2-1-2021

Complete versus subtotal resection of paediatric craniopharyngioma

Wardah Pervez

Saqib Kamran Bakhshi

Farhan A. Mirza

Muhammad Shahzad Shamim

Complete versus Subtotal Resection of Paediatric Craniopharyngioma

Wardah Pervez[1], Saqib Kamran Bakhshi[2], Farhan A. Mirza[3], Muhammad Shahzad Shamim[4]

Abstract

Optimal management of craniopharyngioma is challenging and requires multidisciplinary efforts for a successful outcome. The debate over radical versus a conservative resection followed by radiotherapy, continues. Current literature reports no difference in disease freedom and overall survival between the two surgical viewpoints, and favours conservative resection and radiotherapy, leading to lower morbidity and superior functional outcomes.

Keywords: Paediatric craniopharyngioma, extent of resection, radiotherapy

Introduction

Craniopharyngioma is a benign, epithelial tumour, or as some authors have mentioned, an embryonic malformation in the sellar/suprasellar location. It is predominantly seen in children and adolescents[1] and comprises around 10% of all paediatric central nervous system tumours.[2] The close proximity of craniopharyngioma to eloquent structures such as pituitary gland, optic chiasm, hypothalamus and carotid arteries, makes surgical resection as well as adjuvant radiation challenging.[3]

Significant controversy mars the optimal treatment strategy for managing craniopharyngioma. Even though these are non-malignant lesions, recurrence or disease progression often blemishes the aim of long-term disease control. Gross total resection (GTR) reduces the risk of recurrence and has traditionally been proposed as the best strategy.[4,5] But with the high rates of endocrinological and visual complications associated with GTR the pendulum has swung towards subtotal resection (STR) followed by radiotherapy.[5,6] However, the intellectual impedance, mental retardation, and risk of secondary tumours induced by radiotherapy has again lead surgeons and oncologists to rethink their approach towards this lesion. This has been further supported by the advent of extended endonasal approaches (EEA) providing a direct approach to these challenging tumours, without transgression of normal brain.

[1,2,4]Section of Neurosurgery, Aga Khan University Hospital, Karachi-Pakistan;
[3]Kentucky Neuroscience Institute (KNI), Department of Neurosurgery, University of Kentucky, Lexington, KY, USA.
Correspondence: Muhammad Shahzad Shamim. e-mail: shahzad.shamim@aku.edu

During last few decades, our understanding of the pathogenesis and natural history of craniopharyngioma has improved immensely. Although extensive literature has been published and surgeons have been modifying their approach to these tumours, there is still a lack of consensus on the standard treatment for craniopharyngiomas in children.[7] We have reviewed the existing literature published on the surgical management of craniopharyngioma, and have aimed to summarize the available recommendations for an ideal approach to these tumours.

Review of evidence

Recent advancement in surgical techniques, including increased use of endoscopic approach, and imaging technology has reduced the overall risk of surgical complications during resection of craniopharyngioma. It would be fair to say that trans-nasal endoscopic approach or EEA is now the standard for these tumours. Other common surgical approaches include microscopic and endoscope-assisted trans-cranial approaches, as well as trans-ventricular endoscopic approaches.[8,9]

Surgical Approach

The goal of surgery in most cases is to relieve hydrocephalus and/or decompress the optic apparatus. The surgical approach should be tailored to the morphology of the lesion. Predominantly cystic tumours extending into the third ventricle are amenable to a trans-ventricular endoscopic approach for cyst decompression and biopsy. This can be accompanied by insertion of an Ommaya reservoir for repeat cyst drainage if it enlarges and becomes symptomatic. This also allows instillation of chemotherapeutic agents directly into the cyst. Solid tumours in the sellar/suprasellar region with superior extension into the third ventricle are generally amenable to extended endonasal approaches which provide the most direct corridor to tumour resection. Tumours predominantly in the third ventricle with limited suprasellar presence are difficult to access via the trans-nasal route and often require pituitary transposition maneuvers. Advanced maneuvers may be required to mobilize the pituitary gland and protect the chiasm if the tumour is completely retro-chiasmatic. These tumours are better served with a trans-callosal approach or a sub-frontal translaminar terminalis approach. Regardless of approach, the tumour-hypothalamic plane should be respected at all costs to minimize hypothalamic transgression.

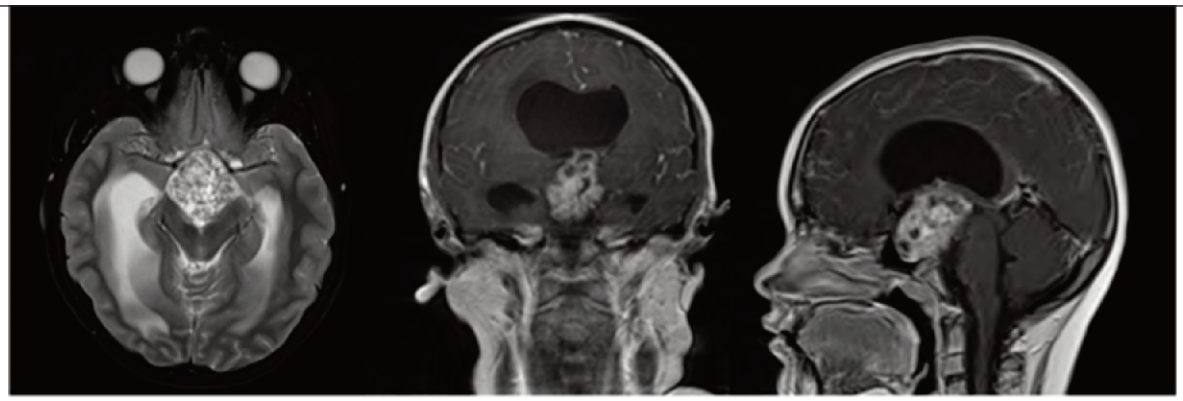

Figure-1: T2 weighted axial section, and T1-post contrast coronal and sagittal sections showing a predominantly solid, heterogenously enhancing sellar and suprasellar craniopharyngioma which is extending into the sphenoid sinus and 3rd ventricle, and is causing hydrocephalus.

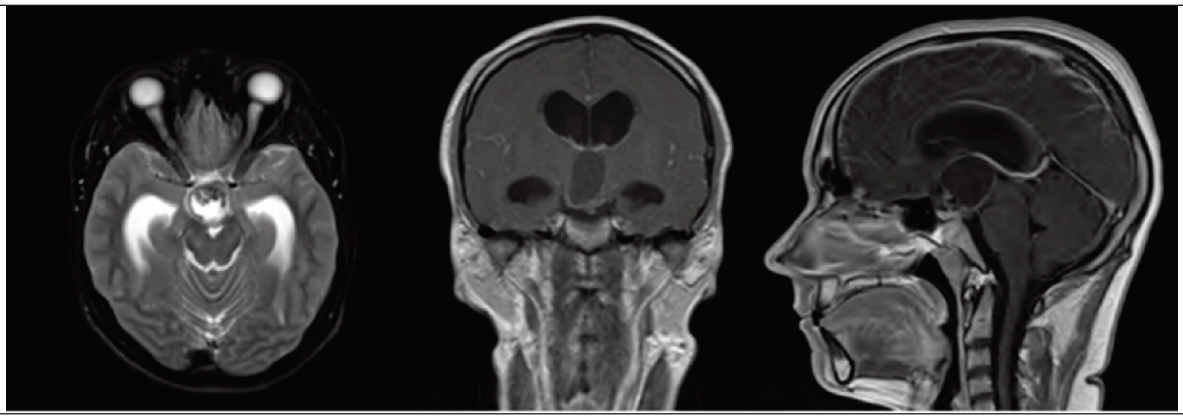

Figure-2: T2 weighted axial section, and T1-post contrast coronal and sagittal sections showing a predominantly cystic, sellar and suprasellar craniopharyngioma which is ring enhancing and has a small enhancing solid component. It is displacing the floor of 3rd ventricle, and is causing hydrocephalus.

Extent of Resection

Sarkar et al. retrospectively studied 37 children (mean age 10 years and median follow-up of 79 months) 16 (43.2%) underwent GTR, 6 (16.2%) had near-total resection and 15 (40.5%) had STR.[10] Extent of resection was defined on post-op MRI done 3-6 months after surgery, and patients with STR had also received adjuvant radiotherapy.[10] Children with STR and radiotherapy had a better survival as compared to those who underwent radical surgery, and the extent of resection did not influence long-term functional outcome.[10]

Vile et al. studied 75 paediatric craniopharyngioma cases treated at their hospital over a 20 year period.[11] Detailed endocrinological, visual, psychological, neurological, hypothalamic and radiological evaluations were completed in all patients pre- and post-operatively. GTR was achieved in 29 (39.7%) children while 44 (60.3%) had STR. Attempts to remove the tumour adherent to the hypothalamus resulted in greater hypothalamic dysfunction after surgery. STR was associated with less post-operative morbidity and there was no statistically significant difference in the tumour recurrence rate between patients who achieved GTR versus those who achieved STR with adjuvant radiotherapy. In a follow up study by the same group, 59 children were treated with a less aggressive approach with STR/cyst aspiration followed by radiotherapy.[12] This study proposed that use of image guidance and endoscopy decreased surgical morbidity, and targeted radiotherapy using proton-beam resulted in better functional outcomes as compared to the earlier cohort study performed at their center. This study also noted that children who present at a younger age (≤ 5 years) are poised to develop higher long-term morbidity. As radiotherapy cannot be safely administered in this young group, they are at a higher risk of tumour recurrence therefore more radical surgical resection should be considered if safely possible.[12]

In another series of 28 children, Liu et al. reported GTR in 11 (39%), STR in 14 (50%), and biopsy in 3 (11%) patients.[13] Seven patients with STR and 2 patients with GTR received adjuvant radiotherapy, whereas the 3 patients who had

undergone biopsy, received intra-cystic interferon therapy via Omaya reservoir. In their cohort, extent of resection alone was not a significant predictor of survival, however, patients who had received adjuvant radiotherapy had better progression free and overall survival, as compared to those who underwent surgery alone or surgery with interferon therapy.

In another series of 25 children 21 (84%) underwent GTR. They had proposed GTR to be associated with more favourable outcome.[14] However, in an extensive systematic review of 377 patients, Clark et al. concluded that STR and radiotherapy offers similar tumour control rate as GTR, and therefore, should be the aim of surgical treatment.[15]

Conclusion

Craniopharyngioma should be considered and treated as a chronic disease. Goal of treatment should be aimed at maximizing preservation of pituitary, hypothalamic, and visual function. Current evidence predominantly suggests that the extent of surgical resection does not predict progression free survival or overall survival in paediatric craniopharyngioma, as GTR and STR with radiotherapy have similar survival outcomes. Aggressive resection carries a higher risk of morbidity and thus, should be reserved for only those cases where the tumour is easily resectable and the tumour-hypothalamic plane can be preserved. In younger children in whom radiotherapy is not a feasible option, maximizing safe resection and use of intra-cystic therapy should be considered.

References

1. DiPatri AJ, Prabhu V. A history of the treatment of craniopharyngiomas. Childs Nerv Syst. 2005;21:606-21.
2. Soldozy S, Yeghyayan M, Yağmurlu K, Norat P, Taylor DG, Kalani MY, Jane JA, Syed HR. Endoscopic endonasal surgery outcomes for pediatric craniopharyngioma: a systematic review. Neurosurg. Focus. 2020;48:E6.
3. Karavitaki N, Cudlip S, Adams CB, Wass JA. Craniopharyngiomas. Endocr. Rev. 2006;27:371-97.
4. De Vile CJ, Grant DB, Kendall BE, Neville BG, Stanhope R, Watkins KE, Hayward RD. Management of childhood craniopharyngioma: can the morbidity of radical surgery be predicted? J Neurosurg. 1996;85:73-81.
5. Stripp DC, Maity A, Janss AJ, Belasco JB, Tochner ZA, Goldwein JW, Moshang T, Rorke LB, Phillips PC, Sutton LN, Shu HK. Surgery with or without radiation therapy in the management of craniopharyngiomas in children and young adults"Int. J. Radiat. Oncol. Biol. Phys. 2004;58:714-20.
6. Yang I, Sughrue ME, Rutkowski MJ, Kaur R, Ivan ME, Aranda D, Barani IJ, Parsa AT. Craniopharyngioma: a comparison of tumor control with various treatment strategies. Neurosurg. Focus.. 2010;28:E5.
7. Clark AJ, Cage TA, Aranda D, Parsa AT, Sun PP, Auguste KI, Gupta N. A systematic review of the results of surgery and radiotherapy on tumor control for pediatric craniopharyngioma Childs Nerv Syst.. 2013;22:231-8.
8. Wijnen M, van den Heuvel-Eibrink MM, Janssen JA, Catsman-Berrevoets CE, Michiels EM, van Veelen-Vincent MC, Dallenga AH, van den Berge JH, Van Rij CM, van der Lely AJ, Neggers SJ. Very long-term sequelae of craniopharyngioma. Eur J Endocrinol. 2017;176:755-67.
9. Park HR, Kshettry VR, Farrell CJ, Lee JM, Kim YH, Won TB, Han DH, Do H, Nyguist G, Rosen M, Kim DG. Clinical outcome after extended endoscopic endonasal resection of craniopharyngiomas: two-institution experience. World Neurosurg.. 2017;103:465-74.
10. Sarkar S, Chacko SR, Korula S, Simon A, Mathai S, Chacko G, Chacko AG. Long-term outcomes following maximal safe resection in a contemporary series of childhood craniopharyngiomas. Acta Neurochir . 2020;19:1-1.
11. De Vile CJ, Grant DB, Kendall BE, Neville BG, Stanhope R, Watkins KE, Hayward RD. Management of childhood craniopharyngioma: can the morbidity of radical surgery be predicted? J Neurosurg. 1996;85:73-81.
12. Marcus HJ, Rasul FT, Hussein Z, Baldeweg SE, Spoudeas HA, Hayward R, Thompson D, Grieve JP, Dorward NL, Aquilina K. Craniopharyngioma in children: trends from a third consecutive single-center cohort study. J Neurosurg: Paed. 2019;25:242-50.
13. Liu AP, Tung JY, Ku DT, Luk CW, Ling AS, Kwong DL, Cheng KK, Ho WW, Shing MM, Chan GC. Outcome of Chinese children with craniopharyngioma: a 20-year population-based study by the Hong Kong Pediatric Hematology/Oncology Study Group. Childs Nerv Syst. 2020;36:497-505.
14. Bidur KC, Prasad DU. Outcome following surgical resection of craniopharyngiomas: A case series. Asian J. Neurosurg.2017;12:514.
15. Clark AJ, Cage TA, Aranda D, Parsa AT, Sun PP, Auguste KI, Gupta N. A systematic review of the results of surgery and radiotherapy on tumor control for pediatric craniopharyngioma. Childs Nerv Syst. 2013;29:231-8.