Craniopharyngioma outcome

see also Hypothalamic obesity

Studies investigating long-term health conditions in patients with craniopharyngioma are limited by short follow-up durations and do generally not compare long-term health effects according to initial craniopharyngioma treatment approach. In addition, studies comparing long-term health conditions between patients with childhood- and adult-onset craniopharyngioma report conflicting results [1].

These tumors are histologically benign, corresponding to WHO grade I and are usually not life threatening. Benign lesions in the brain are considered cured when amenable to surgical resection, however even gross total resection of craniopharyngioma is not always curative and may be associated with significant additional morbidities [2].

Tumor size, hypothalamic involvement (HI), and obesity are associated with reduced overall survival (OS) and functional capacity (FC).

The overall survival rates are high (92%) but occurrences of reduced quality of life are also high. Recurrences after complete resection and progressions of residual tumor after incomplete resection are frequent postsurgical events. Because irradiation is efficient in preventing tumor progression, appropriate timing of postsurgical irradiation is currently under investigation in the randomized multinational trial KRANIOPHARYNGEOM 2007 that analyzes quality of life as primary endpoint.

CPs have the highest mortality of all pituitary tumours. The standardised overall mortality rate varies from 2.88 to 9.28 in cohort studies. Adults with CP have a 3-19-fold higher cardiovascular mortality in comparison to the general population. Women with CP have an even higher risk. The long-term morbidity is substantial with hypopituitarism, increased cardiovascular risk, hypothalamic damage, visual and neurological deficits, reduced bone health and reduction in quality of life and cognitive function [3].

Patients treated for childhood craniopharyngioma often develop hypothalamic obesity (HO), which has a huge impact on the physical condition and quality of life of these patients.

Severe obesity is associated with pathological eating behavior/disorders in craniopharyngioma patients. As these disorders are not disease-specific, risk factors for hypothalamic obesity should be the focus of further craniopharyngioma research [4].

Treatment for HO thus far has been disappointing, and although several different strategies have been attempted, all interventions had only transient effects [5].

Seventy-eight patients (67% White; 41 males, 37 females; mean age 10y 8mo, SD 3y 11mo, range 6-20y) with craniopharyngioma were assessed for tumor extent and diabetes insipidus. All patients underwent overnight polysomnography and multiple sleep latency tests after surgical resection. Executive functioning was assessed using parent-reported measures. Patients and their parents completed measures of HRQoL. None had a history of previous radiation therapy.

Path analysis was used to test hypothesized relations while controlling for demographic and disease characteristics. Analyses revealed poorer parent-reported HRQoL among young people with greater

executive functioning symptoms (estimate -0.83; p<0.001). Direct and indirect effects were found among diabetes insipidus, executive functioning, and parent-reported HRQoL. Diabetes insipidus directly predicted greater global executive functioning impairment (estimate 5.15; p=0.04) and indirectly predicted lower HRQoL through executive functioning impairment (estimate -4.25; p=0.049). No significant effects were found between excessive daytime sleepiness, tumor hypothalamic involvement, diabetes insipidus, executive functioning, and patient-reported HRQoL.

These findings suggest that young people with craniopharyngioma presenting with diabetes insipidus may benefit from targeted neurocognitive and psychosocial screening to inform interventions [6].

Recurrence

see Craniopharyngioma recurrence.

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