Pediatric craniopharyngioma

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Craniopharyngiomas (CP) are rare malformation tumors. Clinical presentation and outcome of pediatric patients with CP regarding age at diagnosis is unclear.

Multicenter observational retrospective cohort studies

A Multicenter national study aimed to describe the characteristics of patients with childhood-onset craniopharyngioma and to analyze factors that impair quality of life (QoL) in this population.

This study included patients treated between 2008 and 2022, from 2 to 25 years of age diagnosed with craniopharyngioma. QoL was assessed once during patient's follow-up by age-adapted versions of Pediatric Quality of Life Inventory (PedsQL) questionnaire.

Sixty-six patients were included. Median age at diagnosis was 5 years (interquartile range [IQR]: 3-8), while median follow-up was 7.4 years (IQR: 2.8-9.7). Most craniopharyngioma were suprasellar (93.9%), and 59.7% had hypothalamic involvement (HI). All patients underwent surgery, 44.4% received radiotherapy, and 23.6% intracystic therapy. Most frequent long-term complications were visual deficit (72.7%) and endocrine impairment (94.5%). Patients exhibited hypothyroidism requiring hormone replacement (92.4%), hypocortisolism (80.3%), diabetes insipidus (86.4%), and/or growth hormone therapy (50%). When parents evaluated QoL, PedsQL median score was 53.8 points out of 100 (IQR: 41-71.6). Higher scores were noted when patients assessed their own QoL (median score 64.8 [IQR: 57.3-81.8]), observing statistically significant differences ($p = .019$). QoL was impaired by repeated surgeries ($r = -.44$; $p = .014$), HI (median score 51.5 [IQR: 39-63.8] vs. 76.4 [59-84.8]; $p = .001$), radiotherapy (median score 51.9 [IQR: 38.1-61.3] vs. 63.8 [IQR: 49-82.5]; $p = .02$) and longer follow-up ($r = -.3$; $p = .01$).

Most patients had significant comorbidities and low overall QoL scores, which was mainly affected by repeated surgery, radiation, and hypothalamic involvement. This reflects the need for further

research and intensified studies of systemic therapy/alternate strategies to broaden the standard-of-care options, so that treatment-related sequalae can be avoided [1)]

Case series

conducted a retrospective data collection of all pediatric patients (<18 years) treated for a skull base lesion at the Division of Pediatric Neurosurgery, University Children's Hospital Basel, Switzerland, between 2015 and 2021. Descriptive statistics and a systematic review of the available literature were additionally conducted.

Results: We included 17 patients with a mean age of 8.92 (±5.76) years and nine males (52.9%). The most common entity was sellar pathologies (n = 8 47.1%), with craniopharyngioma being the most common pathology (n = 4, 23.5%). Endoscopic approaches, either endonasal transsphenoidal or trans ventricular, were used in nine (52.9%) cases. Six patients (35.3%) suffered from transient postoperative complications, while in none of the patients, these were permanent. Of the nine (52.9%) patients with preoperative deficits, two (11.8%) showed complete recovery and one (5.9%) partial recovery after surgery. After screening 363 articles, we included 16 studies with a total of 807 patients for the systematic review. The most common pathology reported in the literature confirmed our finding of craniopharyngioma (n = 142, 18.0%). The mean PFS amongst all the studies included was 37.73 (95% CI [36.2, 39.2]) months, and the overall weighted complication rate was 40% (95% CI [0.28 to 0.53] with a permanent complication rate of 15% (95% CI [0.08 to 0.27]. Only one study reported an overall survival of their cohort of 68% at five years.

Conclusion: This study highlights the rarity and heterogeneity of skull base lesions in the pediatric population. While these pathologies are often benign, achieving GTR is challenging due to the deep localization of the lesions and eloquent adjacent structures, leading to high complication rates. Therefore, skull base lesions in children require an experienced multidisciplinary team to provide optimal care [2)].

The objectives of the study were to identify preoperative prognostic factors in patients with craniopharyngiomas and to develop a risk-based treatment algorithm.

The authors reviewed data obtained in a retrospective cohort of 66 children (mean age 7.4 years, mean follow-up period 7 years) who underwent resection between 1984 and 2001. Postoperative recurrence rates, vision status, and endocrine function were consistent with those reported in the literature. The postoperative morbidity was related to hypothalamic dysfunction. The preoperative magnetic resonance imaging grade clinically assessed hypothalamic function, and the surgeon's operative experience ($p = 0.007$, $p = 0.047$, $p = 0.035$) significantly predicted poor outcomes. Preoperative hypothalamic grading was used in a prospective cohort of 22 children (mean age 8 years, mean follow-up period 1.2 years) treated between 2002 and 2004 to stratify patients according to whether they underwent gross-total resection (GTR) (20%), complete resection avoiding the hypothalamus (40%), or subtotal resection (STR) (40%). In cases in which residual disease was present, the patient underwent radiotherapy. There have been no new cases of postoperative hyperphagia, morbid obesity, or behavioral dysfunction in this prospective cohort.

For many children with craniopharyngiomas, the cost of resection is hypothalamic dysfunction and a poor QOL. By using a preoperative classification system to grade hypothalamic involvement and stratify treatment, the authors were able to minimize devastating morbidity. This was achieved by

identifying subgroups in which complete resection or STR, performed by an experienced craniopharyngioma surgeon and with postoperative radiotherapy when necessary, yielded better overall results than the traditional GTR [3].

To date, however, the Puget system has not been externally validated.

A panel of 6 experts, consisting of pediatric neurosurgeons and pediatric neuroradiologists, graded 30 preoperative and postoperative MRI scans according to the Puget system. Interrater reliability was calculated using Fleiss' κ and Krippendorff's α statistics.

Interrater reliability in the preoperative context demonstrated moderate agreement (κ = 0.50, α = 0.51). Interrater reliability in the postoperative context was 0.27 for both methods of statistical evaluation.

Interrater reliability for the system as defined is moderate. Slight refinements of the Puget MRI grading system, such as collapsing the 3 grades into 2, may improve its reliability, making the system more generalizable [4]

The aim of this cohort study was to determine clinical presentation and outcome in these patients diagnosed at different ages at diagnosis.

Design: Seven hundred nine patients diagnosed with CP were recruited 1999-2021 in HIT-Endo, KRANIOPHARYNGEOM 2000/2007/Registry 2019 and prospectively observed.

Methods: Age at diagnosis was categorized as infants&toddlers (<2y), early childhood (2-<6y), middle childhood (6-<12y), and early adolescence (12-<18y). Overall and event-free survival (EFS), functional capacity (FMH), and quality of life (QoL) (PEDQOL) were assessed.

Results: Severe obesity (BMI >3SDS) was prevalent in 45.4% at the last visit. Lower EFS but better QoL was observed in children with age at diagnosis <6y compared to ≥ 6y. Reduced functional capacity percentiles were associated with increased BMI-SDS at last visit (rho=-0.125, 95% CI [-0.21; -0.04]) and age at diagnosis <2y. Posterior hypothalamic involvement (HI) and hypothalamic lesion (HL) are independent risk factors for reduced event-free survival (HR=1.59, 95% CI [1.12; 2.26]) and obesity at last visit (OR=2.94, 95% CI [1.73; 5.08]). Age at diagnosis did not contribute to severe obesity and reduced QoL.

Conclusions: Diagnosis of CP at an age <6 may help patients adapt early to disabilities, but may lead to a higher probability of CP relapse. Not age at diagnosis but posterior HL may be the contributing factor to severe obesity and a reduced QoL [5].

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