



Craniopharyngioma – a childhood and adult disease with challenging characteristics

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Edited by:

Shlomo Melmed, Burns and Allen Research Institute, USA

Craniopharyngiomas are rare embryogenic malformations of the sellar area with low-grade histological malignancy. Despite high survival rates (87–95% in recent series), quality of life is frequently impaired in long-term survivors due to sequelae caused by the anatomical proximity of the tumor to the optic nerve, pituitary gland, and hypothalamus – in many cases even entanglement with the hypothalamus (Cohen et al., 2011; Müller, 2011; Zoicas and Schöfl, 2012). One of the most serious quality of life complications of craniopharyngioma is hypothalamic obesity or so-called “hypothalamic syndrome,” a problem that can manifest before and/or after treatment. Novel insights into mechanisms of neuroendocrine satiety regulation and the pathogenic relevance of the autonomous nervous system are expected to facilitate future therapeutic approaches for hypothalamic syndrome (Elfers and Roth, 2011; Lustig, 2011; Roth, 2011).

For the time being, current treatment options for craniopharyngioma patients suffering from hypothalamic syndrome are limited, the most effective to date being bariatric surgery. Bariatric surgery is tolerable and effective in weight-reduction for severely obese adult craniopharyngioma patients (Bingham et al., 2012), but considered intrusive and therefore controversial for younger patients. A substantial improvement in prognosis of craniopharyngioma will require the development of risk adapted neurosurgical (Flitsch et al., 2011; Bartels et al., 2012; Puget, 2012; Trippel and Nikkhah, 2012) and radiooncological (Kortmann, 2011) treatment strategies in a multidisciplinary approach. Recent multicenter cooperation in this area has already led to beneficial results.

The consequences of both the surgical treatment and post-surgical management of the disease are as complicated and hypothalamic-intertwined as the tumors themselves. Risk adapted surgical strategies at initial diagnosis should aim at a maximal degree of resection, respecting the integrity of optical and hypothalamic structures in order to prevent severe sequelae and therein minimize consequences that could negatively exacerbate patient quality of life. Because initial hypothalamic tumor involvement typically has an *a priori*, life-long effect on the clinical course (Müller, 2011) experienced by the patient, childhood, and adult onset craniopharyngioma should be recognized as chronic diseases requiring constant monitoring of the consequences and developing medical

resources for treatment in order to provide not only optimal quality of life for patients, but also to garner additional information with the intent of minimizing what at present are severe consequences of both the disease and its treatment.

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Received: 21 May 2012; accepted: 31 May 2012; published online: 04 July 2012.

Citation: Müller HL (2012) Craniopharyngioma – a childhood and adult disease with challenging characteristics. *Front. Endocrinol.* 3:80. doi: 10.3389/fendo.2012.00080
This article was submitted to *Frontiers in Pituitary Endocrinology*, a specialty of *Frontiers in Endocrinology*.

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