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EDITORIAL

Craniopharyngiomas: a life-changing tumor

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Craniopharyngioma is the most common lesion of the hypothalamic and suprasellar region in childhood with a frequent compromised pituitary function. The reported rates for deficits of anterior pituitary hormones and for the antidiuretic hormone (ADH) are 38–95 and 6–38%, respectively [1].

The optimal treatment of patients with craniopharyngioma remains controversial. The quality of life should be considered as a clinically important endpoint in patients, who currently experience good overall survival rates, regardless of the degree of surgical resection. Radical resection is associated with the best outcome in terms of survival and recurrence-free survival [1]. Nevertheless, aggressive behavior, location, involvement of critical structures, tumor size, calcifications, and patient age may limit the extent of resection [2, 3]. Long-term sequelae reduce the quality of life in 50% of long-term survivors, notably obesity and neurobehavioral impairment due to hypothalamic involvement and iatrogenic induced lesions.

Surgery can also carry significant morbidity in terms of visual, hypothalamic, and endocrinological disturbances [3]. The frequency of individual hormone deficits ranges from 88 to 100% for growth hormone, 80 to 95% for gonadotropins, 55 to 88% for adrenocorticotrophic hormone, 39 to 95% for thyroid-stimulating hormone, and 25 to 86% for ADH. Moreover, at least three pituitary hormone deficiencies have been reported in 54–100% of patients undergoing surgery for craniopharyngioma [3]. In our series recovery from a preoperative pituitary deficit never occurred after surgery and the preservation of a normal urinary concentrating ability after surgery occurred in 33.3% of cases. Interestingly, 6.4% of the patients with preoperative diabetes insipidus regained a normal urinary concentrating capacity [3].

The treatment of patients with craniopharyngiomas changed during the decades and it shifted from attempting a total removal to a less aggressive surgery [4]. Radical resection is considered the primary therapy of choice. Nevertheless, the gross total removal of craniopharyngiomas in reported series is extremely variable and the reported rate of critical hypothalamic adherence or infiltration is up to 26.8 % of cases [4]. The definition of such a relationship between the tumor and the hypothalamus on preoperative magnetic resonance imaging (MRI) is strongly recommended to plan the treatment strategy [5]. The involvement of mammillary bodies, the hypothalamic hyperintensity, and retrochiasmatic tumor extension have proved to be useful to define the hypothalamus invasion [5]. The relationship between obesity and hypothalamic damage has been documented in few studies of hypothalamic lesions in man [6]. In the largest series postoperative obesity after surgery, was reported in 15–52% of cases. No reports are available both on the rate of new-onset obesity after surgery and on the normalization of body mass index (BMI) in craniopharyngioma patients with BMI before surgery above or below the normal values [4, 7].

Impairment of hypothalamic functions may result in hyperphagia with obesity, disorders of thirst and water balance, cognitive impairment, disorders of temperature control, and sleep [3–5, 7, 8]. Overall, postoperative obesity is reported in 26–61% of the patients after surgery alone or combined with radiotherapy [3]. Some authors [3] showed

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that postoperatively assessed destruction of the wall of the third ventricle at MRI correlated positively with weight gain at follow-up.

Other authors analyzed the weight increase in relation to the preoperative hypothalamic impairment to find prognostic factors and suggested a grading system of hypothalamic involvement by a radiological score. Moreover, it has been reported that hypothalamic signal changes in T2-weighed MRI and the irregular contrast enhancement predicted the hypothalamic involvement. It seems evident that the chance of hypothalamic damage increases in tumors extended beyond the mammillary bodies [8]. These data highlight the usefulness of a topographical classification of craniopharyngiomas to predict the risks of hypothalamic injury associated with tumor resection and a classification based on tumor relationships to the walls of the third ventricle was proposed [4]

Factors associated with hypothalamic morbidity are young age and, hypothalamic symptoms at presentation, signs of hypothalamic invasion on neuroimaging, a vertical diameter greater than 3.5 cm from the midline, attempts to remove adherent tumor, multiple surgical procedures for recurrence, and a radiation dose greater than 51 Gy delivered to the hypothalamus [9].

The worsening of the cognitive function in patients with craniopharyngioma contributes to poor academic performance, and impaired quality of life in 40% of cases in a 10-year follow-up. It has been reported that 16% of the adults and 26% of the children did not achieve an independent living status with a poor social integration and work or school performance [59]. In another study, the cumulative probability of permanent motor deficits was 11%, epilepsy 12%, psychological disorders requiring treatment 15%, and total dependency for daily activities 9% at 10-years follow-up [1]. Some authors reported that 47% of patients experienced long-term psychosocial disability, while 49% had neurological morbidity such as concentration deficits, personality changes, epilepsy, or memory loss [10]. In our experience this last aspect has a great relevance, and deserves a dedicated surgical approach without callosotomy and without dissection of deep structures involved in memory functions [7].

Data on the treatment options with the least impact on neurobehavioral outcome are still lacking and the assessment of the strategy providing the best functional outcome is difficult. Aggressive surgical resection was associated with a better neuropsychological performance, while others reported that patients treated by conservative surgery with adjuvant irradiation experienced the most consistent return to school and achievement of tertiary education or employment [4]. Some authors compared neurophysiologic results in patients treated either by aggressive, or conservative surgery with or without adjuvant radiotherapy. They found no differences between the two groups in terms of probability of new postoperative morbidities such as hyperphagia, epilepsy, dependency for basal daily activities, and deterioration of work or school status [1]. Extent of surgery and radiation dose seems to affect long-term cognitive function mostly in pediatric population. Tumor location, time to presentation, number of surgical procedures, hydrocephalus influenced baseline and longitudinal intelligence quotient [4]. Based on most recent literature results accepted for publication in this journal, it is strongly advisable to organize a multidisciplinary team of expert neurosurgeons, endocrinologists, and radiation oncologists to discuss treatments combinations and adopt specific strategies for children with craniopharyngioma.

Compliance with ethical standards

Conflict of interest The authors declare that they have no competing interests.

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