Quality of Life in Children Treated for Craniopharyngiomas

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Abstract

Craniopharyngiomas are common but complex paediatric brain lesions that present interesting management challenges. Quality of life is an important consideration while choosing management options. In this review, we have discussed the existing literature on various aspects of quality of life in patients treated for craniopharyngioma, assessed by variety of measurement tools.

Keywords: Craniopharyngioma; Parasellar lesion; Sellar lesion; Quality of life; Hypothalamic lesion

Introduction

Craniopharyngiomas are rare embryonic malformations of sellar and parasellar areas of low histological malignancy, thought to be derived from Rathke's pouch epithelium. The disease is treated with surgery and often adjuvant treatment such as chemotherapy and radiotherapy. Due to the location of pathology and its proximity to vital structures, both the growth of CP as well as the treatment, are associated with high risks for disturbing important neurological and endocrine functions such as visual impairment, stroke, obesity, pituitary insufficiency, etc., thus impairing the quality of life (QoL) in long-term survivors. In this review, we have described how the CP and its treatment may affect patients' QoL despite good disease control.

Review of literature

We have presented the literature, grouping the various outcomes related to childhood onset craniopharyngioma and its treatment into five major categories; obesity, visual and neurological outcomes, endocrine imbalance, psychological and psychosocial outcomes, and other determinants of QoL. There is a large body of literature on this topic, and we have only discussed a few publications for each outcome. Some other notable references on the topic are mentioned in the Table.

I. Obesity

Hypothalamic involvement (HI) either due to the CP, or its treatment has significant effect on long term prognosis. Childhood onset CPs with HI were associated with overall reduced survival and increased Body Mass Index (BMI) at diagnosis and follow up when compared with CPs without HI. Children with hypothalamic lesion extending into the nuclei of posterior hypothalamus, frequently develop hypothalamic obesity (HO) syndrome which is characterized by hyperphagia, diminished satiety, decreased physical activity and consequential obesity. HO risk assessment can be done using hypothalamic lesion scoring (HLS) system which can predict the risk for developing HO and may allow early initiation of intervention strategies. After resection of the tumour, patients with childhood onset CP had 50% increased rate of obesity. Obese patients are at further increased risk of developing subsequent atherosclerotic cardiovascular diseases, metabolic syndromes and type 2 diabetes mellitus (DM).[1]

II. Visual and neurological outcomes

Optic neuropathy may be observed in children with CP despite few cases presenting with visual symptoms.[2] Longstanding papilloedema, direct compression due to tumour, radiotherapy and handling during surgery can all injure the optic pathway, although, direct compression is described as the major cause of visual loss.[3] Pre-chiasmatic tumours and critical pre-operative visual impairment are risk factors for post-surgical visual disturbances. Surgery via trans-sphenoidal route has shown better ophthalmological outcomes, although it can be carried out in only a select group of patients.[4] Prevalence of persistent neurological deficits is reported at around 8%, but may rise up to 36% for larger tumours. Treatment related neurological deficits include cranial nerve injury, stroke, and epilepsy.[5]

III. Endocrine imbalance

Around 40-80% children present with at least one hormonal deficiency at the time of diagnosis. Diabetes insipidus (DI) is found in 17-27% of patients.[6,7] Tumour extension and involvement of hypothalamic-pituitary axis also affect the risks of post-operative endocrine imbalance.[5] Hoffman et al., assessed long term endocrine status of 46 patients with childhood onset CP, and found that all patients were using some form of hormone replacement. Ninety three percent of these patients were on 1-desamine-8-D-arginine vasopressin (DDAVP) replacement, 89% cortisone, 83% thyroid, 30% sex hormones and 20% were on growth hormone

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Table: Studies on childhood onset craniopharyngioma and its treatment.

	Author	Outcomes assessed	Scale used
1	Srinivasan et al[1]	Obesity	BMI-SD, Percentage body fat formula[a]
2	Hoffman et al[6]	Intellectual, Emotional function	Full scale intelligence quotient scores.
3	Memmesheimer et al[10]	Psychological well-being	HADS[b], World Health Organization well-being index (WHO-5)
4	Sterkenburg et al[11]	Obesity, Functional capacity, Psychosocial status, QoL	BMI-SD, EORTC-QLQ-C30[1] questionnaire, MFI-20[2]
5	Kendall et al[15]	Obesity	BMI-SD, MRI-score
6	Riva et al[16]	Neurological, neurocognitive and behavioural development	WCST[c], Benton visual retention test, Trail making test, Raven's test.
7	Muller et al[17]	Functional capacity, Obesity	BMI-SD, FMH[d]
8	Ozyurt et al[18]	Memory, Executive functions	Emotional face recognition task, functional MRI (fMRI)

a Percentage body fat = DXA measured fat/ DXA measured soft tissue plus bone mineral content; b HADS (Hospital Anxiety and Depression Scale); c WCST: Wisconsin Card Scoring Test; d FMH: Fertigkeitenskala Munster/Heidelberg.

replacements.[6] One of the early manifestation of paediatric CP includes impaired growth, which often occurs years before diagnosis. At the time of diagnosis, growth hormone (GH) insufficiency has been recorded in 26-75% of CP patients. On the other hand, GH deficiency was also present in 70-92% of CP patients after treatment.[7,8] The percentage of post-operative DI is between 40-93% while transient post-surgical DI develops in up to 80-100% of total cases.[9]

IV. Psychological and psychosocial wellbeing

Memmesheimer et al., predicted loss of concentration, visual field defects, variable BMI and female sex as strongest predictors for depression in patients treated for CP. For anxiety, BMI, negative life incidents and sleepiness were considered the main predictors.[10] Sterkenburg et al., investigated psychosocial aspects such as education, job, marriage, offspring, and having a driver's license in 89 CP patients and reported that only 2 of the 89 patients surviving to adulthood had offspring (both without HI).[11]

V. Other determinants of QoL

Impaired hypothalamic-pituitary axis (HPA) in patients with childhood onset CP predispose them to QoL consequences that are more severe than adult onset CP, because they haven't established their career, professional lives and family yet. Various studies report some clinical parameters such as female sex, recurrent surgery, obesity, visual field defects and radiotherapy to have adverse effects on QoL. Majority of young adults with childhood onset CP have difficulty developing relationships, separating from their parents and may prefer continuing family support over an independent life.[10] Several studies suggest that the neurobiological cause of the low QoL is involvement of hypothalamus.[7,11] The most common issues in paediatric patients' everyday life include incapability to control emotions, low self-confidence, problems in pursuing education and unease related to physical appearance. Other hardships these patients face include decreased mobility, less self-care, depression, anxiety, withdrawal and somatic complaints such as pain.[12] Neurocognitive deficits such as reduced attention span, memory related problems, sluggish cognitive speed and behavioural instability are considered as precursors to weak academic performance. Considering that intellectual stability has been described in around 82% of patients, visual memory is till defective despite normal visual spatial functioning.[13] Mehren et al., suggested that childhood onset CP patients are at increased risk for apathy, a condition associated with reduced activities of daily living and poor treatment outcomes due to lack of compliance. Apathetic symptoms vary in individuals depending on the tumour extension and involvement of motivational brain circuits.[14] However, interventions such as efficient psychosocial assistance and self-care guidance may decrease the severity of disease.[10] The importance of improved diagnostic facilities, new techniques and early treatment has also been emphasized as it may improve the chances of better QoL in these patients.

Conclusion

Quality of life in craniopharyngioma patients is significantly impaired even after surgical resection and besides a close endocrine follow up, requires physical, psychological and psychosocial rehabilitation.

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