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CASE SERIES OF CRANIOPHARYNGIOMA: MANAGEMENT ANALYSIS & EPIDEMIOLOGY STUDY AT MADRAS MEDICAL COLLEGE – INSTITUTE OF NEUROSURGERY

Neurosurgery

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ABSTRACT

Introduction: Debate continues as to the optimal treatment for craniopharyngioma; radical surgical resection or partial resection followed by radiotherapy. Radical surgical resection may be complicated by intraoperative injury to surrounding structures and stormy postoperative hormonal problems. This study aims to examine the result of safe maximal surgical resection. **Methodology:** Retrospective study of all histopathologically proven craniopharyngiomas who had undergone surgical resection over an almost 4-year period was included. Data were collected reviewing demography, clinical presentation, hormonal dysfunction, extent of resection, and visual deterioration. Outcome was measured in terms of Glasgow Outcome Scale and recurrence. **Results:** Of 41 patients, 20(48.8%) were male and 21(51.2%) were female. Age of patients ranged from 4 to 59 years with a mean of 15.9 years. Thirty-seven patients (90%) had headache, 32 patients (78%) had visual disturbances, 23 patients (56%) had vomiting, and 10 patients (24%) had convulsions. Six patients (15%) had memory and sleep disorders and three patients of those >15 years (12.5%) had amenorrhea/sexual dysfunction. Patients who had surgery followed by radiotherapy had better prognosis, so also those aged 18 or less compared to older, males better than females and those without headache had better prognosis, though not statistically significant. **Conclusions:** Gross total excision if judiciously decided intraoperatively has a favourable outcome with acceptable morbidity. Patient has better prognosis who has surgery with radiotherapy.

KEYWORDS

Craniopharyngioma, extent of resection, outcome

INTRODUCTION

Craniopharyngiomas (cranio = skull, pharynx = throat, and oma = tumor) are benign, slow-growing, locally invasive intracranial tumors that can generate considerable morbidity, and recurrences are often difficult to manage. Reliable morphologic criteria for accurately predicting the clinical outcome of these tumors are lacking. It has challenged the neurosurgeon and his skills for many years. These tumors have a very intricate relation with the hypothalamus, pituitary stalk, and optic apparatus, which have caused excessive problems regarding optimal management. In spite of its recognition in the late 1800's, no clear consensus is reached with regard to its treatment. Philosophy of treatment is not only different among different surgeons but also in the span of a surgeons career. Computerized tomography (CT) and magnetic resonance imaging (MRI) are the investigations of choice, but today, the best imaging tool is MRI. Many people with craniopharyngioma go on to live uncompromised or relatively uncompromised lives. This is not always the outcome. Early detection and better therapeutic techniques are still required and must remain a priority. Total tumor removal is still the ideal form of treatment but should be attempted in situations when all the important adjacent structures can be safeguarded. Total tumor removal should be confirmed by a 3 month's postoperative CT scan and residual tumor should be treated with radiation therapy. Conservative removal of tumor is done when the cerebrovascular structures are at danger during surgery. The aim is to evacuate the distended cyst, excise as much as is absolutely safe and decompress the optic apparatus and hypothalamus. Residual tumor is irradiated. Karavitaki et al. [1] reported the 10 years survival rates of 77% to 100% after partial resection and subsequent radiotherapy and 81.3%–100% after radiologically confirmed total removal. Data on the treatment option with the most favorable impact on survival are not consistent. Most studies[1,2] suggest that postsurgical follow-up should be planned in 1–2 weeks for all patients and radiotherapy should start radiation within 3 weeks of surgery. Patients should be seen every 3 months for the first postsurgical year, every 6 months in the 2nd year, and 3rd years and yearly thereafter and should have a brain MRI/CT scan for comparison with previous films. In Madras medical college, Institute of neurosurgery, no study has been done specifically to determine the incidence, management, and outcome of various treatment modalities on craniopharyngiomas, hence this study aims to generate useful data to improve the management of these patients.

METHODOLOGY

Study design

This is a descriptive hospital-based retrospective and prospective study to enable the researcher to get the required sample size due to the few number of patients.

Study setting

The study was conducted at Institute of neurosurgery -MMC, neurosurgical ward and histological results were retrieved at the pathology department and patients' files were retrieved from the records departments.

Study population

The cases treated from January 2017 to January 2021 at the above-mentioned units with a diagnosis of craniopharyngioma were recruited to the study and cases were followed for the duration of 3 and 6 months after treatment. Records of retrospective clients were retrieved and great effort used to follow-up and interview patients taking part in the study. The Karnofsky score, visual and motor deficits on admission; 1 week, 3 months, and 6 months postoperatively were recorded. Assistance of the ophthalmologists was required to assess visual acuity

Inclusion/exclusion criteria

Inclusion criteria

All patients with confirmed diagnosis and admitted with cranio pharyngioma in any department during the study period within the INS- MMC Patients who gave a voluntary informed consent.

Exclusion criteria

Patients outside the above inclusion criteria Patient not operated within INS-MMC.

Data collection procedure

The information was collected using a data collection sheet (questionnaire). The same questionnaire was used for both the retrospective and prospective parts of the study. Demographic data including the age and sex were collected. The presenting symptoms, signs, and investigations done including laboratory and radiological prior to intervention and comorbidity factors such as hypertension, diabetes, and others were recorded. Preoperative care, treatment modalities, and complications after treatment were documented. Scoring of retrospective clients as per the Karnofsky and Lansky Scores was determined from the patient's file before admission, postoperatively and findings recorded in the file during the follow-up visits were used, and every effort was attempted to follow-up the patients at the neurosurgical clinic. The contacts recorded in the file were sought if the patients would have absconded to attend the follow-up clinics. Scoring of prospective patients as per the Karnofsky score was determined by the researcher on admission, 1-week and 3-months postoperatively, and the patients were followed up for a maximum period of 6 months.

OBSERVATION AND RESULTS

Incidence

The study assessed 41 patients who had been managed as cases of

craniopharyngiomas from 2014 to 2018. Twenty-nine (71%) of the study participants were assessed prospectively and 12 (29%) were assessed retrospectively, over the study period of 2014–2018. There were 577 tumors treated at INS, of which 41 (7.17%) were craniopharyngiomas. Figure 1 shows the proportion of brain tumors that were cases of craniopharyngioma treated at INS over the period.

Demographics

There were 20 (48.8%) male and 21 (51.2%) female among the study participants. The mean age was 18.3 years with a mode of 5 and a standard deviation of 15.9 years.

Table 1: Demographics				P of difference
Characteristic (%)	Retrospective (%)	Prospective (%)	Combined (%)	
Sex Male	14 (48.8)	6 (50.0)	20 (48.8)	0.920
Female	15 (51.7)	6 (50.0)	21 (51.2)	
Total	29 (70.7)	12 (29.3)	41 (100.0)	
Age groups 0-4	1 (3.4)	3 (25.0)	4 (9.8)	0.993
5-14	18 (62.1)	3 (25.0)	21 (51.2)	
15-24	2 (6.9)	2 (16.7)	4 (9.8)	
25-34	3 (10.3)	2 (16.7)	5 (12.2)	
35-44	1 (3.4)	1 (8.3)	2 (4.9)	
45+	4 (13.8)	1 (8.3)	5 (12.2)	
Total	29 (70.7)	12 (29.3)	41 (100)	

Presenting complaints

Thirty-seven patients 90%) had headache, 32 patients (78%) had visual disturbances, 23 patients (56%) had vomiting, and 10 patients (24%) had convulsions. Six patients (15%) had memory and sleep disorders and 3 patients of those >15 years (12.5%) had amenorrhea/ sexual dysfunction, as shown in Figure 3.

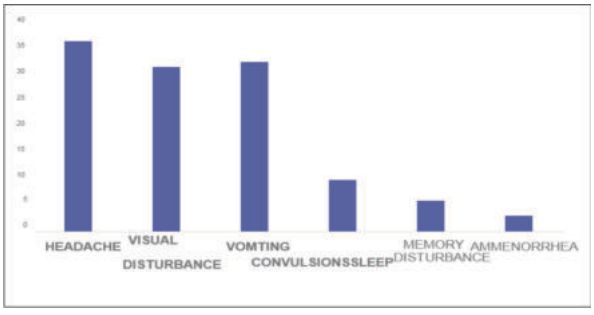


Figure 3

Physical examination findings

Six patients with oculomotor deficit, 3 patients (7%) had abducens nerve palsy, and 1 patient (2%) had vestibule cochlear nerve disorder, Twenty patients had decreased visual acuity and ten patients were totally blind as shown in Figure 4.

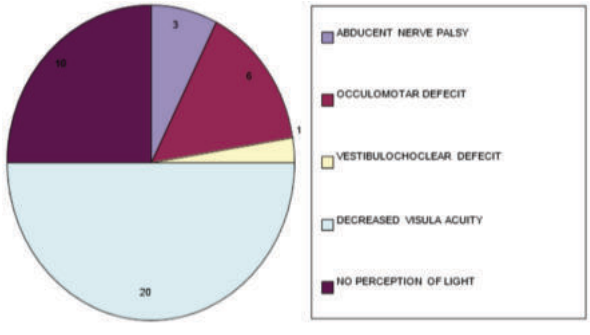


Figure 4

Laboratory investigations

As shown in Table 2, 4 out of 15 (26.7%), 2 out of 5 (40%), and 11 out of 21 (52.4%) had normal prolactin, growth hormone, and cortisol levels, respectively. Nine out of 15 (60.0%) patients had hyperprolactinemia and 13.3% had hypoprolactinemia, while three out of five patients (60%) had low growth hormone levels. Ten out of 21 patients (47.6%) had hypocortisolemia as shown in Table 2.

Table 2
Treatment

Investigations	Value
Prolactin levels Normal	4
Lower	2
High	9
Total	15
Growth hormone levels Normal	2
Lower	3
Total	5
Cortisol levels Normal	11
Total	21
Lower	10

Forty-one patients were done CT-scanning and MRI. Twenty-six (63.4%) and 15 patients (36.6%) had suprasellar and sellar with suprasellar extension on radiological imaging, respectively, as shown in Figure 5. Forty-one patients were done craniotomies. Twenty-three (57.5%) of the patients had surgery alone and seventeen (42.5%) underwent radiotherapy in addition to surgery. Thirty-seven (91.0%), three (7.5%), and one (2.5%) of the patients had partial excision/decompression, biopsy-only, and complete excision, respectively as show in Figure 6, during craniotomy and 23 (57.5%), underwent additional surgical procedures (ventriculoperitoneal shunt) to relieve hydrocephalus. Six patients had two craniotomies done due to confirmed tumor recurrence on follow-up

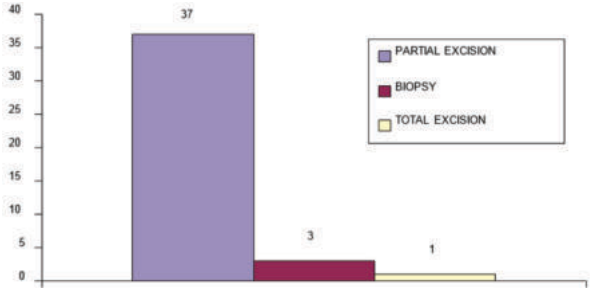
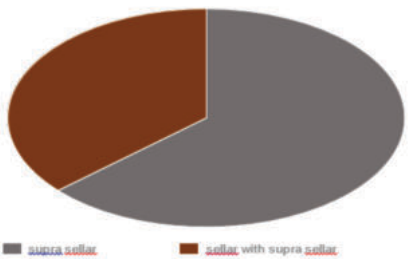


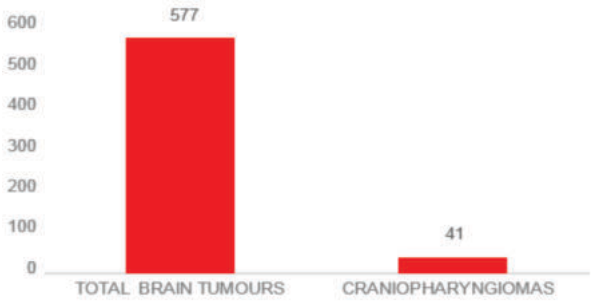
Figure 6: Type of procedure

DISCUSSION

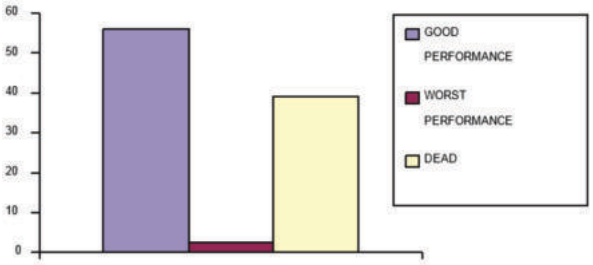
In a study at KNH by Mahindu,[2] the relative incidence of craniopharyngioma was reported as 5.8%. In this study, the relative incidence is 7.1% which is different with the incidence reported by Cushing[3] and Ruberti[4] of 4.4%–4.6%.. If the statistics from the rest of the country were considered, the results could have been different. There was no gender difference in the study participant. Mcdonald et al. [5] reported in their study that there is no variation by gender or race and Shin et al. [6] reported the same findings. Hoffman et al. [7] from the hospital for sick children in Toronto in a study of total excision of craniopharyngioma in 50 children noted that the tumor was more common in boys than girls (56%, 44%), and this is supported by Nagpal[8] and Baskin and Wilson[9] (68%, 32%) and (54%, 36%), respectively. The age at diagnosis varies widely and cases have been reported in fetuses and in the elderly. It may occur at any age but most commonly presents in childhood or adolescence. A bimodal distribution by age with peak incidence rates in children (aged 5–14 years) and among older adults (aged 65–74) was noted.[1] Fahlbusch et al. [10] in Europe also noted a bimodal distribution with peak incidence at 15–20 years and another at 50–55 years. In this study, the tumor tends to occur in the young age groups of 0–14 years and 15–24 years. There is no bimodal distribution noted in this study. Kendall-Taylor et al. [11] reported the peak age at onset as 15–20 years



Radiological location of tumour



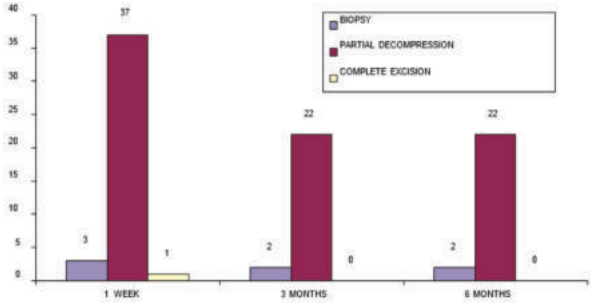
Proportion of tumours that were craniopharyngiomas



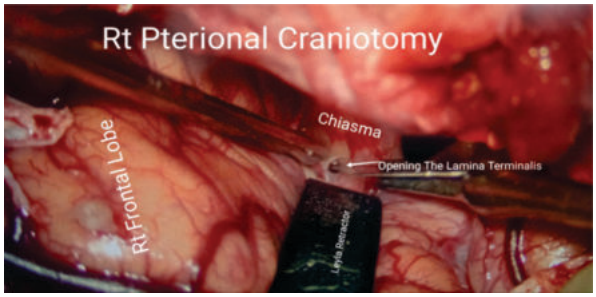
Outcome in relation to all type of treatment

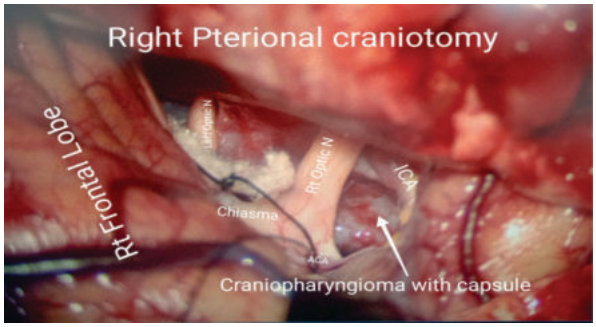
CONCLUSIONS

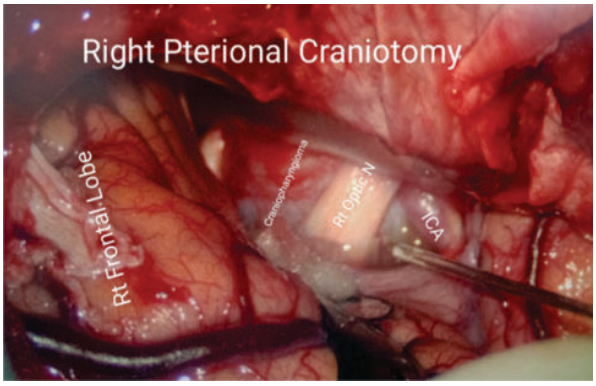
We believe that total removal of craniopharyngiomas is possible in selected cases, and it has been our experience in adults that this cannot often be achieved. A subtotal removal followed by radiation therapy has been a satisfactory approach in our hands, and with modern microsurgical techniques as well as with the advantages of modern radiotherapeutic techniques, this approach has led to remission in approximately 90% of our patients. While our follow-up period is admittedly short and it will be important to continue to follow these patients over the next several decades, it appears that with continuing and careful endocrine replacement therapy and monitoring, these patients can resume a fully functional life with few limitations.

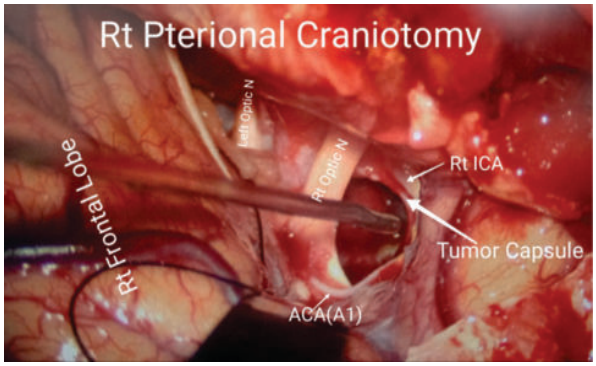


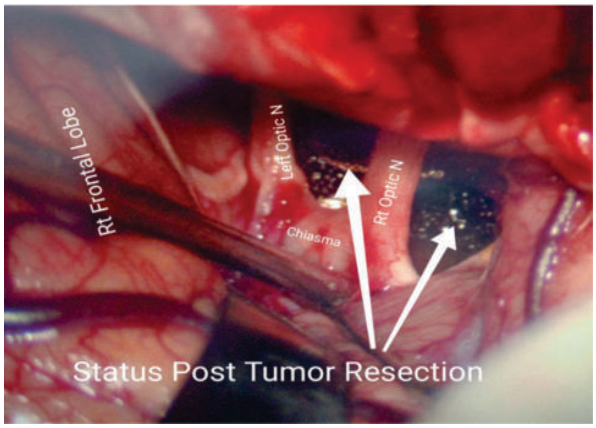
Outcome in relation to the type of surgery

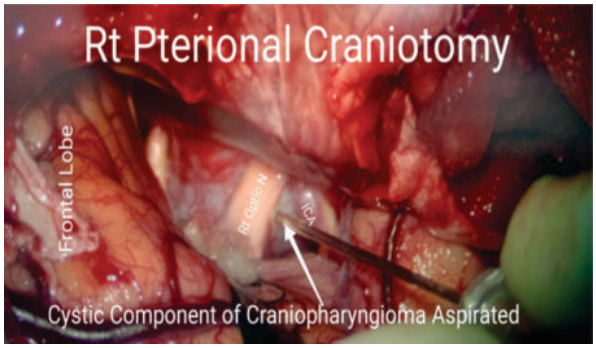


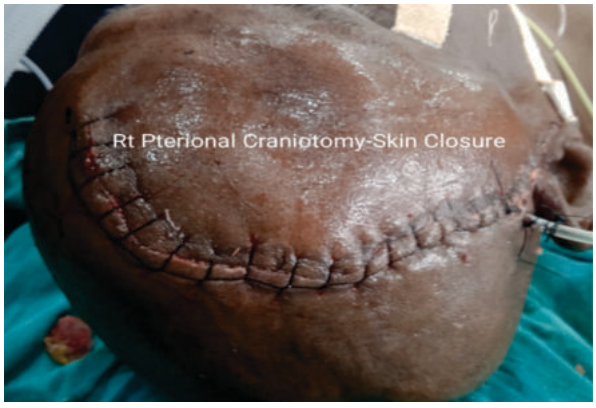












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