History before Diagnosis in Childhood Craniopharyngioma: Associations with Initial Presentation and Long-term Prognosis

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Introduction

Childhood craniopharyngiomas (CP) are often diagnosed after long duration of history (DOH). Tumor size, hypothalamic involvement (HI), and obesity are associated with reduced overall survival (OS) and functional capacity (FC). The effect of DOH and specific symptoms in history on presentation at initial diagnosis and long-term prognosis are unknown.

Patients and Methods

Histories of 411 CP patients recruited in HIT Endo, KRANIOPHARYNGEOM 2000 were retrospectively evaluated for DOH, symptoms and characteristics. The effect of specific manifestations and DOH on clinical presentation and tumor characteristics at time of initial CP diagnosis and long-term outcome were analyzed.

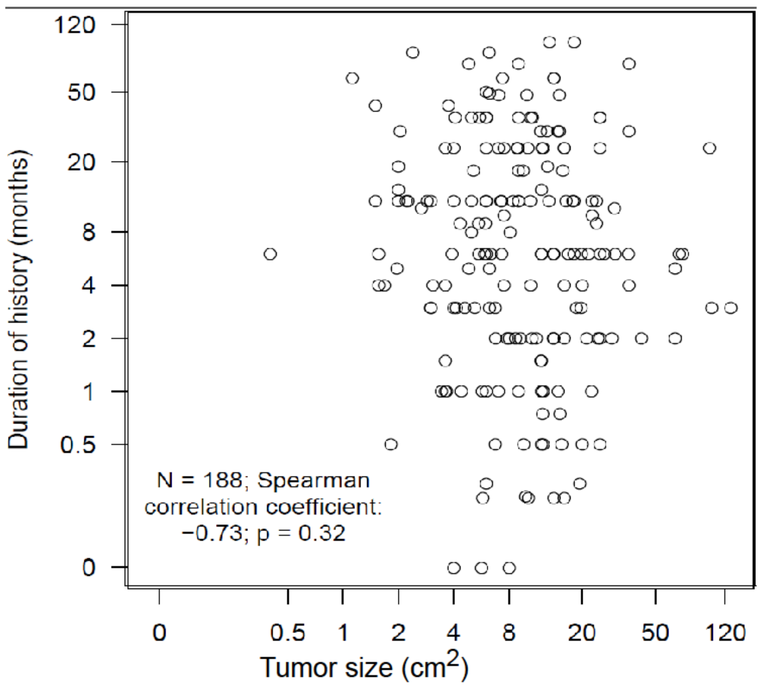

Figure 1A) For the whole CP patient group, no significant correlation was detected between tumor size at initial diagnosis and duration of history before initial CP diagnosis.

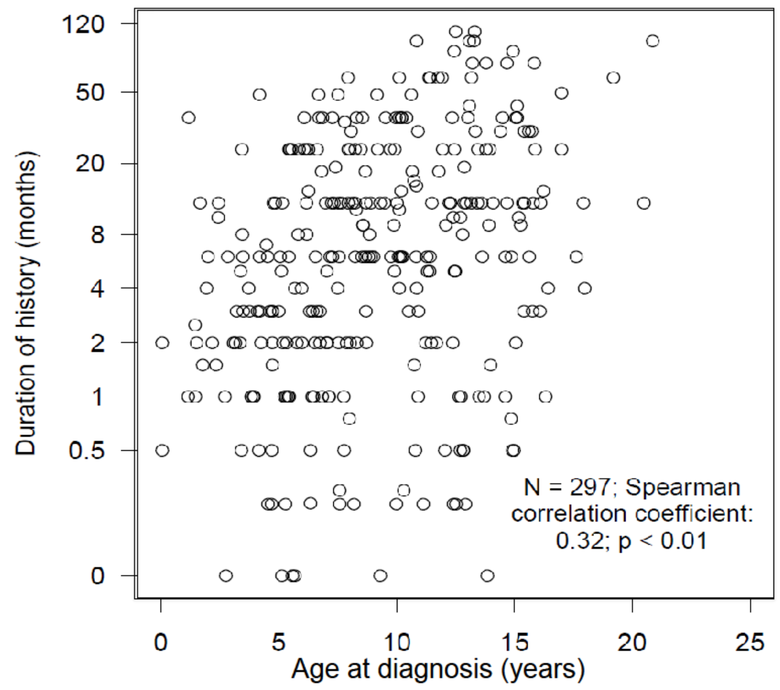

Figure 1B) Duration of history before initial CP diagnosis was positively correlated with patients' age at initial diagnosis.

Table 1: Duration of history before initial diagnosis of childhood-onset CP in 411 patients in regard to patients' characteristics at the time of initial diagnosis and at last visit. BMI SDS according to the references of Rolland-Cachera et al.

Clinical criteria		N (yes/no)	Duration of history (mo) in pts with criteria	Duration of history (mo) in pts without criteria	p
Visual impairment at diagnosis	(y/n)	161/130	6 (0.0–108.0)	10 (0.0–108.0)	0.14
Hydrocephalus at diagnosis	(y/n)	136/126	6 (0.0–108.0)	12 (0.3–96.0)	0.01
Hypothalmic Involvement	(y/n)	23/99	6 (0.3–108.0)	5 (0.3–108.0)	0.33
Tumor location	Intrasellar	5	4 (0.0–12.0)		0.278
	Suprasellar	61	9 (0.0–84.0)		
	Intra+Supra	169	6 (0.0–96.0)		
Tumor size (cm²) at initial diagnosis	≤ 20	172	6 (0.0–96.0)	6 (0.0–96.0)	0.66
	> 20	32	6 (0.0–96.0)	6(0.5–72.0)	
Endocrine deficits at diagnosis	(y/n)	123/167	9 (0.0–108.0)	6 (0.0–96.0)	0.01
Degree of initial tumor resection	incomplete	213	10 (0.0–108.0)	6 (0.0–108.0)	0.07
	complete	136			
BMI > 3 SD at last visit	≤ 3 SD	179	6 (0.0–84.0)	6 (0.0–108.0)	0.35
	> 3 SD	175			
BMI > 4 SD at last visit	≤ 4 SD	220	6 (0.0–84.0)	7 (0.0–108.0)	0.28
	> 4 SD	134			

Results

Median DOH was 6 months (range: 0.1–108 mo) and correlated with age at diagnosis. Tumor size, HI, degree of resection, and BMI at diagnosis were not related to DOH. In multivariate analysis adjusted for age at diagnosis, only hydrocephalus was found to have a significant influence on DOH (p= 0.01). Visual and neurological deficits were associated with larger initial tumor size and impaired 10-yr OS. Weight gain and growth failure were observed with longest DOH. PFS and FC were not related to any specific symptom. Endocrine deficits at diagnosis were associated with long DOH.

Conclusions

CP is frequently diagnosed after long DOH, especially in older children. However, DOH was not associated with tumor size, HI, survival or FC.

Visual and neurological deficits necessitate rapid diagnostic work-up.

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