Original Article

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Surgical Outcomes of Pediatric Craniopharyngiomas in Nepal: A Prospective Observational Study

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Abstract

Background: Craniopharyngiomas are benign tumor of skull base which often follows an aggressive clinical course. Adamantinomatous subtype are frequently observe in age group 5-15 years. Currently, treatment strategy has relied exclusively on surgery and radiotherapy.

Materials and Methods: All pediatric Craniopharyngiomas operated at National Neurosurgical Referral Center, Bir hospital from Nov 2021 to Dec 2023 were included in this study.

Results: A total of 17 patients underwent surgical excision of craniopharyngioma. The male to female ratio was 4.6:1. The mean age of the patients was 10.88 ± 4.18 (Range 2 to 16 years). The most common presenting symptoms were headache (64.7%) followed by visual disturbances.12 patients exhibited preoperative hypothalamic pituitary axis dysfunction including central hypothyroidism (35.6%), central diabetes insipidus (29%), growth hormone deficiency (35.6%), adrenocortical insufficiency (23.5%). The Gross total resection was achieved in 6 patients (45%) while subtotal resection was done in remaining 11 patients (55%). Modified Rankin scale score (mRS) on discharge was favorable outcome (mRS 0,1,2,3) in 75% and unfavorable outcome (mRS 4,5) in 19%. There was one mortality. The overall rate of postoperative Diabetes insipidus (DI) was 35.3 % (6/17), visual deterioration (6%), meningitis (11.7%) behavioral disturbances (6%). On 6 month follow up MRI, recurrence was noted in 5 cases, 3(50%) in cases with gross total resection and 2(18.2%) in subtotal resection with adjuvant radiotherapy group.

Conclusion: We conclude that Surgical excision of pediatric craniopharyngioma caries a substantial risk of postoperative hypothalamic pituitary dysfunction which significantly impair the quality of life and functional recovery of these patients. A high index suspicion of hypothalamic involvement by tumor and its preservation can improvise patient functional outcome. Postoperative radiotherapy is advised in patient with subtotal resection to improve recurrence free survival.

Keywords: Pediatric, Craniopharyngioma, surgery, radiotherapy

INTRODUCTION

Craniopharyngiomas are benign, extra-axial tumor of skull base which often follows an aggressive clinical course. It was originally described by Harvey Cushing in 1932 as a most baffling problem in neurosurgery.[1] It constitutes 1.4 to 4 % of pediatric brain tumors.[2] Erdheim embryogenetic theory explains the origin of craniopharyngioma in children from epithelial cell rests of vanishing hypophyseal duct and its organization in sellar-suprasellar region.[3]

Adamantinomatous Craniopharyngioma (ACP) and Papillary Craniopharyngioma (PCP) are two subtypes distinguished by their histological and specific molecular signatures.[4] ACP displays a bimodal age distribution with peaks between the ages of 5–15 years and 45–60 years.[2] Conversely, PCPs are almost exclusively encountered in adults. They have varied presentation depending upon the epicenter of tumor, mass effect and associated hypothalamic pituitary dysfunction.

Currently, treatment strategy has relied exclusively on surgery and radiotherapy.[5] The gross total resection(GTR) is achievable in 70-90% of cases , however it is associated with significant postoperative morbidity and mortality owing to visual disturbance, hypothalamic dysfunction and panhypopituitarism. Subtotal resection(STR) with postoperative radiotherapy could achieve a similar recurrence free survival as GTR (14-37% vs 0-52%) with significantly low postoperatively morbidity.6 However, radiotherapy in children's is associated with risk of neurocognitive dysfunction, vasculopathy and secondary brain

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tumour.[7]

In this study, We have assessed the effect of extent of resection on relapse , endocrinological, visual and cognitive outcomes in 17 consecutive pediatric craniopharyngioma cases.

Methodology

From Nov 2021 to Dec 2023, all patients treated for Craniopharyngioma at Department of Neurosurgery, Bir hospital were included in this study. Patient with age>18, those who underwent reservoir placement/ biopsy, re-excision, uncertain histological diagnosis, or lost to follow up were excluded.

Clinical evaluation

A detail interview with neurological examination was performed and details were noted in preformed proforma. The demographic profile, clinical presentation, presence of visual dysfunction and performance status were documented.

Endocrinological evaluation

Endocrinological evaluation included preoperative and postoperative determination of anterior pituitary hormone profile, Serum Na+, serum osmolarity.

Radiological evaluation

All patients were initially evaluated with Non contrast Computed tomography (NCCT) head scan. Further tumor details and proximity to nearby critical neurovascular structures were obtained with contrast enhanced Magnetic Resonance Imaging (MRI) of Brain.

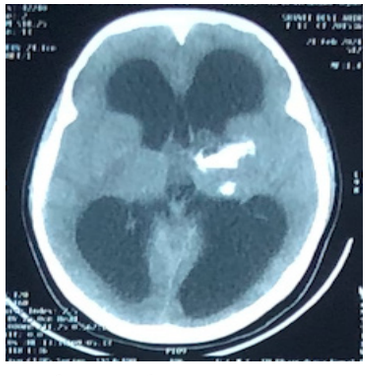


Figure 1.Ct scan of craniopharyngioma

A)Non contrast axial CT scan showing third ventricular mass with gross calcifications and gross hydrocephalus

Operative procedure

All operation were performed by single surgeon via transcranial approach or endoscopic endonasal transsphenoidal approach. The operative corridor was determined by location of tumor, size and visual status. A Standard Pterional craniotomy with subfrontal, subtemporal and transylvian approach were employed for suprasellar tumor with frontal, middle fossa and posterior extension respectively. Postoperatively patient was managed in ICU and NCCT head is done on following day.

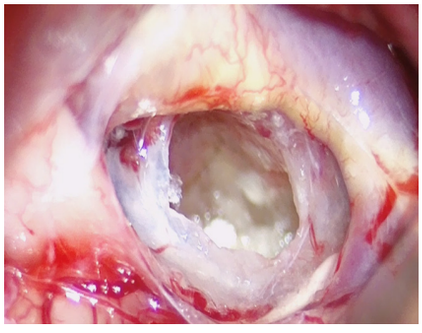


Figure 2. Optico-Carotid triangle

It illustrates the adherence of tumor to adventitia of carotid and optic nerve sheath

Follow up

Patient were followed up postoperatively for 12 months. MRI brain with contrast was done postoperatively at 3 and 6 months to determine residual tumor or recurrence.

Data analysis

All data were entered in preformed proforma. Data analysis was performed with IBM SPSS statistics for windows version 22.

Results

A total of 17 patients underwent surgical excision of craniopharyngioma over a duration of 2 years. Among them, 82% were male and 18% were female with male to female ratio of 4.6:1. The mean age of the patients was 10.88 ± 4.18 (Range 2 to 16 years).

The presenting symptoms are summarized in Table 1. The most common presenting symptoms were headache (64.7%) followed by visual disturbances. Two patients had sudden onset altered consciousness and was bought in emergency. Other symptoms were irritability, seizure, excessive weight gain and developmental delay. The duration of symptoms ranged from 1 month to 6 months. Visual field charting was done in all patient which showed findings as illustrated in Table 2. While on preoperative hormonal analysis, 12 patients exhibited hypothalamic pituitary axis dysfunction including hyperprolactinemia (55%), central hypothyroidism (35.6%), central diabetes insipidus (29%), growth hormone deficiency (35.6%), adrenocortical insufficiency (23.5%).

Table 1: Clinical Features

Symptoms	Number	Percentage (%)
Headache	11	64.7
Diminution of vision	8	47
Weakness of lower limbs	3	17
seizure	2	11.7
Altered sensorium	2	11.7
Vomiting	8	47
Weight gain	2	11.7
Short stature	2	11.7

Table 2. Visual abnormalities

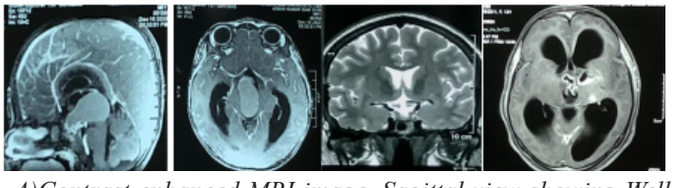
Visual field defect	Percentage (%)
Bitemporal hemianopia	24
Incongruous Homonymous Hemianopia	11.7
Unilateral blindness	11.7
Junctional Syndrome	5.8

All the patient underwent surgical excision. Gross total resection was achieved in 6 patients (45%) while subtotal resection was done in remaining 11 patients (55%). All patients who underwent subtotal excision were subjected to adjuvant conformal radiotherapy.

Table 3.Illustration of location of craniopharyngioma and surgical approach

Location	Surgical approach	Number
Sellar	Endoscopic Endonasal Transsphenoidal	2
Suprasellar		
Prechiasmatic	Pterional /Transsylvian	9
Retrochiasmatic	Pterional/Subtemporal	4
Third ventricular	Interhemispheric Transcallosal	2

Figure 3.Sites of Origin of Craniopharyngioma



A)Contrast enhanced MRI image, Sagittal view showing Well defined cystic craniopharyngioma centered within the sellar-suprasellar region with compression of optic chiasm and third ventricle superiorly

B)Axial section of same, showing Retro chiasmatic extension to intercrural cistern

C)T2 Coronal MR image showing Cysto-solid craniopharyngioma centered in Sella turcica

D)T1 PG contrast image showing intraventricular craniopharyngioma

The overall rate of postoperative Diabetes insipidus (DI) was 35.3 % (6/17). 35% of patient with normal preoperative thyoid function became hypothyroid. In one patient visual acuity deteriorated from preoperative finger count status to blindness. Two patient developed meningitis one of which died on 10th postoperative day. One patient developed behavioral disturbances.

Table 4.Postoperative complications

Complications	Number	Outcome
Vision loss	2	1 improved
Meningitis	2	1 died
Hypothyroidism	6	
Diabetes insipidus	6	2 permanent

Intraoperatively two cases of solid craniopharyngioma were noted whereas rest were cystosolid. Histopathology reports revealed Adamantinomatous features in all cases.

The duration of hospital stay was 14± 5.3 days. Modified Rankin scale score (mRS) on discharge was favorable outcome (mRS 0,1,2,3) in 75% and unfavorable outcome (mRS 4,5) in 19%. There was one mortality.

On 6 month follow up MRI, recurrence was noted in 5 cases, 3(50%) in cases with gross total resection and 2(18.2%) in subtotal resection with adjuvant radiotherapy group. No specific radiation induced toxicity was noted except for regional brain edema.

Discussion

Craniopharyngioma are WHO grade I tumor centered over the sellar- suprasellar region. It develops from the embryonic remanent of rakthe cleft cyst or from metaplasia of stalk cells. It accounts for 54 % of pediatric suprasellar tumors. It is more common in male within the age group of 5 - 10 years. The mean age of presentation in our study was 10.88 ± 4.18 years which is similar to findings of Rachel et al.[8] The nonspecific symptomatology associated with CPs could be the reason for delayed presentation in our settings.

In our study the most common presenting complain was headache (67%) followed by visual disturbances (47%) endocrinopathy(70%) similar to the findings of muller et al.[9] The mean duration of symptoms was 4 weeks to 24 weeks whereas Zucchini et al reported symptoms duration up to 2 years before presentation.[10]

Childhood onset craniopharyngioma are more frequently associated with endocrine dysfunction than in adults. Recent meta-analysis has shown endocrinopathy as high as 80-90% in pediatric CPs.[11-13] In our study, 70.5% had pituitary dysfunction among which hypothyroidism and growth hormone deficiency were most frequent. Furthermore, Diabetes insipidus in perioperative period prolonged the surgical recovery. Unfortunately, this hormonal deficiency persisted in postoperative period with substantial impairment in their quality of life.

Surgical approach was transcranial in 88.3% and endoscopic transsphenoidal in 11.7%. The gross total resection was achieved in 45% whereas subtotal resection was done in 55% with postoperative adjuvant radiotherapy. STR with adjuvant radiotherapy achieved better tumoral control (82%) versus 50% with GTR. In a Duff et al series of 121 CPs recurrence rate of patient undergoing GTR ,STR, STR+RT was 18.2%,50% and 0% respectively.[14] Both the study emphasize the role of adjuvant radiotherapy to decrease tumor recurrence.

The postoperative Diabetes insipidus and hormonal disturbances were more in patient undergoing gross total resection. This finding highlights the inadvertent injury to the hypothalamic structure with radical surgery. Safe subtotal resection with postoperative radiotherapy is advisable to prevent postoperative morbidity and recurrence. Proton bean radiation therapy is a recent advancement to address the toxicity associated with conventional radiotherapy.[8] Ommaya reservoir placement with cystic fluid aspiration and intracavitary Interferon alpha therapy can delay disease progression and offer a protracted time to definitive surgery or radiotherapy in cystic craniopharyngioma.[15]

Conclusion

Craniopharyngioma are uncommon brain tumor with varied clinical presentation. Surgical excision is still a standard of care. However, postoperative hypothalamic pituitary dysfunction significantly impair the quality of life and functional recovery of these patients. A high index suspicion of hypothalamic involvement by tumour and its preservation with meticulous surgical technique and adjuvant radiotherapy can improvise patient functional outcome with excellent long term recurrence free survival.

Limitations

This is a single center study with small sample size and limited follow up. Further multicenter randomized controlled studies evaluating the role of extend of resection in craniopharyngioma in children with or without adjuvant radiotherapy are encouraged to justify our results.

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