Hydrocephalus and Hypothalamic Involvement in Pediatric Patients with Craniohypophyseal Glioma or Cysts of Rathke’s Pouch: Impact on Long-term Prognosis

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Introduction

Pediatric patients with sellar masses such as craniohypophyseal glioma (CP) or cyst of Rathke’s pouch (CRP) frequently suffer disease- and treatment-related sequelae. We analyzed the prognostic relevance of initial hydrocephalus (HY) and hypothalamic involvement (HI) on long-term survival and functional capacity (FC) in pediatric CP or CRP.

Subjects and methods

Using retrospective analysis of patient records, presence of initial HY or HI was assessed in 177 pediatric patients (163 CP, 14 CRP). Twenty-year overall survival (OS) and progression-free survival (PFS), functional capacity, and body mass index (BMI) were analyzed with regard to initial HY, degree of resection, or HI.

Results

105 patients (103/163 CP, 2/14 CRP) presented with initial HY and 96 presented with HI. OS, PFS, and FC were not affected by HY at initial diagnosis. Initial HI (96/177) had major impact on long-term prognosis. HI was associated with lower OS (0.84±0.04), lower FC (p=0.003), and higher BMI at dgx and last follow-up (p=0.000) when compared to pts. without HI (OS: 0.94±0.05). PFS was not affected by HI or degree of resection.

Conclusion

Initial HY has no impact on outcome in patients with sellar masses. OS and FC are impaired in survivors presenting with initial HI. PFS is not affected by HY, HI, or degree of resection. Accordingly, gross-total resection is not recommended in sellar masses with initial HI to prevent further hypothalamic damage.

Characteristics of 177 pediatric patients with sellar masses (163 craniohypophyseal glioma, 14 cysts of Rathke’s pouch) diagnosed 1966-2001 and recruited in HIT Endo. HY hydrocephalus, HI hypothalamic involvement, VP ventriculo-peritoneal, FMI+ ability scale, BMI body mass index. *p < 0.05; **p < 0.01; ***p < 0.001.

Overall survival and hypothalamic involvement

20-yr overall survival and progression-free survival rates of 177 patients (163 CR, 14 CRP) diagnosed between 1966 and 2001 related to MRI findings of hypothalamic involvement (HI) at the time of primary diagnosis.

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