms the cornerstone of treatment, craniopharyngiomas may necessitate a multimodal approach(Figure 8). Radiation therapy, either conventional or stereotactic, plays a role in managing residual tumors or cases where complete resection is not achievable. However, the decision to employ radiation involves careful consideration of potential long-term effects, especially in pediatric cases. Surgical excision followed by external beam irradiation, in cases of residual tumor, is the main treatment option.[8,9]

Chemotherapy, often in the form of intratumoral delivery, has been explored in certain instances. Interferon-alpha, for example, has shown promise in limiting tumor growth, though its use remains investigational. These adjuvant therapies aim to enhance the effectiveness of surgery and mitigate the risk of recurrence, particularly in cases where achieving gross-total resection proves challenging.[10]

Despite advancements in surgical techniques and adjuvant therapies, managing craniopharyngiomas remains complex. The proximity of these tumors to critical structures like the hypothalamus poses inherent challenges,[11] often resulting in postoperative complications (Figure 4). Diabetes Insipidus, hormonal imbalances, and neuropsychological deficits may occur, emphasizing the need for a multidisciplinary approach involving endocrinologists, neurosurgeons, and other specialists.

Craniopharyngioma management extends beyond the immediate postoperative period, necessitating long-term follow-up. Surveillance for recurrence, monitoring endocrine function, and addressing potential late effects of radiation therapy are integral components of comprehensive care. Additionally, understanding the impact of these interventions on the patient's quality of life is crucial. Long-term studies assessing cognitive function, psychosocial well-being, and overall quality of life contribute to refining treatment strategies and tailoring postoperative support. Ongoing research endeavors focus on unraveling the molecular and genetic underpinnings of craniopharyngiomas. Exploring targeted therapies and understanding the tumor's microenvironment may pave the way for more personalized treatment approaches. Advances in imaging modalities, such as functional MRI and positron emission tomography (PET), aim to enhance preoperative planning and intraoperative guidance, further refining surgical outcomes.

As we navigate the complexities of craniopharyngioma management, ongoing collaboration between clinicians, researchers, and patients is crucial. Advancements in diagnostics, surgical techniques, and adjuvant therapies offer hope for improved outcomes and enhanced quality of life for individuals grappling with these intricate tumors. The journey to unraveling the mysteries of craniopharyngiomas continues, with each discovery contributing to more effective treatments and better long-term prognoses.

Conclusion: Craniopharyngiomas, though benign tumors, exhibit malignant behavior, making their management always a significant challenge.Craniopharyngioma patients may remain asymptomatic until they acquire very large size and can present with features of raised intracranial pressure(ICP), visual disturbances and endocrinopathy. While there are multimodal treatment options, surgery stands as the cornerstone in managing this tumor

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