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Case Series

Clinico-epidemiological profile and treatment outcome of craniopharyngioma: a case series

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ABSTRACT

Craniopharyngioma is a rare type of benign (non-cancerous) brain tumor that primarily affects children and young adults. It arises from remnants of the Rathke's pouch, which is an embryonic structure that forms during the development of the pituitary gland. Craniopharyngiomas typically develop near the pituitary gland, in the area known as the sellar region. These tumors can cause various symptoms depending on their size and location, including hormonal imbalances, vision problems, headaches, and growth abnormalities. The management of craniopharyngioma involves a multidisciplinary approach, including surgery, radiation therapy, and hormone replacement therapy. Long-term outcomes can be influenced by the tumor's size, location, and the treatment modality used. Therefore, early recognition, accurate diagnosis, and appropriate treatment are crucial for optimizing the quality of life and long-term prognosis for individuals with craniopharyngioma. A detailed review of a number of craniopharyngioma cases is presented in this case series publication, with an emphasis on the clinical presentation, diagnostic methods, therapeutic options, and long-term results. This study's goal is to give doctors and researchers who work with patients who have this difficult malignancy useful information.

Keywords: Craniopharyngioma, Surgery, Radiation

INTRODUCTION

A relatively benign tumour of the central nervous system (CNS), craniopharyngiomas are uncommon. It is a partially cystic embryonic abnormality that can affect the sellar/parasellar area and cause a variety of symptoms, including headaches, nausea, vomiting, vision difficulties, and endocrine problems. For the medical professionals that treat it, such as those in neurosurgery, neuro-ophthalmology, neurology, endocrinology, and paediatrics, it poses a unique difficulty. The tumour's capacity to cling to its surrounding surfaces presents a hurdle. It is known for its high rates of recurrence as a result, making it notoriously difficult to manage.[1-4] Craniopharyngioma has an incidence of 0.5 to 2 cases per million persons per year. Craniopharyngiomas can be recognized at any age. It is generally considered a paediatric disease accounting for 1.2 to 4% of all intracranial tumours; however, approximately half of the craniopharyngiomas are diagnosed in adults. It has a classical bimodal age distribution, with an increased incidence rate in 5 to 14 years and 50 to 74 years of age. There is no statistical difference in the incidence based on gender, race, and geographical location. The diagnosis of childhood-onset CP is often made late, frequently years after the initial manifestation of symptoms, with a clinical picture characterized by symptoms of increased intracranial pressure (e.g., nausea and headache) at the time of diagnosis. Further primary manifestations are endocrine deficits.[5-9]

Craniopharyngioma has a very high recurrence rate of approximately 50%. It also has high survival rates (83% to

96% five-year survival and 65% to 100% 10-year survival) but also carries similar rates of morbidity, with almost all patients developing some sequelae. Surgical intervention is indicated to confirm the diagnosis, tumours causing neurologic deficits, pituitary dysfunction, and hydrocephalus. The most common surgical approaches are endoscopic endonasal transsphenoidal (EET) or transcranial, depending on the location of craniopharyngioma. Extension of resection is a matter of debate. Gross total resection has been associated with an increased incidence of post-surgical deficits, with no change in the recurrence rates. Radiation treatment is used in patients with residual disease or to prevent recurrences. Radiation therapy includes conventional external radiotherapy, proton beam therapy, stereotactic radiotherapy, radiosurgery, and brachytherapy. The goal of radiotherapy is to decrease tumour burden while protecting essential neural structures. Specific Gy doses have been designated for every radiation modality. Multiple reports have suggested decreased mortality with slightly reduced morbidity following radiation therapy. Despite this, radiation therapy has not been proven to reduce the recurrence rate. Therefore, it continues to be an adjuvant modality to neurosurgical intervention. Intracystic therapy is primarily used for purely cystic craniopharyngioma. Toxic substances like radioactive isotopes, bleomycin, interferon-alpha are used in intracystic therapy to produce tumour fibrosis and sclerosis. This method has been reported to produce significant cyst shrinkage, but there is limited data on its use and support. A disadvantage of this option is that severe neurotoxicity can occur in some cases due to cystic leakage of the sclerosing substance.[10-14] The aim of our study was to study the clinico-epidemiological profile and treatment outcome of craniopharyngioma.

CASE SERIES

This retrospective study was carried out at the radiation oncology department of the Sher-I-Kashmir Institute of Medical Sciences in Soura, Srinagar, India. The 12 cases of craniopharyngioma identified between January 2019 and December 2021 have all pertinent clinical and epidemiological information available. Treatment, overall survival, and follow-up data were examined together with clinical and epidemiological characteristics. The data were first keyed into a Microsoft excel spreadsheet and cleaned for any inaccuracies. Statistical analysis was done using IBM statistical package for the social sciences (SPSS) Statistics for Windows from IB Corp. (released 2020, Version 27.0. Armonk, NY, USA). Categorical variables were shown in the form of frequencies and percentages.

In the current study, a total of 12 patients were analysed, 8 were children and 4 adults. The gender distribution in the children's group was balanced (3 females, 3 males), however there was a small male predominance in the adult group (4 male cases, 2 female cases). Headache, vision issues, neurological symptoms (convulsions, diplopia, loss of conscience, vertigo), problems with memory, focus, and attention, and irregular sleep patterns were the most prevalent symptoms that led to a diagnosis. Others included pituitary insufficiency symptoms like growth arrest, delayed puberty.

Of the 12 patients, 4 (33.3%) got surgery alone, 8 (66.7%) underwent surgery followed by adjuvant radiation treatment (RT). Most of these patients who underwent surgery and radiotherapy afterward only had tumour decompression. The radiation was given 4-6 weeks after surgery by conformal technique to a dose of 54 Gy in 30 fractions @1.8Gy per fraction. In all patients that received adjuvant radiotherapy tumour control was better. The overall survival was 90% and 80% at 5 and 10 years respectively (Table 1). Three individuals experienced recurrence after an average of 2-3 years and underwent re-surgery. Following surgery, visual problems did not significantly improve. In both adults and children, treatment outcomes and side effects were comparable. None of the patients received systemic therapy.

Table 1: Clinical profile of patients.

Parameters	N (percentage)
Adults	4 (33.3)
Children	8 (66.6)
Surgery alone	4 (33.3)
Surgery + radiation	8 (66.7)
Overall survival at 5 years	90
Overall survival at 10 years	80

DISCUSSION

Although craniopharyngiomas are histologically benign, they are usually infiltrative neoplasms and are intricately associated with the optic structures, hypothalamus and the circle of Willis. These tumours have a bimodal age distribution and occur most commonly among patients who are 5 to 14 and 50 to 74 years old. Because of the many different presenting symptoms and potential complications associated with the various treatment options, we advocate for prospective trials that take into consideration not only control rates, but quality of life issues.

Neurosurgical intervention is the initial treatment of choice but the recommended extent of tumour resection is still debated. CRF remain a surgical challenge because of the tumour dimensions and structure as well as the adherence to neighbouring neurovascular structures. Craniotomy is frequently used in most series, the transsphenoidal approach being reserved for small intrasellar or infradiaphragmatic tumours.[15,16]

The rate of gross tumour resection (GTR) has dramatically grown globally over the past several decades as a consequence of considerable advancements in imagistic methodologies and neurosurgical procedures. Parallel to this, perioperative mortality fell.[17] However, the newly

published outcomes are still incredibly diverse (because to variation in tumour dimensions/structure/invasion as well as the neurosurgical team's experience). Between 17 and 89% of GTRs have been reported.[18,19] Subtotal resection (STR), often followed by adjuvant postoperative radiation, accounts for the majority of the remaining cases. Because of the much-decreased recurrence rate compared to those with subtotal resection followed up and monitored conservatively, this strategy is advised by several experts. Naturally, these comments cannot be converted into therapeutic recommendations in the absence of randomised research, but to date subtotal resection with adjuvant radiotherapy appears to offer a reasonable chance of cure with lower recurrence rates and relatively lower morbidity.

The results of the various series of fractionated radiotherapy in the era of CT-based treatment planning with or without MRI fusion show high control rates of 92.0% to 100.0%, albeit with short follow-up (1.3–8.2 years). It should be noted that high control rates were noted in the series reported by Merchant et al despite 40 of 88 patients treated (45.5%) receiving surgery described as "minimal to none" and that the extent of resection was not related to PFS.[20]

Modern imaging, utilizing computed tomography (CT) simulation with magnetic resonance imaging (MRI) fusion, has allowed for better delineation of intracranial tumour volume. This has allowed for expansion of the gross tumour volume by 5 mm to cover microscopic disease (i.e., clinical target volume) with an additional expansion margin of 3 mm to account for daily setup (planning tumour volume). This technique has been found to yield very high rates of local control with short-term follow-up.[21] Intracavitary bleomycin has been reported in small patient series. Although the majority of cystic lesions will decrease in size with therapy, drug leakage into surrounding tissues has been associated with serious events and fatal effects such as hypothalamic damage, hearing loss, cerebrovascular events, blindness, and brain edema. Interferon alpha has also been used for intracystic treatment with low rates of morbidity. Although high response rates have been noted, only short-term follow-up is available.[22-25]

In this study overall survival. The overall survival was 90% and 80% at 5 and 10 years respectively. Local control was better in patients who received adjuvant radiation. None of the patients received chemotherapy.

CONCLUSION

The highest chance of a cure is achieved by total excision of the tumour; however, this should not be done at the price of substantial functional impairment. Adjuvant radiation improves local control, especially in patients with persistent illnesses following tumour decompression.

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